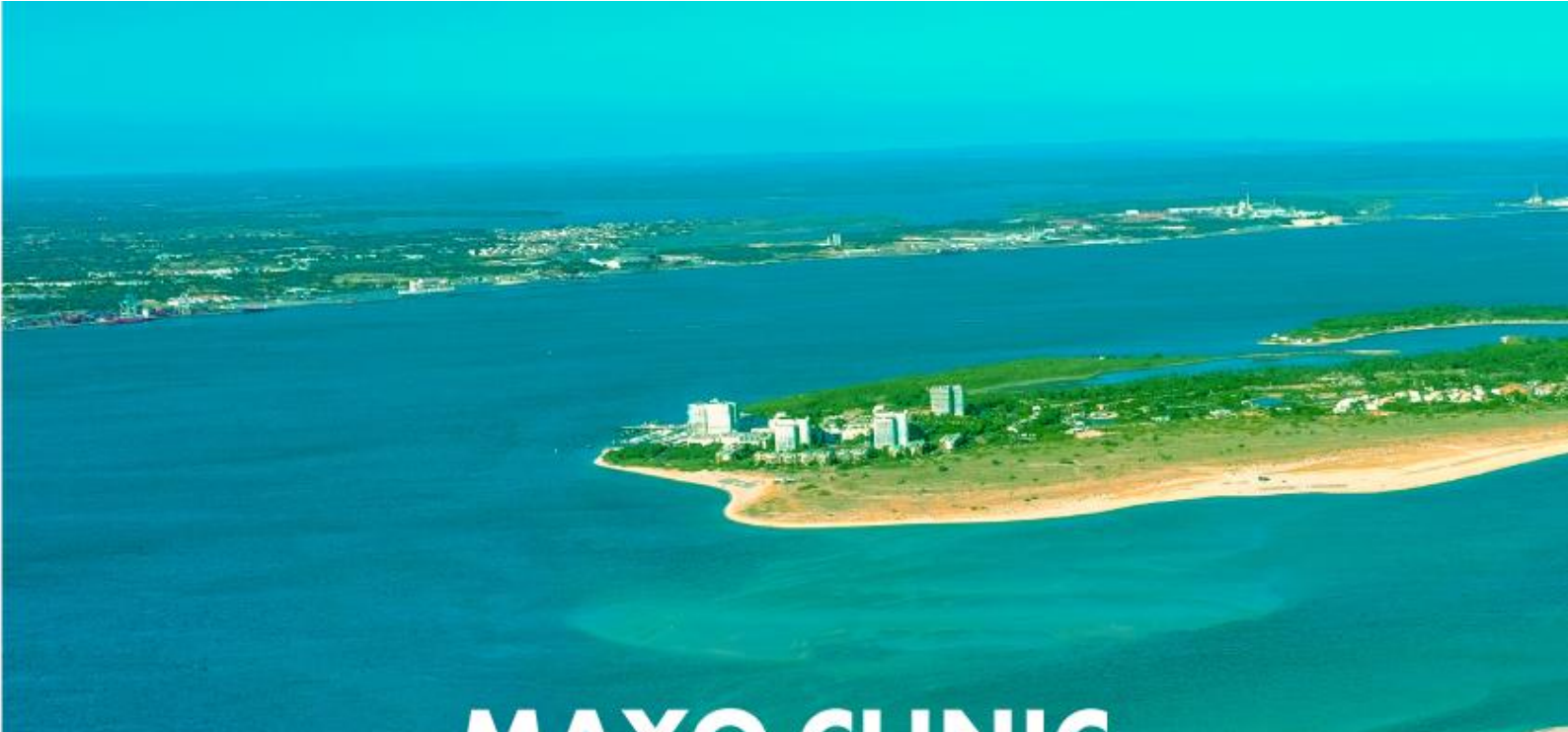

MAYO CLINIC NEPHROLOGY UPDATE

27th Annual Symposium on Renal Diseases

SCIENTIFIC EXCELLENCE IN A UNIQUE SETTING

June 26-27, 2026 | The Anchor Troia, Portugal

ABSTRACT BOOK

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ABSTRACTS

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OC 39

LIFE-THREATENING INFLUENZA A-ASSOCIATED THROMBOTIC MICROANGIOPATHY AND HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS: A RARE CASE WITH SUCCESSFUL HEMATOLOGIC AND RENAL RECOVERY

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Unidade Local de Saúde de Coimbra

Category: Glomerular diseases

Background

The coexistence of thrombotic microangiopathy (TMA) and hemophagocytic lymphohistiocytosis (HLH) is rare, particularly when triggered by Influenza A. This infection may induce both entities through excessive immune activation, complement dysregulation, and endothelial injury, leading to severe multiorgan dysfunction.

Case description

A 22-year-old previously healthy man presented with fever, odynophagia, myalgias, and cola-colored urine. He was hemodynamically stable. Laboratory evaluation revealed anemia, thrombocytopenia, elevated lactate dehydrogenase and bilirubin, schistocytes on peripheral smear, negative direct antiglobulin test, normal ADAMTS13, marked hyperferritinemia, hypofibrinogenemia, elevated inflammatory markers, and non-oliguric acute kidney injury (Table 1). Imaging showed hepatosplenomegaly. A diagnosis of TMA was established, and an HScore of 181 indicated a high probability of HLH. Influenza A was identified as the trigger.

The patient was admitted to the intensive care unit due to multiorgan involvement. Oseltamivir, antibiotics, and anakinra were initiated immediately, and eculizumab was started on day 3. Renal function worsened, requiring two hemodialysis sessions. Further evaluation revealed nephrotic-range proteinuria (protein/creatinine ratio 20,110 mg/g) and hypoalbuminemia (2.8 g/dL). Autoimmune, monoclonal, and additional infectious causes were excluded (Table 1).

After infection control, corticosteroid therapy (prednisolone 1 mg/kg/day) was started for nephrotic syndrome. Kidney biopsy showed membranoproliferative glomerulonephritis with TMA lesions, low-level immunoglobulin IgG, IgM and complement C3 deposition, and diffuse podocyte effacement on electron microscopy. Complement genetic testing identified a previously undescribed heterozygous CFHR4 missense variant, possibly conferring susceptibility to atypical hemolytic uremic syndrome (aHUS).

During follow-up, inflammatory markers and HLH resolved, allowing discontinuation of anakinra. The patient remained dialysis-independent with progressive renal recovery. At discharge, he remained on eculizumab and prednisolone. Persistent proteinuria (3,600 mg/24h) after 10 weeks of steroids led to rituximab (1 g, two doses, 15 days apart), followed by rapid steroid tapering. At last follow-up, 4 weeks after the second rituximab dose, while on prednisolone 10 mg and eculizumab, serum creatinine was 1.87 mg/dL, serum albumin was normal, and 24-hour proteinuria had decreased to 1,768 mg.

Discussion and learning point

This case illustrates Influenza A as a rare trigger of concomitant TMA and HLH in a healthy young adult with an unknown CFHR4 variant. Histology demonstrated overlapping immune complex deposition, endothelial injury, and podocytopathy. The interplay between interferon-driven inflammation and complement activation likely represents a shared pathogenic mechanism linking HLH and TMA in viral infections.

A tailored multidisciplinary approach combining anakinra for HLH, eculizumab for TMA, and corticosteroids plus rituximab for persistent nephrotic syndrome resulted in HLH resolution, dialysis independence, and significant renal recovery. This case underscores the importance of early recognition and combined immunomodulatory therapy in life-threatening virus-associated thromboinflammatory syndromes.

Table 1: Initial diagnostic work-up.

On admission

Variable	Result	Normal range	Variable	Result	Normal range
Hemoglobin (g/dL)	11.5	13.5 – 17.5	Sodium (mmol/L) / Potassium (mmol/L)	134 / 3.7	136 – 146 / 3.5 – 5.1
MCV (fl) / MCH (pg)	79.0 / 28.4	80 – 100 / 26 – 34	Total protein (g/dL) / Albumin (g/dL)	6.3 / 3.9	6.6 – 8.3/ 3.5 – 5.2
White-cell count ($x10^3$/ mm^3)	6.9	4 - 10	Total bilirubin (mg/dL) / Direct bilirubin (mg/dL)	3.5 / 0.6	<1.2 / <0.5
Platelet count ($x10^3$/mm^3)	4	150 - 450	Alkaline phosphatase (U/L)	119	30 – 120
Peripheral blood smear	Presence of schistocytes	< 2 schy	Aspartate aminotransferase (U/L) / Alanine aminotransferase (U/L)	334 / 125	<35 / <45
Prothrombin time (sec)	14.8	9.4 - 12.5	Lactate dehydrogenase (U/L)	3,568	<248
Partial-thromboplastin time (sec)	35.9	23.4 - 35.4	Creatine kinase (U/L)	462	30 – 170
Fibrinogen (mg/dL)	192.0	200 - 500	Triglycerides (mg/dL)	176	43.8 - 195.1
D-Dimer (ng/mL)	>150,000	<500	Ferritin (ng/mL)	56,632.6	30 - 300
Haptoglobin (g/L)	0.66	0.3 - 2	c-Reactive protein (mg/dL) / Procalcitonin (ng/mL)	16.22 / 21.4	< 0.5 / 0-0.5
Direct antiglobulin test	Negative	Negative	Urinalysis • Proteins (mg/dL) • Leukocytes (/HPF) • Red blood cells (/HPF)	 >300 11 55	 <5 <2
Urea (mg/dL)/ Creatinine (mg/dL)	61.0 / 3.56	7.9 - 20.9 / 0.72 - 1.18	Urinary protein to creatinine ratio (mg/g) / Urinary albumin to creatinine ratio (mg/g)	20,110 / 16,425	< 0.2 / < 0.03

Additional studies

Variable	Result	Normal range
ADAMTS13 activity (%)	73%	40 – 130
Complement component 3 (g/L) / Complement component 4 (g/L)	0.86 / 0.20	0.82 - 1.85 / 0.15 - 0.53
Serum cryoglobulins	Negative	
Auto-immune antibodies [a]	Negative	
Serum protein electrophoresis, immunoglobulins, serum and urinary free light chains	Irrelevant changes	
Infectious disease serology [b]	Negative	
Blood and urine cultures	Negative	
Abdominal ultrasound and abdominopelvic Computed Tomography	Hepatomegaly (20cm) and splenomegaly (13cm) with no other relevant changes.	
Transthoracic echocardiography	Irrelevant changes	

a. Anti-nuclear antibodies (ANA), anti-dsDNA antibodies, antineutrophil cytoplasmic antibodies (ANCA), anti-glomerular basement membrane (MBG) antibodies, anti-THSD7A and anti-PLA2R antibodies, Anti-Sm, anti-Ro (SSA), anti-La (SSB), anti-RNP, anti-Scl-70 and anti-Jo-1 antibodies.

b. Human immunodeficiency virus, hepatitis A, B, C and E, Cytomegalovirus, Herpes simplex virus, Epstein-Barr virus, Bartonella henselae, Coxiella burnetii, Toxoplasma gondii, Leptospira, Treponema pallidum, Rickettsia conorii.

OC 84
PARANEOPLASTIC IMMUNOGLOBULIN A VASCULITIS – A MULTIDISCIPLINARY APPROACH

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Category: Systemic diseases

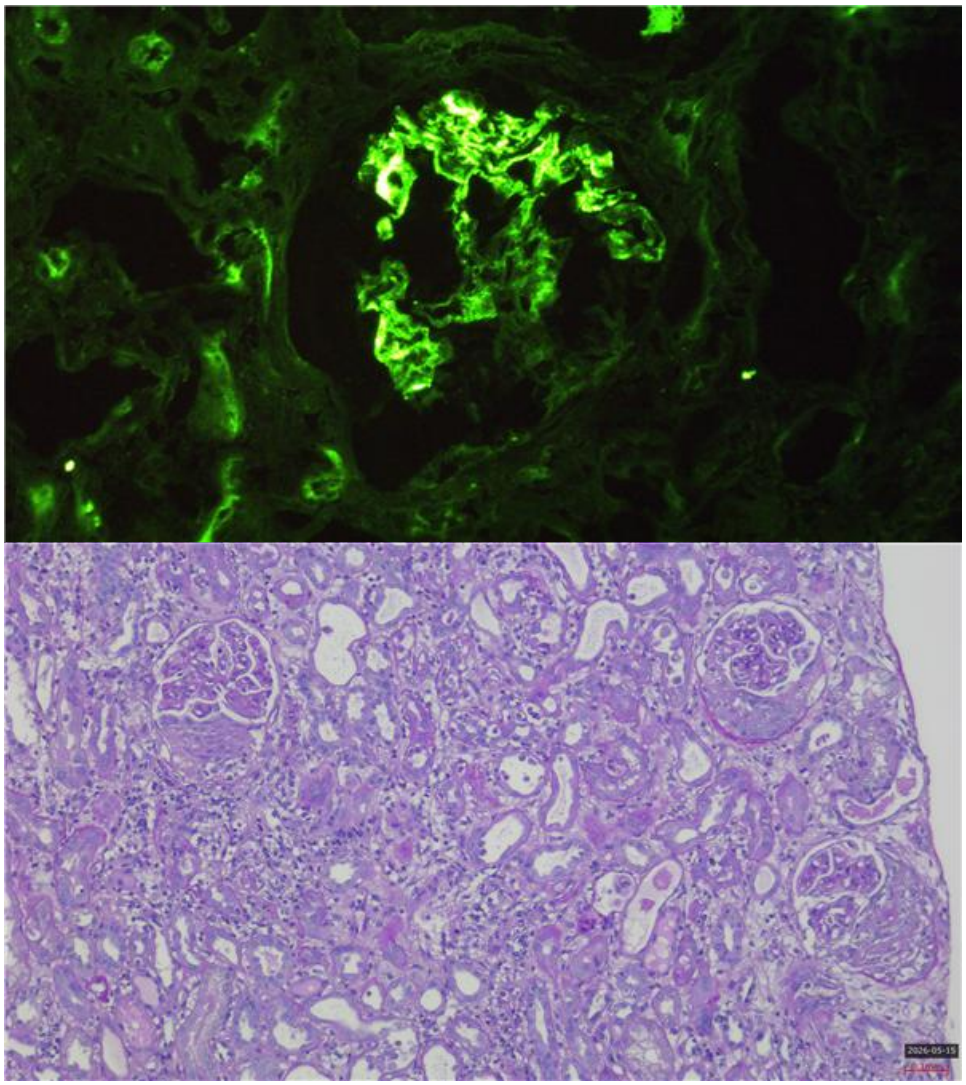
Background: Immunoglobulin A (IgA) vasculitis is rare in adults. Its presentation as a paraneoplastic syndrome is exceptionally uncommon. This case highlights the diagnostic challenge of paraneoplastic IgA vasculitis with renal involvement and underscores the importance of multidisciplinary management.

Case description: An 82-year-old woman with stage IVA colon adenocarcinoma with hepatic metastases, diagnosed in 2024, underwent right hemicolectomy followed by adjuvant chemotherapy with folinic acid, 5-fluorouracil, irinotecan, and bevacizumab from January to June 2025. Shortly thereafter, she developed bilateral leg edema, unresponsive to diuretic. There was a rapid decline in renal function (creatinine from 0.61 mg/dL to 2.60 mg/dL in less than four weeks) and hypoalbuminemia (29.2 g/L). Over the following days, she developed dysuria and macroscopic hematuria, with deterioration of kidney function (creatinine 3.37 mg/dL), erythrocyturia (1968/µL), leukocyturia (33/µL), and nephrotic range proteinuria (albumin-to-creatinine ratio was 14 g/g and protein-to-creatinine ratio 20 g/g). She was admitted to the Nephrology ward for acute kidney injury with hematoproteinuria.

During hospitalization, petechiae were noted. There were no symptoms of respiratory infection; immunologic studies (C3 and C4; anti-nuclear, anti-dsDNA, anti-glomerular basement membrane, anti-neutrophil cytoplasmic and anti-phospholipase A2 receptor antibodies; cryoglobulin test; circulating immune complexes assay; and schistocyte evaluation) and viral serologies were unremarkable. Serum protein electrophoresis showed hypogammaglobulinemia. Urine sediment revealed dysmorphic erythrocyturia, and 24-hour urinary protein and albumin were 7.4 g and 4.8 g, respectively. Kidney biopsy showed small-vessel vasculitis with crescentic membranoproliferative glomerulonephritis (Figure 1), fibrinoid necrosis, and IgA (Figure 2) and C3 mesangial/parietal deposits on immunofluorescence. In the presence of metastatic disease, paraneoplastic IgA vasculitis with rapidly progressive glomerulonephritis was diagnosed; bevacizumab was a possible contributing factor to the nephrotic-range proteinuria.

Renal function deteriorated, with a peak creatinine of 6.64 mg/dL and oligoanuria, necessitating dialysis initiation. No immediate oncologic therapeutic options were available, and after multidisciplinary discussion involving Nephrology, Oncology, and Surgery, immunosuppressive therapy with corticosteroids and cyclophosphamide was initiated. After five cycles of cyclophosphamide, renal function improved (creatinine 2.2 mg/dL), allowing dialysis withdrawal and resumption of chemotherapy, including bevacizumab. By April 2026, creatinine had further improved to 1.5 mg/dL, with concomitant oncologic response, and proteinuria of 537 mg/g.

Discussion and learning points: Paraneoplastic IgA vasculitis should be considered in adults with systemic and renal manifestations, mainly if there is active malignancy. Exclusion of other etiologies and kidney biopsy are key for diagnosis. When immediate treatment of the underlying neoplasm is not feasible, carefully balanced immunosuppression may be kidney- and life-saving. This case illustrates the importance of early recognition and multidisciplinary collaboration in managing rare but potentially reversible paraneoplastic renal syndromes.



OC 20
PEROXIDASIN (PXDN) EXPRESSION IN KIDNEY BIOPSIES OF PATIENTS WITH PATHOGENIC OR HYPOMORPHIC GLA GENE VARIANTS

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KEYWORDS: Fabry disease, Peroxidasin, Chronic kidney disease

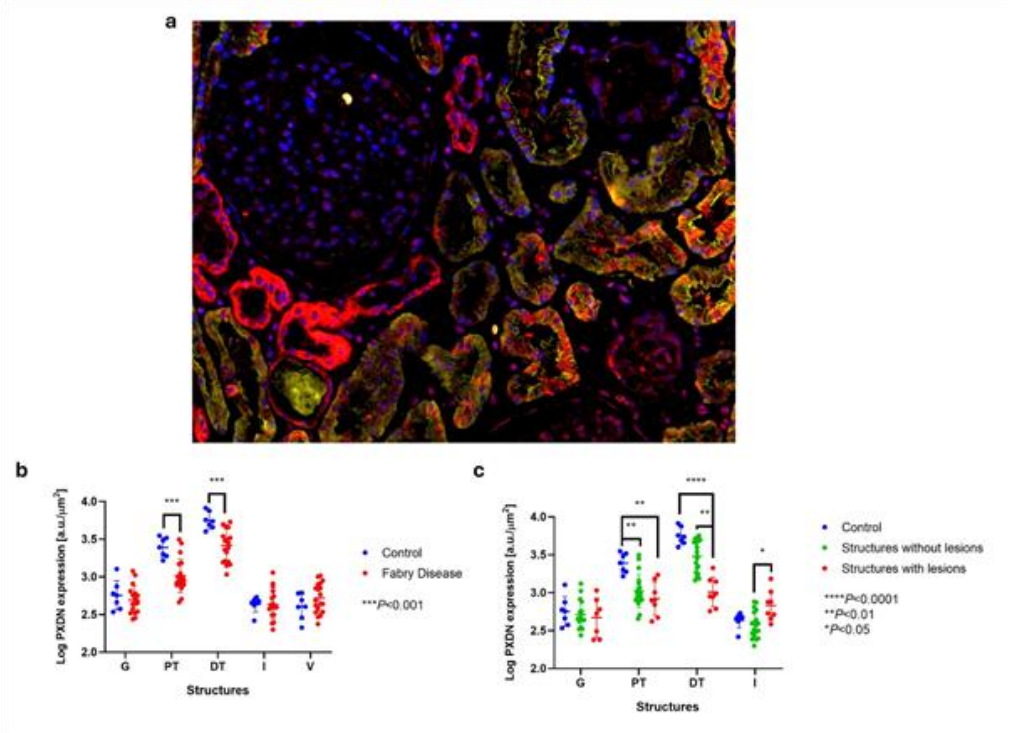
BACKGROUND: Peroxidasin (PXDN) is an enzyme of the peroxidase family essential for extracellular matrix formation and tissue development, with increased expression reported in a transforming growth factor beta (TGF-β)-dependent murine model of kidney fibrosis. Fabry disease (FD) is a rare X-linked lysosomal disorder caused by deleterious variants in the alpha-galactosidase gene (*GLA*). In FD, globotriaosylceramide accumulation in the kidney is associated with increased TGF-β release and epithelial-to-mesenchymal transition, promoting

overexpression of pro-fibrotic mediators and progression to renal fibrosis. Here, we characterize PXDN expression in kidney biopsies (KBx) from patients with *GLA* gene variants (*GLA*-GV).

MATERIALS AND METHODS. Formalin-fixed, paraffin-embedded KBx samples from patients with GLA-GV (n=19) were retrieved from the Department of Anatomic Pathology, São João University Hospital (Porto, Portugal). Clinical and demographic data are summarized in Table 1. Control samples consisted of percutaneous KBx with normal histology (n = 7), obtained from Haukeland University Hospital, University of Bergen (Norway). Periodic acid–Schiff (PAS) staining was performed using standard protocols. PXDN expression was assessed by fluorescence immunohistochemistry (anti-PXDN antibody, Abbexa #abx101906), and proximal tubules were identified by anti-CD15 staining (Ventana/Roche, #760-2504). Whole-slide images were acquired using a Nanozoomer S60 Digital Slide Scanner (Hamamatsu Photonics, Hamamatsu, Japan). Kidney cortex compartments (glomeruli, tubules, vessels, interstitium) were manually segmented in QuPath (https://qupath.github.io/) using brightfield images of fluorescence slides, with PAS sections serving as topographic references to confirm histomorphology, including glomerulosclerosis, tubular atrophy and interstitial fibrosis. Fluorescence intensity was quantified in defined regions of interest using ImageJ/Fiji after rolling ball background subtraction. Statistical analysis employed Mann-Whitney and Kruskall-Wallis tests with Dunn's multiple comparisons (GraphPad Prism 8.0.2).

RESULTS: PXDN was detected in all cortex kidney compartments (Figure 1a), with highest expression in tubules. Compared with controls, PXDN expression was significantly reduced in the tubular compartment of *GLA*-GV patients (Figure 1b). Within the *GLA*-GV group, atrophic tubules showed further reduction in PXDN levels, reaching statistical significance in the distal tubules. Conversely, PXDN expression was increased in the fibrotic interstitium (Figure 1c).

DISCUSSION AND CONCLUSION: These findings implicate PXDN in the pathological processes underlying tubular atrophy and interstitial fibrosis in *GLA*-GV. This study identifies a previously unrecognized role for PXDN in disease progression, supporting further investigation into its mechanistic and potential therapeutic relevance.



OC 30
Utility of the ex vivo endothelial C5b-9 deposition assay in patients with lupus nephritis

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Methods:

In a cohort of biopsy-proven LN (N=14), sera obtained at the time of diagnostic kidney biopsy were tested in an *ex vivo* assay measuring C5b-9 deposition on cultured human microvascular endothelial cells (HMEC-1) by immunofluorescence. Results were dichotomized as positive vs negative according to the predefined positivity threshold used in the dataset (>1.5 increase vs negative control). Patients were compared across demographics and comorbidities; renal parameters (serum creatinine, eGFR, proteinuria, albuminuria, serum albumin, haematuria and urinary erythrocyte dysmorphisms); systemic lupus activity (SLEDAI, anti-dsDNA); biopsy findings (ISN/RPS class; activity and chronicity indices); and complement profile (C1q, C1 inhibitor, C2, C3, C4, C5, soluble C5b-9, factor B, and biopsy C3/C1q deposits).

Results: 29% of patients were ex vivo C5b-9 positive. The most prominent association was with systemic disease activity: SLEDAI was significantly higher in C5b-9–positive vs negative patients (31.25±5.74 vs 13.10±3.54; p=0.006); however, anti-dsDNA did not separate groups (382.28±330.22 vs 346.83±283.15; p=0.224[MJSR1]). [JCC2] Renal function was comparable between groups (serum creatinine 0.77±0.23 vs 0.78±0.34 mg/dL; p=0.8; eGFR 86.25±7.50 vs 84.60±15.09 mL/min; p=0.9[MJSR3] 43). Proteinuria tended to be higher in positives (3.16±3.98 vs 1.12±0.87 g/24h; p=0.6) with numerically higher albuminuria (4852±5351.8 vs 1458.5±2173mg/g creatinine; p=0.2) and slightly lower serum albumin (3.10±0.48 vs 3.79±0.68 g/dL; p=0.086). Haematuria was more frequent in positives (100% vs 50%; p=0.2) and erythrocyte dysmorphisms were numerically more common (75% vs 30%; p=0.2). Histological classification did not differ. Biopsy activity index showed no differences in activity among C5b-9–positive cases (3.00±2.45 vs 6.6±3.2; p=0.075)[JCC4] , while chronicity index was similar (3.5±2.65 vs 2.3±1.25; p=0.5). Conventional complement measures showed limited discrimination: C3 (58.9±16.93 vs 87.04±27.55 mg/dL; p=0.11), C4 (10.05±5.32 vs 14.81±7.58 mg/dL; p=0.3), and soluble C5b-9 (1461.3±677.4 vs 1408.71±403.54 ng/mL; p=0.5).

Conclusions: In this LN cohort, *ex vivo* endothelial C5b-9 deposition identified a subset of patients with markedly higher systemic lupus activity, while standard complement biomarkers (C3/C4 and soluble C5b-9) and renal function measures showed limited discrimination. Future studies with large sample size are of interest in LN and *ex vivo* C5b-9 functional activity.

ABSTRACTS

POSTERS

POSTERS - CLINICAL CASE

PO 04
MEDIUM CUT-OFF HEMODIALYSIS IN MYELOMA CAST NEPHROPATHY: INSIGHTS FROM A SINGLE-CENTER CASE SERIES

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ULS Médio Tejo

Category: Acute Kidney Injury

Background:

Myeloma cast nephropathy (CN) is a major cause of dialysis-dependent acute kidney injury (AKI) driven by high circulating free light chains (FLC). Medium cut-off (MCO) membranes allow enhanced removal of middle molecules, including FLC, but clinical experience in real-world settings remains limited.

Case description:

We report a series of six patients with dialysis-dependent AKI due to presumed or biopsy-proven CN treated in a single center using a standardized protocol combining anti-myeloma therapy and MCO hemodialysis (Table 1).

Baseline FLC levels ranged from 1,550 to 30,000 mg/L. A marked early decline in circulating FLC was observed in most patients during the initial intensive phase of MCO therapy (Figure 1), with four patients achieving reductions greater than 80%. Despite this consistent biochemical response, renal outcomes were heterogeneous. Three patients recovered kidney function, while the remaining did not. Notably, lack of renal recovery was observed in patients with delayed initiation of chemotherapy or treatment interruptions due to infectious complications, despite significant FLC reduction. In contrast, patients who received early and uninterrupted chemotherapy were more likely to recover renal function.

Discussion and learning points:

This case series illustrates the real-world implementation of MCO hemodialysis in patients with CN and highlights the variability of renal outcomes despite effective FLC reduction. Although MCO hemodialysis was associated with consistent and substantial FLC reduction and was well tolerated, renal recovery remained heterogeneous across patients. Notably, patients who did not recover kidney function were those in whom the combined therapeutic approach was delayed or interrupted, mainly due to infectious complications, despite significant biochemical response. These findings support the integration of MCO hemodialysis with anti-myeloma therapy, suggesting that a timely and coordinated approach may be important to optimize renal outcomes. Further prospective studies are needed to better define the clinical impact of extracorporeal FLC removal.

PO 05
WHICH CAME FIRST: TUMOR LYSIS SYNDROME OR RENAL FAILURE IN A HIGH TUMOR BURDEN LYMPHOMA PATIENT

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Category: Systemic diseases

Keywords: tumor lysis syndrome, renal failure, lymphoma

Background:
Tumor lysis syndrome (TLS) is an oncologic emergency caused by rapid cellular breakdown leading to electrolyte and metabolic disturbances. Although commonly associated with cytotoxic therapy, spontaneous TLS may occur in malignancies with high tumor burden. Differentiating TLS from renal failure–induced metabolic abnormalities is challenging due to overlapping biochemical findings.

Case description:

A 53-year-old male with a history of hypertension presented with abnormal outpatient laboratory results and one week of flank pain. He reported urinary dribbling, progressive lower extremity swelling, and a 35–40 lb unintentional weight loss over six months.

Laboratory evaluation revealed severe renal dysfunction (creatinine 7.6–8.4 mg/dL), blood urea nitrogen 75 mg/dL, hyperkalemia, hyperphosphatemia, hyperuricemia (14.3 mg/dL), elevated lactate dehydrogenase, and pancytopenia.

Computed tomography demonstrated a large retroperitoneal mass with extensive lymphadenopathy and hepatosplenomegaly (Figure 1). Given concern for TLS, the patient was treated with rasburicase, aggressive intravenous hydration, and close metabolic monitoring.

Excisional lymph node biopsy confirmed follicular lymphoma, grade 1–2, with immunophenotype positive for CD20, CD10, BCL2, and focal BCL6 expression (Figure 2).

Discussion and learning points:

This case highlights the diagnostic overlap between TLS and renal failure. Renal failure, particularly from obstructive uropathy, can produce hyperkalemia and hyperphosphatemia. However, marked hyperuricemia and elevated lactate dehydrogenase favor TLS physiology.

The patient likely had dual pathology: high tumor burden leading to spontaneous TLS and concurrent renal impairment limiting clearance of metabolic byproducts. This bidirectional interaction amplifies metabolic derangements and worsens renal injury.

Importantly, TLS should not be excluded based on tumor type alone. Although follicular lymphoma is typically indolent, tumor burden rather than proliferation rate is a critical determinant of TLS risk.

PO 06

BRIDGING NEPHROLOGY AND BIOENGINEERING: A DIALYSIS-AWARE CLOSED-LOOP STRATEGY FOR GLYCEMIC INSTABILITY IN ESRD

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Category: Systemic diseases

Keywords: ESRD, hemodialysis, glycemic variability

Background:
Glycemic control in end-stage renal disease (ESRD) is complex due to reduced renal insulin clearance, impaired gluconeogenesis, and altered counterregulatory responses. Hemodialysis (HD) introduces rapid glucose shifts, increasing glycemic variability and risk of hypoglycemia during HD sessions. Continuous glucose monitoring (CGM) studies demonstrate disproportionate hypoglycemia on HD days and hyperglycemia on non-HD days.

Case description: A 74-year-old male with ESRD on maintenance HD and type 2 diabetes mellitus (T2DM) presented with altered mental status and severe hyperglycemia (>400 mg/dL). He had four admissions for diabetic ketoacidosis (DKA) within six months. A consistent pattern of hypoglycemia during HD days and hyperglycemia on non-HD days was observed. Laboratory findings confirmed DKA. He was treated with intravenous insulin and supportive care with clinical improvement. Despite resolution of the acute episode, significant glycemic instability persisted, suggesting inadequacy of conventional insulin regimens in this setting.

Discussion and learning points: This case highlights a high-risk ESRD phenotype characterized by HD-associated glycemic variability and recurrent DKA. HD alters glucose homeostasis through insulin removal, dialysate composition, and metabolic shifts, leading to fluctuating insulin requirements. Standard markers such as hemoglobin A1c (HbA1c) are unreliable in ESRD, making CGM a superior monitoring modality.
Emerging evidence supports automated closed-loop insulin delivery systems in dialysis patients, demonstrating improved time-in-range and reduced hypoglycemia, with adaptive insulin reduction during HD. However, glucose-only systems may not adequately predict metabolic decompensation such as DKA.
We propose a dialysis-aware, dual-analyte closed-loop system integrating CGM with adjunct analyte monitoring (e.g., ketones or insulin levels). Such a system would incorporate HD timing, provide adaptive insulin modulation, and include ESRD-specific safety constraints.

PO 07
Granulomatous Interstitial Nephritis in Sarcoidosis: A Multisystem Presentation

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Background

Granulomatous interstitial nephritis (GIN) is a rare cause of acute kidney injury (AKI), most commonly associated with infections, drugs, or systemic diseases such as sarcoidosis. Renal involvement is uncommon but may lead to severe and potentially reversible kidney dysfunction if promptly recognized and treated.

Case description
A 38-year-old previously healthy man, with no regular medication, presented to the emergency department with a 2-week history of nausea and vomiting, associated with abdominal pain, asthenia, and productive cough. He also reported a 4 kg weight loss over one month.

On admission, he was hemodynamically stable except for mild tachycardia, and physical examination was unremarkable. Laboratory tests showed hemoglobin 12.4 g/dL, leukocytes 5.16×10?/L, platelets 104×10?/L, creatinine 5.7 mg/dL (baseline 0.7), urea 153 mg/dL, normal electrolytes and CRP 2.57 mg/dL. ACE levels were elevated (146 U/L). Immunological workup revealed normal immunoglobulins, a normal kappa/lambda ratio, negative electrophoresis and immunofixation, normal complement levels, and no evidence of autoimmunity.

Infectious workup was negative, including serologies for HIV, HBV, HCV, CMV, and EBV. Extensive microbiological investigation excluded *Mycobacterium tuberculosis*, zoonotic infections, leptospirosis, and enteric pathogens.

Chest-abdomen-pelvis CT demonstrated bilateral pulmonary infiltrates, mediastinal and hilar lymphadenopathy, and hepatomegaly. Bronchoalveolar lavage revealed a CD4/CD8 ratio of 5:1. Bone marrow study, including immunophenotyping and biopsy, showed no evidence of lymphoproliferative disease. Liver biopsy was unremarkable, while gastric biopsy revealed non-necrotizing granulomatous gastritis.

During hospitalization, the patient developed progressive AKI requiring initiation of hemodialysis. Renal biopsy demonstrated extensive lymphoplasmacytic interstitial infiltration with tubular destruction and early granuloma formation, along with features of acute tubular necrosis. Immunofluorescence was negative. These features were consistent with granulomatous interstitial nephritis.

Given the multisystem involvement—constitutional symptoms, cytopenias, hepatosplenomegaly, pulmonary infiltrates, elevated ACE levels, and GIN—a diagnosis of systemic sarcoidosis was considered. The patient was treated with intravenous methylprednisolone pulses (1 g for 3 days), followed by oral prednisolone.

Clinical evolution was favorable, with gradual recovery of renal function. Hemodialysis was discontinued after approximately six weeks. At discharge, serum creatinine stabilized at 2.0–2.5 mg/dL under corticosteroid therapy.

Discussion and learning point
This case highlights severe multisystem sarcoidosis presenting with dialysis-dependent AKI due to GIN. Diagnosis required exclusion of infectious, autoimmune, and hematologic causes. Early corticosteroid therapy enabled significant renal recovery. Renal biopsy remains crucial in atypical presentations.

PO 09
WORSENING CHRONIC KIDNEY DISEASE AS AN ALERT TO OTHER ETIOLOGIES – A CASE REPORT

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ULS Santa Maria

Category Glomerular diseases

Background

Patients with chronic kidney disease (CKD) may develop additional renal disorders over time, accelerating progression or altering its course. Regular follow-up, including urinary sediment and proteinuria, is essential for screening these changes.

Case description

A 62-year-old caucasian man with CKD (baseline creatinine 1,7 mg/dL), long-standing poorly controlled hypertension, and obesity presented with five months of progressive leg edema, shortness of breath, fatigue and foamy urine. Lab revealed renal function declined over eight months, with serum creatinine rising to 2.55 mg/dL, with nephrotic-range proteinuria (uRAC 3992 mg/g), mild microscopic hematuria, hemoglobin 10.6 g/dL, calcium 9.7 mg/dL, and albumin 3.7 g/dL. Further workup demonstrated faint serum lambda light chain monoclonal band, Bence Jones lambda in urine, and markedly abnormal free light chain ratio (0.001). Kidney biopsy showed global nodular glomerulosclerosis with acellular PAS-positive nodules, interstitial fibrosis, and tubular atrophy; congo red staining was negative; immunofluorescence was negative, but immunohistochemistry revealed lambda light chain deposits in glomerular nodules (figure 1). Findings were consistent with lambda-type light chain deposition disease. Renal function stabilized under antiproteinuric and antihypertensive therapy, and he was referred to Hematology for a bone marrow biopsy and initiation of clone-directed chemotherapy.

Discussion and learning point

This case emphasizes the importance of evaluating worsening renal function and increasing proteinuria in CKD patients, as these changes may reveal a treatable underlying disorder. Early recognition of a monoclonal gammopathy, particularly in older adults, is critical, since timely diagnosis and clone-directed therapy can significantly improve renal outcomes and overall survival.

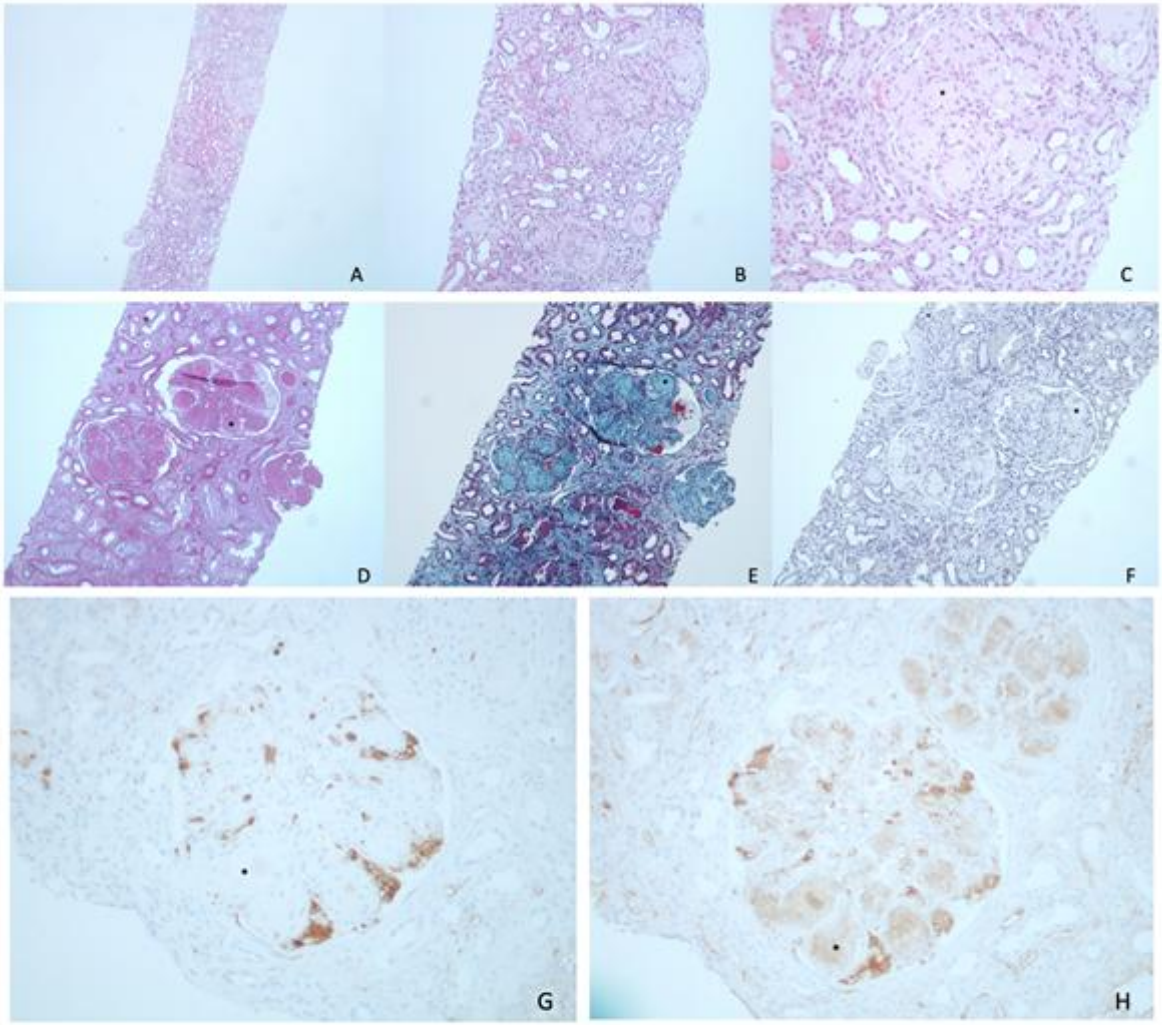

Figure 1 A – HE 40x; B – HE 100x; C - HE 200x; D – PAS 100x; E – Masson's Trichrome (MT) 100x; F – Congo Red (CR) 100x; G – Immunohistochemistry for kappa light chains 200x; H – Immunohistochemistry for lambda light chains 200x

(*) – Presence of global nodular glomerulosclerosis, with formation of acellular nodules (A–C). The nodules stain with PAS and Masson's Trichrome (D) and do not stain with Congo Red (F); in the immunohistochemical study, restriction of lambda light chains of immunoglobulins is observed (G).

PO 10
LUPUS NEPHRITIS – A SPECTRUM OF SUSCEPTIBILITY, CLINICAL PRESENTATION AND HISTOPATHOLOGY

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Category: Systemic diseases

Background: Systemic Lupus Erythematosus (SLE) is significantly more prevalent in females, with males often presenting a more aggressive clinical phenotype. Karyotype 46, XX in phenotypically male individuals (De la Chapelle syndrome) is an exceedingly rare condition that may influence autoimmune susceptibility due to X-chromosome gene dosage. Furthermore, viral

triggers such as severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) infection have been implicated in the onset or flare of glomerulonephritis. We report a case of rapidly progressive Lupus Nephritis (LN) in a male patient with 46, XX karyotype.

Case description: A 41-year-old male with a past medical history of infertility (karyotype 46, XX) presented with a 1.5-year history of peripheral edema and petechiae following SARS-CoV-2 infection. Laboratory evaluation revealed rapid progressive kidney injury, with serum creatinine increasing from 0.9 to 3.3 mg/dL within three months. He exhibited nephrotic-range proteinuria (urinary protein-to-creatinine ratio [PCR] 4.11 g/g), hematuria, and severe anemia (Hemoglobin [Hb] 8.2 g/dL). Immunological studies showed positive antinuclear antibodies (1/1280), positive anti-double-stranded DNA (372 IU/mL) and anti-Smith antibodies, and low serum C3 and C4 complement protein. Renal biopsy identified 13 glomeruli, 10 of which demonstrated crescents (6 cellular, 4 fibrocellular) and fibrinoid necrosis. Immunofluorescence (IF), repeated on both frozen and paraffin sections, was negative. Electron microscopy (EM) confirmed subendothelial and mesangial electron-dense deposits, confirming the diagnosis of Class IV LN (ISN/RPS 2018 classification) with an activity index of 14/24 and a chronicity index of 3/12. Induction therapy was initiated with methylprednisolone pulses, followed by oral prednisolone, and mycophenolate mofetil.

The patient developed severe bone marrow toxicity (Hb 6.3 g/dL, Leukopenia 2,500/µL), requiring treatment to be switched to cyclophosphamide and supportive care with Granulocyte-Colony Stimulating Factor injections. Clinical remission was subsequently achieved, and maintenance therapy with azathioprine was started. At the most recent follow-up, 2 years after diagnosis, the patient presents stable renal function (creatinine 1.1 mg/dL), resolved proteinuria (PCR 0.4 g/g), and normalized serum complement levels.

Discussion: This case illustrates that genetic background plays a pivotal role in lupus nephritis, emphasizing how X-chromosome gene dosage may increase SLE susceptibility. It also highlights that negative IF findings do not exclude a diagnosis of LN; instead, the integration of clinical context, pathology findings and laboratory workup help establish the diagnosis. EM is crucial for confirming the presence of deposits that may remain undetected by conventional immunofluorescence, considering limitations in technique sensitivity or technical factors. Additionally, this case underscores one of the serious adverse effects of immunosuppressive therapy.

PO 11
ADULT-ONSET SEMAPHORIN-3B-ASSOCIATED MEMBRANOUS NEPHROPATHY: A DIAGNOSTIC PITFALL RESOLVED BY ANTIGEN-BASED CLASSIFICATION

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Background: Semaphorin-3B (SEMA3B)–associated membranous nephropathy (MN) is a recently described, antigen-defined subtype of MN, predominantly reported in pediatric patients, but also in young adults. Cases of post-kidney transplant recurrent SEMA3Bassociated have also been reported in both children and adults.

Case Presentation: We report a 40-year-old woman who presented with nephrotic syndrome and was concurrently diagnosed with celiac disease. Kidney biopsy demonstrated stage I membranous nephropathy with IgG-dominant immune deposits and negative serum anti-PLA2R antibodies. Despite a gluten-free diet, optimized supportive therapy, and corticosteroid treatment, nephrotic-range proteinuria persisted. Rituximab therapy resulted in complete clinical remission.

Subsequent laser microdissection and mass spectrometry analysis of the kidney biopsy identified SEMA3B as the target antigen, establishing the diagnosis of SEMA3B-associated MN. 9

Discussion: This case highlights that adult-onset SEMA3B-associated MN can clinically mimic secondary disease in the presence of autoimmune comorbidities. Antigen identification by mass spectrometry was decisive in establishing diagnostic correlation while supporting effective immunosuppressive therapy. These findings underscore the clinical value of antigen-based classification in membranous nephropathy.

Learning points:

- Adult-onset Semaphorin-3B–associated membranous nephropathy is rare and may associate with different underlying diseases.
- Antigen identification by laser microdissection and mass spectrometry helped frame diagnostic uncertainty considering current antigen-based classification.
- Rituximab induced complete remission after failure of conservative measures and corticosteroid therapy.

Figure 3. Clinical course over time.

Longitudinal evolution of serum creatinine, serum albumin, and proteinuria, illustrating progressive reduction in proteinuria and normalization of albumin levels following rituximab therapy.

Figure 4. Laser microdissection and mass spectrometry showing high spectral counts for SEMA3B in the index case (N321). Also shown are complement factors C3, C5, and C9 indicating activation of complement pathways. Five control cases (Ctl 03, Ctl 04, Ctl 106, Ctl 108, Ctl 111) are also shown. The control cases are normal pre transplant kidney biopsies.

PO 12
Late-Onset cblC Disease Presenting as Chronic Thrombotic Microangiopathy

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ULS São João

Category: Glomerular diseases

Background: Cobalamin C (cblC) deficiency, due to biallelic MMACHC variants, is a rare disorder of intracellular cobalamin metabolism. While typically diagnosed in infancy, late-onset presentations may involve predominantly renal manifestations, particularly thrombotic microangiopathy (TMA), posing significant diagnostic challenges.

Case Description: A 21-year-old woman with a 3-year history of hypertension and persistent proteinuria was referred for nephrology evaluation. Laboratory findings showed subnephrotic proteinuria (up to 3.2 g/day), mild renal impairment (sCr 1.4mg/dL) and disproportionate to kidney function anemia (Hb 10.1 g/dL), without hematuria. Evaluation for secondary hypertension and an autoimmune panel, including antiphospholipid antibody screening, was unremarkable, and complement levels were within normal limits.

Kidney biopsy revealed a membranoproliferative pattern with C3 predominance, mesangial and endocapillary hypercellularity, and double-contour formation. Chronic vascular lesions (arteriolar

hyalinosis and hyperplastic arteriolosclerosis) were prominent. Electron microscopy showed no deposits.

Given the role of the complement system in the disease pathogenesis, genetic testing was performed, identifying compound heterozygosity for two pathogenic MMACHC variants (c.271dup (p.Arg91Lysfs*14) and c.565C>A (p.Arg189Ser)). Biochemical evaluation confirmed marked hyperhomocysteinemia (163 µmol/L) and elevated urinary methylmalonic acid (569 µmol /mmol of Cr), establishing the diagnosis of cblC deficiency. Notably, serum vitamin B12 levels were normal. Treatment with hydroxocobalamin, betaine and folate was initiated, along with dietary protein restriction.

Discussion and learning points: This case illustrates a late-onset cblC deficiency presenting as chronic kidney disease with features mimicking C3 glomerulopathy. The absence of overt acute TMA lesions and predominance of chronic vascular changes may delay recognition. Severe hyperhomocysteinemia represents a critical diagnostic clue and should prompt metabolic and genetic evaluation in young patients with unexplained TMA-like or C3-dominant lesions. cblC deficiency should be considered in the differential diagnosis of chronic TMA and C3-dominant glomerulopathy, as early diagnosis enables targeted therapy and may improve renal outcomes.

PO 13
TARGETED COMPLEMENT INHIBITION WITH IPTACOPAN IN C3 GLOMERULOPATHY: A SUCCESSFUL CASE REPORT

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Category Glomerular diseases

Background

C3 glomerulopathies (C3G) are rare complement-mediated kidney diseases caused by persistent dysregulation of the alternative complement pathway, leading to predominant C3 deposition and chronic inflammatory injury. Clinical presentation is heterogeneous and includes proteinuria, hematuria, hypertension, hypocomplementemia, and variable loss of kidney function. Conventional management, based on renin–angiotensin–aldosterone system (RAAS) blockade and non-specific immunosuppression, frequently fails to achieve durable disease control. Growing understanding of C3G pathophysiology has shifted therapeutic paradigms toward targeted complement inhibition, particularly at the level of factor B.

Case description

A 14-year-old male presented in 2017 with nephrotic syndrome, macroscopic hematuria, hypertension, and low serum complement levels, with preserved kidney function. A positive antistreptolysin O titre supported an initial diagnosis of post-streptococcal glomerulonephritis. However, persistent nephrotic-range proteinuria prompted kidney biopsy, which revealed membranous and endocapillary proliferative glomerulonephritis with dominant C3 deposition (Figure 1). Genetic testing identified a heterozygous C3 variant (c.2203C>T, p.Arg735Trp), supporting a diagnosis of C3G. Treatment with corticosteroids, mycophenolate mofetil, and RAAS blockade resulted in incomplete and unstable disease control. After more than four years of follow-up, a relapse with nephrotic-range proteinuria occurred following SARS-CoV-2 infection. Repeat kidney biopsy in 2025 demonstrated active C3G. Given refractoriness to conventional immunosuppression, therapy with the oral factor B inhibitor iptacopan was initiated, leading to a rapid and sustained reduction in proteinuria within one month, with 0.735 g/day at last follow-up, inactive urinary sediment and stable kidney function (Figure 2).

Discussion and learning point

C3G poses significant diagnostic and therapeutic challenges due to histopathological overlap with post-infectious and immune-complex glomerulonephritis, particularly in the presence of triggering infections. This case highlights the importance of reassessing diagnosis in patients with persistent proteinuria and underscores the value of immunofluorescence and genetic testing in identifying complement-driven disease. Patients with genetically mediated alternative pathway dysregulation often respond poorly to non-specific immunosuppression, as illustrated by the clinical course in this case, which provides real-world evidence of the limitations of conventional therapy in genetically driven C3G. Factor B inhibition directly targets the amplification loop of the alternative pathway, providing a rational therapeutic strategy. The prompt and clinically meaningful reduction in proteinuria after iptacopan initiation reinforces the central role of alternative pathway dysregulation in C3G and supports its efficacy in suppressing complement overactivation and controlling disease activity. This case underscores the value of personalized complement-targeted therapy in relapsing or treatment-refractory disease.

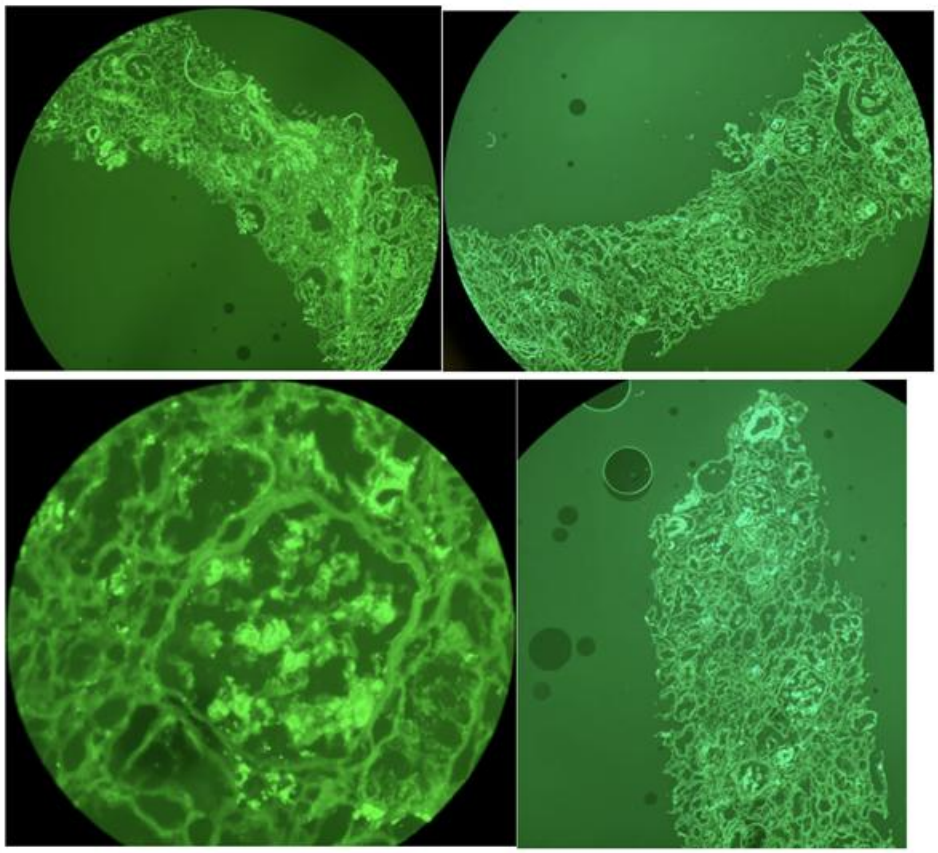

Figure 1: Immunofluorescence microscopy strongly positive for C3 deposits.

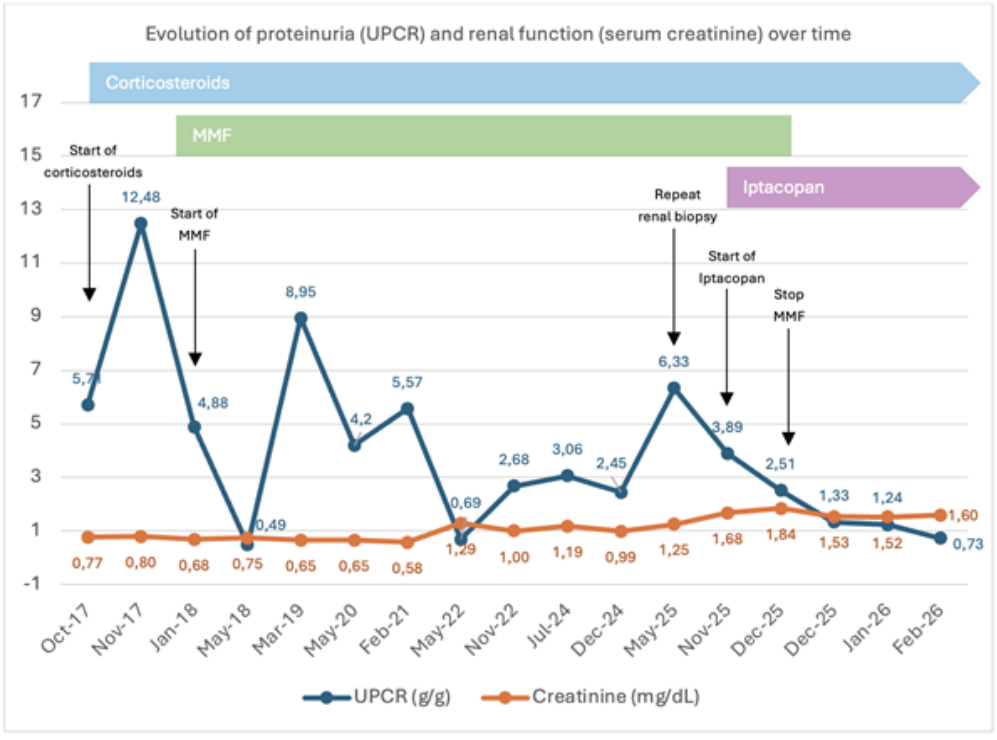

Figure 2: Evolution of proteinuria and serum creatinine since beginning of follow-up.

PO 14
CROSSLINKING MONOCLONALITY AND CRYOGLOBULINEMIA: A CASE OF MGRS-ASSOCIATED GLOMERULONEPHRITIS

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Keywords: MGRS, type 1 cryoglobulinemia, glomerulonephritis.

Background:

Monoclonal gammopathy is characterized by the presence of a monoclonal immunoglobulin (Ig) in the plasma, urine, or both, secreted by clonal plasma cells or B lymphocytes. When end-organ damage is present, the clinical syndrome is classified according to the affected organ system, such as monoclonal gammopathy of renal significance (MGRS).

Case description

A 56-year-old woman with recently diagnosed dyslipidemia and hypertension was referred to nephrology with a 3-month history of peripheral edema and 3+ proteinuria on urine dipstick. On examination, blood pressure was 161/100 mmHg, heart rate 85 bpm, and lower limb edema was noted, with no skin lesions.

On admission, creatinine was 0.8 mg/dL, albumin was 31.1 g/L, and cholesterol was 379 mg/dL. Automated urinary sediment showed erythrocyturia (2010/uL). The urine protein-to-creatinine ratio was 11.2 g/g and the albumin-to-creatinine ratio 7892 mg/g. Complement factors were suppressed with C3 of 72 mg/dL, C4 of 11 mg/dL and C1q of 14 mg/dL. Serum immunofixation revealed a monoclonal IgM-Kappa (κ) band. Serum free light chain (FLC) κ and lambda (λ) were 2.85 mg/dL and 1.58 mg/dL, respectively, with a κ/λ FLC ratio of 1.80 (reference range: 0.26-1.65). Rheumatoid factor was undetectable. Autoimmune and viral serologic screening f was negative. The bone marrow biopsy had <10% plasma cells.

A kidney biopsy was performed, which showed diffuse mesangial and endocapillary hypercellularity, with a lobulated glomerular appearance, and focal double contours of the capillary loops, consistent with a membranoproliferative glomerulonephritis (GN). Sclerosed or crescentic glomeruli were not identified. Numerous glomerular intraluminal pseudothrombi were present, staining red on trichrome and positive for Periodic acid-Schiff (PAS). Congo red staining was negative. Immunofluorescence demonstrated granular staining of the capillary wall, mesangial deposits, and pseudothrombi for C3 (1+), C1q (1+), and IgM (1+), with light chain restriction characterized by κ positivity (3+) and negative λ. IgG and IgA were negative.

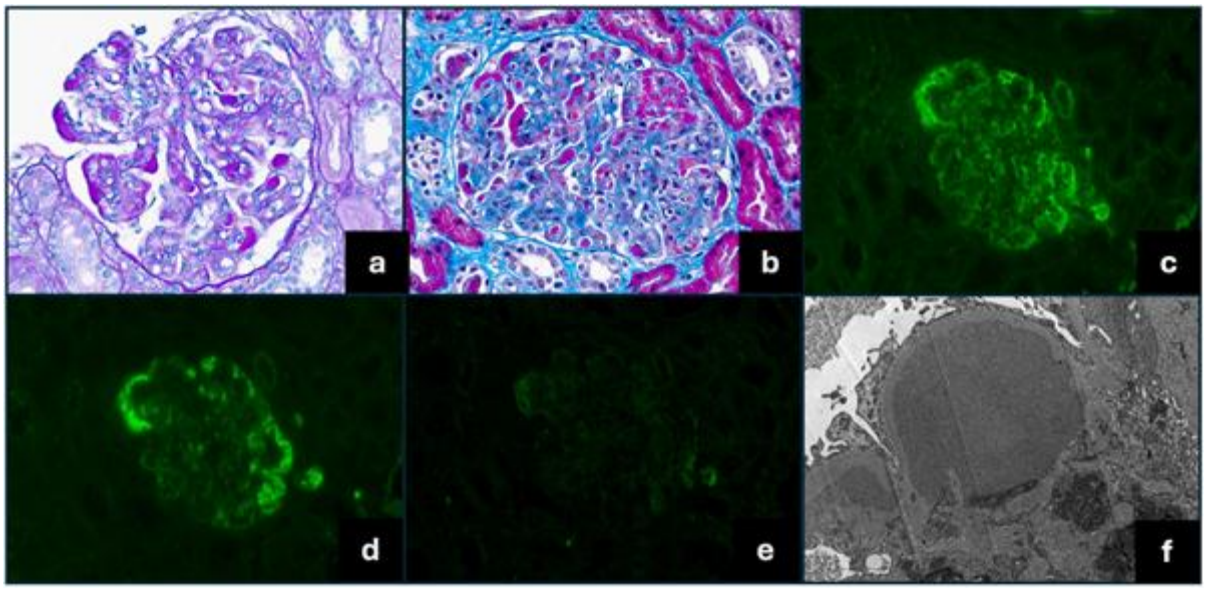
Electron microscopy revealed numerous large electron-dense subendothelial and intraluminal deposits, focally organized into fibrils and curved microtubules (**Figure 1**). The findings were consistent with type 1 cryoglobulinemic glomerulonephritis, according to the Renal Pathology Society/International Kidney and Monoclonal Gammopathy Research Group consensus. Serological testing for cryoglobulin detection was performed and yielded negative results.

Following multidisciplinary discussion, a treatment regimen consisting of corticosteroids, rituximab, and cyclophosphamide was initiated because of the diagnosis of IgMκ MGRS.

Discussion and learning points

This case highlights the importance of early recognition of MGRS as a clinically significant entity that warrants treatment even in the absence of diagnostic criteria for haematologic malignancy. Although serum cryoglobulin testing was negative, the kidney biopsy findings fulfilled the recently proposed criteria for type 1 cryoglobulinemic glomerulonephritis, emphasizing the central role of kidney pathology in establishing the diagnosis. MGRS frequently leads to progressive renal injury and recurrence after transplantation; therefore, early recognition is crucial to prevent irreversible damage.

Figure 1: Light microscopy (x200), showing a glomerulus with endocapillary hypercellularity and numerous pseudothrombi in PAS (a) and trichrome (b) staining. (c) Immunofluorescence demonstrated IgM staining of capillary wall deposits and pseudothrombi. Immunofluorescence revealed (d) kappa light chain restriction (negative lambda in (e)). (f) Electron microscopy revealed organized fibrillary and microtubular deposits (x8000).



PO 15
OXALIPLATIN-INDUCED FANCONI SYNDROME PRESENTING AS SEVERE METABOLIC ACIDOSIS AND HYPOKALEMIA: A CASE REPORT

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Category: Nephrotoxicity

Key-words: oxaliplatin, acute kidney injury, hypokalemia, renal tubular acidosis

Background

Oxaliplatin is a third-generation platinum derivative considered less nephrotoxic than cisplatin. Oxaliplatin is frequently used as an adjuvant and palliative chemotherapy to treat various gastrointestinal tumors, namely colorectal and gastric cancer. Oxaliplatin may induce renal

tubular vacuolization, acute tubular necrosis, renal tubular acidosis, and acute kidney injury, which occur within days of exposure.

Case description

We report a case of a 68-year-old woman who presented to the oncology day hospital for a scheduled chemotherapy session, presenting with a one-week history of anorexia, reduced urine output, and dysuria, two weeks after receiving oxaliplatin-based chemotherapy.

Her premorbid history was significant for hypertension, dyslipidemia, and chronic kidney disease secondary to diabetes mellitus (baseline serum creatinine of 1.2 mg/dL). She had previously been diagnosed with invasive gastric adenocarcinoma and was receiving adjuvant FOLFOX chemotherapy — comprising oxaliplatin, 5-fluorouracil (5-FU), and folinic acid — having completed three cycles, the last of which occurred two weeks prior.

Initial laboratory work-up revealed worsening renal function (serum creatinine of 2.5 mg/dL) and severe hypokalemia (1.5 mmol/L), prompting potassium replacement and urgent referral to the emergency department.

On admission, vital signs revealed a blood pressure of 139/52 mmHg, heart rate of 78 bpm, and peripheral oxygen saturation of 98% on room air. The patient was tachypneic, with an otherwise unremarkable physical examination. Arterial blood gas demonstrated severe metabolic acidosis (pH of 7.19, PaCO2 of 17 mmHg, HCO3 of 6.5 mEq/L). Serum lactate was mildly elevated at 2.8 mmol/L. Hyperchloremia was noted (113 mEq/L). Her urine pH was 5.0. Additional biochemistry revealed hypophosphatemia (2.6 mg/dL) and hypocalcemia (corrected calcium 7.7 mg/dL, albumin 3.1 g/dL), completing a biochemical picture consistent with proximal renal tubular acidosis (type 2, pRTA) / Fanconi's syndrome.

The patient was admitted and managed with intravenous fluid resuscitation, electrolyte replacement, and alkali therapy. Oxaliplatin was discontinued. Renal function recovered substantially, with serum creatinine improving to 1.4 mg/dL at discharge, and normalization of electrolytes and acid-base status.

Discussion and learning points

This case illustrates a rare but clinically significant complication of oxaliplatin — Fanconi syndrome, a generalized proximal tubular dysfunction characterized by urinary wasting of bicarbonate, phosphate, uric acid, glucose, and amino acids. The constellation of severe metabolic acidosis, hypokalemia, hypophosphatemia, hypocalcemia, and hyperchloremia in the setting of recent oxaliplatin exposure is highly consistent with this diagnosis.

While oxaliplatin is traditionally regarded as less nephrotoxic than cisplatin, cumulative tubular toxicity should not be underestimated, particularly in patients with pre-existing chronic kidney disease, as in this case. Underlying renal vulnerability likely lowered the threshold for tubular injury.

PO 16
FROM CHRONIC INFLAMATION TO ORGAN DYSFUNCTION: A CASE OF AA AMYLOIDOSIS

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Unidade Local de Saúde de Santo António

Category | Background | Case description | Discussion and learning point

Background

AA amyloidosis is a rare complication of persistent chronic inflammation, resulting from the systemic deposition of amyloid protein derived from serum amyloid A. Diagnosis requires a high degree of clinical suspicion and the exclusion of hematological, hereditary, and senile causes.

Case Description

A 48-year-old woman with a history of hepatitis C virus infection, intravenous drugs use, active smoking, and recurrent deep vein thrombosis was admitted with generalized edema, anorexia and weight loss. She had multiple venipuncture scars. In the laboratory workup an inflammatory syndrome (CRP 33 mg/dL; ESR 134 mm/h), proteinuria 11 g/g creatinine, albumin 2.6 g/dL and severe renal failure (creatinine 6–10.6 mg/dL) were identified. There was no information regarding the baseline kidney function. Abdominal ultrasound demonstrated small kidneys with loss of corticomedullary differentiation, hepatosplenomegaly, and splenomegaly with a spleen measuring 20 cm. Transthoracic echocardiography revealed moderate-to-severe left ventricular hypertrophy with an interventricular septum measuring 15 mm and a posterior wall thickness of 11 mm, with preserved systolic function.

Immunological studies excluded monoclonal gammopathy, vasculitis (negative ANCA PR3/MPO), autoimmunity (negative ANA, anti dsDNA and C1q), and cryoglobulinemia. Elevated serum amyloid A levels were observed.

The nephrotic syndrome aggravated during the hospitalization and kidney dysfunction worsened with need of hemodialysis initiation.

Taking into account the nephrotic syndrome and the chronic intravenous drugs use the major clinical suspicion was an AA amyloidosis.

The diagnosis was confirmed by minor salivary gland biopsy, showing amyloid deposits positive for Congo red staining with apple-green birefringence under polarized light (Figures 1,2). Immunohistochemistry is not yet available.

Fig.?2 (Congo Red, 10x) – Positive Congo Red staining, showing amyloid deposits with a periductal and perivascular distribution pattern

Fig.?1 (Congo Red, polarized light, 40x) – Amyloid deposit showing apple-green birefringence under polarized light, with a periductal distribution

Discussion

This case illustrates an aggressive AA amyloidosis case with multisystem involvement affecting the kidneys, liver, spleen and heart. Probably this patient had already an established chronic kidney disease due to amyloidosis, since the kidneys were small, which is unusual in acute amyloidosis cases. The usefulness of minor salivary gland biopsy as a less invasive diagnostic method compared with kidney biopsy is highlighted in this case. Mass spectrometry could be useful for confirming the amyloid deposit type, but the clinical suspicion indicates an AA amyloidosis.

Learning Points

• AA amyloidosis should be considered in patients with nephrotic syndrome and persistent chronic inflammation.

• Exclusion of other amyloidosis causes is essential.

• Minor salivary gland biopsy is very sensitive and can avoid kidney biopsy risks.

Keywords: AA amyloidosis; nephrotic syndrome; end-stage renal disease

PO 17
Silent Endocarditis with a Renal First Manifestation

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Background

Membranoproliferative glomerulonephritis (MPGN) secondary to infective endocarditis remains an underrecognized entity and may present with predominant renal involvement before cardiac or infectious symptoms become clinically apparent. Bartonella species are a rare but increasingly recognized cause of culture-negative endocarditis, frequently identified only after extended diagnostic workup.

We describe a case in which renal pathology provided the key diagnostic clue to uncover afebrile prosthetic valve endocarditis due to Bartonella, substantially altering management and preventing prolonged inappropriate immunosuppression.

Case description

A 66-year-old man with hypertension, type 2 diabetes and bioprosthetic aortic valve replacement (2020) presented with one-month history of constitutional symptoms, palpable purpura, anemia, rapidly progressive renal failure (serum creatinine 2.82 mg/dL) and *de novo* hematuria and proteinuria. Epidemiologic exposure included close contact with three domestic cats. At admission he was afebrile, had no cardiac murmurs and inflammatory markers were not elevated. Initial workup showed normocytic anemia, thrombocytopenia, low complement (C3 and C4), positive ANA (1:160) but negative anti-dsDNA, ANCAs, antiphospholipid antibodies and cryoglobulins. Viral serologies were negative. Urine protein-to-creatinine ratio was 1.4 g/g. Serial blood cultures were negative. A vasculitic process was considered and high-dose glucocorticoids were initiated.

Renal biopsy revealed proliferative glomerulonephritis with endocapillary hypercellularity and neutrophilic infiltration, including focal cellular crescents. Immunofluorescence showed immune complex deposition with C3 dominance. These findings prompted reconsideration of the initial diagnosis and triggered cardiac investigation. Transesophageal echocardiography demonstrated a vegetation on the bioprosthetic valve, consistent with probable infective endocarditis in a culture-negative setting. PET-18F-FDG subsequently demonstrated focal uptake on the aortic prosthesis, corroborating the diagnosis. Empirical ampicillin, vancomycin and gentamicin were initiated and immunosuppression was tapered.

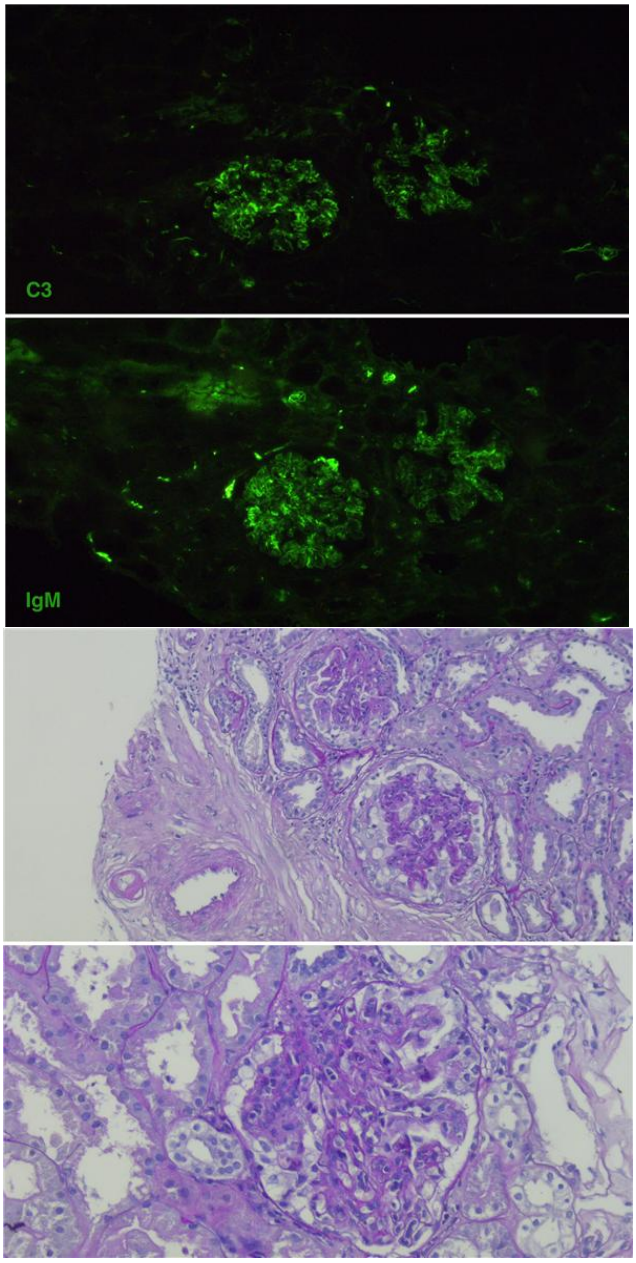
Further etiologic testing revealed negative serology for Coxiella, Legionella, Brucella, Mycoplasma and Aspergillus, but markedly elevated IgG titers for Bartonella henselae (1:1280) and Bartonella quintana (1:640), consistent with Bartonella prosthetic valve endocarditis. Antibiotic therapy was adjusted to doxycycline and rifampin for six weeks.

Progressive renal failure led to uremic symptoms and the need for hemodialysis. At reporting, the patient remains dialysis-dependent but maintains preserved diuresis, and future renal recovery remains uncertain as antibiotic therapy is still ongoing.

Discussion and learning point

This case highlights the diagnostic value of renal biopsy in atypical glomerulonephritis and underscores that immune-complex MPGN with C3-dominant deposition and prominent neutrophilic infiltration should raise suspicion for infective endocarditis, even in afebrile, culture-negative patients. Identifying Bartonella infection was essential because treatment depended on prolonged antibiotic therapy rather than immunosuppression, which could otherwise worsen renal and clinical outcomes. Clinicians should maintain a high index of suspicion for occult infection in culture-negative glomerulonephritis, particularly in patients with prosthetic valves and relevant epidemiologic exposures.

Although uncommon, renal-first presentation of infective endocarditis represents an important diagnostic challenge, emphasizing the need to recognize subtle clinicopathologic clues that can expedite diagnosis and optimize outcomes.



PO 18
DIFFERENTIAL DIAGNOSIS OF IgA NEPHROPATHY IN AN ELDERLY WOMAN

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Background: Glomerulonephritis with dominant IgA deposition presents a diagnostic challenge, particularly in elderly patients with comorbidities and recent infectious contexts, where IgA-dominant infection-related glomerulonephritis (IgADIRGN) and secondary causes of IgA nephropathy (IgAN) need to be considered. The concomitant presence of monoclonal gammopathy adds diagnostic complexity, necessitating exclusion of monoclonal gammopathy of renal significance. The risks of immunosuppression must always be weighed against the benefits,

particularly in elderly patients. We present a case illustrating these diagnostic and therapeutic challenges.

Case description: An 83-year-old woman with hypertension, diabetes, and baseline sCr 0.9 mg/dL presented with two weeks of lower limb oedema, petechial rash, and oliguria. She denied fever or symptoms of localising infection. BP was 160/80 mmHg. Labs revealed sCr 1.73 mg/dL, UACR 5600 mg/g, dysmorphic haematuria, and 1.5 g/24h proteinuria. Immunofixation revealed monoclonal IgG-Lambda; free light chains were elevated with normal ratio. C3, C4, ANCA, ANA, ENA, anti-dsDNA, anti-PLA2R, anti-THSD7A, ASO, cryoglobulins and viral/syphilis screens were negative. A CT scan three months prior showed cavitary pneumonia, which was treated. During admission, sCr peaked at 3.77 mg/dL, without hydroelectrolytic imbalances. Bone marrow biopsy excluded monoclonal clones. Kidney biopsy (7 glomeruli; 4 sclerosed) showed mesangial hypercellularity, segmental sclerosis, one cellular crescent, and ~30% tubular atrophy and inflammation. Congo red was negative; immunohistochemistry confirmed no light chain restriction, and immunofluorescence showed IgA 2+ and C3 2+ (mesangial and capillary wall); IgG, IgM, fibrinogen, and C1q were negative, and Kappa/Lambda were equivocal (+/-). IgAN was diagnosed, oral prednisolone (1 mg/kg/day) commenced, and a new chest and abdomen CT and PET scans were ordered to rule out other secondary causes of IgAN. She was discharged with sCr 1.68 mg/dL, awaiting imaging, with out-patient follow-up.

Discussion: The immunofluorescence pattern of dominant mesangial and capillary wall IgA 2+ with codominant C3 2+ is the hallmark of IgAN. Light microscopy findings were consistent with a MEST-C classification of M1, E0, S1, T1, C1. The absence of endocapillary hypercellularity is notable, as this feature is present in 40-80% of IgADIRGN cases and is a key histological discriminator between IgAN and IgADIRGN. Normal serum C3 and C4 levels, the abscence of liver disease, inflammatory bowel disease, coeliac disease and negative viral serologies support primary IgAN, but imaging is still pending for other causes, such as paraneoplastic. The temporal gap since the patient's pneumonia and absence of active infection at presentation weaken the case for an infection-related trigger. IgA vasculitis is also a likely diagnosis as the patient presented with petechial rash and rapid decline of kidney function, despite not having gastrointestinal symptoms. The monoclonal IgG-Lambda paraprotein was considered an incidental monoclonal gammopathy of undetermined significance, as the bone marrow as clear and kidney biopsy showed no light chain restriction. For treatment, the use of oral corticosteroids was supported by the TESTING trial and the KDIGO 2025 guideline for patients at high risk of progression, such as the presence of a cellular crescent and rapidly declining kidney function, despite the risks associated with the patient's advanced age.

Learning-points: This case highlights the diagnostic challenge of a mesangioproliferative GN with dominant IgA deposition. Thorough investigation to rule out secondary causes is essential. While the decision to start immunosuppression must be weighed against the potential risk of infection, the severity of this case warranted the use of corticosteroids, which have resulted in significant improvement thus far.

PO 24
AN UNUSUAL FACE OF IGA NEPHROPATHY: PRIMARY IGA NEPHROPATHY WITH A MEMBRANOPROLIFERATIVE PATTERN

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Background

IgA nephropathy is the most common primary glomerulonephritis and is characterized by mesangial IgA deposition with variable mesangial proliferation. However, atypical histopathological patterns are increasingly recognized. Among these, a membranoproliferative

(MPGN) pattern is rare and poses as a diagnostic challenge, as it is more commonly associated with secondary etiologies. Its identification in the context of IgA-dominant deposition should therefore prompt a comprehensive etiological workup.

Case description

A 65-year-old woman with no significant past medical history, except for a remote positive antinuclear antibody screen (1:160), presented to the emergency department with a two-week history of new-onset hypertension, and visual disturbances. She reported a self-limited episode of acute gastroenteritis three weeks prior, followed by macroscopic hematuria.

Physical examination was unremarkable, except for a blood pressure of 188/72 mmHg.

Laboratory evaluation revealed acute kidney injury, with a serum creatinine of 2.4 mg/dL (baseline 0.7 mg/dL). Urinalysis showed an active sediment, and 24-hour urine collection demonstrated proteinuria of 2.9 g (1 g of albuminuria). Serum IgA levels were elevated, while complement levels were normal.

Serologic testing for autoimmune disease, including ANA, antidsDNA, ANCA, rheumatoid factor, was negative. Antiphospholipid testing revealed positivity for anti-β2 glycoprotein I antibodies (IgM high titer, IgG low titer), with negative anticardiolipin and lupic anticoagulant antibodies. Serum and urine protein electrophoresis and immunofixation were negative, and the serum free light chain ratio was normal.

Given the presence of a nephritic syndrome, a kidney biopsy was performed. Light microscopy revealed 12 glomeruli, all showing diffuse endocapillary hypercellularity composed of mononuclear cells, along with segmental duplication of the glomerular basement membrane. The interstitium showed edema with mononuclear infiltrate, and the tubules exhibited features of acute tubular injury with numerous intratubular erythrocytes and red blood cell casts.

Due to a MPGN pattern and while immunofluorescence (IF) was being processed, complementary study was broadened: infectious screening, including hepatitis B and C, HIV, Mycoplasma, Coxiella, and tuberculosis, was negative. Blood cultures were sterile, and transthoracic echocardiography showed no evidence of endocarditis. Complement functional and regulatory studies, as well as cryoglobulins, were unremarkable.

IF demonstrated dominant IgA deposition (+++), with co-dominant C3 staining (++) in the mesangium and capillary walls. No significant staining for IgG, IgM, or C1q was observed. There was no light chain restriction.

Based on the IF, an IgA-dominant nephropathy was considered. In the absence of clinical or laboratory evidence of secondary causes of IgA deposition, and given the absence of active infection, both secondary IgA nephropathy and infection-related IgA-dominant glomerulonephritis were excluded; therefore, primary IgA nephropathy with MPGN features was considered.

Oral steroids along with antiproteinuric measures were initiated, with a reduction in serum creatinine (2.8 mg/dL to 1.2 mg/dL) and proteinuria (3.5 g/g to 0.3 g/g). Hematuria also improved.

Discussion and learning points

IgA nephropathy typically shows mesangial IgA deposition; a MPGN pattern is rare and raises concern for secondary etiologies. In this case, the MPGN features prompted an extended workup, which excluded infection-related glomerulonephritis, autoimmune disease, liver disease, and monoclonal gammopathy. The presence of IgA-dominant deposits with C3 involving both mesangial and capillary compartments suggests enhanced complement activation and a more aggressive phenotype, associated with heavier proteinuria and worse renal outcomes compared to classical IgA nephropathy.

PO 26
DARATUMUMAB FOR EARLY RECURRENT PRIMARY FSGS AFTER KIDNEY TRANSPLANTATION

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ULSSM

Keywords: FSGS recurrence, kidney transplantation, daratumumab

Category: Glomerular diseases

Background

Primary focal segmental glomerulosclerosis (FSGS) recurrence after kidney transplantation remains a major cause of early graft dysfunction and loss, with limited response to standard therapies such as plasma exchange (PEX) or rituximab. Increasing evidence suggests that long-lived plasma cells and circulating permeability factors contribute to disease pathogenesis, which may explain incomplete responses to B-cell depletion alone. Targeting CD38-expressing plasma cells with daratumumab has emerged as a promising strategy in refractory cases, particularly when used early after recurrence.

Case description

A 26-year-old male with end-stage kidney disease due to childhood-onset steroid-dependent and later steroid-resistant primary FSGS, with negative genetic testing, underwent deceased-donor kidney transplantation in December 2025. The early post-transplant course was complicated by delayed graft function and nephrotic-range proteinuria within the first week, consistent with early disease recurrence.

Allograft biopsy performed on day 9 demonstrated segmental podocyte hypertrophy and extracapillary hypercellularity, without evidence of rejection (g0, t0, v0), C4d deposition, polyomavirus infection or immune deposits on immunofluorescence, suggesting recurrent podocytopathy. A second biopsy performed four months later, together with electron microscopy findings of diffuse podocyte foot process effacement involving >80% of the glomerular surface in the absence of electron-dense deposits or ultrastructural features of antibody-mediated rejection, confirmed recurrent FSGS.

The patient was treated with intensive PEX, intravenous immunoglobulin and rituximab (1 g, two administrations 14 days apart), without sustained response. Despite these interventions, proteinuria remained in the nephrotic range, with 24-hour urinary protein excretion up to 20.8 g. Due to persistent nephrotic syndrome and graft dysfunction, temporary dialysis was required, followed by recovery of renal function.

Given refractory disease, daratumumab (1800 mg subcutaneously) was initiated on April 7, 2026. A marked decrease in proteinuria was observed, with albumin-to-creatinine ratio decreasing from a peak of 13,970 mg/g to 2,386 mg/g within nine days. At last follow-up, approximately three weeks after treatment, 24-hour urinary protein excretion was 2.6 g, consistent with partial remission. Graft function remained stable, with serum creatinine around 1.4 mg/dL (Figure 1).

Discussion and learning points

This case highlights a clinically meaningful response to daratumumab in early, biopsy-proven recurrent primary FSGS after kidney transplantation, refractory to conventional therapies. The reduction in proteinuria supports the hypothesis that plasma cells contribute to disease activity and may represent a key therapeutic target not addressed by anti-CD20 therapies. These findings align with emerging evidence demonstrating that anti-CD38 therapy can reduce proteinuria and decrease dependence on PEX in similar patients, particularly when introduced early in the disease course. Although complete remission was not achieved at last follow-up, partial remission with preservation of graft function represents a relevant clinical benefit in this high-risk setting. Early integration of plasma cell-targeted therapy may improve outcomes and warrants further prospective evaluation.

Figure 1. Evolution of proteinuria, serum albumin, and serum creatinine following deceased-donor kidney transplantation (KTx) in a patient with early recurrent FSGS treated with daratumumab. Shaded bands indicate HD periods. Post-daratumumab: best response shown (day 9).

Abbreviations: FSGS, focal segmental glomerulosclerosis; HD, hemodialysis; IVIg, intravenous immunoglobulin; KTx, kidney transplantation; PEX, plasma exchange; RTX, rituximab.

PO 27
DIAGNOSIS AND DISEASE PROGRESSION EVALUATION OF MONOCLONAL GAMMOPATHIES IN PATIENTS WITH CHRONIC KIDNEY DISEASE

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Background: The study of monoclonal gammopathies is a complex process that can be hindered by the presence of concomitant chronic kidney disease, as its diagnostic criteria often overlap with changes resulting from this condition—namely renal dysfunction and anemia—or may be masked by dysregulation of the calcium-phosphate metabolism.

Case description: We describe the case of a 52-year-old male with hypertension, referred to the Nephrology appointment for worsening renal function (serum creatinine of 4.97 mg/dL) and

nephrotic-range proteinuria (protein/creatinine ratio of 3.7g/g) without associated nephrotic syndrome. Renal echography revealed atrophic kidneys, so renal biopsy could not be performed. The etiological study conducted showed no significant changes, particularly in serum protein electrophoresis. Due to progressive worsening of renal function, he was started on hemodialysis in the same year. Three years later, in an evaluation for determination of the possibility of kidney transplantation, a monoclonal Immunoglobulin G (IgG) Lambda spike was detected, prompting evaluation by the Hematology department. Laboratory tests, conditioned by terminal renal disease, showed mild hypercalcemia and anemia, with a Pathological/Non-Pathological (P/NP) free light chain ratio of 2. A body CT scan did not show any suspicious lytic lesions. A bone biopsy revealed 40% plasma cells, leading to a diagnosis of monoclonal gammopathy of undetermined significance (MGUS), and a decision was made to continue monitoring, using as baseline calcium and hemoglobin values those prior to the detection of the change in serum protein electrophoresis. In the following two years, the condition progressed, with a progressive increase in IgG levels (from 2290 to 3890mg/dL) and the P/NP free light chain ratio (from 2 to 8). At this time, he was also evaluated by a dentist for a suspected unrelated dental abscess, but the observation was compatible with a non-infectious lesion. A CT scan of the oral cavity showed thickening of the gingival soft tissues with erosion of the adjacent dental arch and reactive submandibular lymphadenopathy, with a focal bone lesion observed in the body of C3. A biopsy of the oral lesion revealed a neoplastic mass consisting of CD138+ CD56+ plasma cells with Lambda restriction. Given the progression of the disease, the diagnosis of multiple myeloma was made, and the patient was started on supportive treatment along with prednisolone and dara-VTd (daratumumab, bortezomib, thalidomide, and dexamethasone), resulting in improved laboratory findings and a significant reduction in the size of the oral mass.

Discussion and learning point: This case highlights the challenge that is the diagnostic and decision of surveillance versus treatment options in monoclonal gammopathies in terminal renal patients, whose diagnostic criteria, disease severity, and progression are consistently altered. These patients must therefore be meticulously re-evaluated to ensure timely treatment of multiple myeloma, with the therapeutic plan being a critical topic for multidisciplinary team discussion.

PO 28
IDIOPATHIC COLLAPSING FOCAL SEGMENTAL GLOMERULOSCLEROSIS - A DIAGNOSIS AND TREATMENT CHALLENGE

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Background: Glomerular diseases are conditions difficult to manage due to the complexity of their pathophysiology and response to therapy. The collapsing variant of focal segmental glomerulosclerosis (FSGS) stands out due to its greater severity at presentation and its tendency to progress to chronic renal dysfunction, usually associated with viral infections - typically Human Immunodeficiency Virus (HIV), hepatitis C, parvovirus B19, Epstein-Barr Virus (EBV), and Cytomegalovirus (CMV) - or medications. Nevertheless, there are cases of idiopathic collapsing FSGS.

Case description: We report the case of a 67-year-old caucasian woman, with hypertension and no other significant personal medical history. She was referred to the Nephrology appointment for suspected nephrotic syndrome, with 9g of proteinuria in a 24-hour urine sample. Anti-proteinuric therapy was optimized, and a renal biopsy was performed, revealing a collapsing FSGS pattern, with 30% interstitial fibrosis.The etiological study showed negative serologies for HIV, Parvovirus B19, hepatitis B and C, CMV, and EBV. There was no history of interferon, pamidronate, tyrosine kinase inhibitors, or other drugs typically associated with this disease. As such, she started high-dose prednisolone (80mg/day, equivalent to 1mg/kg/day) along with calcium and vitamin D supplementation, with progressive improvement of the condition, allowing for gradual corticosteroid tapering. Considering the patient’s age, bone densitometry was performed, which revealed osteoporosis of the lumbar spine, and the corticosteroid dose was reduced progressively until discontinuation. The patient remained in partial remission for about 3 years, at which point

there was a recursion of the nephrotic syndrome. At this stage, considering the osteoporosis and degree of interstitial fibrosis, a decision was made to initiate prednisolone at a lower dose (0.5mg/kg/day) along with mycophenolate mofetil (MMF). After starting this treatment regimen, there was complete remission of nephrotic syndrome, but the patient developed left shoulder pain with functional impairment, and avascular necrosis of the humeral head was radiographically confirmed. In this context, corticosteroid therapy was discontinued, and treatment with MMF alone was maintained, with clinical stability to this day.

Discussion and learning point: This case is noteworthy for the diagnosis of idiopathic collapsing FSGS in an elderly patient with no comorbidities or medications associated with the disease, also highlighting the importance of careful monitorization of the secondary effects of the medications used in its treatment.

PO 29
CONFLICTING TREATMENTS: MANAGEMENT OF LUPUS NEPHRITIS AND HIV

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Background: The management of lupus nephritis remains a significant challenge which requires careful consideration of the multiple immunosuppressive treatments available, each presenting a different adverse effect profile and potential drug interactions. This complexity is amplified in patients with comorbidities, namely Human Immunodeficiency Virus (HIV) infection receiving antiretroviral therapy, where pharmacological interactions must be carefully weighed.

Case description: We describe the case of a 31-year-old woman with a personal history of HIV infection on quadruple therapy with abacavir (300mg/day), ritonavir (100mg/day), rilpivirine (25mg/day), and darunavir (800mg/day). Her obstetric history includes three pregnancies with the last pregnancy terminated at 10 weeks due to suspected lymphoproliferative disease. She was referred to the Nephrology appointment for nephrotic-range proteinuria (6g/24h), which later progressed to nephrotic syndrome. The etiological study revealed positivity for anti-nuclear antibodies and anti-dsDNA antibodies, with mild complement consumption. A renal biopsy was performed, revealing lupus nephritis with a membranous pattern and some mesangial proliferation, consistent with lupus nephritis class V + II. She was started on therapy with methylprednisolone pulses followed by prednisolone, and mycophenolate mofetil (MMF) after pregnancy was excluded.

This therapy was not well tolerated due to gastrointestinal side effects, and was therefore switched to mycophenolic acid. Initially there was a good clinical response, but nephrotic syndrome recurred. As a result, tacrolimus was introduced, initially at 1mg twice daily. A constant supratherapeutic tacrolimus level was observed analytically, which was explainable by the interaction with ritonavir. The tacrolimus dose was progressively reduced to 0.5mg once a week, with good tolerance and maintenance of tacrolimus levels within therapeutic range. Since then, the patient’s renal function has remained stable, with a complete remission of nephrotic syndrome.

Discussion and learning point: This case highlights the complexity of managing patients with lupus nephritis associated with multiple comorbidities and the medications necessary for its control. Antiretroviral therapy, while essential for managing HIV patients, carries the risk of various drug interactions, which must be considered when selecting the most appropriate therapy. Regular monitoring of these patients is crucial to ensure the control of drug doses, tolerance, and efficacy of the proposed therapeutic regimens.

PO 31
THROMBOTIC MICROANGIOPATHY AS THE INITIAL PRESENTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS: A CASE REPORT

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Unidade Local de Saúde de Trás-os-Montes e Alto Douro

Category - Systemic diseases

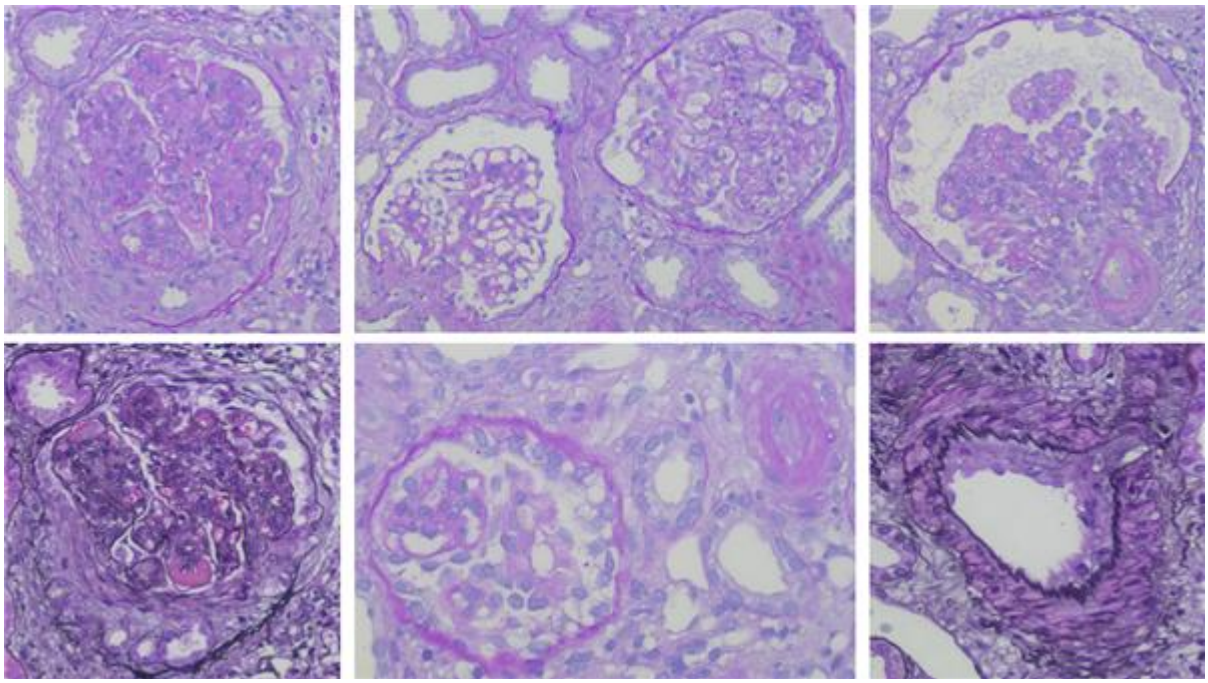
Background
Renal thrombotic microangiopathy (TMA) occurs in 1–4% of lupus nephritis (LN) and is associated with more severe disease and poorer renal outcomes. Systemic TMA secondary to systemic lupus erythematosus (SLE) is rare (1.0% in a retrospective cohort of lupus patients), with only 8% of these presenting as the initial manifestation of SLE. Despite plasma exchange and standard immunosuppression for lupus nephritis, mortality remains high (32–52%). In SLE, TMA with preserved ADAMTS13 activity and negative antiphospholipid antibodies frequently corresponds to complement-mediated TMA due to genetic or acquired functional defects in the regulation of the alternative complement pathway.

Case description
A 22-year-old woman with no prior kidney disease presented with a one-month history of progressive oedema, haemoptysis, fever, abdominal pain and diarrhea. On admission, she was hypertensive (blood pressure 155/100mmHg) with signs of fluid overload.
Laboratory evaluation revealed microangiopathic haemolytic anaemia (haemoglobin 6.6 g/dL, schistocytes 7%, undetectable haptoglobin and negative coombs test), thrombocytopenia (88×10?/L) and acute kidney injury (creatinine 3.3 mg/dL), with proteinuria and active urinary sediment. Imaging showed pericardial and pleural effusions and bilateral pulmonary infiltrates. An intermediate PLASMIC score (5) prompted treatment with high-dose corticosteroids, therapeutic plasma exchange and caplacizumab.
The patient developed multiorgan dysfunction including lethargy, respiratory failure, acute left ventricular systolic dysfunction with elevated cardiac biomarkers and oliguria. Bronchoscopy excluded active alveolar haemorrhage. The ADAMTS13 activity was preserved (63%), excluding thrombotic thrombocytopenic purpura. Immunological evaluation showed marked hypocomplementaemia (reduced C3, C4 and C1q) and high-titre antinuclear and anti-double stranded DNA antibodies, consistent with SLE, with negative antiphospholipid antibodies. Infections were excluded.
Complement inhibition with eculizumab and immunosuppression with cyclophosphamide were initiated as induction therapy. Kidney biopsy confirmed TMA associated with diffuse proliferative LN – class IV (Figure 1). Genetic testing for atypical haemolytic uraemic syndrome was negative. Despite renal and haematological improvement with platelet count normalization after 4 weeks of effective treatment, the patient developed fatal catastrophic multifocal intracranial haemorrhage.

Discussion and learning point
Complement-mediated TMA in LN does not respond well to plasma exchange or immunosuppression with glucocorticoids and cyclophosphamide and may be best treated with terminal complement blockade. In a systematic review of 30 patients treated with eculizumab for TMA secondary to SLE, 93% achieved a favourable response. However, in our case, the disease followed a fatal course despite multimodal therapy, including complement inhibition.
In addition, complete remission rates in LN remain approximately 44%, highlighting the need for new therapeutic strategies. Given the role of complement dysregulation in LN pathogenesis, adjunctive complement inhibition may benefit patients with refractory disease; further studies are required.

Figure 1: Kidney biopsy showing features of thrombotic microangiopathy and diffuse proliferative lupus nephritis (class IV) on light microscopy. Immunofluorescence demonstrated granular deposition of IgG ++, C3 ++, C1q ++, kappa ++ and lambda ++.

Keywords
Systemic lupus erythematosus, Thrombotic microangiopathy, Complement inhibition



PO 32
HEMOPERFUSION AS AN ADJUNCT TO HEMODIALYSIS IN HYPERTRIGLYCERIDEMIA-ASSOCIATED PANCREATITIS WITH ACUTE KIDNEY INJURY

Vanessa Yvonne Emano- Abregana MD; Kristine May Valmoria M.D, FPCP, DPSN

CAGAYAN DE ORO POLYMEDIC PLAZA

Acute pancreatitis is a potentially fatal inflammatory disorder most commonly caused by gallstones and alcohol use. Hypertriglyceridemia accounts for up to 10% of cases and is associated with more severe disease and systemic complications, including acute kidney injury (AKI). The interplay of hypovolemia and an unchecked inflammatory cascade contributes to multiorgan dysfunction and increased mortality. Hemoperfusion, an extracorporeal blood purification modality, has emerged as a potential adjunct by adsorbing circulating cytokines and inflammatory mediators while helping maintain acid base balance.

This is a case of a 56-year-old Muslim female admitted due to acute change in sensorium preceded by a one-week history of fever, abdominal pain, and body weakness, later accompanied by jaundice and melena. On admission, she was awake but combative, later developed hypotension, oliguria and subsequently intubated. Laboratory evaluation revealed leukocytosis (14.2 ×10?/L) with neutrophilic predominance (74.9%), hemoglobin 104 g/L, and platelets 210 ×10?/L. Serum lipase was nine times markedly elevated (1306.50 U/L) with hypertriglyceridemia (517.06 mg/dL). Severe renal impairment was evident (creatinine 9.53 mg/dL, eGFR 4 mL/min/1.73m², BUN 148.96 mg/dL), consistent with AKI Stage 3. Liver tests showed cholestatic injury, with total bilirubin 9.54 mg/dL and alkaline phosphatase 321.7 U/L, while procalcitonin was

markedly elevated (34.22). Abdominal CT demonstrated biliary sludge without gallstones or pancreatic mass.

In the setting of refractory metabolic acidosis and volume overload, the patient underwent urgent hemodialysis combined with hemoperfusion. Notably, after two sessions, there was rapid hemodynamic stabilization, resolution of encephalopathy, and restoration of urine output. Continued renal support facilitated recovery of kidney function, allowing discontinuation of dialysis. At discharge, creatinine had decreased to 2.12 mg/dL, with a concomitant reduction in triglyceride levels to 252.50 mg/dL.

This case highlights how hemoperfusion may offer meaningful benefit in severe acute pancreatitis complicated by multiorgan dysfunction. The patient's rapid improvement after initiation of therapy suggests a potential role for early cytokine removal in altering the course of disease. While evidence remains limited, cases like this add to the growing knowledge and warrant further investigation on this emerging therapy.

PO 33
SEVERE ACUTE KIDNEY INJURY IN MULTIPLE MYELOMA WITH PULMONARY PLASMACYTOMA: A MONOCLONAL GAMMOPATHY OF RENAL SIGNIFICANCE SPECTRUM PRESENTATION

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Unidade Local de Saúde de Viseu Dão-Lafões

Category | Acute Kidney Injury

Background

Acute kidney injury (AKI) is a frequent and severe complication of multiple myeloma (MM), commonly related to light chain-mediated tubular injury. Extramedullary plasmacytomas are uncommon and may obscure the initial diagnostic approach, particularly when presenting as pulmonary masses.

Case description

We report a 76-year-old man presenting with asthenia, normocytic anemia, and rapidly progressive AKI requiring urgent kidney replacement therapy (KRT). Laboratory workup showed serum creatinine 6.8 mg/dL, hemoglobin 8.9 g/dL, ionized calcium 1.35 mEq/l, and proteinuria of 1.2 g/24h. Serum immunoglobulins revealed markedly elevated immunoglobulin G (IgG) 7005 mg/dL and suppressed immunoglobulin M (IgM) <21 mg/dL. Serum free light chains showed lambda predominance (4214 mg/L) with a markedly abnormal ratio. Serum protein electrophoresis and immunofixation confirmed a monoclonal IgG lambda component.

Given the high tumor burden and need for urgent therapy, kidney biopsy was not performed, and a presumptive diagnosis of light chain cast nephropathy was established. Bone marrow biopsy demonstrated extensive plasma cell infiltration. Oncologic fluorescence in situ hybridization (FISH) identified TP53 deletion in 40% of nuclei, consistent with high-risk cytogenetics. Imaging revealed extensive osteolytic bone disease and a peripheral pulmonary mass in the right lower lobe. Histopathological examination of the lung lesion confirmed extramedullary plasmacytoma with 93% plasma cells.

Treatment with bortezomib and dexamethasone was initiated promptly, with daratumumab added from the second cycle. The patient showed progressive renal recovery, allowing discontinuation of KRT after two months.

Discussion and learning point

This case illustrates a classic but severe presentation of MM with dialysis-dependent AKI, most consistent with light chain cast nephropathy, where kidney biopsy may be safely deferred in the presence of typical clinical and laboratory features. The coexistence of extramedullary pulmonary plasmacytoma is rare and may mimic primary lung malignancy, requiring histological confirmation. High-risk cytogenetics, including TP53 deletion, are associated with aggressive disease and poorer prognosis. Early recognition and rapid initiation of clone-directed therapy with proteasome inhibitors are critical to maximize the likelihood of renal recovery, and may be independent of anti-CD38 or other complementary directed therapies.

PO 34
CAKUT – WHEN TO SUSPECT AND SEARCH FOR GENETIC CAUSES

Joana Abreu; Adriana Dias; Catarina Veiga; Joaquim Milheiro; João Melo; Ana Rodrigues; Sérgio Lemos

Unidade Local de Saúde de Viseu Dão-Lafões

Category: Systemic diseases

Background: Congenital anomalies of the kidney and urinary tract (CAKUT) account for 20–30% of all anomalies identified in the prenatal period. Among them is unilateral renal agenesis (URA) - the congenital absence of renal parenchymal tissue, whose incidence is about 0.05%. Its causes include genetic and environmental factors.

Case description: We present a 29-year-old woman with congenital right-sided URA and history of 5 pregnancies, 2 of which ended in first-trimester spontaneous abortions and 1 preterm delivery at 36 weeks with early neonatal death due to bilateral renal agenesis. She had a family history of end-stage chronic kidney disease (CKD) in her mother, presumed secondary to hypertensive nephroangiosclerosis.

Review of her obstetric history highlighted the presence of anhydramnios and non-visualized kidneys on prenatal diagnosis (PND) ultrassound resulting in an early neonatal death. In that context, chorionic villus sampling was performed, identifying a heterozygous mutation in the GREB1L gene, classified as a pathogenic variant with autosomal dominant inheritance. She was subsequently referred for a Genetics consultation, where maternal inheritance was confirmed.

Currently, she reports recurrent urinary tract infections (UTIs), approximately 4/year, with a serum creatinine (sCr) of 0.5 mg/dL, remaining otherwise asymptomatic.

Discussion and learning point: This case paradigmatically illustrates the importance of performing genetic testing in selected patients with CAKUT. The patient had an apparently isolated congenital renal malformation, stable and without significant functional impairment, which often leads to a conservative approach without genetic investigation. However, her obstetric and family history raised the possibility of an unrecognized familial transmission.

The mutation identified in the GREB1L gene is associated with a variable phenotype. There's evidence of incomplete penetrance with some affected individuals presenting with bilateral renal agenesis, others with URA compatible with life, and others with milder manifestations such as vesicoureteral reflux.

This case raises the hypothesis that the previous spontaneous abortions may have corresponded to fetuses with more severe forms of the same condition, undiagnosed. Thus, if genetic investigation had been performed earlier—namely after the diagnosis of URA in the patient or due

to the adverse obstetric history—it would have been possible to establish a diagnosis sooner, provide appropriate genetic counseling, and offer targeted PND. Also, the outcome of the most recent pregnancy could have been anticipated, allowing informed decision-making at earlier stages.

This case therefore reinforces the need for a more systematic approach to genetic investigation in patients with CAKUT, even in apparently isolated and benign forms. The identification of a mutation has implications not only for the patient but also for the family, enabling targeted screening, risk stratification, and informed reproductive planning.

In the absence of a specific treatment for the genetic condition, the implementation of nephroprotective measures is essential. In this case, that includes the prevention of recurrent UTIs and monitoring of sCr and proteinuria, with appropriate management as needed.

PO 35
NEPHROTIC SYNDROME TWO MONTHS POSTPARTUM IN A WOMAN WITH PRE-ECLAMPSIA: MEMBRANOUS NEPHROPATHY

Joaquim Milheiro; Catarina Veiga; Adriana Dias; Joana Abreu; João Melo; Cátia Pêgo; Carla Lima; Sérgio Lemos

Unidade Local de Saúde de Viseu Dão-Lafões

Category | Glomerular diseases

Background |

Membranous nephropathy (MN) is a leading cause of nephrotic syndrome in adults and may be primary or secondary. In postpartum patients, the differential diagnosis is broadened by pregnancy-related immune shifts, recent infections, thyroid disease, and the need to consider secondary triggers carefully. Establishing the underlying cause is essential for targeted management.

Case description |

A 26-year-old woman presented with progressive edema developing over one week, two months after induced delivery at 38 weeks due to de novo pre-eclampsia. Two weeks prior to edema onset, her 2-year-old child had an upper respiratory tract infection of presumed viral origin. Initial evaluation showed nephrotic syndrome with proteinuria of 10.6 g/day, serum albumin 1.7 g/dL, hemoglobin 16.9 g/dL, serum creatinine 0.7 mg/dL, total cholesterol 780 mg/dL, high-density lipoprotein 57 mg/dL, low-density lipoprotein 675 mg/dL, triglycerides 240 mg/dL, thyroid-stimulating hormone 19 mIU/L, free thyroxine 0.8 ng/dL, and normal sedimentation rate. Autoimmune testing was negative except for anti-thyroid peroxidase antibodies (125 IU/mL). Anti-phospholipase A2 receptor (anti-PLA2R) antibodies were negative. Kidney biopsy confirmed membranous nephropathy with unremarkable light microscopy. The patient showed strong preference for alternative medicine and was reluctant about biological therapies. After discussion of treatment options, she was started on tacrolimus in addition to supportive treatment. Total remission was achieved by month 2, with proteinuria decreasing to 150 mg/day.

Discussion and learning points

This case illustrates the diagnostic overlap between postpartum renal disease and pre-eclampsia-associated kidney injury. However, the nephrotic-range proteinuria, severe hypoalbuminemia, marked hyperlipidemia, preserved kidney function, and biopsy-proven MN support a primary glomerular process rather than isolated residual pre-eclampsia. Anti-PLA2R negativity requires careful consideration of secondary MN, but the absence of systemic inflammatory markers,

negative autoimmune screening, and lack of an alternative secondary cause favor PLA2R-negative primary MN. The preceding viral exposure in a household contact may represent an immune trigger, although causality cannot be established.

Management was additionally complicated by the patient's preference for alternative medicine and reluctance to receive biological therapies, underscoring the importance of shared decision-making and clear counseling. Tacrolimus offered a practical non-biological treatment option in this context. This case underlines the difficulty in classifying membranous nephropathy as primary versus secondary in a woman with autoimmune thyroiditis and past pre-eclampsia, and highlights the need to tailor therapy to both disease severity and patient-related barriers to treatment.

PO 36
AN UNEXPECTED CAUSE OF HEMATOPROTEINURIA AND RENAL ASYMMETRY

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Unidade Local de Saúde de Viseu Dão-Lafões

Category: Systemic Diseases

Background: Hematoproteinuria is a frequent reason for Nephrology referral. Although it raises suspicion of glomerular etiology, rare causes should not be overlooked as they may require specific diagnostic tests and treatments.

Case description: 20-year-old woman, followed in Pediatric Nephrology after an isolated episode of macroscopic hematuria and proteinuria (1 g/g) at age 15 after streptococcal-negative tonsillitis. Her renal function was normal and the initial workup revealed no abnormalities except for a slight reduction in right kidney parenchymal-sinus differentiation. During follow-up, she maintained persistent microscopic hematuria and occasional episodes of right lumbar pain, without proteinuria. No history of recurrent urinary tract infections or nephrolithiasis was noted.

At age 17, renal ultrasound raised suspicion of left-sided Nutcracker Syndrome (NCS), showing narrowing of the left renal vein (LRV) at the level of its crossing with the superior mesenteric artery (SMA) and an aorto-mesenteric angle of less than 45°, findings later confirmed by CT angiography, with evidence of collateral circulation.

Imaging studies consistently showed renal asymmetry, with a smaller right kidney (9.6 cm) compared to the left (11.7 cm). MAG3 renal scintigraphy confirmed functional asymmetry: 63.6% contribution from the left kidney, 36.4% from the right.

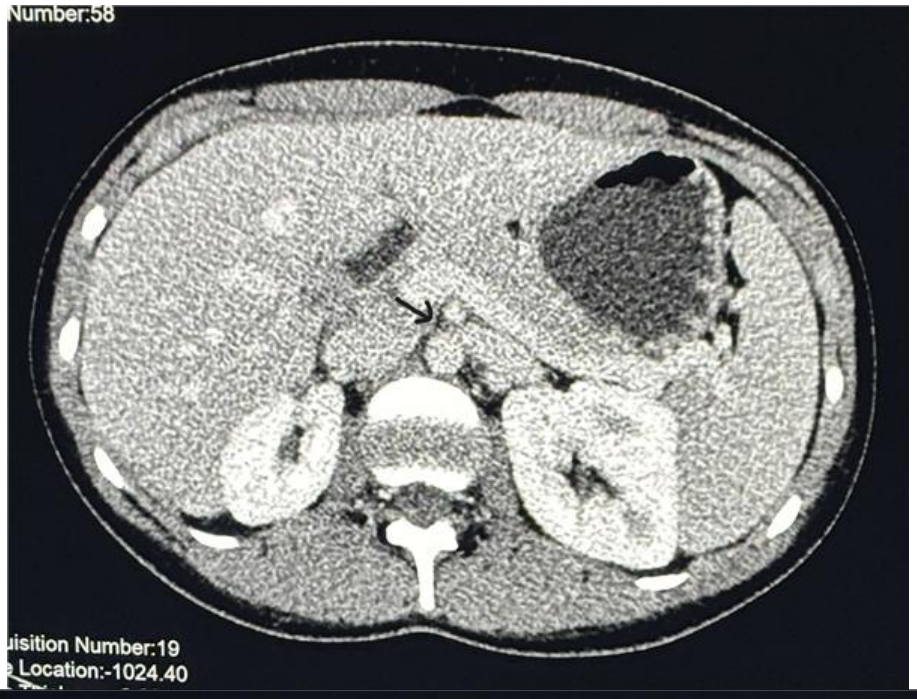
Alternative causes of renal asymmetry were excluded, namely right renal artery stenosis and antiphospholipid syndrome. Heterozygous mutation of the MTHFR gene and mild protein S deficiency (46%) were detected, without indication for anticoagulation.

Discussion and learning points: NCS is a rare vascular condition more common in children, usually resolving with growth. It refers to compression of the LRV between the aorta and the proximal portion of the SMA, leading to varying degrees of extrinsic stenosis of the LRV, which may be asymptomatic but can also cause microscopic or macroscopic hematuria, proteinuria, hypertension, lumbar pain, dyspareunia, dysmenorrhea and pelvic varices. Renal dimensions are usually preserved. When present, morphological changes typically involve slight enlargement of the left kidney due to venous congestion, although renal atrophy may rarely occur in chronic cases. Thus, the contralateral renal asymmetry described in this case is an unusual finding and prompted investigation of other overlapping conditions, such as stenosis or thrombosis of the right renal vessels.

An alternative explanation that cannot be excluded is congenital unilateral renal hypodysplasia, whose coexistence with NCS may be incidental or suggest a common genetic basis not yet fully understood. Another hypothesis to consider is overlap with IgA nephropathy.

Undiagnosed or untreated NCS carries risks of chronic kidney disease due to prolonged LRV hypertension and potential thrombosis. In this case, with documented functional renal asymmetry, appropriate management is especially important.

NCS treatment may be conservative in children or in patients with mild symptoms, or more invasive, involving surgical or endovascular interventions in more symptomatic or refractory cases. Currently, the patient has a serum creatinine of 0.6mg/dL, protein/creatinine ratio of 0.138 g/g, and erythrocyturia of 20/µL, remaining under follow-up by Vascular Surgery.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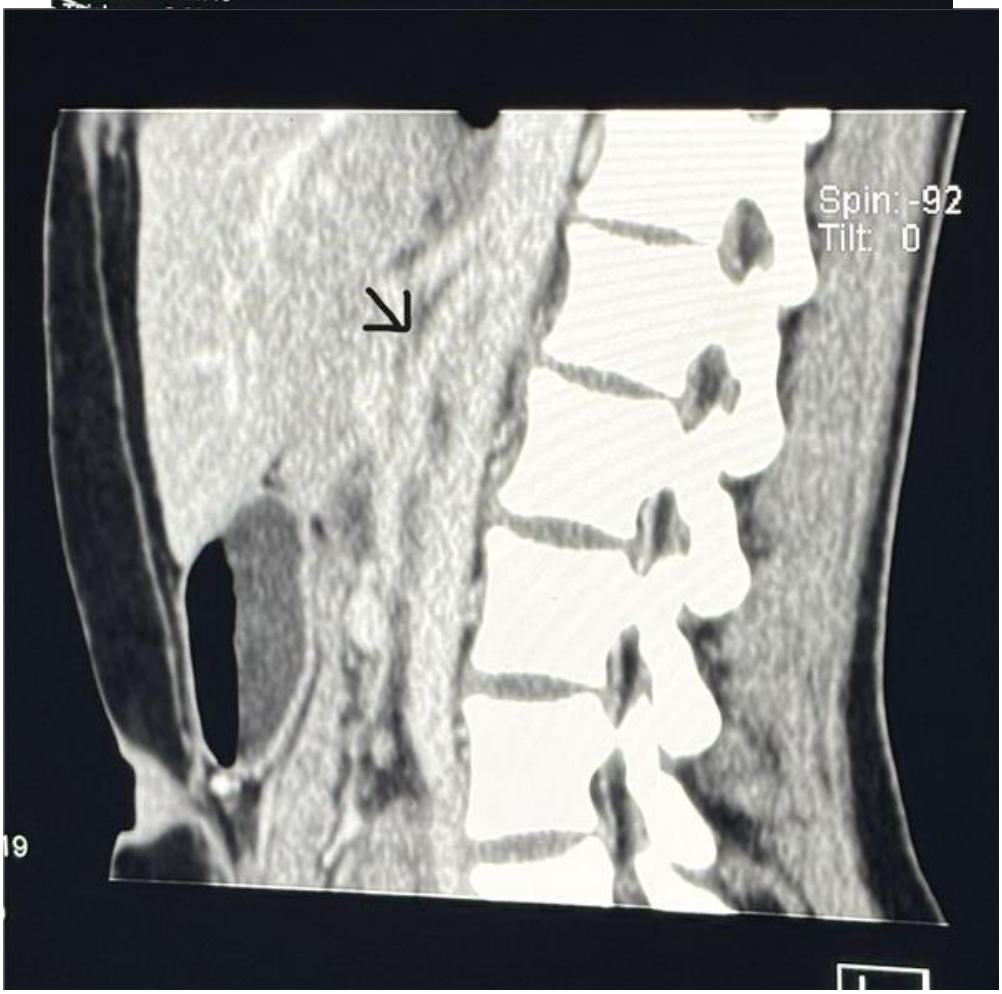
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PO 37
UNMASKING LUPUS NEPHRITIS IN SLE WITH CRYOGLOBULINAEMIA

Helena de Oliveira; Pedro Ribeirinho Soares; Bruno Besteiro; Yeika Larrinaga; Jorge Almeida; Fernando Friões

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Background: Systemic lupus erythematosus (SLE) is a chronic multisystem autoimmune disease characterised by autoantibody production and immune complex deposition, with frequent renal involvement that significantly impacts prognosis. Lupus nephritis (LN) may be clinically silent and requires a high index of suspicion, even in the absence of overt renal dysfunction. Although vasculitis can occur in SLE, cryoglobulinaemic vasculitis represents a less common but relevant association, particularly when overlapping with renal disease.

Case description: This case concerns a 55-year-old patient with a history of arterial hypertension, admitted due to new-onset cutaneous lesions (digital purpura, auricular acrocyanosis, and lower limb petechiae) evolving over one week, alongside bicytopenia (anaemia — direct Coombs positive for anti-IgG, without haemolysis — and thrombocytopenia) under investigation. Anamnesis did not reveal significant findings initially, however retrospective review revealed a three-year history of arthralgia and recent weight loss, with prior immunological studies showing ANA (1:640, speckled) and anti-dsDNA positivity. Extensive evaluation excluded neoplastic and infectious causes. Immunological reassessment confirmed ANA and anti-dsDNA positivity, along with strongly positive anti-nucleosome and anti-histone antibodies. Complement levels remained normal, and rheumatoid factor was negative. Given cold-induced exacerbation of skin lesions, cryoglobulins were tested and found to be positive. A diagnosis of SLE-associated cryoglobulinaemic vasculitis was established, and immunosuppressive therapy with prednisolone (1 mg/kg/day) and rituximab was initiated, resulting in marked improvement in cutaneous and haematological manifestations. Despite preserved renal function, further evaluation revealed active urinary sediment with dysmorphic erythrocytes and non-nephrotic proteinuria (1.95 g/24 h, including 700 mg of albumin), raising suspicion for underlying renal involvement. Renal biopsy performed on day 13 confirmed lupus nephritis class II+V (ISN/RPS), with an activity index of 0/24. No additional immunosuppressive therapy was introduced. The patient was discharged with planned follow-up in both Nephrology and Autoimmune Disease clinics.

Discussion: This case underscores the importance of systematic renal evaluation in SLE, even in the absence of clinical renal impairment. The detection of dysmorphic haematuria and proteinuria was pivotal in identifying subclinical lupus nephritis which may otherwise remain unrecognised. The coexistence of cryoglobulinaemic vasculitis adds further complexity, although secondary causes were excluded. Notably, normal complement levels did not preclude active systemic disease. Early recognition of renal involvement allowed appropriate histological classification and risk stratification, guiding management. The favourable response to corticosteroids and rituximab highlights the role of B-cell–targeted therapy in SLE with vasculitic and haematological manifestations, while reinforcing the need for ongoing renal surveillance.

PO 38
FAMILY SCREENING REVEALS VARIABLE RENAL EXPRESSIVITY IN SDCCAG8-RELATED DISEASE

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Unidade Local de Saúde de Viseu Dão-Lafões

Category | Glomerular diseases

Background |

Syndromic kidney disease can be difficult to recognize when the renal phenotype is unusual. In ciliopathies and other inherited disorders, glomerular disease is less typical than structural urinary tract abnormalities, making genetic and histologic correlation essential.

Case description |

We report a 51-year-old woman referred to nephrology for chronic kidney disease of unclear cause. She had previously been labeled as having Usher syndrome because of retinitis pigmentosa and sensorineural hearing loss, without prior genetic confirmation. Family history was notable for parental consanguinity and multiple affected relatives with cardiac disease, congenital malformations, and congenital deafness. Kidney biopsy was performed and showed focal segmental glomerulosclerosis, perihilar variant. This atypical renal presentation prompted genetic testing, which identified a pathogenic SDCCAG8 variant, c.397G>T, consistent with Bardet-Biedl syndrome type 16 and Senior-Løken syndrome type 7. Subsequent testing in one of her six siblings revealed the same mutation, although that sibling had no renal manifestations. All first-degree relatives underwent serum creatinine and proteinuria screening, which was unremarkable; the patient remained the only family member with renal involvement.

Discussion and learning point

This case expands the renal phenotype associated with SDCCAG8-related ciliopathy. Although kidney disease is expected in syndromic disorders, it usually presents as congenital urinary tract malformations, cystic disease, or tubulointerstitial nephropathy rather than perihilar FSGS. The absence of identifiable secondary causes for FSGS, together with the syndromic background and consanguineous family history, supports a monogenic cause. The identification of the same variant in an unaffected sibling and the absence of kidney abnormalities in screened relatives highlight variable penetrance and incomplete expressivity, which can delay recognition of inherited disease. This case reveals the importance of genetic testing and family screening when kidney disease is unexplained and the phenotype is atypical, as molecular diagnosis can guide prognosis, surveillance, and genetic counseling.

PO 40
ONE TECHNIQUE TO CHANGE IT ALL: IMPACT OF MASS SPECTROMETRY IN DIAGNOSIS AND MANAGEMENT OF AMYLOIDOSIS

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Amyloidosis encompasses several disorders characterized by tissue deposition of misfolded proteins which lead to multiorgan damage. Diagnosis requires biopsy for amyloid typing, essential for guiding treatment. Conventional techniques, such as immunofluorescence (IF) and immunohistochemistry (IHC), have limited sensitivity and specificity in amyloid subtyping. In contrast, proteomic analysis by high-resolution mass spectrometry (MS) is highly sensitive and specific but remains limited in routine clinical practice due to restricted availability. We present a case series of six patients who underwent tissue biopsy in order to characterize the role of MS in establishing and/or confirming amyloid subtype in a series of challenging amyloidosis cases, and its impact on subsequent patient management.

Pt 1: A 78 year-old woman presented with nephrotic syndrome [urinary albumin-creatinine (uACR) ratio 3597 mg/g]. Dysproteinemia work-up was unremarkable. Kidney biopsy demonstrated amyloid deposits, classified as AA amyloid in IHC. No underlying cause for AA amyloidosis was found, the sample was sent for MS. MS revealed AL λ amyloidosis, prompting a change in treatment regimen.

Pt 2: An 86 year-old woman presented with nephrotic syndrome (uACR 7400 mg/g). An IgM λ clone was identified in plasma, although kidney biopsy showed amyloid without light-chain restriction. The patient refused bone marrow biopsy. MS confirmed AL λ amyloid, leading to the initiation of targeted therapy.

Pt 3: A 61 year-old man with peripheral neuropathy and cardiac involvement, with positive family history for ATTR amyloidosis and positive genetic testing, underwent salivary gland biopsy prior to tafamidis start. Amyloid deposits were identified, with negative IHC and IF results. MS revealed AL λ amyloid, triggering additional investigation; therapeutic regimen is under multidisciplinary discussion.

Pt 4: A 68 year-old man presented with chronic kidney disease (SCr 1,65 mg/dl) and subnephrotic proteinuria. An IgG λ monoclonal protein was identified in plasma. Bone marrow biopsy revealed 11% plasmocytes. In the absence of myeloma-defining events, a diagnosis of indolent myeloma was established and a watchful waiting strategy was adopted. Given persistent proteinuria, kidney biopsy was performed, and revealed Congo red–positive amyloid deposits with negative IHC and IF. MS identified AL λ amyloidosis, leading to initiation of targeted therapy.

Pt 5: A 78 year-old man presented with acute kidney injury requiring haemodialysis (serum creatinine (SCr) at diagnosis 11,5 mg/dL). An IgA κ monoclonal protein was identified in plasma, along with 26% plasmocytes in the bone marrow, establishing an IgA κ multiple myeloma diagnosis. Abdominal fat biopsy demonstrated amyloid deposits without light-chain restriction. MS revealed AL λ amyloid, discordant with the clone initially identified. However, due to rapid clinical deterioration, a decision was made not to initiate therapy.

Pt 6: A 77 year-old woman under follow-up for MGUS IgA λ, presented with de novo nephrotic syndrome (uACR 8053 mg/g) and hypoalbuminemia (sAlb 2.9 g/dl). There was no change in sCreat levels. Bone marrow biopsy showed 9,4% plasma cells; no amyloid deposition was observed. Abdominal fat biopsy revealed amyloid deposits, with inconclusive IHC and IF tests. Only after MS was performed, it was possible to establish a definitive diagnosis of AL amyloidosis and to initiate targeted therapy.

This case series demonstrates that MS resulted in a more precise diagnosis, promoting therapeutic changes with significant impact in patient prognosis. Although its availability remains limited, the authors advocate for MS use in amyloidosis classification when such is not achieved by IHC nor IF and/or clinical context is not explained by the initial typing by other techniques

N	1	2	3	4	5	6
Age at diagnosis (years)	78	86	61	68	78	77
Gender	F	F	M	M	M	F
Clinical presentation	Nephrotic syndrome	Nephrotic syndrome	Peripheral neuropathy	Chronic kidney disease	Acute kidney injury	Nephrotic syndrome
SCr at diagnosis (mg/dL)	1,88	2,48	1,05	1,65	11,5	1,1
uACR at diagnosis (mg/g)	3597	7401	3,7	1168	-	8053
dFLC	28,2	3,7	-	11,6	220,1	54.7
% plasma cells in bone marrow	2,4	-	-	11	26,4	9,4
Clinical diagnosis	AA amyloidosis	-	ATTR amyloidosis	Indolent multiple myeloma	IgA κ multiple myeloma	IgA λ MGUS
Biopsy specimen	Kidney	Kidney	Salivary Gland	Kidney	Abdominal Fat	Abdominal Fat
Congo red staining	+	+	+	+	+	+
IHC	AA Amyloid	-	-	-	-	-
IF	-	IgM ++	-	-	-	-
Amyloid subtype in MS	λ AL	λ AL	λ AL	λ AL	λ AL	λ AL
Pre-MS treatment	0	BDR	Tafamidis	0	0	0
Post-MS treatment	Dara-VCD	BDR	Tafamidis	Dara-VRD	0	Dara-VCD
Death	No	No	No	No	Yes	No

BDR - bortezomib, rituximab and dexamethasone; Dara-VCD - daratumumab, bortezomib, cyclophosphamide, dexamethasone; Dara-VRD - daratumumab, bortezomib, lenalidomide, dexamethasone; dFLC - difference between involved and uninvolved free light chains; F – female; IF - immunofluorescence; IHC - immunohistochemistry; M – male; MGUS- Monoclonal Gammopathy of Undetermined Significance; MS - mass spectrometry; SCr - serum creatinine; uACR - urinary albumin-creatinine ratio;

PO 41
A Diagnostic Challenge: Erdheim–Chester Disease Mimicking Acute Pyelonephritis

Mariana Morais; Marta Margarido; Ana Rita Ramos; Filipa Trigo; Marisa Roldão; Cátia Figueiredo; Rachele Escoli; Ivan Luz; Paulo Santos

Unidade Local de Saúde do Médio Tejo

Background

Erdheim–Chester disease is a rare, and probably subdiagnosed, non-Langerhans cell histiocytosis characterized by multiorgan xanthogranulomatous infiltration. It is a multisystem disease with potential involvement of the neurological and renal systems. Clinical presentation is heterogeneous and may mimic infectious, inflammatory, or neoplastic processes, often leading to delayed diagnosis.

Case description

An 81-year-old man with previously normal renal function and history of type 2 diabetes mellitus, hypertension, dyslipidemia and rheumatic polymyalgia was admitted to the emergency department with a 2-day history of diarrhea, vomiting and anorexia. Blood analysis revealed increased inflammatory parameters and acute kidney injury (Table 1). Abdominopelvic computed tomography scan showed bilateral renal pelvic ectasia and perirenal fat stranding (Figure 1-a), with a striated-type nephrogram, suggestive of acute pyelonephritis. The patient developed urosepsis, with urine culture positive for Enterococcus faecalis, and was started on piperacillin–tazobactam. The infection resolved, however, renal dysfunction persisted (Table 1). Following antibiotic therapy, renal and bladder ultrasound remained abnormal, demonstrating edematous changes in the perirenal space and mild edema of the perivesical adipose tissue. During hospitalization, the patient developed cognitive impairment. Brain magnetic resonance imaging revealed subacute infarcts with hemorrhagic transformation of the parieto-occipital and bilateral temporal junctions, along with chronic infarct sequelae. Renal biopsy was performed, showing histiocytic infiltration extending from the capsule to the

deep cortex, with medullary polymorphonuclear infiltrate and microabscess formation, extensive tubulitis with polymorphonuclear invasion (Figure 1-b,c,d). Findings were suggestive of Erdheim–Chester disease (CD68+, factor XIIIa+, S100−, CD1a−). At the time of discharge, renal dysfunction persisted (Table 1) with progression to stage 4 chronic kidney disease during follow-up. Five months later, the patient died due to a stroke. No skeletal involvement was identified on plain radiography; whole-body metabolic imaging (FDG-PET/CT) had been requested but was not performed prior to the patient's death.
The advanced stage at diagnosis and rapid clinical deterioration limited timely multidisciplinary evaluation and molecular testing, which are currently essential to guide targeted therapy in Erdheim–Chester disease.

Discussion and learning point

This case underscores the diagnostic challenge of Erdheim–Chester disease, whose presentation often mimics common infectious or inflammatory conditions, contributing to its likely underdiagnosis. Persistent renal dysfunction, despite appropriate therapy, and evidence of multisystem involvement should raise suspicion for this entity. Early recognition through tissue biopsy is crucial given the potential for progressive multiorgan involvement.

PO 42
A TUMOUR IN DISGUISE: AA AMYLOIDOSIS AS THE FIRST SIGN OF RENAL CELL CARCINOMA

Manuel Lopes da Silva; Inês Alves Gaspar*; Carolina Branco; Rui Nogueira; Natacha Rodrigues; Estela Nogueira; Sofia Jorge; José António Lopes; Cláudia Costa

ULS Santa Maria

Background:

AA amyloidosis is a systemic disorder characterized by the extracellular deposition of amyloid A fibrils in multiple organs, with the kidney representing the most frequently involved organ. It arises as a complication of sustained chronic inflammation, from a broad spectrum of underlying conditions, including autoimmune diseases, chronic infections and malignancy, that contribute to persistently elevated serum amyloid A (SAA) protein levels. Given the variety of possible causes, it can pose a challenging diagnosis and management relies primarily on targeted treatment of the underlying disease.

Case description:

A 58-year old caucasian woman, with no relevant prior medical history, presented to the emergency department with a 4-day history of asthenia, anorexia, diarrhoea, nausea, vomiting, and progressive bilateral lower limb oedema. Physical examination showed pallor, hypotension and anasarca. Laboratory workup revealed nephrotic-range proteinuria (11 g/24h), severe hypoalbuminaemia (serum albumin 1.1 g/dL), microcytic anaemia (Hb 10 g/dL), elevated inflammatory markers (C-reactive protein 14.4 mg/dL, Erytrocite Sedimentation rate 120 mm/h), and markedly elevated SAA (471 mg/L). Immunological workup, serum and urine immunofixation, and infectious serologies were unremarkable. Abdominal CT showed a left renal mass (7.7 cm) with features suspicious for malignancy. Abdominal fat biopsy was positive for amyloid A and P components. A presumptive diagnosis of AA amyloidosis secondary to renal carcinoma was established. Echocardiography revealed left ventricular hypertrophy and autonomic dysfunction with orthostatic hypotension was present.

After discussion with the Urology team, the patient was submitted to radical left nephrectomy, which confirmed a clear cell renal cell carcinoma, pT3a, with renal vein branch invasion. Renal biopsy demonstrated glomerular and vascular amyloid deposits, with positive

immunohistochemistry for amyloid A and P components, confirming secondary renal AA amyloidosis.

Following nephrectomy, SAA levels normalised (471 to 19 mg/L) and proteinuria declined. However, the patient developed KDIGO stage 3 acute kidney injury, requiring initiation of haemodialysis.

Discussion and learning points:

AA amyloidosis can present as a paraneoplastic syndrome associated with renal cell carcinoma, in which nephrectomy represents the therapeutic cornerstone. The marked reduction in proteinuria and SAA levels following nephrectomy confirmed the causal relationship between the tumour and the amyloid formation process. Nevertheless, renal and overall prognosis in these patients remains poor, reinforcing the need for early diagnosis to prevent further organ damage.

PO 43
FROM SILENT PLAQUE TO SYSTEMIC STORM: A CASE REPORT

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ULS Santa Maria

Category: Acute kidney injury

Background: Cholesterol crystal embolism (CCE) is a systemic condition caused by embolization of cholesterol crystals from unstable atherosclerotic plaques to small- and medium-sized arterioles, leading to ischemia and inflammation, most commonly affecting the kidneys, skin, gastrointestinal tract, and central nervous system. It classically occurs after vascular interventions, anticoagulation or thrombolysis. Rarely, spontaneous CCE occurs, particularly in patients with diffuse atherosclerosis and risk factors such as hypertension, smoking, and diabetes. Diagnosis is often challenging, due to its heterogeneous presentation and absence of a clear precipitating event, with tissue biopsy remaining the gold standard. Prognosis is poor, with about one-third of patients progressing to end-stage kidney disease and significant long-term mortality.

Case description: A 57-year-old Caucasian man with a history of hypertension, dyslipidemia, obesity, tobacco use, inchemic heart failure, diffuse atherosclerotic disease without prior vascular interventions, and chronic kidney disease without regular follow-up (baseline serum creatinine [sCr] 1.6 mg/dL), presented to the emergency department with a four month history of epigastric pain, vomiting, and non-bloody diarrhea, associated with a 10 kg weight loss. He denied oliguria or use of non-steroidal anti-inflammatory drugs. He had been on anticoagulation therapy and statins for the preceding six months, and pantoprazole had been prescribed two weeks prior. On examination, he had cold extremities, livedo reticularis involving the dorsum, and limbs, and two ulcers on the dorsum of the right foot (Figure 1). Laboratory tests showed hemoglobin 11.4 g/dL, leucocytosis 18.7 × 10?/L with neutrophilia and eosinophilia, platelets 373 × 10?/L, sCr 6.11 mg/dL, metabolic acidosis, and LDL cholesterol 48 mg/dL. Computed tomography angiography excluded large vessel renal ischemia or obstructive uropathy. Kidney ultrasound showed normal-sized kidneys with reduced corticomedullary differentiation. Urinalysis showed mild proteinuria,uACR 287 mg/g anduPCR 734 mg/g, and microscopic hematuria. Further workup was negative for autoimmune disease, dysproteinemia and infections. Acute kidney injury was initially attributed to volume depletion, with a possible contribution from pantoprazole-induced interstitial nephritis. Following supportive treatment and medication withdrawal, there was a transient improvement in kidney function (KF), with a nadir sCr of 4.51 mg/dL and resolution of metabolic acidosis. Given the incomplete improvement of KF, a kidney biopsy was performed, demonstrating cholesterol crystal clefts within occluded interlobar arteries, along with marked arteriolosclerosis and multilayering of the internal elastic lamina and intimal fibrosis. (Figure 2) KF

continued to decline, and renal replacement therapy was started and the patient discharged dialysis-dependent.

Discussion and learning point: This case highlights the systemic nature of CCE, affecting the kidneys, skin, and gastrointestinal tract. Furthermore, it underscores that CCE can occur spontaneously, in the absence of an identifiable trigger, which contributes to frequent diagnostic delay. CCE should be considered in patients with diffuse atherosclerosis, even when lipid levels are controlled and no recent intervention has occurred. Early recognition is critical, although therapeutic options remain limited and largely supportive. Despite appropriate management, the disease often follows a progressive course, as seen in our case, culminating in dialysis dependence. This case draws attention to spontaneous CCE as an underrecognized cause of multisystem disease and demonstrates the importance of recognizing subtle clinical features to facilitate an earlier diagnosis.

Figure 1 - Livedo reticularis of the dorsum, arms and legs and dorsum ulcers of the right foot

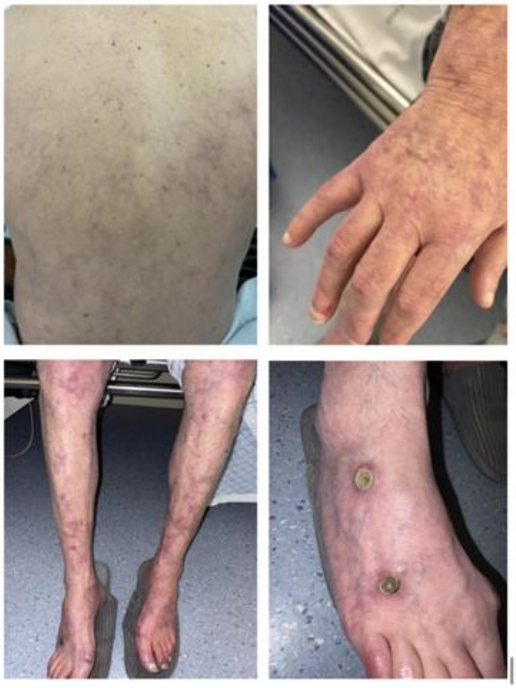
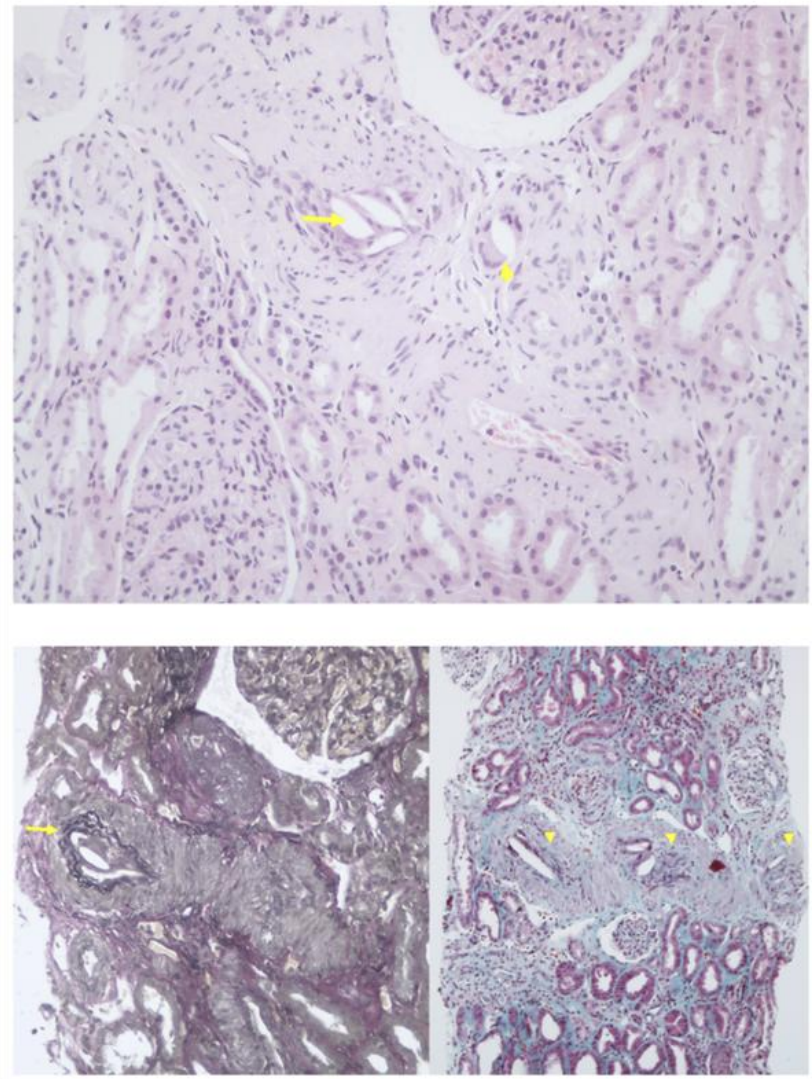


Figure 2 - Kidney biopsy findings



PO 44
WHEN THE URINARY SEDIMENT DOES NOT MATCH THE BIOPSY: DIFFUSE LUPUS NEPHRITIS WITH RENAL THROMBOTIC MICROANGIOPATHY

Catarina Bico Filipe; Ana Rita Almeida; Sara Frazão de Brito; Catarina Teixeira; Adriana Fernandes; Sónia Silva

Unidade Local de Saúde de Loures-Odivelas

Background: Microscopic examination of urinary sediment is a key diagnostic tool in glomerular disease. In proliferative lupus nephritis, dysmorphic erythrocytes and acanthocytes are usually observed.

Case description: A 51-year-old African male with a 10-year history of uncontrolled hypertension and 1-year of unintentional weight loss (8%), presented to the emergency department with dyspnea, orthopnea and lower limbs edema with approximately one-week evolution. At admission the patient had a blood pressure of 250/100 mmHg, was polypneic, requiring supplementary oxygen and lung auscultation revealed bilateral crackles.

Laboratory workup revealed: normocytic anaemia (haemoglobin of 9.4 g/dL, MCV of 94 fL) with a positive direct Coombs test, no thrombocytopenia (platelets 284x10^9/L), LDH elevated (477

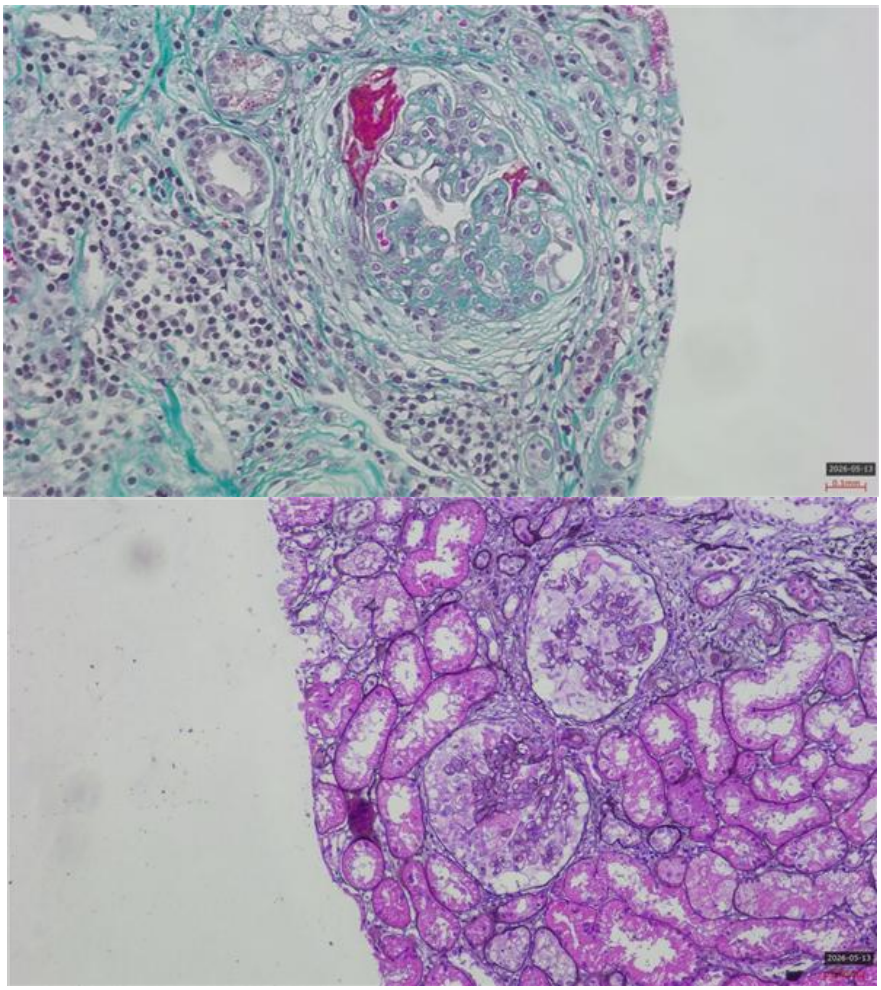
U/L), but without hyperbilirubinemia; acute kidney injury (creatinine of 1.83 mg/dl), urine albumin-creatinine ratio of 622mg/g and urine protein-creatinine ratio of 1069 mg/g). Initial urinalysis showed haematuria (206 erythrocytes/µL) and mild leukocyturia. Microscopic urinary sediment demonstrated predominantly isomorphic erythrocytes. Urine culture was negative. Renal ultrasound demonstrated diffuse increase in echogenicity of the renal parenchyma.

IGRA test was positive and thoraco-abdominal-pelvic CT demonstrated bilateral apical micronodularities and traction bronchiectasis, there was no mention of adenopathies, hepatomegaly or splenomegaly and no sign of pyelocaliceal dilatation or urothelial cancer. Peripheral blood immunophenotyping showed no expression of lymphoproliferative disorders.

Autoimmune testing revealed ANA 1:640 with positivity for anti-SSA, anti-SSB, anti-Ro52, anti-Sm, and RNP/Sm antibodies, with low C3 and negative anti-dsDNA. Lupus anticoagulant, anticardiolipin antibodies and anti-beta-2-glycoprotein were negative. Protein electrophoresis revealed a peak in the gamma region. Serum IgG was elevated; free light chains were elevated but with a normal ratio. Bence Jones was negative. Serum immunofixation did not reveal a monoclonal component. Bone marrow aspirate revealed normal cellularity with 3% of plasma cells, however bone biopsy identified 20% of plasma cells.

During hospitalization the patient had various complications: aggravating anaemia, which motivated the administration of intravenous immunoglobulin; two episodes of acute pulmonary edema and sepsis originating from phlebitis, with methicillin-susceptible *Staphylococcus aureus* isolation in blood cultures for which he was given flucloxacillin. During this period creatinine increased to a maximum of 4.3 mg/dl, but after sepsis recovery and volume overload control, kidney function recovered to the baseline. Since kidney disease's etiology was still unknown, renal biopsy was performed. Histology revealed diffuse proliferative lupus nephritis (class IV) with high activity index (11/24) and mild chronicity (3/12), associated with renal thrombotic microangiopathy.

Discussion and learning points: This case highlights a striking urinary sediment–histology discrepancy in severe proliferative lupus nephritis. These findings suggest that innocent urinary sediment does not exclude severe immune-mediated glomerulopathy, reinforcing the critical role of renal biopsy in patients with unexplained kidney injury.



PO 46
FOCAL SEGMENTAL GLOMERULOSCLEROSIS IN A YOUNG AFRICAN WOMAN WITH LUPUS: LUPUS PODOCYTOPATHY OR APOL1 NEPHROPATHY?

Micael Pompermayer; Lígia Ribeiro; Maria Marques; João Grilo; Raquel Chorão; Catarina Santos; Rui Filipe; Joana Coutinho

Unidade Local de Saúde de Castelo Branco

Background: Focal segmental glomerulosclerosis (FSGS) in patients of African ancestry with systemic lupus erythematosus (SLE) presents a diagnostic and therapeutic challenge, as it may reflect lupus podocytopathy (LP) or APOL1 high-risk variant–associated nephropathy. Both can present with nephrotic syndrome and minimal or absent immune deposits on immunofluorescence, making distinction difficult. However, they differ in pathophysiology, prognosis, and response to therapy. Electron microscopy and APOL1 genotyping are critical for diagnosis and guide treatment decisions.

Case description: A 30 year-old Angolan woman presented with influenza A infection, left pleural effusion, bilateral peripheral edema, acute kidney injury (serum creatinine 1.6 mg/dL; baseline 0.80 mg/dL) and hypertension. Medical history included hypertension and treated malaria infection. Renal ultrasound report described normal kidneys and no signs of obstructive uropathy. Laboratory evaluation revealed hemoglobin 7.2 g/dL with direct Coombs-positive test, low haptoglobin and elevated LDH. Erythrocyte sedimentation rate >140 mm/h, normal platelets and

albumin of 2.2 g/dL. Urinalysis revealed nephrotic range proteinuria (6.6 g/g) with microscopic hematuria. Immunological workup was positive for antinuclear antibodies (anti-ribosomal, anti-nucleosome, anti-SSA/SSB, anti-Sm, anti-RNP), increased anti-dsDNA, hypocomplementemia (low C3, C4, C1q), and negative ANCA, anti-GBM, anti-PLA2R and antiphospholipid antibodies. Serologies for hepatitis C (HCV) and human immunodeficiency virus (HIV) were negative. Hepatitis B surface antigen (HBsAg) was positive with undetectable viral load due to possible past infection. Kidney biopsy (8 glomeruli) demonstrated features of chronic kidney injury with focal segmental glomerulosclerosis, including 37.5% global sclerosis, one segmental sclerotic lesion, Bowman's capsule synechiae, along with 15% interstitial fibrosis and mononuclear infiltrate. Immunofluorescence demonstrated segmental IgM deposition without a full-house pattern. Treatment included intravenous methylprednisolone (500 mg for 3 days), followed by oral prednisolone (initially 60 mg/day, tapered to 20 mg/day over 4 months), mycophenolate sodium (360 mg twice daily), and hydroxychloroquine (400 mg/day), in addition to supportive therapy (empagliflozin, enalapril and nifedipine). At 4 months, kidney function improved (creatinine 0.86 mg/dL) with partial remission of proteinuria (1.86 g/g). APOL1 genotyping and electron microscopy are pending.

Discussion and learning point: This case highlights the challenge of distinguishing lupus podocytopathy from APOL1-associated FSGS in patients of African ancestry with SLE. Despite clinical and histological overlap, these entities differ in mechanisms and therapeutic responsiveness. The absence of a full-house immunofluorescence pattern argues against classical lupus nephritis, yet does not exclude other lupus-related podocytopathies. Electron microscopy and APOL1 genotyping are essential to refine diagnosis and guide therapy. While initial immunosuppression led to partial remission, uncertainty regarding long-term management underscores the need to balance timely nephroprotective therapy with cautious escalation of immunosuppression pending further data.

PO 47
MPO-ANCA VASCULITIS MIMICKING PYELONEPHRITIS: A CASE OF RAPIDLY PROGRESSIVE GLOMERULONEPHRITIS

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ARS Algarve

Category: Systemic diseases

Keywords:

ANCA - associated vasculitides, MPO-ANCA, rapidly progressive glomerulonephritis, pyelonephritis-like presentation

Background:

Antineutrophil cytoplasmic antibody (ANCA)–associated vasculitides are rare autoimmune conditions that cause inflammation of blood vessels and may present as rapidly progressive glomerulonephritis. Early recognition is essential to prevent irreversible kidney damage.

Case description:

A 76-year-old woman with a mechanical aortic valve on warfarin and hypertension was admitted to the Emergency Department with lumbar pain, hematuria, nausea, and fever. Laboratory evaluation revealed severe acute kidney injury (serum creatinine 9.6 mg/dL from baseline 0.9 mg/dL), metabolic acidosis, hyperkalemia, and elevated inflammatory markers. Urinalysis showed hematuria and proteinuria. Abdominal computed tomography showed kidneys of normal size with

preserved parenchymal thickness, no evidence of obstructive uropathy, a 41 mm septated cyst in the lower pole of the left kidney, and a small cortical lesion with macroscopic fat suggestive of angiomyolipoma (Figure 1). Given the clinical presentation and inflammatory profile, acute pyelonephritis was first suspected, empirical antibiotic therapy was initiated. Blood and urine cultures were negative. However, despite appropriate antimicrobial treatment, renal function failed to improve, leading to reconsideration of the initial diagnosis. Further immunological work-up revealed positive myeloperoxidase ANCA (MPO-ANCA), supporting ANCA-associated vasculitis. The patient also presented with supratherapeutic international normalized ratio (INR 9.1), consistent with warfarin-associated coagulopathy, and microcytic anemia requiring transfusion. Immunosuppressive treatment was initiated with high-dose corticosteroids followed by Rituximab. Renal biopsy was not performed due to patient refusal and increased hemorrhagic risk in the setting of coagulopathy. Renal function and electrolyte disturbances progressively improved during hospitalization, with serum creatinine decreasing to 4.0 mg/dL at discharge (Figure 2). The patient was discharged stable with corticosteroid taper and planned rituximab maintenance.

Discussion and learning point:

This case shows a severe presentation of MPO-ANCA vasculitis with rapidly progressive glomerulonephritis and pyelonephritis-like presentation. Failure of renal function to improve despite appropriate antibiotic therapy and the absence of microbiological confirmation were fundamental in redirecting the diagnostic work-up toward ANCA-associated vasculitis. In the absence of histological confirmation, diagnosis relied on clinical, serological, and radiological findings. Renal biopsy remains the diagnostic gold standard but may be limited by patient preference and bleeding risk. Early initiation of immunosuppression should be considered when clinical suspicion is high. Despite marked initial renal impairment, partial renal recovery was achieved, underscoring the importance of timely recognition and multidisciplinary management.

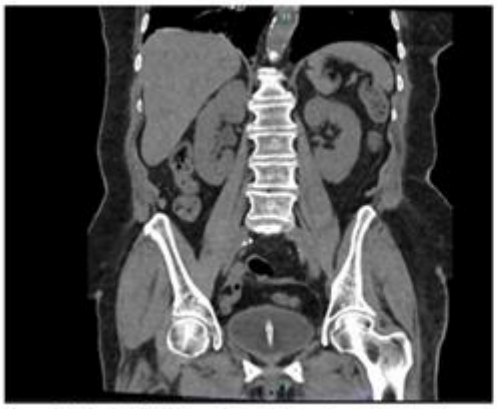

Figure 1. Abdominal CT kidneys of normal size and parenchymal thickness, no evidence of obstructive uropathy, a 41 mm septated cyst in left kidney and angiomyolipoma

Figure 2. Temporal changes in laboratory parameters during hospitalization

Parameter	Admission	Peak	Discharge
Creatinine (mg/dL)	9.6	10	4
Hemoglobin (g/dL)	8.5	9.4	8.8
INR	9.1	1.2	N/A
C- reactive protein (mg/L)	142	160	3
Potassium (mmol/L)	6.3	5.1	3.9
pH	7.283	7.38	N/A
HCO3 (mmol/L)	15.8	21	23
Blood urea nitrogen (mg/dL)	107	108	85

PO 48
A RARE CASE OF POST-TRANSPLANT RECURRENCE OF ANCA-ASSOCIATED VASCULITIS

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Category: Glomerular Diseases.

Keywords: Kidney transplant, ANCA-associated vasculitis, recurrence.

Background: Kidney transplantation (KT) is safe and optimal for those reaching end-stage kidney disease (ESKD) due to ANCA-associated vasculitis (AAV) with glomerulonephritis (AAV-GN). Recurrent disease in the allograft is rare in the current era of modern post-transplant immunosuppression. Both early and late recurrences have been described and appear to behave differently: early recurrence manifests aggressively, while late recurrence is usually indolent and progressive. We report a rare case of late AAV recurrence with severe pulmonary-renal syndrome including de novo diffuse alveolar hemorrhage (DAH).

Case Description: A 53-year-old white male was diagnosed with CKD of unknown etiology at age 37 (2008). He rapidly progressed to ESKD requiring dialysis shortly after the first Nephrology appointment. Deceased-donor KT occurred in 2013, with induction performed using anti-thymocyte globulin protocol and no complications until hospital discharge, showing normal allograft biopsy 14 days post-transplant. Creatinine (Cr) remained stable at 1.5mg/dL until 2017, when nephritic syndrome with AKI (Cr 2.8mg/dL) motivated kidney graft biopsy, revealing 2/13 globally sclerosed (GS) glomeruli and 10% interstitial fibrosis/tubular atrophy (IFTA) without evidence of crescents or fibrinoid necrosis. He was treated with prednisone 20 mg daily tapered over 12 months, partially improving to Cr 2.0mg/dL. In March 2024 (11 years post-KT), he presented with pulmonary-renal syndrome requiring dialysis. ANCA-MPO were positive on workup. Allograft biopsy revealed 7/11 GS glomeruli, as well as one with a cellular crescent (Figure 1A), negative immunofluorescence (including C4d), one artery with fibrinoid necrosis (Figure 1B), and 75% IFTA. Steroids, rituximab and plasmapheresis enabled dialysis-free discharge. Two months later, the patient died of cerebral fungal infection and hemorrhagic stroke.

Discussion and learning points: In this case of post-KT late AAV recurrence, we observed an insidious progressive course over several years - challenging to diagnose despite graft biopsy - culminating in a rare, sudden-onset organ-threatening pulmonary-renal syndrome, including de novo alveolar hemorrhage. A retrospective analysis revealed pre-transplant ANCA positivity that had not been integrated into the initial clinical assessment. This highlights that presumed ESKD of unknown etiology may mask underlying AAV, which carries a persistent risk of post-transplant recurrence despite modern-era post-KT immunosuppression. As AAV recurrences are highly heterogeneous, multidisciplinary care is essential to balance immunosuppression with infectious risk. Improved prognostic markers are needed to personalize long-term follow-up and better calibrate therapeutic intensity in these high-risk patients.

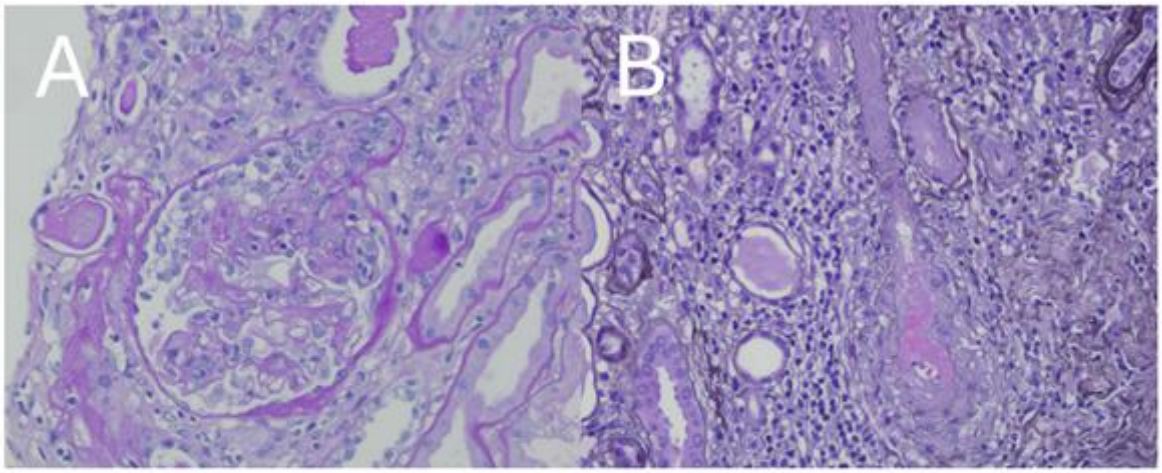

Figure 1. Second graft biopsy. **(A)** PAS stain, 400x magnification, showing the sole glomeruli with a cellular crescent and **(B)** Jones Methenamine Silver stain, 400x magnification: an oblique section of a small-sized artery shows strong, pink-colored fibrinoid necrosis of the inner arterial layers.

PO 50
Not all that lools like a Vasculitis is a Vasculitis: a Hypertensive Thrombotic Microangiopathy in Disguise

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Unidade Local de Saúde de Viseu Dão-Lafões

KEY-WORDS: Thrombotic microangiopathy; Hypertension; Pulmonary–renal syndrome

BACKGROUND:
Concurrent pulmonary and renal involvement is most commonly associated with small-vessel vasculitis and anti-glomerular basement membrane disease. However, alternative etiologies may mimic this clinical pattern, creating significant diagnostic uncertainty and a risk of inappropriate therapeutic decisions.

CASE DESCRIPTION:

A 52-year-old man with a history of untreated hypertension and no regular medical follow-up presented with progressive dyspnea and asthenia. On admission, he was lethargic, with marked hypertension (197/107 mmHg), hypoxemia, and diffuse crackles on pulmonar auscultation. Laboratory evaluation revealed severe acute kidney injury (creatinine 20 mg/dL; urea 534 mg/dL), anemia (7.3 g/dL), thrombocytopenia (60.0 ×10?/L), elevated lactate dehydrogenase (1027 U/L), and increased inflammatory markers (white blood cell count 14.2 ×10?/L, C-reactive protein 25.43 mg/dL and procalcitonin 27.87 ng/mL). Arterial blood gas analysis showed metabolic acidosis (pH 7.24; HCO3? 11.5).

Initial evaluation revealed schistocytes on peripheral blood smear and low haptoglobin, consistent with microangiopathic hemolytic anemia. In this context, complement-mediated thrombotic microangiopathy was suspected, with a possible infectious trigger, and empirical antibiotic therapy was initiated. Chest computed tomography demonstrated extensive bilateral ground-glass opacities with areas of consolidation, suggesting pulmonary involvement and a pulmonary–renal phenotype, without clinical evidence of alveolar hemorrhage.

Further investigation showed normal ADAMTS13 activity, excluding thrombotic thrombocytopenic purpura. Complement levels and autoimmune studies, including ANCA and anti-glomerular basement membrane antibodies, were negative.

The patient required intensive care unit admission, renal replacement therapy with ultrafiltration, and aggressive blood pressure control. Renal ultrasound demonstrated features of chronicity, including reduced corticomedullary differentiation and multiple bilateral lithiasic foci and cortical cysts, leading to the decision not to pursue kidney biopsy. Additional evaluation confirmed longstanding uncontrolled hypertension with evidence of end-organ damage, including hypertensive cardiomyopathy and retinopathy. No infectious cause or alternative etiology of thrombotic microangiopathy was identified.

A diagnosis of hypertensive thrombotic microangiopathy was established. Rapid resolution of hematological abnormalities was observed following blood pressure control and supportive therapy, without the need for immunosuppressive or complement-targeted treatment. However, the patient remained dialysis-dependent at discharge, with no recovery of renal function, reflecting advanced structural kidney damage.

DISCUSSION AND LEARNING POINTS:

Early recognition is essential to guide appropriate management and avoid unnecessary or potentially harmful therapies.

Clinical evolution demonstrates a key dissociation between the reversibility of the microangiopathic process with blood pressure control and supportive therapy, and the absence of renal recovery, reflecting advanced structural kidney damage and poor prognosis, already suggested by imaging findings of chronicity. These findings emphasize the central role of blood pressure control in the management of hypertensive thrombotic microangiopathy, as well as the importance of early diagnosis.

PO 53
A Diagnostic Odyssey: Recurrent Non-PTH-Mediated Hypercalcemia With Acute Kidney Injury

Cláudia Coelho; Catarina Oliveira Silva; Johanna Viana; Joana Costa; Eduardo Cruz; Joana Medeiros; Renata Carvalho; Bárbara Oliveira; Sofia Homem de Melo Marques; Rui Silva; Raquel Vaz; Catarina Teixeira; Rui Costa

Unidade Local de Saúde de Braga

Background

Non–PTH-mediated hypercalcemia represents a significant diagnostic challenge and encompasses malignancy-associated hypercalcemia, granulomatous diseases, vitamin D disorders, and drug-related causes. Severe hypercalcemia can induce afferent arteriolar vasoconstriction, nephrogenic diabetes insipidus, and acute kidney injury (AKI), creating a self-perpetuating cycle of renal dysfunction and metabolic derangement. Renal sarcoidosis is an uncommon manifestation of systemic sarcoidosis and may present as granulomatous tubulointerstitial nephritis or hypercalcemia-related renal impairment. This case illustrates the complexity of diagnosing recurrent severe non–PTH-mediated hypercalcemia associated with AKI in an elderly patient with multiple comorbidities.

Case description

A 79-year-old man with hypertension, type 2 diabetes mellitus, dyslipidemia, hyperuricemia, overweight, and prior tobacco use was admitted for severe AKI (serum creatinine [sCr] 11.7

mg/dL, with a prior sCr of 1.54mg/dL) and hypercalcemia (13.7 mg/dL). He reported unintentional weight loss, fatigue and anorexia. Urinalysis revealed leukocyturia, microscopic hematuria, glucosuria, and tubular proteinuria (urine protein/creatinine ratio of 2g/g; albumin/creatinine ratio of 100mg/g) . PTH was suppressed, while PTH-related peptide and 1,25-dihydroxyvitamin D levels were normal. Extensive malignancy (including cranial, toracic, abdominal and pelvic CT and cystoscopy), infectious (including viral serologies and IGRA), and autoimmune workup was negative, with imaging showing only mediastinal and bilateral hilar lymphadenopathies. Protein electrophoresis and immunofixation was negative and ACE was normal. Renal function improved with hypercalcemia correction and volume repletion, without dialysis, and the patient was discharged (with a sCr of 3.2 mg/dL) to complete etiological workup in an outpatient setting.

The patient was then readmitted with recurrent severe non–PTH-mediated hypercalcemia and non-oliguric AKI (sCr 6.02 mg/dL). At this time, ACE was mildly elevated and immunoelectrophoresis was performed, showing a small IgG kappa monoclonal peak, with a normal serum free light chain ratio. Renal biopsy revealed chronic granulomatous tubulointerstitial nephritis, with non-necrotizing epithelioid granulomas, multinucleated giant cells, moderate to marked interstitial fibrosis and tubular atrophy, and no immune deposits. Subsequent bronchofibroscopy with EBUS demonstrated non-necrotizing epithelioid granulomas and excluded malignancy, supporting the diagnosis of sarcoidosis. Corticosteroid therapy with prednisolone 60 mg/day was initiated, with progressive tappering, resulting in sustained normocalcemia and partial renal recovery (sCr 3.0 mg/dL at 3.5 months).

Discussion and learning point

This case highlights the importance of a structured differential diagnosis in severe non–PTH-mediated hypercalcemia with AKI, with malignancy remaining the leading concern. Granulomatous diseases, particularly sarcoidosis, should be considered when hypercalcemia coexists with lymphadenopathy, tubular proteinuria, and constitutional symptoms, even in the absence of markedly elevated ACE levels, given its limited sensitivity and specificity, with early recognition being crucial, as timely corticosteroid treatment may prevent irreversible renal damage.

The identification of a monoclonal IgG kappa peak further complicated the diagnostic process. However, in older adults, monoclonal gammopathy of undetermined significance is frequent and should not preclude parallel investigation of other etiologies. Therefore, renal biopsy was essential to confirm granulomatous interstitial nephritis, exclude monoclonal immunoglobulin–related renal disease, evaluate chronicity for prognostication, and guide immunosuppressive therapy.

PO 56
AN UNEXPECTED DIAGNOSIS OF ANTI–GLOMERULAR BASEMENT MEMBRANE DISEASE IN A PATIENT PRESENTING WITH METFORMIN-ASSOCIATED LACTIC ACIDOSIS AND ACUTE KIDNEY INJURY

Eva Nunes; Maria Pestana; Ana Vida; Pedro Vieira; Gil Gomes Silva

SESARAM

Category: Acute kidney injury

Keywords: Acute kidney injury, Anti-glomerular basement membrane disease, Crescentic glomerulonephritis

Background:
Acute kidney injury (AKI) in elderly patients is often multifactorial and frequently attributed to dehydration, infection, or drug toxicity. However, lack of renal recovery should prompt investigation of less common etiologies, even when apparent triggers are present.

Case Presentation:

A 76-year-old Caucasian male with a history of hypertension, type 2 diabetes mellitus treated with metformin, and benign prostatic hyperplasia presented to the Emergency Department with recurrent vomiting and nausea. Laboratory tests and arterial blood gas analysis revealed severe anuric AKI (creatinine 13.8 mg/dL), hyperkalemia (potassium 8.4 mEq/L), and lactic acidosis (pH 6.9; lactate 9.2 mmol/L). Abdominal computed tomography showed kidneys of normal size and appearance.

Despite initial medical management, urgent renal replacement therapy was initiated. A presumptive diagnosis of metformin-associated lactic acidosis (MALA) with prerenal AKI due to prolonged volume depletion was made. However, due to persistent renal dysfunction without recovery, a renal biopsy was performed. Histological examination demonstrated diffuse necrotizing crescentic glomerulonephritis (100% crescents) with linear IgG staining along the glomerular basement membrane consistent with anti–glomerular basement membrane (anti-GBM) disease and associated acute tubular necrosis.

Serological testing confirmed markedly elevated circulating anti-GBM antibodies (>1000 U/mL). Antinuclear antibodies, anti–double-stranded DNA antibodies, antineutrophil cytoplasmic antibodies (ANCA), and serologies for human immunodeficiency virus (HIV), hepatitis B, and hepatitis C were negative. Complement levels were within normal limits. Given the extensive crescent formation and absence of pulmonary hemorrhage, immunosuppressive therapy was not initiated. At present, the patient remains dependent on maintenance hemodialysis.

Discussion and Learning Points:

This case highlights the importance of maintaining a broad differential diagnosis in AKI. Although common contributing factors such as MALA and dehydration were initially identified, the absence of renal recovery was a key clinical indicator warranting further investigation with kidney biopsy. Anti-GBM disease is a rare but severe condition that may be overlooked when more common causes of AKI coexist, particularly in elderly patients. While no direct causal relationship between metformin use and anti-GBM disease has been established, the profound metabolic disturbance associated with lactic acidosis may act as a potential trigger in immunologically susceptible individuals. Early recognition is critical, although prognosis remains poor in cases with advanced histological damage, as illustrated by this patient’s outcome.

PO 58
MULTIPLE RENAL LESIONS ASSOCIATED WITH LAMBDA LIGHT CHAINS: DEPOSITION DISEASE, TUBULOINTERSTITIAL NEPHRITIS AND INCIPIENT CAST NEPHROPATHY

Eva Nunes; Ana Vida; Pedro Vieira; Maria Pestana; Gil Gomes Silva

SESARAM

Category: Systemic diseases

Keywords: Multiple myeloma, light chain deposition disease, tubulointerstitial nephritis

Background:
Monoclonal gammopathy of renal significance (MGRS) encompasses a spectrum of disorders in which monoclonal proteins cause organ damage and may represent an early manifestation of multiple myeloma (MM). Light chain–mediated renal injury typically presents with a single predominant histological pattern; however, the simultaneous occurrence of multiple lesions is rare. We describe an unusual case combining light chain deposition disease (LCDD), light chain–associated tubulointerstitial nephritis (TIN), and incipient cast nephropathy—an association scarcely reported in the literature.

Case Description:
A 52-year-old woman with a history of monoclonal gammopathy of undetermined significance (MGUS), followed by Hematology since 2016, presented with new-onset hypertension, macroscopic hematuria, lower limb edema, and progressive fatigue. These symptoms developed after a respiratory infection treated with a one-week course of amoxicillin/clavulanic acid. Laboratory evaluation revealed acute kidney injury, with serum creatinine rising from a baseline of 0.6 mg/dL to 1.5 mg/dL, along with active urinary sediment characterized by dysmorphic erythrocyturia (34–55%). Proteinuria was 900 mg/24 hours. The erythrocyte sedimentation rate was elevated (130 mm/h). Serum studies showed an IgG/lambda monoclonal component of 28 g/L and a kappa/lambda free light chain ratio of 0.1. A kidney biopsy was performed and demonstrated intense linear staining for lambda light chains in tubular basement membranes and glomeruli, consistent with LCDD. Additionally, there was an interstitial inflammatory infiltrate with lambda restriction, supporting a diagnosis of light chain–associated TIN. Proteinaceous casts in distal tubules with predominant lambda expression indicated incipient cast nephropathy.

Treatment was initiated with prednisolone (1 mg/kg), and the patient was referred to Hematology. A myelogram revealed 10.4% plasma cell infiltration. Based on renal involvement and bone marrow findings, a diagnosis of MM was established. The patient was treated with lenalidomide, bortezomib, daratumumab, and dexamethasone. Eight months after diagnosis, she underwent autologous stem cell transplantation. On follow-up, renal function improved significantly, with serum creatinine decreasing to 0.82 mg/dL and resolution of proteinuria.

Discussion and Learning Points:

This case illustrates an atypic presentation of LCDD, revealing also multiple toxicity patterns considering light chain involvement. Kidney biopsy was key to establish the diagnosis and allow targeted treatment. A critical point was the differential diagnosis of TIN; although antibiotic use suggested drug-induced etiology, lambda chain restriction in the infiltrate and the association with LCDD confirmed monoclonal nature of the lesion.

PO 59
When MPO-ANCA rises without clinical relapse: limitations of biomarker-driven decision-making in eosinophilic granulomatosis with polyangiitis

Adriana Moita Dias; , Catarina Maia Veiga; Joaquim Milheiro ; Joana M. Abreu; Joana Figueiredo; André Ferreira; Inês Cunha; Tânia Couto Sousa; Sérgio Lemos

Unidade Local de Saúde de Viseu Dão-Lafões

KEYWORDS:
Eosinophilic granulomatosis with polyangiitis; ANCA; Rituximab

BACKGROUND:
Assessing disease activity in eosinophilic granulomatosis with polyangiitis (EGPA) remains challenging, particularly under rituximab therapy, where discordance between biomarkers and clinical status may occur.

CASE DESCRIPTION:
A 68-year-old woman with a history of systemic sclerosis with pulmonary involvement and no prior renal disease presented with 15 days of asthenia and exertional dyspnea, with normal blood pressure (132/77 mmHg). Initial investigations revealed markedly elevated serum troponin (1025.74 ng/L), serum creatinine of 1.4 mg/dL, and C-reactive protein of 16.53 mg/dL. Electrocardiogram, chest CT angiography, and renal ultrasound were unremarkable. She was admitted to the Cardiology department with suspected myocarditis versus cardiac involvement related to systemic sclerosis.

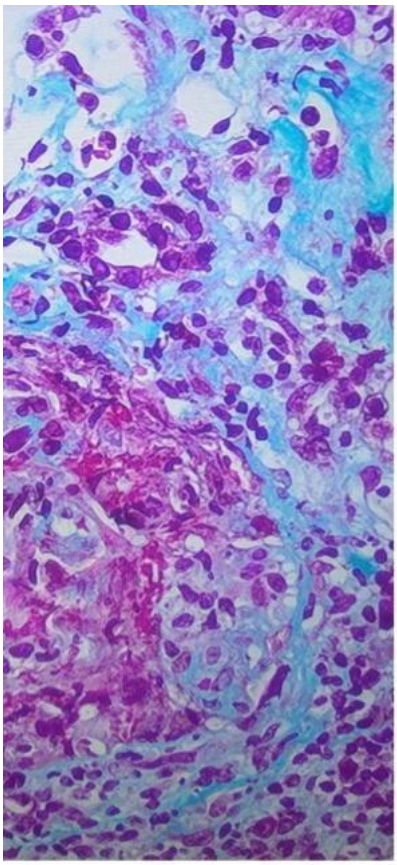
During hospitalization, she developed recurrent fever without evidence of infection, persistent asthenia, and exertional intolerance, with sustained troponin elevation. Laboratory evaluation showed eosinophilia (1.6 × 10?/L) and acute kidney injury (peak creatinine 1.7 mg/dL), with mild proteinuria (370 mg/24h) and no hematuria. Immunological workup revealed anti-myeloperoxidase (MPO)-ANCA positivity (160 U/mL). Transthoracic echocardiography was normal. Cardiac magnetic resonance imaging demonstrated non-ischemic late gadolinium enhancement without evidence of active myocardial inflammation, consistent with prior myocardial involvement.

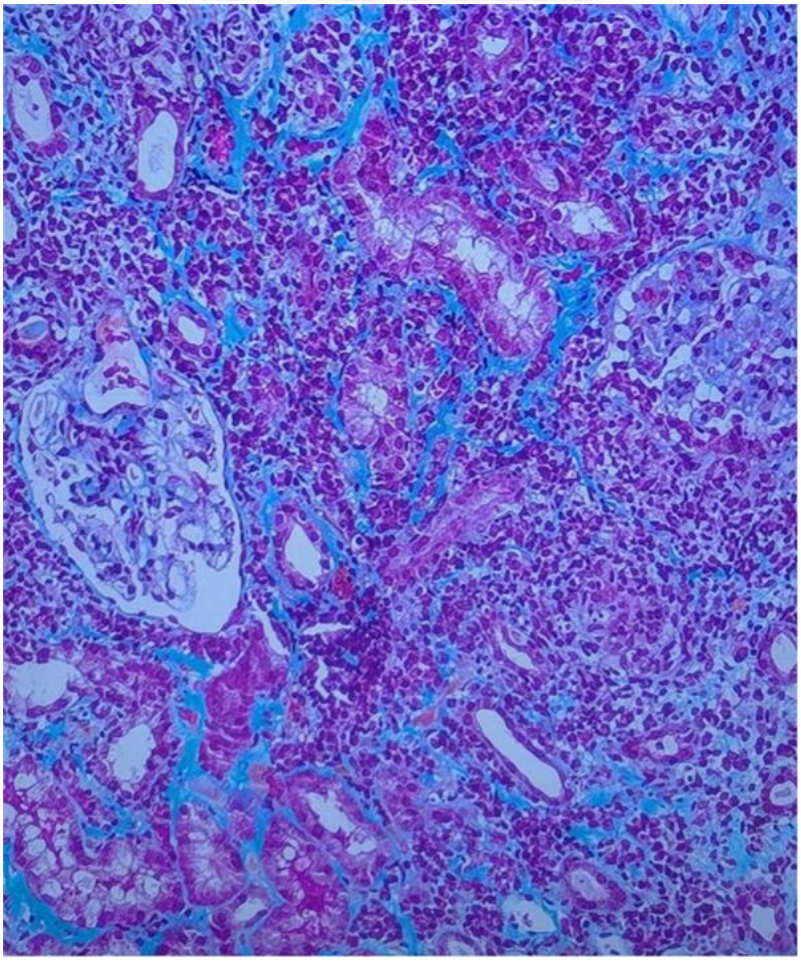
Kidney biopsy (10 glomeruli) showed pauci-immune necrotizing glomerulonephritis with fibrinoid necrosis and cellular crescents in 2 glomeruli (Figure 1), along with marked eosinophil-rich interstitial inflammatory infiltrate involving approximately 40% of the cortex, tubulitis, and acute tubular necrosis (Figure 2). Immunofluorescence was negative, consistent with a pauci-immune process. These findings supported a diagnosis of EGPA with multiorgan involvement, compatible with ANCA-associated glomerulonephritis (Berden classification: focal class).

Treatment with rituximab and corticosteroids induced clinical and biochemical remission, with normalization of troponin levels (13.8 ng/L), improvement of inflammatory markers, and stabilization of renal function (creatinine 1.3 mg/dL). During follow-up over several months, while on maintenance rituximab, a progressive and sustained rise in MPO-ANCA titers (up to 19 U/mL) was observed despite persistent clinical remission, stable renal function, and no new organ involvement. The patient remains under close monitoring, and the decision to re-administer rituximab is guided by immunological assessment, including lymphocyte count, rather than ANCA trends alone.

DISCUSSION AND LEARNING POINTS:
This case challenges biomarker-driven clinical decision-making in EGPA. Cardiac magnetic resonance imaging highlights that organ involvement may persist as a marker of prior injury in the absence of active inflammation. Concurrently, rising MPO-ANCA titers during sustained clinical remission illustrate that these biomarkers may not reliably reflect disease activity.

Uncritical reliance on ANCA trends to guide therapy may lead to inappropriate treatment escalation. This case underscores the need for integrated clinical assessment and cautions against biomarker-driven management strategies that may expose patients to unnecessary treatment and potential iatrogenesis.





PO 60
PLA2R-POSITIVE MEMBRANOUS NEPHROPATHY ASSOCIATED MANTLE CELL LYMPHOMA

Eva Nunes; Maria Pestana; Ana Vida; Pedro Vieira; Gil Gomes Silva

SESARAM

Category: Glomerular diseases

Keywords: Membranous nephropathy, Phospholipase A2 receptor, Mantle cell lymphoma

Background: Membranous nephropathy (MN) is a leading cause of nephrotic syndrome in adults. Although MN may occur as a paraneoplastic syndrome—most often linked to solid tumors and less frequently to lymphomas—the presence of Phospholipase A2 receptor (PLA2R) antibodies typically supports a diagnosis of primary MN. However, even in a presumptive primary MN malignancy cannot be definitively excluded at presentation or during follow-up.

Case description: A 50-year-old caucasian male with a history of asymptomatic celiac disease and active smoking presented with foamy urine and lower limb edema for over 3 months. Laboratory evaluation showed preserved renal function (creatinine 0.89 mg/dL), severe hypoalbuminemia (albumin 17 g/dL) and hypercholesterolemia (total cholesterol 405 mg/dL). Urine analysis suggested a nephrotic range proteinuria with 4+ proteinuria which was confirmed in a 24h urine analysis (30gr/day). Renal ultrasound showed normal sized kidneys. A kidney biopsy was performed revealing IgG, kappa and lambda granular capillary deposits suggestive of membranous nephropathy. IF staining of PLA2R was negative in the glomerular capillary wall. Serological testing revealed circulating anti-PLA2R antibodies (titer 1:50) with normal antinuclear antibodies, anti-neutrophil cytoplasmic autoantibodies and anti-glomerular basement membrane antibodies. Thyroid function and viral serologies were unremarkable. Regardless of serum PLA2r antibodies positivity the patient was submitted to cancer screening exams which showed no abnormalities. Primary MN was presumed and treatment with renin–angiotensin–aldosterone system inhibition was started with ramipril along with empagliflozin, furosemide, atorvastatin, enoxaparine and sodium restriction was initiated. Due to persistent nephrotic syndrome, tacrolimus was initiated, resulting in rapid reduction of proteinuria. After 3 months, anti-PLA2R antibodies became undetectable and tacrolimus was continued for 14 months. 3 months after discontinuation, the patient developed generalized lymphadenopathy and splenomegaly. Inguinal lymph node biopsy confirmed mantle cell lymphoma. Chemotherapy (R-DHAP, R-CHOP, and ibrutinib) was initiated. Renal function progressively declined without recurrence of proteinuria. The patient died due to sepsis.

Discussion and learning points: The clinical course of this patient raises important questions regarding the etiology of membranous nephropathy (MN) and its potential association with malignancy. Although anti-PLA2R positivity, negative initial cancer screening, and response to tacrolimus supported a diagnosis of primary MN, the subsequent diagnosis of mantle cell lymphoma (MCL) shortly thereafter raises the possibility that MN represented an early paraneoplastic manifestation of an underlying, initially subclinical malignancy. This case aligns with emerging evidence suggesting that the incidence of cancer may be increased in patients with PLA2R-associated MN compared to the general population. These findings highlight the importance of continued long-term oncologic surveillance in patients with PLA2R-related MN.

PO 61
ANTI-C1Q VASCULITIS: WHEN COMPLEMENT UNMASKS GLOMERULAR DISEASE

Marta Margarido; Mariana Morais; Rita Ramos; Filipa Trigo; Marisa Roldão; Cátia Figueiredo; Inês Duarte; Ivan Luz; Rachele Escoli; Paulo Santos

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Category:

Glomerular Diseases

Background:

Anti-C1q vasculitis is a rare immune complex-mediated small vessel vasculitis characterized by chronic urticaria, hypocomplementemia, and anti-C1q antibodies, with possible systemic involvement including renal disease.

Case description:

A 47-year-old woman with antiphospholipid syndrome and overweight was referred to Nephrology in February 2025 for impaired renal function. She had no family history and was asymptomatic. One month later, she developed pruritic, non-scaling cutaneous lesions unresponsive to antihistamines and topical corticosteroids, along with asthenia and abdominal pain.

Laboratory evaluation showed worsening renal function (serum creatinine (SCr) peak 2.5 mg/dL), active urinary sediment with dysmorphic hematuria, subnephrotic proteinuria (urine protein/creatinine (UPC) ratio peak 642 mg/g), anemia (hemoglobin nadir 7 g/dL) and thrombocytopenia (100×10?/L). Complement levels were markedly decreased (C3 39.3 mg/dL, C4 8.4 mg/dL, C1q 12.1 mg/dL, CH50 14.9 U/mL), ANA was positive (1:160), and anti-dsDNA was negative.

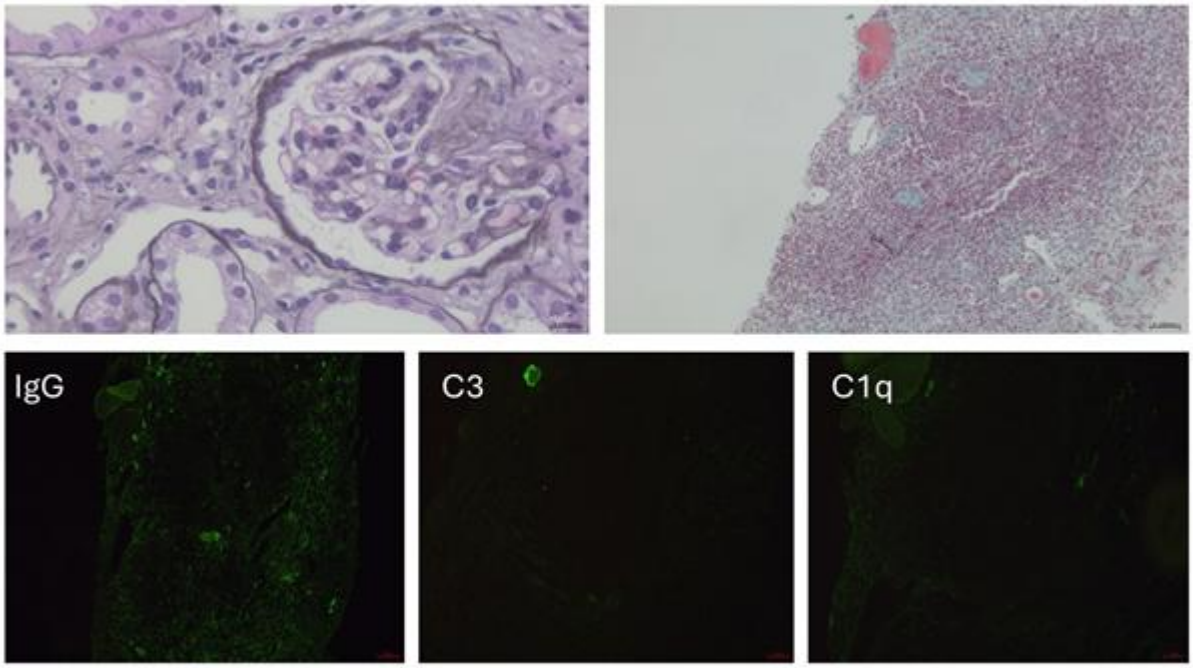
Given rapid renal deterioration with systemic features, she was admitted for further evaluation. Persistent hypocomplementemia, along with positive anti-C1q antibodies, was documented. Kidney biopsy revealed immune complex-mediated membranoproliferative glomerulonephritis, with marked interstitial inflammatory infiltrate with granulomatous features, tubulitis, and vasculitis involving a medium-sized artery. Immunofluorescence showed IgG, C3, and C1q deposits in the interstitium and vessel walls, without glomerular deposits.

High-dose corticosteroid therapy was initiated, with progressive clinical and laboratory improvement, including resolution of cytopenias and skin lesions, normalization of complement levels, and renal recovery (SCr 1.0 mg/dL; UPC ratio 143 mg/g), with only mild residual active urinary sediment at one year follow-up with low-dose corticosteroid maintenance therapy.

Discussion and learning point:

This case illustrates the diagnostic complexity of complement-mediated disease with systemic involvement. The combination of hypocomplementemia, including C1q, positive anti-C1q antibodies, and clinicopathological findings supported the diagnosis despite heterogeneous renal histopathology.

It underscores the importance of integrating clinical, serological, and histopathological data in patients with hypocomplementemic glomerulonephritis. Early immunosuppressive therapy was associated with significant clinical and laboratory improvement, highlighting the potential reversibility of organ involvement when promptly recognized.



PO 62
CRESCENTIC GLOMERULONEPHRITIS AS A RARE PRESENTATION OF AA AMYLOIDOSIS: WHEN THE KIDNEY BIOPSY REVEALS THE UNDERLYING DISEASE

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AA amyloidosis is a systemic disorder caused by the extracellular deposition of fibrils derived from serum amyloid A, an acute-phase reactant whose sustained elevation, driven by chronic inflammation, is the central pathogenic event. The kidney is the most frequently involved organ, typically presenting with proteinuria progressing to nephrotic syndrome. Glomerular crescents are a non-specific lesion seen across many glomerular diseases and are not classically considered a feature of renal amyloidosis, although rare reports describe their occurrence superimposed on amyloid deposits.

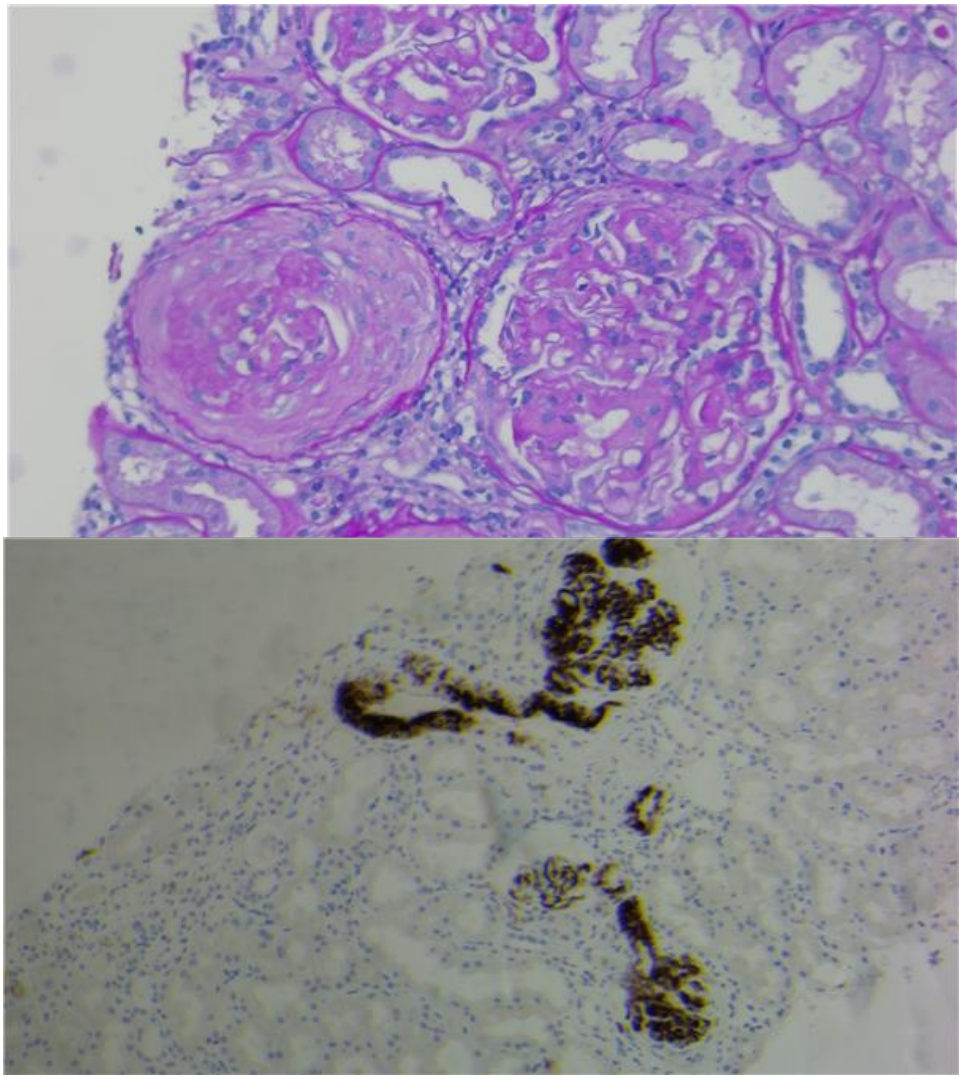
A 73-year-old man was referred for evaluation of severe albuminuria (urine albumin/creatinine ratio 2.2 g/g) with preserved kidney function (serum creatinine 1.0 mg/dL). His history was notable for long-standing hypertension, previous moderate-to-severe psoriasis, post-infectious bronchiectasis, and several years of progressive arthralgia of the hands and knees, managed only with non-steroidal anti-inflammatory drugs. Laboratory work-up serum albumin 2.4 g/dL, microscopic hematuria (>50 erythrocytes/HPF), normocytic anemia (hemoglobin 9.8 g/dL), with negative autoantibody, monoclonal-protein, and infectious screens. During follow-up, the patient developed a pulmonary embolism attributed to nephrotic syndrome; an extensive evaluation excluded malignancy. Kidney biopsy revealed global sclerosis in approximately one-third of glomeruli, fibrocellular crescents in two of the remaining glomeruli, and mesangial expansion by amorphous, PAS- and silver-negative material (Figure 1). Congo red staining demonstrated apple-green birefringence under polarized light in glomeruli and vascular walls, with immunohistochemical positivity for amyloid A protein (Figure 2). Immunofluorescence was negative for immunoglobulins, complement factors, and light chains. Interstitial fibrosis involved approximately 20% of the cortical area.

The diagnosis of AA amyloidosis prompted a directed search for an underlying inflammatory cause. Although bronchiectasis and previous psoriasis were considered, a focused clinical re-evaluation uncovered chronic inflammatory polyarthralgia of the hands and feet with wrist and dorsal-hand swelling, ultimately leading to a new diagnosis of seropositive rheumatoid arthritis. Treatment was initiated with prednisolone 5 mg daily, methotrexate 15 mg weekly, and the interleukin-6 receptor inhibitor tocilizumab 162 mg subcutaneously weekly.

This case illustrates two clinically relevant points: (i) AA amyloidosis may unmask a previously unrecognized inflammatory rheumatic disease, underscoring the need for a thorough search for an underlying inflammatory driver whenever this diagnosis is established; and (ii) crescent formation can complicate amyloid deposition and should not divert attention from the underlying systemic process. Preserved kidney function and limited interstitial fibrosis at diagnosis, combined with effective suppression of serum amyloid A through interleukin-6 receptor blockade, may favor a better long-term renal outcome.

Figure 1. Periodic acid–Schiff (PAS) stain demonstrating a fibrocellular crescent and an adhesion of the glomerular tuft.

Figure 2: Immunohistochemistry revealing Amyloid A protein positivity in glomeruli and vascular walls.



PO 63
Anti-nephrin antibodies: a revolutionary discovery

Inês Costa Clemente; Liliana Gonçalves Cunha; Bruno Rodrigues; Ana Azevedo; Catarina Abrantes; Catarina Marouço; João Bernardo; Joel Ferreira; Miguel Petrucci; Patrícia Valério; Teresa Stock Cunha

Unidade Local de Saúde do Estuário do Tejo

Category: Glomerular diseases

Background:

Minimal Change Disease (MCD) is a primary podocytopathy and a leading cause of nephrotic syndrome in both children and adults. Histologically, it is characterized by diffuse podocyte foot process effacement on electron microscopy, in the absence of significant abnormalities on light microscopy or immunofluorescence. Recent evidence has identified circulating anti-nephrin autoantibodies as a key pathogenic and diagnostic marker in MCD.

Case presentation:

We present a 50-year-old male, born in Mozambique, with a past medical history for non-Hodgkin Lymphoma in remission and Oropharyngeal Squamous Cell Carcinoma recently treated and in remission, presented to the Emergency Department with a one-month history of progressive lower limb and facial edema, associated with foamy urine.

On admission, laboratory evaluation revealed acute kidney injury, AKIN stage 2, with a serum creatinine level of 1.91 mg/dL (baseline 0.74 mg/dL), blood urea of 298 mg/dL, and severe hyperkalemia (6.70 mmol/L). Urinalysis showed marked proteinuria (≥300 mg/dL). Further investigation demonstrated nephrotic-range proteinuria (urine protein-to-creatinine ratio (PCR) of 16 g/g), accompanied by severe hypoalbuminemia (serum albumin 0.88 g/dL) and marked hypercholesterolemia (total cholesterol 435 mg/dL). The autoimmune workup was negative. Two weeks prior, the patient underwent a positron emission tomography (PET) with no evidence of oncologic and hematologic disease recurrence. The renal biopsy showed: 12 glomeruli with no significant structural abnormalities on light microscopy. Immunofluorescence microscopy demonstrated fine granular (“dust-like”) IgG staining along podocyte foot processes. The diagnosis of MCD was made, and he was treated with oral prednisolone 1mg/Kg/day. Testing for circulating anti-nephrin antibodies was positive.

Despite two months of full-dose corticosteroid therapy, the patient developed severe steroid-refractory nephrotic syndrome, with a serum creatinine level of 2.58 mg/dL, a protein-to-creatinine ratio (PCR) of 8.87 g/g, and a serum albumin level of 0.60 g/dL. A repeat PET scan again demonstrated no evidence of active oncologic or hematologic disease. Rituximab therapy was subsequently initiated – two doses of 1000mg administered two weeks apart. The patient also developed *Klebsiella pneumoniae* bacteremia and received targeted antibiotherapy and two doses of endovenous immunoglobulins (IG) due to low baseline IgG levels. Due to severe hypogammaglobulinemia we had prophylactic amoxicillin. After six weeks of treatment with rituximab with no response, tacrolimus was initiated, with a subsequent favorable response in few weeks with a complete remission after four months. Anti-nephrin antibodies were re-measured during follow-up while the patient was under treatment, showing a progressive decline in their levels.

Discussion:

Anti-nephrin antibodies represent a revolutionary discovery because of their promising non-invasive biomarker for diagnosis, assessment of activity of the disease, and monitoring the

response of the treatment in MCD. In this specific case, anti-nephrin antibodies played a significant role in establishing the second line treatment in a severe case, reinforcing the importance of this novel biomarker, and highlighting its potential as an important step toward precision medicine, with the capacity to improve clinical decision-making and patient outcomes.

Learning points:

This case shows, in clinical practice, the importance of anti-nephrin antibodies in the management of refractory Minimal Change Disease.

PO 64
Hidden Cryoglobulinemia in Monoclonal Gammopathy of Renal Significance: The Diagnostic Value of Kidney Biopsy

Diogo Borges; Rafael Terêncio; Mariana Pais; Catarina Cardoso; Maria Rita Dias; Cristina Vilela

Unidade Local de Saúde Almada-Seixal, Almada, Portugal

Category: Systemic diseases

Background:

Monoclonal gammopathy of renal significance (MGRS) can cause diverse renal lesions, including glomerular injury mediated by monoclonal immunoglobulin deposition. Type II cryoglobulinemia associated with IgM monoclonal gammopathy is a rare cause of MGRS and may present with systemic vasculitic manifestations and progressive kidney dysfunction.

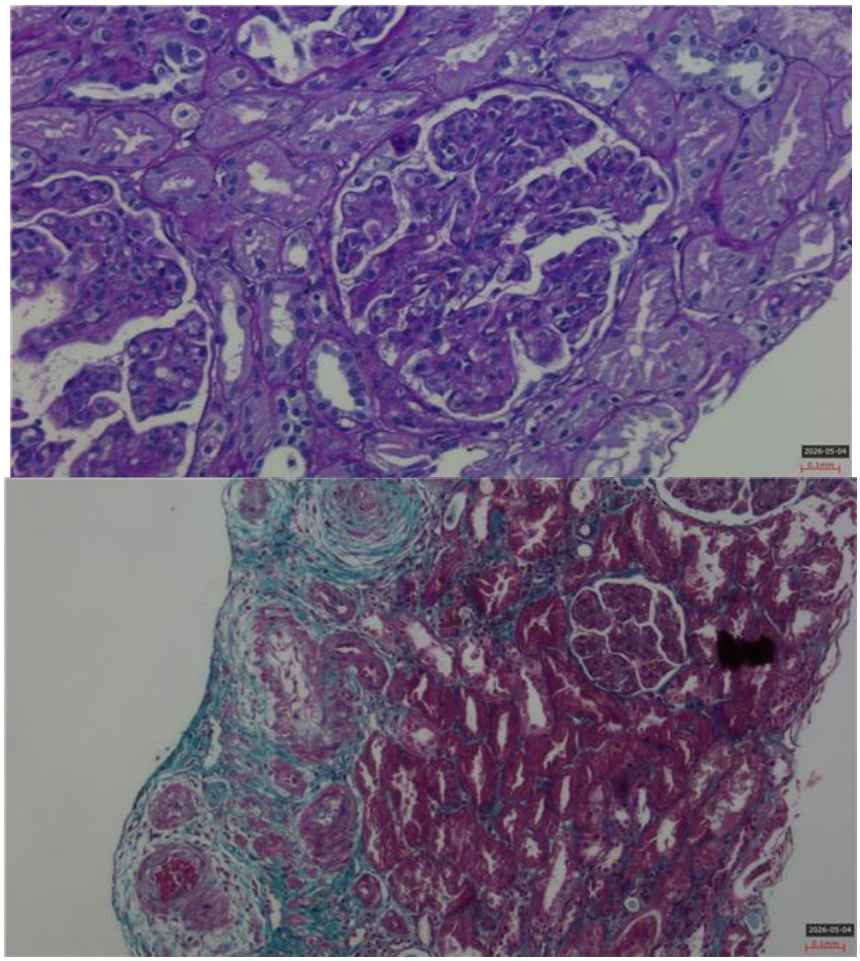
Case description:

A 75-year-old Caucasian female with a history of hypertension, paroxysmal atrial fibrillation, heart failure with preserved ejection fraction, dyslipidemia and prior ischemic presented with fatigue, lower limb edema, purpuric skin lesions and lower extremity paresthesias. Initial workup revealed normocytic anemia (hemoglobin 10.7 g/dL), serum creatinine 1.36–1.66 mg/dL, hypoalbuminemia (2.7 g/dL), 24-hour proteinuria of 800 mg, monoclonal IgM/kappa gammopathy, low complement levels (C3 82 mg/dL, C4 0.6 mg/dL), and markedly elevated rheumatoid factor (865 IU/mL). Autoimmune serologies were negative. Although serum cryoglobulins were negative, the clinical picture raised suspicion for systemic cryoglobulinemic disease.

Kidney biopsy showed membranoproliferative glomerulonephritis with intracapillary pseudothrombi, medium-vessel involvement, and immunofluorescence demonstrating mesangial and capillary wall deposits of IgM and kappa light chains, consistent with type II monoclonal cryoglobulinemic glomerulonephritis. Hematologic evaluation excluded overt lymphoma or Waldenström's macroglobulinemia. MGRS-associated cryoglobulinemic vasculitis was assumed. The patient was treated with methylprednisolone pulses followed by rituximab (1 g ×2), with subsequent oral prednisone taper. Supportive therapy and close multidisciplinary follow-up were maintained.

Discussion and learning point:

This case highlights that MGRS may present cryoglobulinemic glomerulonephritis even in the absence of detectable circulating cryoglobulins. Kidney biopsy was essential for diagnosis. Early recognition and multidisciplinary treatment are critical to prevent progressive renal damage and systemic complications.



PO 65
Breaking the Rules of Anti-GBM Disease: High Titers Without Classical Pathology

Silvia Vega Gonzalez

Hospital de Bellvitge

Anti–glomerular basement membrane(anti-GBM) disease is a rare small-vessel vasculitis affecting glomerular and/or pulmonary capillaries. It is characterized by circulating anti-GBM antibodies directed against the noncollagenous (NC1)domain of the α3 chain of type IV collagen in the glomerular and alveolar basement membranes. Around 90% of patients present with renal involvement, while 20–40% have isolated kidney disease and only 10% isolated pulmonary disease. The disease has a bimodal age distribution, affecting younger adults with combined pulmonary–renal involvement and older patients with isolated renal disease more frequently.

Diagnosis relies on kidney biopsy considered the gold standard, typically demonstrating strong linear IgG deposition along the glomerular basement membrane immunofluorescence. Most patients present with rapidly progressive glomerulonephritis (GN) and 40–60% develop pulmonary hemorrhage. Poor renal prognosis is associated with severe renal dysfunction, extensive crescent formation, and dialysis dependence at presentation. Renal recovery in dialysis-dependent patients is uncommon, occurring in only 8–10% of historical cases.

We describe a 32-year-old woman admitted with suspected anti-GBM disease. She presented a 3–4 week history of asthenia and constitutional symptoms without pulmonary manifestations or oliguria. Laboratory tests revealed anemia, acute kidney injury (creatinine 184

mmol/L),microscopic hematuria,minimal albuminuria, strongly positive anti-GBM antibodies(1187), and negative ANCA. ChestX-ray was normal.

An early kidney biopsy was performed and treatment initiated with plasma exchange, corticosteroids, and cyclophosphamide. Histology showed 21 glomeruli, 70–80% partially sclerosed with fibroepithelial crescents and focal necrosis with weak, non-linear IgG expression immunofluorescence along the basement membrane while IgA, IgM, complement, and light chains were negative.

During hospitalization, the patient developed febrile neutropenia secondary to cyclophosphamide and progressive renal failure requiring dialysis. Anti-GBM titers decreased markedly after treatment. Peritoneal dialysis was later initiated, but over follow-up the patient experienced progressive renal recovery, increasing urine output, negativization of anti-GBM antibodies at 3 months, and improvement in kidney function (eGFR 24 ml/min), allowing dialysis discontinuation.

Our case showed a marked discordance between very high anti-GBM antibody titers and the clinical and histological findings. Unlike previously described atypical anti-GBM cases, which are characterized by linear Ig deposition on kidney biopsy without detectable circulating antibodies, our patient had strongly positive circulating anti-GBM antibodies but lacked the typical linear IgG staining pattern and presented with a non-indolent clinical course.

Linear IgG deposition is not specific to anti-GBM disease and may also occur in diabetic nephropathy, monoclonal immunoglobulin deposition disease (MIDD), and fibrillary glomerulonephritis. In our patient, who was young and non-diabetic, additional staining techniques and electron microscopy excluded these alternative diagnoses.

Another peculiarity was the significant renal recovery despite dialysis-dependent renal failure and markedly elevated anti-GBM antibody titers, a finding rarely observed in classical anti-GBM disease. The combination of absent linear IgG deposition, recovery after dialysis, and isolated anti-GBM seropositivity suggests a rare and previously undescribed phenotype, possibly related to non-pathogenic anti-GBM antibodies or an ANCA-negative vasculitic process. Unlike the more commonly reported overlap syndromes with concurrent ANCA positivity, ANCA-negative disease with isolated anti-GBM positivity and non-classical histological findings remains exceptionally rare.

PO 66
BEYOND THE OBVIOUS IN MEMBRANOUS NEPHROPATHY: CLINICAL CHALLENGES IN DIAGNOSIS AND TREATMENT BASED ON A CASE REPORT

Tiago Resende; Roberto Calças Marques; Rodolfo Almeida; Filipa Brito Mendes; Ana Cabrita; Ana Paula Silva

Unidade Local de Saúde do Algarve

Category | Glomerular diseases

Background | Membranous nephropathy (MN) is a leading cause of nephrotic syndrome (NS) in adults. Malignancy-associated MN should be considered in older patients, particularly when clinical or biochemical red flags are present. However, serum anti-phospholipase A2 receptor (PLA2R) antibody positivity strongly supports the diagnosis of primary or PLA2R-associated MN, even when a potential secondary cause is identified. This diagnostic distinction is clinically relevant, as treatment decisions must balance MN activity, kidney prognosis, malignancy status, and immunosuppression-related risks.

Case description | A 62-year-old male was admitted to our ward with progressive NS of unknown origin (peripheral edema, hypoalbuminemia, de novo hyperlipidemia, proteinuria of 18g/24h, see table 1 for laboratory results) along with an estimated glomerular filtration rate (eGFR) below 60mL/min/1,73m^2. A prostate-specific antigen level of 27,6ng/mL was found at admission, which led to suspicion of a prostate cancer and possible secondary MN. Prostate

magnetic resonance imaging was performed and identified a mass, which was later biopsied, confirming Gleason 7 (3+4) prostate adenocarcinoma. A kidney biopsy was performed, confirming the diagnosis of MN. Light microscopy showed glomerular wall thickening and podocyte hypertrophy. Immunofluorescence revealed granular parietal deposits of Immunoglobulin (Ig) G+++, C3+++, kappa+++ and lambda+++ , as well as segmental IgM deposits. Nevertheless, serum anti-PLA2R antibody test was also done, whose positive result, after discharge, established the final diagnosis of primary MN on a patient with a concurrent prostate malignancy. Supportive therapy was started including losartan at the maximum tolerated dose, diuretics and a statin. However, 2 months later the patient was readmitted due to worsening edema (a weight loss of 9,4kg during hospitalization was later reported at discharge, 13,6% of his total weight), hypoalbuminemia of 2,0g/dL, sustained high anti-PLA2R levels, decreasing eGFR and a stabilized proteinuria of 3g/day (table 1). As the patient didn´t achieve complete MN remission and had active malignancy, a decision was made to start immunosuppressive therapy with intravenous rituximab (1000mg on days 1 and 15) to avoid the adverse effects of other immunosuppressive regimens. 2 months after the last rituximab administration, and despite not having started any cancer treatment, peripheral edema has improved, albuminemia has increased by 25%, urinary protein-to-creatinine ratio had decreased by 43% and eGFR had improved by 13% (table 1).

Discussion and learning point | The etiological diagnosis of MN remains challenging, requiring the exclusion of secondary causes but also the screening for primary MN even when a secondary etiology seems likely. Despite not being always required for the diagnosis of primary MN in patients with NS and a positive test for anti-PLA2R antibody, a kidney biopsy might be helpful in patients with decreasing eGFR to better clarify the extent and cause of kidney injury. In this patient with an active malignancy, rituximab presented as a safer alternative to alkylating agents, illustrating how newer immunosuppressive options may allow for tailored therapies that combine desired clinical outcomes with the safest possible adverse reaction profile. Even with an eGFR close to 30mL/min/1,73m2, positive results were obtained with rituximab, especially in albuminemia, proteinuria, eGFR and clinical signs of NS.

PO 67
FULL-HOUSE MEMBRANOUS NEPHROPATHY WITHOUT SLE: LONG-TERM FOLLOW-UP OF FOUR CASES WITH PLA2R TISSUE POSITIVITY

Beatriz Barros; Beatriz Silva; Beatriz Gil de Oliveira Braga; Andreia Campos; Pedro R Pereira; Sofia Oliveira Correia; Josefina Santos

Unidade Local de Saúde de Santo António

Category: GLOMERULAR DISEASES

Background

Membranous nephropathy (MN) with full-house immunofluorescence (IF) is classically associated with lupus nephritis (LN) class V. This finding often triggers investigation and treatment for systemic lupus erythematosus (SLE). However, this pattern may occur in patients without clinical or serological evidence of SLE, creating a diagnostic and therapeutic dilemma.

Case description

We report four cases of biopsy-proven MN with full-house IF. Two patients presented with nephrotic syndrome, while two were asymptomatic with subnephrotic proteinuria. Kidney function was preserved overall, except for one patient. Microhematuria was present in two cases. Complement levels were normal in two patients, with isolated C4 reduction in one and combined C3/C4 reduction in one case. Anti–double-stranded deoxyribonucleic acid antibodies were negative in all patients, and antinuclear antibody was negative in three. None fulfilled 2019 EULAR/ACR classification criteria for SLE at diagnosis.

Kidney biopsy showed a predominantly non-proliferative pattern without crescents. IF demonstrated IgG-dominant full-house staining, with IgG4 positivity in 2 of 3 evaluated cases. Phospholipase A2 receptor (PLA2R) staining was positive in all evaluated patients (3/3), while circulating antibodies were detected in only 1 of 3 evaluated patients. Secondary causes of MN were excluded; THSD7A and other antigens were not assessed.

Treatment strategies varied (supportive therapy, corticosteroids, rituximab, or calcineurin inhibitors). Of the two patients with nephrotic-range proteinuria at baseline, Case 1 achieved partial remission and Case 2 achieved complete remission; Case 3 maintained subnephrotic proteinuria; Case 4 showed progression of proteinuria during the index episode. Two relapses were documented (Cases 1 and 4). No patient developed clinical or serological features of SLE during follow-up (8-181 months). The main clinicopathological features and outcomes are summarized in Table 1.

Discussion and learning points

In MN with full-house IF but no evidence of SLE, reliance on IF alone may lead to misclassification as LN, which can influence treatment decisions. The combination of negative SLE serology, absence of systemic manifestations, predominantly non-proliferative histology, and PLA2R tissue expression is consistent with PLA2R-associated MN.

The coexistence of PLA2R positivity and full-house staining suggests that this pattern is not specific for SLE. Recent studies have identified additional target antigens associated with MN, including EXT1/EXT2, NELL-1, and semaphorin-3B. In particular, EXT1/EXT2-associated MN has been linked to lupus-related MN, even in seronegative cases. Mass spectrometry-based approaches may help define the underlying antigenic profile in these patients.

These cases highlight that full-house IF is not diagnostic of LN and requires clinical and serological correlation. PLA2R tissue positivity may support classification closer to primary MN, even in serum-negative cases. Long-term follow-up remains essential, particularly in young patients with hypocomplementemia at presentation.

Table 1. Case Characteristics and Outcomes

	CASE 1	CASE 2	CASE 3	CASE 4
CLINICAL PRESENTATION				
Age at onset (years)/Sex	68/F	22/M	25/M	5/F
Symptoms	Peripheral edema	Peripheral edema	Asymptomatic	Asymptomatic
Proteinuria (UPCR, g/g creatinine)	9.01	4.93	1.20	0.54
Microhematuria	No	Yes	Yes	No
Serum creatinine (mg/dL)	2.18	0.58	0.92	0.41
IMMUNOLOGY AT PRESENTATION				
ANA (titer)/anti-dsDNA	1:160; Negative	Negative / Negative	Negative / Negative	Negative / Negative
C3/C4	Normal / Normal	Normal / Low	Normal / Normal	Low / Low
PLA2R (serum)	Positive	Negative	Missing	Negative
BIOPSY				
Hypercellularity	No	Mesangial	Missing	Mesangial
Crescents	No	No	No	No
C1q	No	Yes	Yes	Yes
PLA2R (tissue)	Yes	Yes	Missing	Yes
IgG4 (tissue)	Yes	Missing	Yes	No
TREATMENT				
Immunosuppression	Rituximab (2023 and 2025)	Cyclosporine + Glucocorticoids (2026)	None	Glucocorticoids (2011); Cyclosporine + Glucocorticoids (2024)
FOLLOW-UP				
Follow-up time (months)	42	8	175	181
Relapse	Yes (2025)	No	No	Yes (2024)
Creatinine (mg/dL) at last follow-up	1.90	0.61	0.84	0.51
Proteinuria (UPCR, g/g creatinine) at last follow-up	0.86	0.20	0.63	1.78
PLA2R serum antibodies at last follow-up	Negative	Negative	Negative	Negative
C3/C4 at last follow-up	Normal / Normal	Normal / Normal	Normal / Normal	Normal / Normal

Abbreviations: F, female; M, male; UPCR, urine protein-to-creatinine ratio; ANA, antinuclear antibody; anti-dsDNA, anti-double stranded deoxyribonucleic acid; C3/C4, complement components 3 and 4; PLA2R, phospholipase A2 receptor; IgG4, immunoglobulin G4; SLE, systemic lupus erythematosus.

PO 69
Blisters and acute renal failure: unraveling the connection

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Keywords, Acute interstitial nephritis, Erythema multiforme, Sodium-glucose cotransporter 2 inhibitors

Introduction

Acute interstitial nephritis (AIN) is characterized by an inflammatory infiltrate affecting the tubules and interstitium. The classic clinical triad (fever, eosinophilia, and rash) occurs in only 10% of cases. Approximately 70-75% of cases are drug-induced, although associations with autoimmune diseases, neoplasms, and infections are also described.

Case Description

An 81-year-old female with a history of obesity, hypertensive heart disease, and dyslipidemia was recently diagnosed with diabetes mellitus. Ten days prior to admission, she started empagliflozin + metformin 5+1000mg and pregabalin 150mg. She presented to the Emergency Department with painful bullous eruptions on the inner thighs (4-day duration) (Figure 1). On admission, she was normotensive and oliguric. Laboratory tests revealed severe acute kidney injury (AKI) with serum creatinine (sCr) = 7.43 (baseline 0.85mg/dL), elevated creatine kinase (CK) = 4793 U/L, and eosinophilia (600/µL), without leukocytosis or C-reactive protein (CRP) elevation. Urinalysis showed hematoproteinuria and leukocyturia, with a urinary protein-to-creatinine ratio (uPCR) of 838mg/g and urinary albumin-to-creatinine ratio (uACR) 307 mg/g, suggesting non-albuminuric proteinuria. Work up for monoclonal gammopathies and viral serologies (including herpes simplex virus (HSV)) was negative. However, immunofluorescence was positive for perinuclear anti-neutrophil cytoplasmic antibodies (p-ANCA), raising suspicion of ANCA-associated vasculitis. She received methylprednisolone pulses and two sessions of plasmapheresis; however, subsequent enzyme-linked immunosorbent assay (ELISA) for ANCA was negative, making this diagnosis less likely.

Due to the progression of bullous lesions to the wrists and oral mucosa (Figure 1), and negative autoantibody testing (anti-desmoglein 1/3 and anti-bullous pemphigoid (BP) 180/230), a skin biopsy was performed. Histopathology revealed hydropic degeneration and colloid bodies in the basal layer of the epidermis with extensive necrosis and lymphocytic perivascular inflammation, consistent with erythema multiforme (EM) or Stevens-Johnson syndrome (SJS).

Simultaneously, a renal biopsy showed AIN with severe acute tubular injury and necrosis (ATN) affecting <5% of tubules (Figure 2). Minimal chronic changes were noted.

Discussion and Conclusion

The concurrence of severe AKI and bullous dermatological manifestations is a diagnostic challenge requiring the exclusion of systemic vasculitis and autoimmune diseases. The diagnosis of drug-induced AIN and EM was established by temporal and histopathological correlation. Both pregabalin and empagliflozin are described, albeit rarely, as triggers for AIN, highlighting the need for a high clinical suspicion with new medications.

The rhabdomyolysis increased AKI severity through ATN mediated by myoglobinuria.

Immediate drug discontinuation and systemic corticosteroid therapy led to progressive recovery of renal function without the need for renal replacement therapy.

This case emphasizes that the kidneys and skin may serve as simultaneous targets of drug hypersensitivity, requiring an early multidisciplinary approach to optimize outcomes.

PO 70
Coexistence of IgA Nephropathy and AA Amyloidosis in a patient with Crohn's disease

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ULS Amadora-Sintra

Keywords: IgA nephropathy, AA amyloidosis, Crohn disease

Introduction

Crohn disease (CD) is an inflammatory bowel disease characterized by transmural inflammation. Renal involvement occurs in 6% to 23% of patients. The diagnosis remains challenging, requiring

differentiation between secondary manifestations, systemic inflammation-related injury or therapy-induced nephrotoxicity.

Case Description

A 42-year-old male with a history of CD (diagnosed at age 14, status post-ileocecal resection) was referred to Nephrology. He had been on weekly adalimumab since January 2024, alongside mesalazine (5-aminosalicylic acid [5-ASA]), with clinical remission since May 2024.

Renal function declined between May and November 2025, with serum creatinine (sCr) rising from 1.12 to 1.69 mg/dL. Urinalysis showed hematoproteinuria with a urinary protein-to-creatinine ratio (uPCR) of 598.7 mg/g, and urinary sediment showed dysmorphic erythrocytes.

Workup revealed elevated immunoglobulin A (IgA) (761 mg/dL), while complement, antinuclear antibodies (ANA), anti-neutrophil cytoplasmic antibodies (ANCA), and anti-glomerular basement membrane (anti-GBM) antibodies were negative. Viral serologies and monoclonal gammopathy screening were also negative.

Renal biopsy was perfomed with two primary findings:

1. IgA nephropathy (IgAN) – moderate mesangial expansion and proliferation with mild chronic damage. Oxford M1 E1 S1 T0 C1 criteria. [Figure 1a]. Immunofluorescence showed deposits of IgA (4+), C3 (3+), with negative results for other immunoglobulins and complements [Figure 1b].
2. AA amyloidosis – Congo red-positive deposits in the glomeruli and peritubular interstitium [Figure 2].

Discussion

Initial differential diagnoses included 5-ASA-induced acute or chronic interstitial nephritis, a common complication in CD; however, this was histologically excluded. Adalimumab-induced glomerulonephritis was also considered, as anti-tumor necrosis factor (TNF) agents can induce immune-complex deposition.

The renal biopsy revealed IgAN with aggressive histological features, likely secondary to CD activity. Additionally, the presence of AA amyloidosis serves as a definitive biomarker of sustained, subclinical systemic inflammation.

Following a multidisciplinary discussion, CD restaging will be performed to investigate inflammatory activity. Given the renal progression while under TNF-alpha inhibition, the therapeutic strategy was switched to ustekinumab (an interleukin (IL) 12 and 23 agent). This choice also allows for the exclusion of an adalimumab-associated reaction.

Although IL-6 inhibitors (such as tocilizumab) show high efficacy in treating AA amyloidosis, they are not currently indicated for CD. Additionally, nephroprotective therapy was optimized. Due to the presence of histological markers of activity (E1 and C1), targeted-release budesonide was initiated to reduce the mucosal IgA load and prevent irreversible fibrosis.

Conclusion

This case highlights that clinical remission in Crohn disease does not guarantee biological quiescence. Renal biopsy is essential to distinguish between drug-induced injury and systemic manifestations, allowing for targeted therapeutic adjustments to preserve renal function.

PO 72

MONOCLONAL GAMMOPATHY-ASSOCIATED C3 GLOMERULOPATHY PRESENTING AS MEMBRANOPROLIFERATIVE GLOMERULONEPHRITIS: A DIAGNOSTIC CHALLENGE

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Unidade Local de Saúde do Algarve

Category | Background | Case description | Discussion and learning point

Background: C3 Glomerulopathy is a rare complement-mediated glomerular disease caused by dysregulation of the alternative complement pathway. Monoclonal gammopathies may promote complement activation, leading to secondary C3 glomerulopathy even without overt hematologic malignancy. Recognition of this association is clinically relevant, as clone-directed therapy may improve renal outcomes.

Case Description: A 60-year-old man with chronic kidney disease, ischemic heart disease, dyslipidemia, asthma and psoriasis was referred to nephrology due to worsening kidney function. Regular medication included ramipril, atorvastatin and acetylsalicylic acid. Laboratory evaluation revealed serum creatinine of 1.37 mg/dL, estimated glomerular filtration rate of 56 mL/min/1.73m², urine albumin-to-creatinine ratio of 2835 mg/g, subnephrotic proteinuria (3.1 g/24h), microscopic hematuria, iron-deficiency anemia (hemoglobin 8.9 g/dL) and isolated C3 consumption. Remaining autoimmune study was negative. Serum protein electrophoresis identified a monoclonal gamma peak of 1.46g/dL. Kidney ultrasound showed mildly reduced corticomedullary differentiation. The patient reported nonspecific fatigue, without edema or other systemic manifestations. Due to suspected monoclonal gammopathy of renal significance (MGRS), further investigation was performed. Bone marrow aspirate revealed 2.3% plasma cells, while bone biopsy, karyotype and fluorescence in situ hybridization studies showed no evidence of hematologic malignancy. Kidney biopsy identified 23 glomeruli, 6 globally sclerotic, while the remaining showed a membranoproliferative pattern with endocapillary proliferation (Figure 1), including 1 fibrocellular crescent with Bowman capsule rupture. Interstitial inflammatory infiltrate and erythrocyte casts were also observed. Immunofluorescence revealed dominant C3 deposits (Figure 2), and pronase digestion excluded immunoglobulin and light-chain deposition, establishing the diagnosis of C3 Glomerulopathy associated with monoclonal gammopathy. Genetic testing for complement pathway abnormalities showed no alterations. The patient is currently awaiting hematology evaluation for initiation of clone-directed therapy.

Discussion and Learning Points: This case illustrates the association between monoclonal gammopathies and alternative complement pathway dysregulation leading to C3 glomerulopathy. The absence of overt hematologic malignancy may delay recognition of MGRS, particularly when renal manifestations precede clinically significant hematologic disease. The coexistence of isolated C3 consumption, membranoproliferative histologic pattern, and monoclonal gammopathy should raise suspicion for monoclonal gammopathy-associated C3 glomerulopathy, even when bone marrow findings are subtle. Kidney biopsy plays a central diagnostic role. Recent evidence supports clone-directed therapies, including cyclophosphamide-bortezomib-dexamethasone and daratumumab-based regimens, aiming to suppress the pathogenic clone responsible for complement dysregulation and improve renal outcomes.

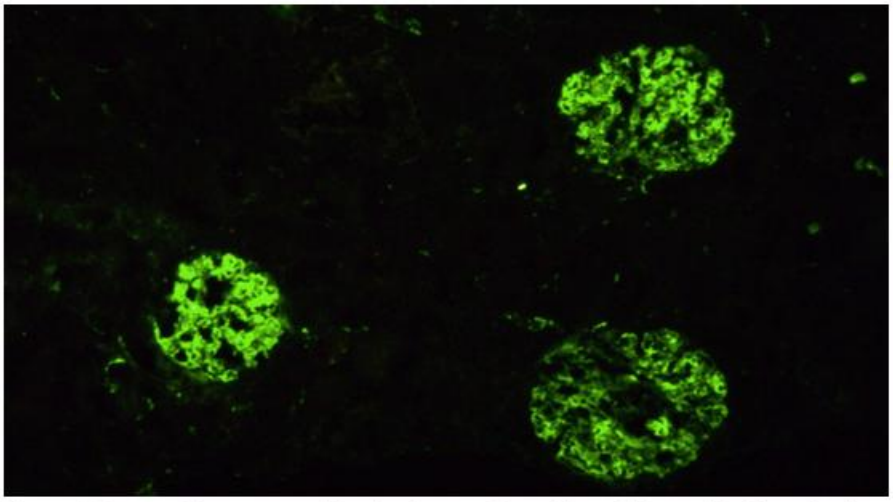

Figure 2: Immunofluorescence showing intense C3 staining with granular deposits along the glomerular basement membrane and mesangium.

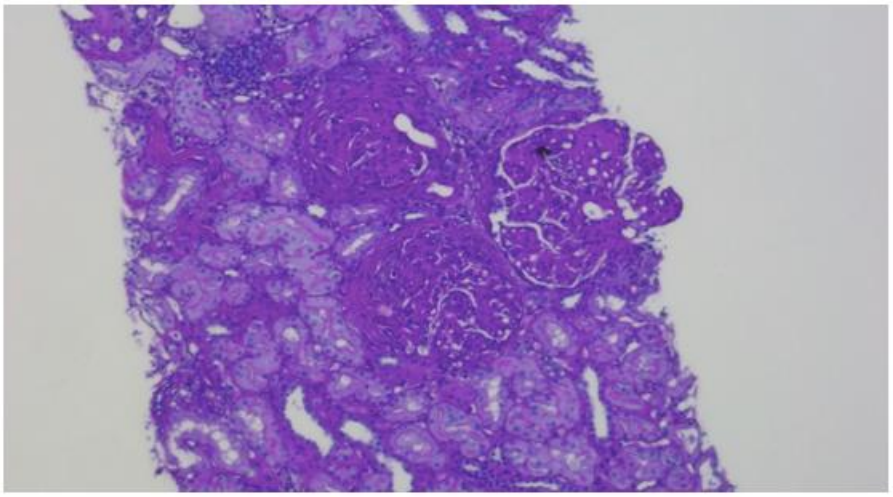

Figure 1: Periodic acid-Schiff stain showing lobulated glomeruli with mesangial expansion, endocapillary proliferation, and double contours of the glomerular basement membrane, consistent with membranoproliferative pattern.

PO 74
AN UNUSUAL CAUSE OF NEPHROTIC-RANGE PROTEINURIA AFTER SEPTIC SHOCK

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Unidade Local de Saúde de São José

Background: AA amyloidosis is a recognized complication of chronic inflammatory diseases, typically presenting as a slowly progressive proteinuric kidney disease. However, acute and massive onset of nephrotic syndrome following a severe inflammatory insult is rarely described. Sustained elevation of serum amyloid A (SAA) during intense systemic inflammation may accelerate amyloid deposition, leading to abrupt and atypical clinical presentations.

Case description: A 70-year-old woman diagnosed with mixed connective tissue disease for over 20 years, previously treated with methotrexate and subsequently on prednisolone and mycophenolate mofetil, with normal baseline kidney function (creatinine 0.76 mg/dL) and no prior significant proteinuria, was admitted into the intensive care unit with invasive pneumococcal disease complicated by septic shock and multiorgan failure.

Her clinical course was marked by respiratory failure requiring non-invasive ventilation and severe anuric acute kidney injury requiring continuous kidney replacement therapy, later transitioned to intermittent hemodialysis. During recovery, after partial tapering of immunosuppression, she developed massive nephrotic-range proteinuria, with an initial protein-to-creatinine ratio (PCR) of 73,390 mg/g, decreasing to 31,217 mg/g on repeat testing, and later quantified as 10 g/24h. This was accompanied by hypoalbuminemia and persistent kidney dysfunction despite recovery of urine output.

Urinalysis revealed adjacent hematuria and leukocyturia, raising suspicion of an autoimmune disease flare. Based on this hypothesis, immunosuppressive therapy was intensified with corticosteroid pulses and increased mycophenolate dosing, without clinical or renal improvement.

Given the atypical clinical course and lack of response to therapy, a kidney biopsy was performed, unexpectedly revealing AA amyloidosis (figure 1), supported by elevated SAA levels (58 mg/L). Despite persistent dialysis dependency, the patient remained clinically stable and was discharged on a maintenance hemodialysis program.

Discussion and learning points: This case highlights an unusual presentation of AA amyloidosis, characterized by the abrupt onset of massive proteinuria following a severe inflammatory event. While AA amyloidosis is typically a chronic and progressive condition, this presentation raises the possibility of an accelerated amyloidogenic process triggered by intense systemic inflammation.

The concept of an “amyloid storm”, analogous to other dysregulated inflammatory responses, may help conceptualize this rapid clinical evolution, although it remains poorly defined.

Clinicians should consider underlying amyloidosis in patients with disproportionate proteinuria emerging after critical illness, particularly in the context of chronic inflammatory disease.

Importantly, maintaining a high index of suspicion can prevent unnecessary or inappropriate therapeutic escalation. In the present case, identification of the underlying process avoided further intensification of immunosuppression, underscoring the importance of recognizing atypical presentations of AA amyloidosis in guiding therapeutic management.

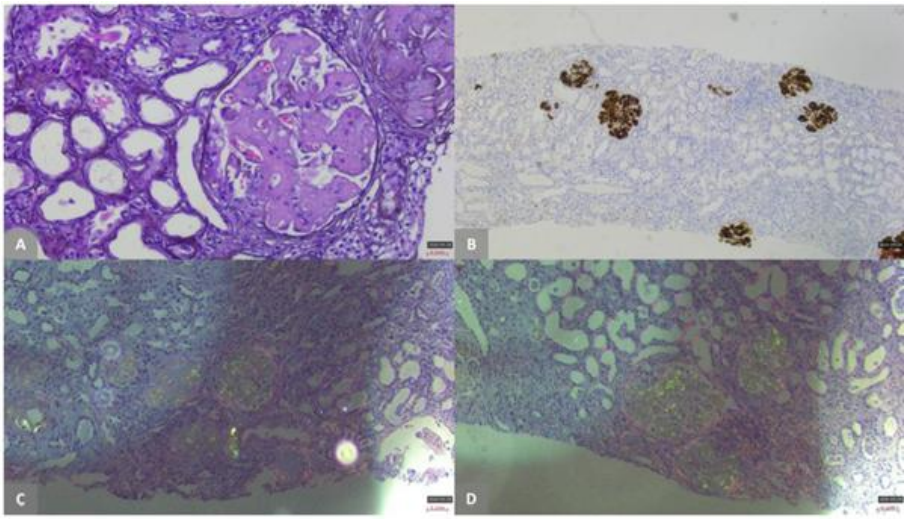

Figure 1. Histological images of kidney biopsy showing amyloid A deposition. (A) PAS stain showing glomerular deposition of PAS-negative amorphous material. (B) Positive immunohistochemical staining for amyloid A. (C/D) Congo red stain viewed under polarized light showing “apple green” birefringence.

PO 75
THE CLUE WAS IN THE URINE: DELAYED DIAGNOSIS OF ETHYLENE GLYCOL POISONING

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Unidade Local de Saúde de São José

Background: High anion gap metabolic acidosis is a life-threatening condition, commonly attributed to lactic acidosis in critically ill patients. However, toxic alcohol ingestion remains an often underrecognized cause. Ethylene glycol is metabolized into glycolic and oxalic acids, leading to severe metabolic acidosis and characteristic calcium oxalate crystal deposition, which can result in acute kidney injury. Initial toxicology screening may not cover ethylene glycol intoxication and clinical presentation may be nonspecific, yet timely recognition is crucial to prevent irreversible organ damage.

Case description: A 69-year-old man with a history of alcohol use disorder, hypertension and heart failure was found unconscious and brought to the emergency department. On admission, he was normotensive but presented with altered mental status and severe metabolic acidosis (pH 6.77, pCO2 18 mmHg, HCO3? 2.6 mmol/L, anion gap 27,4), with lactate level above measurable range (0,1 to 31mmol/L) and mild hyperkalemia (5.3 mmol/L), despite preserved kidney function (creatinine 1 mg/dL).

Initial investigation, including toxicology screening and computed tomography, revealed no clear etiology, and lactic acidosis was presumed as the likely cause. Supportive therapy with fluids and bicarbonate was initiated, and the patient was admitted to the intensive care unit where he was started on continuous kidney replacement therapy (CKRT), resulting in rapid correction of acidemia.

Following discontinuation of CKRT, the patient remained anuric, with progressive kidney dysfunction (creatinine rising from 1 to 5.6 mg/dL); renal ultrasound showed preserved parenchymal thickness but reduced corticomedullary differentiation. After transfer to the nephrology department, manual microscopy of urinary sediment revealed abundant calcium oxalate crystals with distinctive needle-like morphology, raising suspicion for ethylene glycol intoxication (figure 1). Kidney biopsy confirmed extensive intratubular calcium oxalate crystal deposition, associated with acute tubular injury (figure 2).

The patient required intermittent hemodialysis for a month, with subsequent partial kidney function recovery and discharge with a serum creatinine of 2.2 mg/dL. For the following months, he continued to improve until baseline-values (creatinine 1.0 mg/dL).

Discussion and learning points: This case illustrates a delayed diagnosis of ethylene glycol intoxication, initially misinterpreted as severe lactic acidosis. The markedly elevated lactate levels, coupled with nonspecific clinical findings and negative toxicology screening, contributed to this delayed recognition.

Urinary sediment examination was a pivotal diagnostic tool, revealing characteristic calcium oxalate crystals and prompting diagnostic reconsideration, confirmed histologically. This highlights the enduring value of urine microscopy as a low-cost diagnostic technique that may be key in accurate diagnosis.

Notably, despite severe initial presentation and biopsy-proven tubular injury, the patient achieved significant renal recovery, emphasizing that recognition, even if delayed, may still allow for functional improvement.

Clinicians should maintain a high index of suspicion for toxic alcohol ingestion in cases of unexplained severe metabolic acidosis; prompt intervention may prevent irreversible kidney damage and improve outcomes.

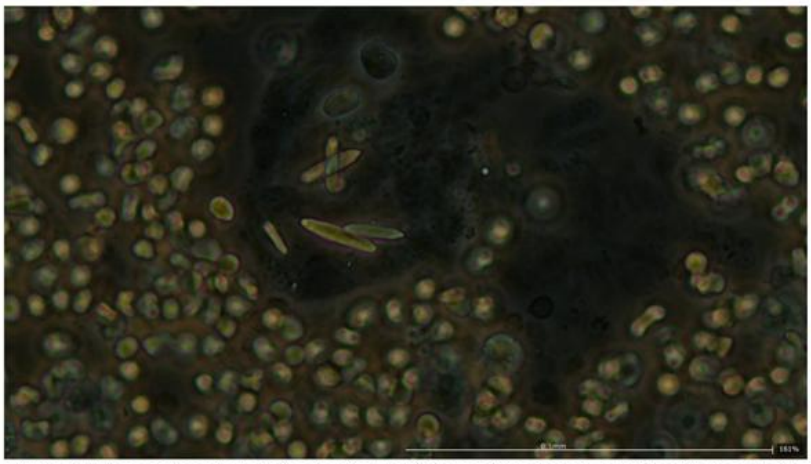

Figure 1. Urine sediment analysis showing multiple calcium oxalate crystals with needle-like morphology.

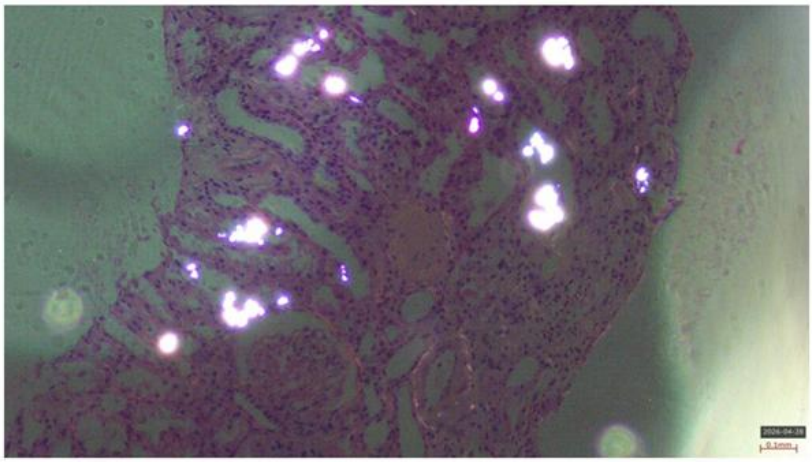

Figure 2. Extensive intratubular calcium oxalate crystal deposition viewed under polarized light.

PO 76
A DIAGNOSIS REVEALED IN THE KIDNEY TRANSPLANT: EARLY RECURRENCE OF C3 GLOMERULOPATHY

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Category: Glomerular diseases

Key words: C3 glomerulopathy, kidney transplantation, recurrence

Background:

C3 glomerulopathy is a rare complement-mediated disease caused by dysregulation of the alternative pathway (AP), leading to glomerular C3 deposition and progressive kidney damage. It is associated with a high recurrence rate after kidney transplantation (KT) and poor graft survival. Diagnosis may be delayed, particularly when the disease has not been confirmed by biopsy.

Case description:

A 61-year-old male with chronic kidney disease of unknown etiology - characterized by long-standing haematuria and proteinuria, including episodes of synpharyngitic macroscopic haematuria -, progressed to kidney failure requiring hemodialysis in 2021. Although IgA nephropathy was suspected, it was never biopsy-confirmed.

He underwent deceased-donor KT in December 2024. The early post-transplant course was complicated by delayed graft function attributed to acute tubular necrosis, tacrolimus toxicity, partial renal vein thrombosis, and pyelonephritis. At discharge, serum creatinine was 3.04 mg/dL, subsequently improving to a nadir of 1.3mg/dL.

During follow-up, new onset erythrocyturia and progressively increasing proteinuria were observed despite stable immunosuppression, negative immunological, including complement and virological workups and normal serum and urinary immunoelectrophoresis. A graft biopsy performed in October 2025 and revealed features consistent with C3 glomerulopathy (Figure 1, Figure 2). Genetic variants and autoantibodies linked to AP hyperactivity were negative.

The patient subsequently developed nephrotic syndrome and progressive graft dysfunction, requiring repeated hospitalizations early 2026, ultimately lead into graft failure and reinitiation of dialysis. Although treatment with iptacopan, a factor B inhibitor was requested, it could not be initiated in time.

Discussion and learning point:

This case highlights the importance of considering C3 glomerulopathy in patients with unexplained chronic kidney disease prior to transplantation, given its high recurrence rates - reported in up to two-thirds of cases - and the associated risk of significant graft loss.

Early post-transplant recurrence should be suspected in patients presenting with new-onset proteinuria and graft dysfunction. Advances in the understanding of complement-mediated pathophysiology have led to the development of targeted therapies, which may play a role in modifying disease progression.

Timely recognition, prompt diagnosis and early initiation of targeted therapy are critical to improving graft outcomes, and long-term prognosis in patients with C3 glomerulopathy.

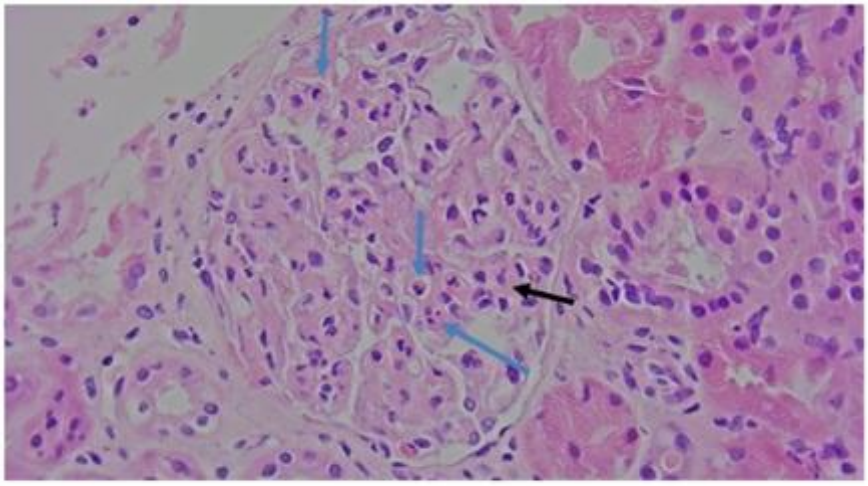

Figure 1: Kidney graft biopsy showing mesangial expansion and proliferation (black arrow) and endocapillary proliferation (blue arrows) (H&E, 400x).

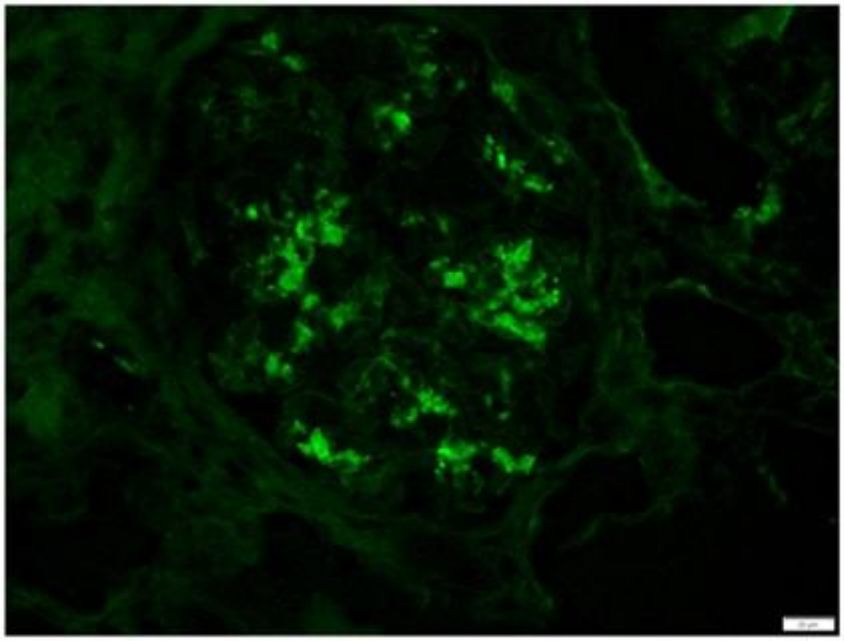

Figure 2: Immunofluorescence showing mesangial C3 stanning.

PO 77
PRIMARY MEMBRANOUS NEPHROPATHY WITH PROGRESSIVE LOSS OF THERAPEUTIC RESPONSE

Margarida Patrocínio Gomes; Leonor Marques Gomes; Rita Theias; André Weigert

ULS Lisboa Ocidental

Category : Glomerular diseases

Background

Primary membranous nephropathy (MN) is the leading cause of nephrotic syndrome in non-diabetic adults. Rituximab is the recommended first-line therapy per Kidney Disease: Improving Global Outcomes (KDIGO) 2021 guidelines, yet up to 30% of patients fail to achieve remission.

Obinutuzumab, a glycoengineered type II anti-CD20 monoclonal antibody with enhanced B-cell cytotoxicity and superior tissue penetration, has emerged as a rescue option in rituximab-resistant MN.

Case description

A 53-year-old male with biopsy-proven anti-phospholipase A2 receptor (anti-PLA2R)-positive primary MN, followed since 2003. A Ponticelli-based regimen was interrupted after one month due to pulmonary toxicity (hemoptysis). Over two decades, he received cyclosporin for more than 14 years and five rituximab courses, achieving documented clinical remissions (complete or partial) with each agent separately at different periods. However, he progressively lost therapeutic response: despite persistent peripheral B-cell depletion (CD19 <5 cells/microL) with successive rituximab courses, anti-PLA2R never reached immunological remission (<2 RU/mL) and proteinuria remained in the nephrotic range.

After the fifth rituximab course (September 2024) and cyclosporin discontinuation, anti-PLA2R declined from 22.8 to 8.1 RU/mL without reaching immunological remission. Obinutuzumab 1000 mg was administered in May 2025 (100 mg day 1 + 900 mg day 2), achieving for the first time documented immunological remission: anti-PLA2R fell to 1.4 RU/mL (August 2025) with sustained CD19 depletion. However, proteinuria persisted at approximately 5000 mg/24h, prompting a second renal biopsy (January 2026).

Histology revealed: membranous nephropathy stage 2-3/4 with IgG4-dominant deposits and strong C4d positivity, confirming primary aetiology; secondary focal segmental glomerulosclerosis (FSGS), likely attributable to prolonged calcineurin inhibitor exposure and metabolic comorbidities; mild-to-moderate acute tubular injury; and moderate chronicity (glomerulosclerosis 29.4%, interstitial fibrosis and tubular atrophy 15-20%).

A second obinutuzumab course was administered in March 2026, combined with low-dose tacrolimus. At last follow-up (May 2026), anti-PLA2R was 2.2 RU/mL (borderline immunological remission), serum albumin had improved to 3.7 g/dL, and proteinuria was 6908 mg/24h.

Discussion and learning point

In previously responsive patients who become refractory to rituximab, true resistance should be defined by persistent anti-PLA2R despite complete peripheral B-cell depletion, not by proteinuria alone.

Obinutuzumab achieved immunological remission where five rituximab courses failed, likely through superior depletion of tissue-resident autoreactive B-cell clones.

Persistent proteinuria despite immunological remission warrants repeat biopsy: secondary FSGS, an immunotherapy-resistant structural lesion accumulating over decades of disease and treatment, was identified as an independent contributor.

Immunological remission precedes clinical remission by 12-24 months; in the presence of established structural damage, proteinuria persistence should not be interpreted as treatment failure.

PO 78
SYPHILIS-ASSOCIATED MEMBRANOUS NEPHROPATHY WITH PLA2R TISSUE POSITIVITY: A CASE REPORT

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| Category|: Glomerular Diseases

|Background|

Syphilis incidence has resurged across Europe since 2015, and renal involvement, while uncommon, is increasingly recognized. Membranous nephropathy (MN) is the most frequent glomerular manifestation of syphilis, mediated by subepithelial immune complex deposition with podocyte and glomerular basement membrane injury. The diagnosis is frequently delayed or missed, as nephrotic presentation may precede or overshadow the classic features of syphilis. Human immunodeficiency virus (HIV) coinfection, which is common in patients with syphilis-related kidney disease, further complicates the diagnostic workup. We report a case of secondary syphilis-associated MN in an HIV-positive patient with a characteristic immunopathological profile and rapid remission following antibiotic therapy alone, and provide a review of the existing literature.

|Case description|

A 52-year-old man with HIV with suppressed viral-load presented with asthenia, weight loss, painful bilateral inguinal lymphadenopathy, and a maculopapular rash involving the trunk, limbs, and soles. Venereal diseases research laboratory (VDRL) testing confirmed secondary syphilis, for which he was treated with penicillin. One week later, he returned with generalized edema, dyspnea, and orthopnea. Renal function was preserved, but evaluation revealed nephrotic-range proteinuria and hypoalbuminemia. Serum levels of C3 and C4 were normal and antinuclear antibodies, and circulating anti-phospholipase A2 receptor (PLA2R) were negative. Urinalysis was unremarkable except for granular casts. Renal biopsy demonstrated diffuse thickening of the glomerular basement membrane (GBM) and spike-like subepithelial projections on light microscopy, with diffuse granular Immunoglobulin (Ig) G and C3 staining on immunofluorescence, compatible with the diagnosis of MN. Immunohistochemistry was positive for PLA2R antigen and IgG, but negative for IgG4 (Figure1). Partial remission occurred one week after symptom onset, two weeks after antibiotic therapy, without immunosuppression.

|Discussion and learning point|

This case illustrates key diagnostic pitfalls in syphilis-associated MN. Tissue PLA2R antigen positivity with absent circulating anti-PLA2R antibodies and IgG4 negativity suggests secondary, rather than primary MN or HIV-associated immune complex disease. Immunohistochemistry for neuron-derived neurotrophic factor (NDNF), a recently described target antigen in syphilis-associated MN, was not performed; retrospective testing of the archived biopsy material would be of interest to confirm the antigen profile. The rapid remission observed following penicillin therapy reinforces the causal link and emphasizes the importance of serological syphilis screening in all patients presenting with nephrotic syndrome, particularly amid rising incidence and in HIV-positive individuals.

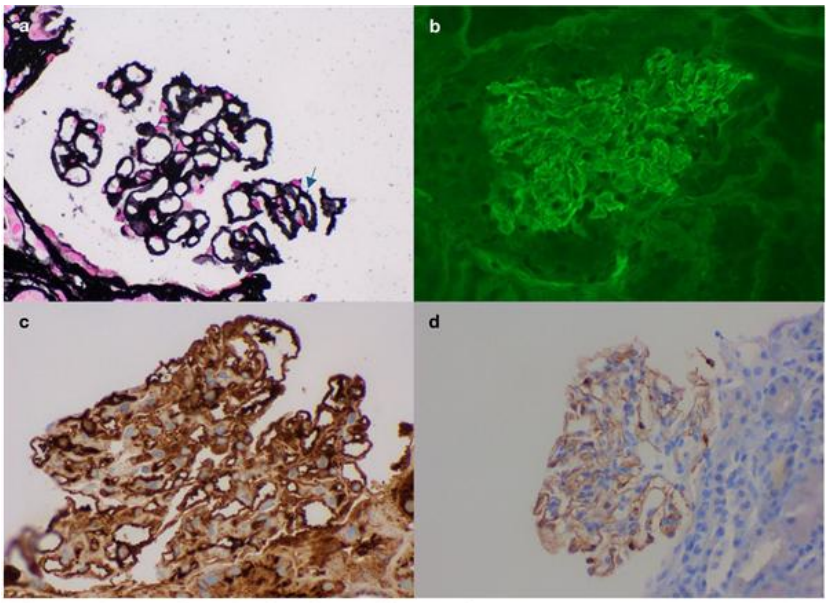

Figure 1. Renal biopsy findings in syphilis-associated membranous nephropathy. (a) Jones silver stain exhibiting diffuse glomerular basement membrane thickening with subepithelial spike formation (arrow; original magnification x600). (b) Immunofluorescence for IgG demonstrating diffuse granular deposits along the glomerular capillary walls (original magnification x600). (c) Immunohistochemistry for IgG showing subepithelial immune complex deposition (original magnification x600). (d) Immunohistochemistry for PLA2R antigen revealing granular positivity along the capillary loops (original magnification x400). PLA2R1, anti-phospholipase A2 receptor.

PO 79
PAROXYSMAL NOCTURNAL HEMOGLOBINURIA AND C3 GLOMERULOPATHY: THE SAME COMPLEMENT DYSREGULATION OR A SIMPLE COINCIDENCE

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Unidade Local de Saúde de Santo António

Background

Paroxysmal Nocturnal Hemoglobinuria (PNH) and C3 glomerulopathy (C3G) are distinct diseases and although complement overactivation is common to both, fluid phase alternative pathway dysregulation (C3G), or terminal pathway dysregulation (PNH) predominates resulting in the very different phenotypes seen in these diseases.

Case description

A 42-years-old male, with known PNH, under Crovalimab (anti-C5 recycling monoclonal antibody) was admitted with severe acute kidney injury, with serum creatinine of 4.53mg/dL and no previously known kidney disease. He had a recent history of presumed gastroenteritis and a hospital admission with parvovirus infection three months prior. There was an aggravation of his chronic pancytopenia – 2.44x10^3/µL leucocytes, hemoglobin of 6.3mg/dL and 73x10^3/µL platelets – and, during hospitalization, the patient required multiple red blood cells transfusions due to ongoing hemolysis. He had macroscopic hematuria, with erythrocytes seen on urinary sediment, non-fully albuminuric proteinuria, with a protein creatinine ratio of 1.6 g/g creatinine. Kidney ultrasound showed normal sized kidneys, with marked echogenicity of the renal parenchyma, with reduced parenchymal-sinus differentiation. There was C3 consumption in the first immunological panel, not seen in the subsequent ones. Diuresis was kept over 2000ml a day,

but the patient continued to experience worsening renal function. The kidney biopsy showed hemosiderin deposits in tubular cells, interstitium with foci of polymorphic inflammatory infiltrate, acute tubular necrosis (ATN), and mesangial proliferative pattern. Immunofluorescence (IF) revealed only mesangial deposits of C3 (+++) (figure 1). The patient was started on corticosteroid therapy and back on crovalimab, hemolysis was controlled and kidney function began slowly improving, with a creatinine of 1.03mg/dL in the last follow up. Complement genetic panel did not show any mutations.

Discussion and learning point

The changes found in the IF were not expected, which raises the possibility of some complement associated mutation or, perhaps, a new association of PNH and C3G. However, this entity is not fully supported by the morphological changes in the kidney biopsy and it usually does not manifest as acute kidney injury. Clinical and pathological integration is essential, as it is clinical surveillance.

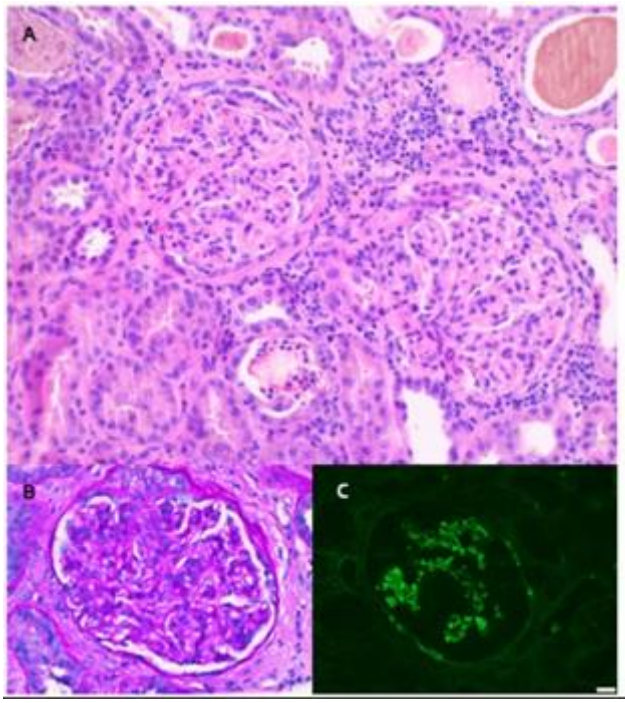


Figure 1: A – Glomeruli with predominantly mesangioproliferative aspects, interstitium with foci of polymorphic inflammatory infiltrate with lymphomononuclear predominance. Intratubular eosinophilic content – erythrocytes. Tubules with appearance of acute tubular necrosis. Brown stippling in tubular cells – hemosiderosis. (HE, 20x); B – Adhesion (synechiae) between the Bowman's capsule and the glomerular tuft (PAS, 40x); C – MesangialC3 deposits (+++) (IF, 40X).

PO 80
OXALATE NEPHROPATHY IN OLDER ADULTS: KIDNEY BIOPSY AS THE KEY TO THREE OCCULT ETIOLOGIES

Inês Gaspar; Marco Morais; Manuel Silva; Cláudia Costa; Rui Nogueira; Carolina Branco; Estela Nogueira; Iolanda Godinho; Paulo Fernandes; Natacha Rodrigues; João Gonçalves; João Boavida; Mafalda Pinho; Sofia Jorge; José António Lopes

ULS Santa Maria

Category: AKI

Background

Acute Kidney Injury (AKI) is a major cause of morbidity and mortality in older hospitalized patients, in whom frailty, multimorbidity, and polypharmacy often complicate the diagnostic approach. In selected cases, kidney biopsy (KB) remains essential to clarify unexplained or progressive AKI. Among the rare causes identified through histopathology is oxalate nephropathy (ON), a crystalline tubulointerstitial disease caused by calcium oxalate deposition, and associated with

frequently occult etiologies. We present a case series of three older adults in whom KB was crucial in uncovering three distinct underlying causes of ON.

Case 1:

An 81-year-old woman was referred to the emergency department (ED) after routine laboratory testing revealed AKI, with urea of 180 mg/dL and a serum creatinine (sCr) of 14.0 mg/dL. Urinalysis was unremarkable, and kidney ultrasound showed no abnormalities. A comprehensive serologic workup for autoimmune, paraproteinemic, and infectious etiologies was negative. KB demonstrated calcium oxalate crystals deposition associated with diffuse acute tubular injury (Figure 1). Further detailed history revealed chronic, unprescribed ingestion of 2000 mg of vitamin C daily. A diagnosis of ON secondary to excessive ascorbic acid intake was established. The patient initiated hemodialysis and remains dialysis-dependent at 6 months of follow-up.

Case 2:

A 75-year-old man presented with fatigue, dyspepsia, and weight loss. Over 6 months, serum creatinine rose from 1.3 to 4.31 mg/dL, associated with normocytic anemia, hyperphosphatemia, and non-anion gap metabolic acidosis. Urinalysis showed sterile leukocyturia and non-nephrotic proteinuria, without hematuria. Kidney ultrasound revealed increased cortical echogenicity and non-obstructive nephrolithiasis. KB demonstrated calcium oxalate crystal deposition, acute tubular injury, and interstitial fibrosis, establishing the diagnosis of oxalate nephropathy. CT revealed a pancreatic mass. 24-hour urine metabolic profile confirmed hyperoxaluria and hypocitraturia, with normocalcemia. Serum amylase and lipase were suppressed and fecal elastase was undetectable, confirming exocrine pancreatic insufficiency in the setting of gastrointestinal malignancy. The patient was started on pancreatic enzyme replacement therapy, hydration, and dietary oxalate restriction, with referral to Oncology. At 1-month outpatient follow-up, KF remained stable (sCr 4.5 mg/dL) with preserved urine output.

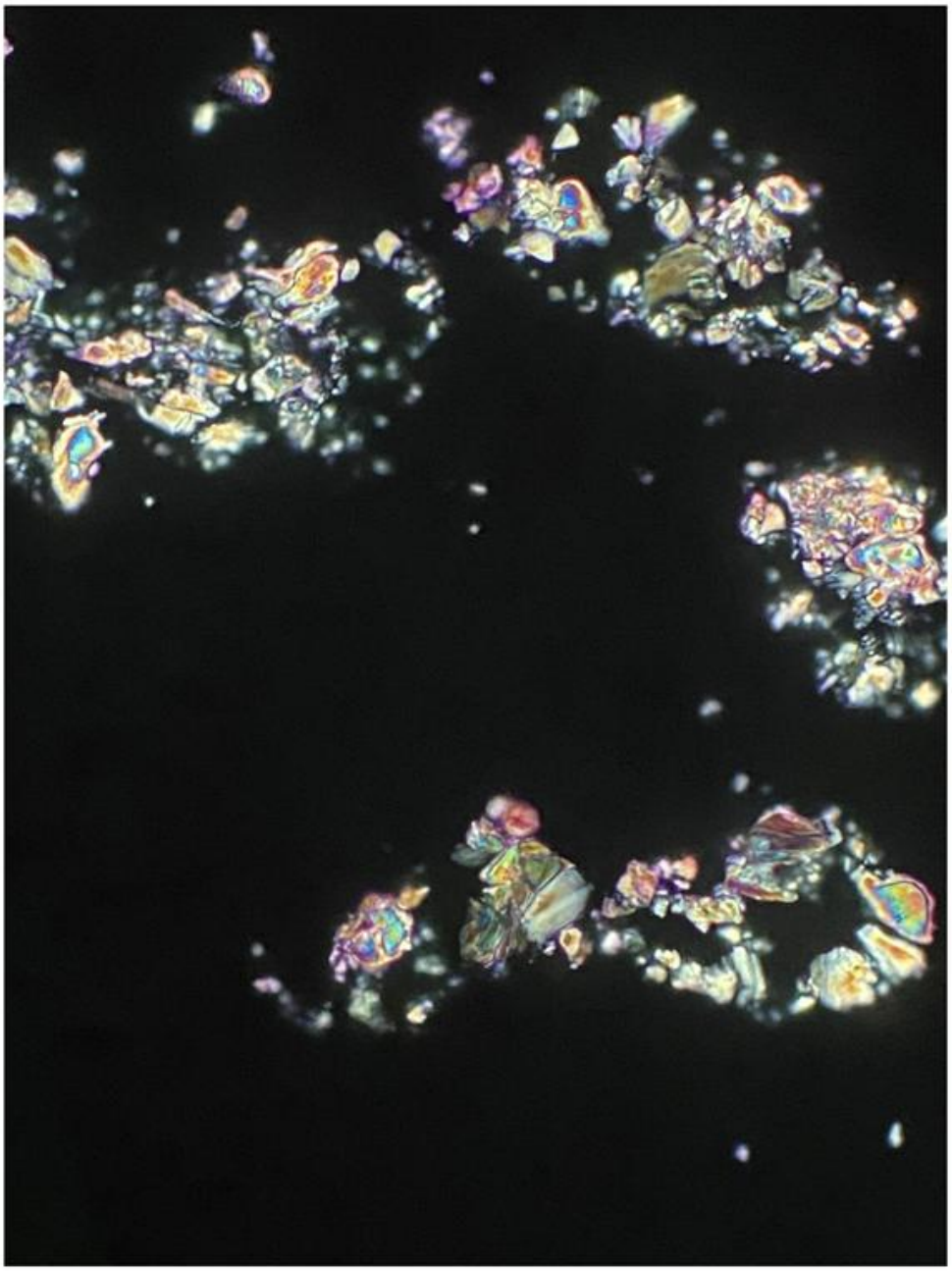
Case 3:

A 71-year-old female with a history of epithelioid angiosarcoma of the face and neck, undergoing second-line chemotherapy with pegylated liposomal doxorubicin (PLD) in the previous 3 months, presented with progressive decline of KF (sCr 1,01 > 4,23 mg/dL) over a period of 3 months. Urinalysis revealed only mild leukocyturia. KB demonstrated diffuse tubular injury with abundant calcium oxalate and calcium phosphate crystals within the tubules. KF continued to deteriorate and the patient initiated hemodialysis. Further investigations showed no evidence of alternative metabolic drivers beside the presence of methoxypolyethylene glycol, an excipient within the PLD formulation. PLD was consequently discontinued, and the patient transitioned to immune checkpoint inhibition, remaining dialysis-dependent at 6 months of follow-up.

Discussion and learning point

Oxalate nephropathy often presents insidiously and may remain clinically silent, making KB a key diagnostic tool. Identified etiologies included iatrogenic vitamin C toxicity, malignancy-associated enteric hyperoxaluria, and suspected toxicity related to polyethylene glycol–based chemotherapeutic agents, with PLD considered the most likely trigger of AKI in our patient. Careful clinicopathologic correlation was essential to uncover the underlying systemic causes and guide targeted management.

Figure 1 - Light microscopy with polarized light



PO 81
BEYOND CLASS V LUPUS NEPHRITIS: PRESUMED LUPUS PODOCYTOPATHY IN RECURRENT NEPHROTIC SYNDROME

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Background

Lupus podocytopathy (LP) is rare renal manifestation of systemic lupus erythematosus (SLE). Kidney biopsy is central to diagnosis. We report the therapeutic challenges of presumed LP in a patient with a contraindication to conventional percutaneous kidney biopsy.

Case description

A 53-year-old man with a 7-year history of SLE, followed by the Internal Medicine department, who was receiving prednisolone (PDN) 10 mg daily. His medical history was notable for two prosthetic cardiac valves, requiring anticoagulation with warfarin.

In April 2025, the patient developed abrupt-onset anasarca and oliguria, without extrarenal SLE activity besides arthritis. Evaluation showed nephrotic syndrome, with serum albumin 2.5 g/dL, total cholesterol 283 mg/dL, 24-hour proteinuria of 10.16g and albuminuria of 5.7g. Serum creatinine was 2.6 mg/dL. Urinary sediment was bland. Additional evaluation showed negative anti-dsDNA, normal C3, C4 and C1q. Renal ultrasound was unremarkable.

Kidney biopsy was not performed because interruption of anticoagulation was considered to carry a high risk of thrombosis of the prosthetic valves. Transjugular kidney biopsy (TKB) was recognized as a potential alternative, but it was unavailable.

Given the abrupt onset of nephrotic syndrome, negative serological markers of SLE activity, and inactive urinary sediment, empirical treatment for presumed LP was initiated with intravenous methylprednisolone (MTP) pulses, followed by oral PDN at 1 mg/kg/day. Mycophenolate mofetil (MMF) was also started at 2g/day. Despite IV diuretics and albumin, he developed anuric acute kidney injury with diuretic-refractory hypervolemia, requiring 3 sessions of hemodialysis.

At discharge, 17 days after initiation of dual immunosuppression (IS), serum creatinine was 0.6 mg/dL, serum albumin was 3.5 g/dL, and proteinuria had decreased to 419 mg/day.

He remained in complete remission for 4 months. In October, nephrotic syndrome recurred while he was receiving PDN 10 mg/day, MMF 2 g/day, and ramipril 1.25 mg/day. Serum albumin was 2.4 g/dL, serum creatinine was 0.8 mg/dL, and 24-hour urine cshowed proteinuria of 17.98 g/day. There was no extrarenal SLE activity, urinary sediment was inactive, complement levels were preserved, and anti-dsDNA antibodies were negative.

Three additional IV MTP pulses were administered, followed by PDN 40 mg/day, while MMF was continued at 2 g/day. Tacrolimus was introduced because of an insufficient reduction in proteinuria.

Five months later, only partial remission had been achieved prompting consideration of treatment escalation.

Discussion

This case illustrates the therapeutic challenges of treating lupus nephritis (LN) in the absence of a histologic diagnosis.

Although class V LN is more common than LP, several clinical features favoured a podocytopathy-like process: abrupt onset of nephrotic syndrome, rapid initial response to high-dose steroids, relapse during steroid tapering and bland urinary sediment.

A TKB would have been valuable, as it could have distinguished LP from class V LN and assessed chronicity, while avoiding the risk of prosthetic valve thrombosis associated with interruption of anticoagulation. This is particularly relevant in LN, where clinicopathological discordance is a well-recognized phenomenon.

No formal treatment guidelines exist for LP. At presentation, MMF was added to glucocorticoids because of extrarenal lupus activity and as a steroid-sparing agent in a patient with significant

corticosteroid burden. At relapse despite dual IS, tacrolimus was added in line with the KDIGO 2021 recommendations for relapsing minimal change disease.

Partial remission despite triple therapy raises the question of treatment escalation. Rituximab would be a logical next step, as it is used in frequently relapsing or steroid-dependent podocytopathies and may also be considered in class V LN with inadequate response to MMF and calcineurin inhibitors.

PO 82
GENETIC DIAGNOSIS IN THE 8TH DECADE: A CLINICAL CASE

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Category: Genetics

Background

Chronic kidney disease (CKD) of genetic origin accounts for approximately 10–20% of CKD cases in adults and may be underdiagnosed, particularly in patients with CKD of unknown etiology. Although genetic CKD typically presents at younger ages, there is no upper age limit for disease onset. Therefore, late-onset genetic kidney diseases should also be considered, especially in individuals with a positive family history or other suggestive clinical features.

Case description

A 76-year-old Caucasian woman was referred to the nephrology clinic for evaluation of CKD stage G3A3 according to KDIGO classification, with proteinuria of 2 g/day and significant albuminuria (urinary albumin-creatinine ratio of 1500 mg/g), associated with a bland urinary sediment and asymptomatic hyperuricemia. Renal ultrasound demonstrated normal-sized kidneys with increased parenchymal echogenicity and multiple cortical and parapelvic cysts measuring 1–2 cm. Investigation for secondary causes of CKD, including autoimmune diseases, viral infections, and dysproteinemia, was negative. Family history was notable for CKD: the maternal grandmother underwent hemodialysis in her late seventies due to CKD of unknown etiology, and the patient's mother had maculopathy without known CKD. No consanguinity was reported. The patient was referred to a nephrogenetics clinic and, following pre-test genetic counseling, underwent genetic testing using a next-generation sequencing panel based on whole-exome sequencing for CKD of unknown etiology, including mitochondrial genome analysis. Genetic testing identified a heterozygous pathogenic variant in the DNAJB11 gene, c.319dup, p.(Ser107Phefs*27), classified as pathogenic according to the criteria of the American College of Medical Genetics and Genomics. To our knowledge, this variant has not been previously described in the literature. The molecular findings were consistent with the clinical phenotype, confirming the diagnosis of autosomal dominant polycystic kidney disease type 6, with or without polycystic liver disease (MIM #618061). The patient was subsequently referred for genetic counseling and screening of at-risk family members, particularly her son, who is of reproductive age.

Discussion and learning point

A genetic etiology should be considered in patients with CKD of unknown cause. Although genetic kidney diseases are more commonly present earlier in life, there is no upper age limit for disease onset. Therefore, genetic testing may also play an important role in the diagnostic work-up of late-onset CKD, as described in disorders such as Fabry disease and autosomal dominant tubulointerstitial kidney disease associated with UMOD variants, as well as in other inherited

kidney diseases, including those related to DNAJB11 pathogenic variants. In addition to improving diagnostic accuracy, establishing a genetic diagnosis may have important clinical implications, including reproductive and family planning counseling, screening for extrarenal manifestations, and optimizing kidney transplantation strategies through appropriate selection of related living donors and assessment of post-transplant disease recurrence risk.

PO 83
IMMUNOTACTOID GLOMERULONEPHRITIS: A RARE GLOMERULAR DISEASE PRESENTING WITH PROTEINURIA IN PREGNANCY

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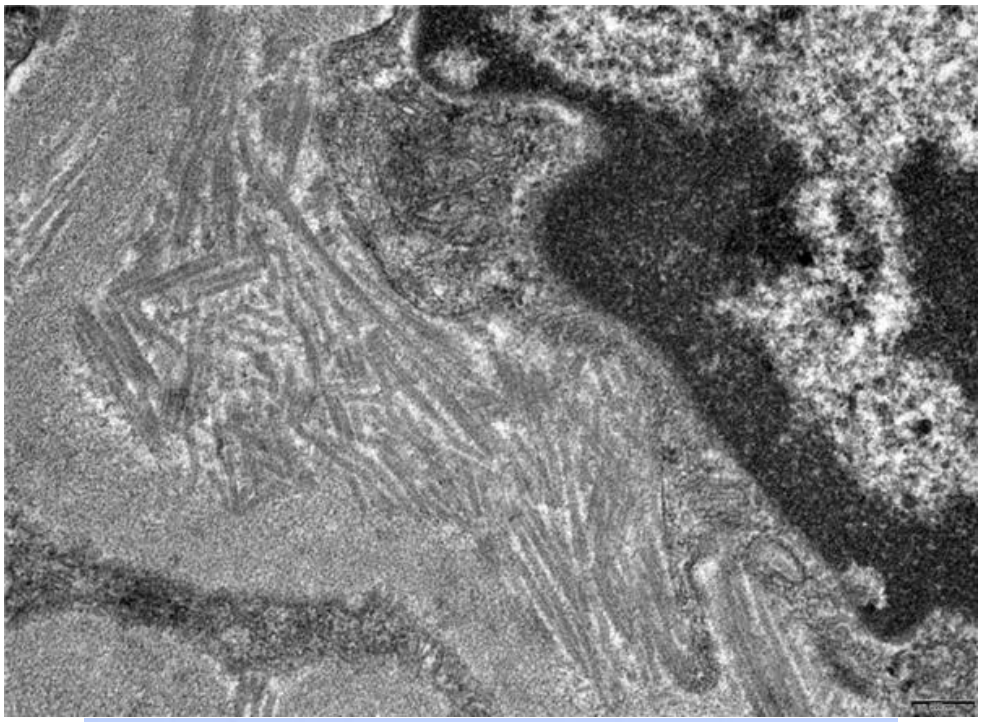
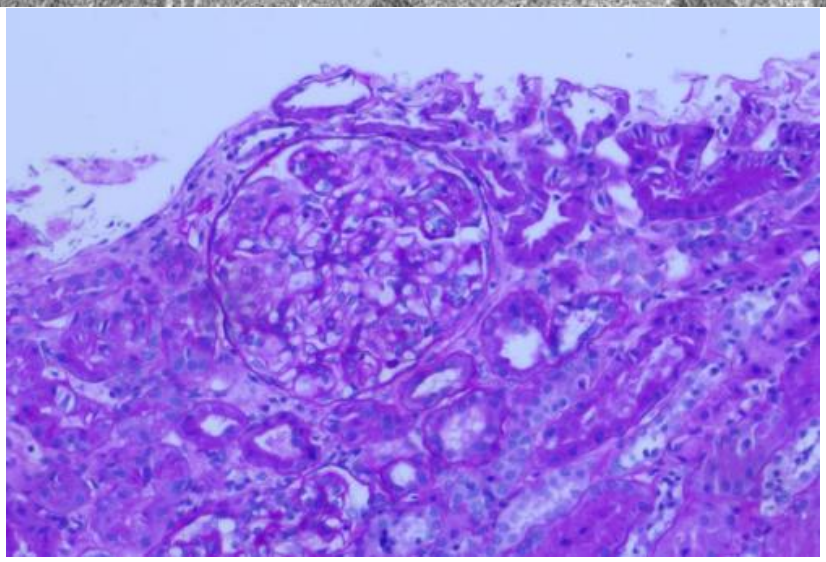
Background: Immunotactoid Glomerulopathy (ITG) is a rare glomerular disease (< 0.06% of native kidney biopsies in some series) affecting mainly adults between 50 and 60 years, generally presenting with nephrotic-range proteinuria, microscopic hematuria, and/or reduced kidney function. An underlying monoclonal gammopathy or lymphoproliferative disorder is identified in up to two-thirds of cases, and treatment directed at the hematologic disease may lead to remission.

Case Description: A 44-year-old woman was referred to nephrology for nephrotic- range proteinuria first detected during pregnancy. The pregnancy was complicated by gestational diabetes with no preeclampsia; delivery was uneventful. Proteinuria persisted two months postpartum, prompting further investigation. Her medical history included Chronic Lymphocytic Leukemia (CLL) under active surveillance, HLA- B27–positive spondyloarthritis, and former smoking. Postpartum evaluation revealed nephrotic proteinuria (5.8 g/day) and intermittent microscopic hematuria (13 erythrocytes per high-power field). Renal function remained preserved. Autoimmune and viral serologies were negative, with no complement consumption. Serum protein electrophoresis showed no monoclonal component, serum immunofixation was positive for IgG/kappa, serum free light chain ratio was within normal range and urinary immunofixation was negative. Kidney biopsy revealed proliferative glomerulonephritis with segmental mesangial and endocapillary hypercellularity (Figure 1). Immunofluorescence showed granular IgG and C3 deposition with kappa light chain restriction. Electron microscopy revealed organized microtubular deposits (> 20 nm) in mesangial, subendothelial, and subepithelial locations with diffuse podocyte foot process effacement (Figure 2), Immunotactoid Glomerulopathy was diagnosed. Following discussion with hematology, she initiated obinutuzumab- venetoclax, with proteinuria regression from 6 g to 1 g/day and microhematuria to 2+/high-power field within four months after treatment initiation. As an adverse effect, she had transient neutropenia that resolved after a G-CSF course.

Discussion and learning points: CLL-associated glomerular disease occurs in less than 1% of CLL cases; the most frequent pattern is membranoproliferative glomerulonephritis (45%), with ITG representing a rarer but well-recognized association. This case is notable for the patient's young age, reinforcing that gammopathy-related renal disease should not be overlooked in younger patients. Kidney biopsy was decisive, especially electron microscopy, confirming ITG and identifying CLL as the likely pathogenic driver, directly guiding treatment. In patients with hematologic malignancies, routine evaluation of renal impairment enables early management before irreversible renal damage occurs. Also, this case highlights the importance of nephrology-hematology collaboration in integrating biopsy findings with hematologic assessment and promptly initiating clone-directed therapy that resulted in significant proteinuria remission.





PO 85
RITUXIMAB: A "GAME CHANGER" IN RELAPSING NEPHROTIC SYNDROME

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Background
Minimal change disease (MCD) is a leading cause of nephrotic syndrome, particularly in children but also in young adults. While glucocorticoids induce remission in most cases, a subset of patients develop frequently relapsing or steroid-dependent disease, with significant treatment-related toxicity. Rituximab has emerged as an effective steroid-sparing option, particularly in difficult-to-control cases.

Case Description

A 26-year-old caucasian man with no relevant past medical history presented in November 2016 with a 2-month history of edema, weight gain and foamy urine. Laboratory evaluation showed preserved kidney function (creatinine 0.9 mg/dL), severe hypoalbuminaemia (1.7 g/dL) and hypercholesterolaemia (315 mg/dL). Antinuclear antibodies were positive (1:640), while the remaining autoimmune and infectious work-up was negative. Urinalysis revealed nephrotic-range proteinuria (urine protein-to-creatinine ratio [UPCR] 4916 mg/g) without haematuria. Kidney ultrasound was unremarkable. Kidney biopsy was performed after starting prednisolone 1mg/kg and showed normal glomeruli on light microscopy with negative immunofluorescence (IF), suggesting MCD; however, electron microscopy (EM) revealed partial podocyte foot process effacement with focal segmental sclerosis-like changes, raising the possibility of evolving FSGS. Genetic testing and malignancy screening were negative. The subsequent course was marked by steroid dependence and multiple relapses. The first relapse (UPCR 7273 mg/g) was complicated by femoropopliteal deep vein thrombosis. PDN was reintroduced (80mg/day) together with anticoagulation, leading to complete remission. In December 2017, proteinuria recurred (2172 mg/g) during steroid tapering, prompting initiation of tacrolimus (TAC) (2mg twice daily). In July 2019, after PDN discontinuation and TAC reduction to 1 mg twice daily, proteinuria increased again (2758 mg/g), requiring TAC dose escalation and PDN reintroduction. A second kidney biopsy, also performed under prednisolone, showed normal light microscopy and negative IF, but again persistent FSGS-like changes on EM. In May 2020, a further relapse occurred after PDN discontinuation. Given the frequently relapsing course and cumulative exposure to steroids and calcineurin inhibitors, rituximab was initiated in 2023 (1g every 6 months for 2 years), allowing TAC withdrawal and leading to sustained complete remission. One year after the last dose, the patient remains in complete remission, with preserved kidney function and minimal proteinuria (UPCR 70 mg/g).

Discussion and learning points

This case highlights the effectiveness of rituximab in achieving sustained remission in steroid-dependent and frequently relapsing nephrotic syndrome refractory to conventional therapies, including calcineurin inhibitors. It also supports its use as a safe steroid- and calcineurin inhibitor–sparing option in adults, even in the presence of heterogeneous histological findings. Therapeutic response remains a key determinant in guiding management, and early use of rituximab may help reduce cumulative exposure to corticosteroids and calcineurin inhibitors, thereby minimizing treatment-related toxicity while improving long-term outcomes in this complex and difficult-to-manage population.

PO 86
Sub-nephrotic range proteinuria and renal insufficiency: a case of Fibrillary Glomerulonephritis

Henrique Gouveia Rodrigues; Miguel Gonçalves; Gil Silva

SESARAM

Background

Fibrillary glomerulonephritis (FGN) is a rare form of glomerulonephritis, accounting for 0.5-1% of native kidney biopsies, often presenting with nephrotic syndrome, microscopic hematuria, and renal insufficiency. It typically affects patients in their 5th–6th decade and frequently progresses to end-stage kidney disease (ESKD).

Case Presentation

We present the case of a previously healthy 36-year-old male bodybuilder referred to the nephrology clinic in late 2025 for microscopic hematuria, sub-nephrotic range proteinuria (urine protein-to-creatinine ratio [uPCR] ~2 g/g), and an elevated serum creatinine (SCr) of 1.9 mg/dL.

The patient reported taking 15 grams of creatine supplements daily for over five years. He specifically denied the use of other dietary supplements, performance-enhancing substances, illicit drugs, as well as any recent use of non-steroidal anti-inflammatory drugs (NSAIDs) or antibiotics. He also reported no recent infections and lacked a personal or family history of kidney disease, gross hematuria, or hearing loss.

Upon clinical evaluation, the patient was entirely asymptomatic. Physical examination was unremarkable, notable only for normal blood pressure and the absence of peripheral edema. An extensive laboratory work-up was also unremarkable: complement levels were normal, and screening for antinuclear antibodies (ANA) and antineutrophil cytoplasmic antibodies (ANCA) was negative. Infectious disease serologies for HIV, syphilis, and hepatitis B and C were non-reactive. Additionally, the serum free light chain ratio was normal, and serum protein electrophoresis revealed no monoclonal spikes. A review of the patient's medical records showed elevated serum creatinine levels (SCr of 1.6 mg/dL) and microhematuria since 2022.

The patient was initiated on standard antiproteinuric therapy with an angiotensin-converting enzyme inhibitor (ACEi) and a sodium-glucose cotransporter-2 inhibitor (SGLT2i). He was also strongly advised to discontinue all dietary supplements. At the one-month follow-up, his SCr had improved to 1.6 mg/dL, and his proteinuria had decreased to a uPCR of ~0.9 g/g; however, microscopic hematuria persisted. Given these findings, a percutaneous renal biopsy was performed.

Light microscopy revealed heterogeneous glomerular mesangial expansion due to infiltrating extracellular material that was Congo red-negative. Immunofluorescence demonstrated focal smudgy mesangial staining for IgG (1+), C3 (1+), and both kappa (1+) and lambda (1+) light chains. Electron microscopy revealed randomly arranged fibrillary deposits with a mean diameter of 14.5 nm within the mesangium, along with segmental effacement of the podocyte foot processes. Based on these histopathological findings, a diagnosis of FGN was established.

FGN generally carries a poor prognosis, with nearly 50% of patients progressing to end-stage kidney disease (ESKD) within two to four years of diagnosis. Management primarily focuses on blood pressure control and proteinuria reduction using ACE inhibitors. While immunosuppression, particularly with rituximab, is sometimes utilized, studies have demonstrated mixed results regarding its efficacy in achieving partial remission.

Conclusion

Given our patient's favorable response to conservative antiproteinuric measures, we opted against initiating immunosuppressive therapy at this time. At four months post-diagnosis, renal function remains stable with no evidence of disease progression, although persistent microscopic hematuria indicates ongoing clinical activity. The purpose of this paper is to report a rare case of FGN presenting with an unusually indolent disease course.

PO 87

TUBEROUS SCLEROSIS COMPLEX WITH HEPATIC AND RENAL ANGIOMYOLIPOMAS: DIAGNOSTIC DELAY, PREVENTED HEPATECTOMY AND EXTRAORDINARY RESPONSE TO EVEROLIMUS

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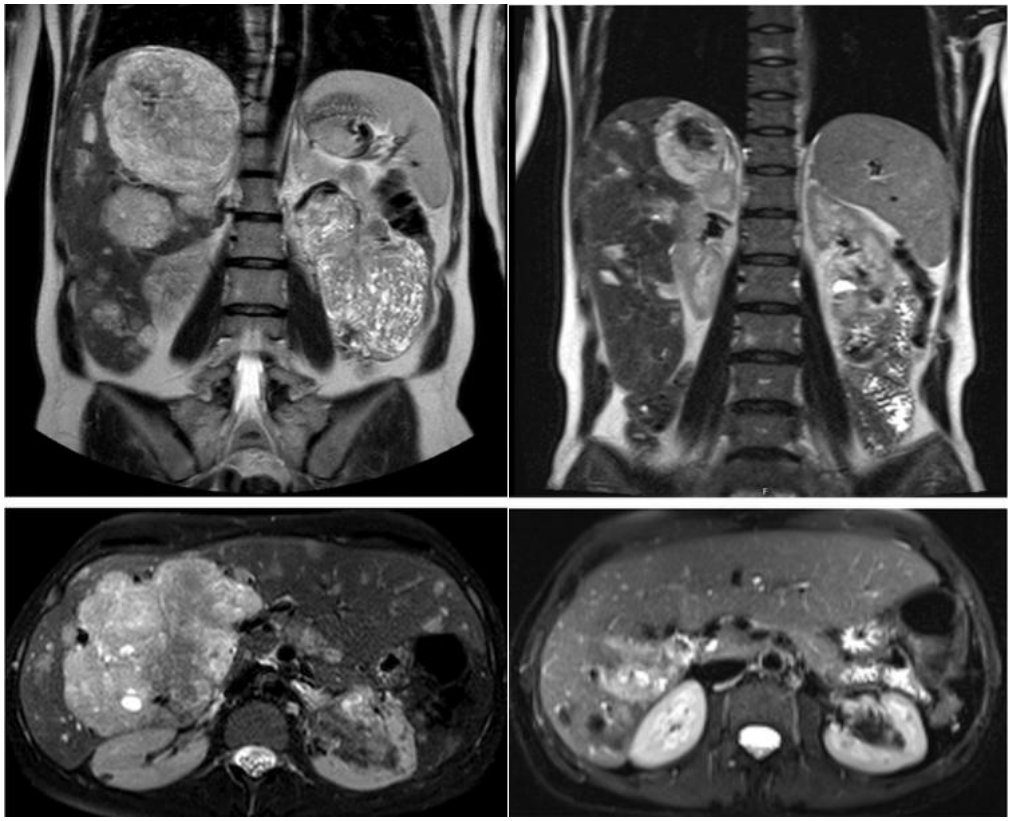
Background: Tuberous sclerosis complex (TSC) is an autosomal dominant disorder caused by pathogenic variants in *TSC1* or *TSC2* genes, leading to mTOR pathway dysregulation and multisystem tumour development. Systemic manifestations include renal and hepatic angiomyolipomas (AMLs), with the primary concern being hemorrhage due to spontaneous rupture. Epithelioid AML may mimic renal cell carcinoma (RCC) on imaging, creating significant diagnostic challenges. Everolimus is the standard of care for TSC-associated AMLs.

Case Description: A 44-year-old woman presented in 2021 with progressive abdominal distension. In September 2024, comprehensive evaluation established the diagnosis of TSC with multi-organ involvement: cutaneous (facial angiofibromas and hypopigmented macules of the lower limbs), pulmonary (multifocal micronodular pneumocyte hyperplasia) and hepatic and renal AMLs. CT showed bilateral renal lesions suspicious for epithelioid AML versus RCC and hepatic lesions compatible with AML, though metastatic disease could not be excluded. Abdominal MRI (October 2024, Figure 1) demonstrated a large 11 cm highly vascular left renal mass with diffusion restriction, extending into the renal sinus and vein, once again raising the possibility of AML vs RCC. Massive hepatomegaly (29.5 cm) with multiple lesions (largest 22 cm) prompted consideration of hepatectomy. Liver biopsy confirmed AML and tissue analysis identified two pathogenic *TSC2* variants not detected in peripheral blood. Accounting for the risk of spontaneous rupture, the patient was being evaluated for selective embolisation of the renal AML. Everolimus was initiated at 5 mg/day in August 2025, subsequently uptitrated to 10 mg/day, with good tolerance. Follow-up MRI (March 2026, figure 2) demonstrated a remarkable radiological response: kidneys normalised in size and the largest renal AML measured 35x30x60mm; there was a reduction of the hepatomegaly (21cm) and all lesions reduced significantly, the two largest at 68x91x88mm and 53x70x61mm. This led to a significant improvement in the patient's symptoms and to the deferral of both hepatectomy and selective embolisation.

Discussion: This case highlights two critical learning points. First, the impact of diagnostic delay: the patient was symptomatic since 2021, TSC was only confirmed in September 2024. Without the correct diagnosis, the patient was heading towards hepatectomy - a major intervention for a benign, medically treatable condition. This highlights the need to consider TSC in adults with bilateral renal and hepatic lesions and the importance of a dedicated multidisciplinary team specialised in hereditary nephropathies. Second, the extraordinary response to mTOR inhibition: within months of starting everolimus, clinically meaningful volumetric reduction was achieved in both renal and hepatic AMLs, eliminating the need for surgical and endovascular procedures.

Figure 1: Abdominal MRI (October 2024), coronal (upper) and axial (lower) sections depicting hepatomegaly (29.5 cm) and hepatic (22 cm) and renal (11 cm) masses

Figure 2: Abdominal MRI (March 2026), coronal (upper) and axial (lower) sections showing reduction of the liver size (21 cm) and of the hepatic (the largest with 91 mm) and renal (35x30x60 mm) masses



PO 88
THE SILENT CULPRIT: A RARE CAUSE OF MEMBRANOPROLIFERATIVE GLOMERULONEPHRITIS

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Category: Glomerular diseases

Background

Membranoproliferative glomerulonephritis (MPGN) is a pattern of glomerular injury often associated with immune complex deposition. When MPGN is characterized by an immunoglobulin-positive pattern, secondary causes such as chronic infections, autoimmune diseases, and monoclonal gammopathies must be excluded.

Case description

We present the case of a 64-year-old male with a 1-year history of cyclic subjective nocturnal fever. His medical history included aortic valve replacement and ascending aortic graft complicated by infective endocarditis two years earlier. Laboratory evaluation showed anemia (hemoglobin 9.3 g/dL), impaired renal function (creatinine 1.7 mg/dL; estimated glomerular filtration rate 45 mL/min/1.73 m^2), low C3 (68 mg/dL), and normal C-reactive protein. Urinalysis revealed microscopic hematuria and proteinuria (albumin-to-creatinine ratio of 3200 mg/g). Blood cultures, viral serologies (HIV, HCV and HBV), autoimmune screening and monoclonal gammopathy workup were negative.

Serology confirmed chronic *Coxiella burnetii* (C. burnetii) infection with elevated phase I and II IgG (1:51200) and IgM (phase I 1:3200; phase II 1:1600), supported by a positive polymerase chain reaction. Transesophageal echocardiography showed no vegetations, but positron emission tomography demonstrated increased uptake along the aorta, suggesting vascular involvement. Treatment with doxycycline and hydroxychloroquine was initiated.

A kidney biopsy was performed (total of 16 glomeruli) and revealed global mesangial and endocapillary hypercellularity with capillary wall thickening and segmental duplication. Two glomeruli showed capillary wall disruption with fibrinoid necrosis and cellular crescents, associated with Bowman's capsule disruption (Figure 1). Immunofluorescence demonstrated positive staining for C3 (++++) in a segmental pseudolinear pattern along the capillary walls and mesangium with rare deposits of IgM, C1q, kappa, and lambda light chains (++/+++) in the capillary walls (Figure 2). The patient was diagnosed with immune complex-mediated MPGN secondary to chronic C. burnetii infection.

Due to marked inflammatory activity on kidney biopsy and worsening renal function despite 3 months of antibiotic therapy, a short course of corticosteroids was added to the treatment regimen.

During follow-up, renal function stabilized (creatinine 1.8 mg/dL), with significant reduction in proteinuria (albumin-to-creatinine ratio decreasing to 200 mg/g), and normalization of complement levels.

Discussion and learning point

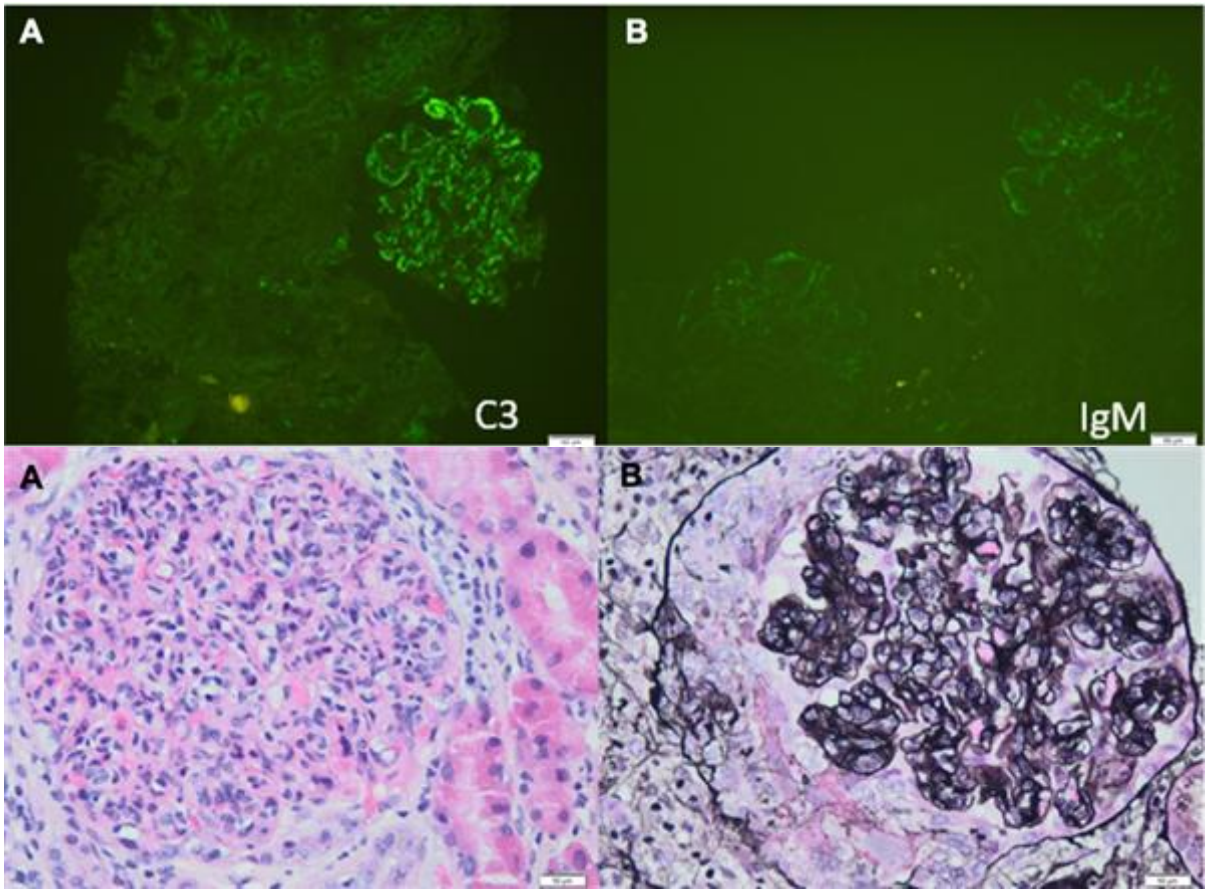
Chronic Q fever is a rare and underrecognized cause of infection-related MPGN, with only a limited number of cases described in the literature. Diagnosis is frequently delayed due to nonspecific symptoms, a prolonged disease course, and the absence of vegetations or positive blood cultures in many cases.

This case highlights the importance of considering C. burnetii infection in patients presenting with MPGN, particularly in suspected culture-negative endocarditis. Early recognition and prolonged targeted antimicrobial therapy are essential to improve renal and overall outcomes. This case also supports the potential role of adjunctive corticosteroid therapy in selected patients, although evidence remains limited.

Keywords: membranoproliferative glomerulonephritis, Q fever, infection-related glomerulonephritis

Figure 1: Light microscopy findings of the kidney biopsy. (A) Periodic acid-Schiff stain showing mesangial and endocapillary hypercellularity. (B) Silver stain demonstrating capillary wall thickening with segmental duplication and a cellular crescent with disruption of Bowman's capsule.

Figure 2: Immunofluorescence findings of the kidney biopsy. (A) Strong C3 staining in a segmental pseudolinear pattern along the capillary walls and mesangium. (B) Weak IgM staining along the capillary walls.



PO 90
CRESCENTIC IGA NEPHROPATHY AFTER KIDNEY TRANSPLANTATION: INSIGHTS FROM THREE CASES WITH ADVERSE GRAFT OUTCOMES

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Category: Glomerular Diseases

Keywords: IgA nephropathy, kidney transplantation, crescentic IgAN

Background: IgA nephropathy (IgAN) recurrence after kidney transplantation is common and usually follows an indolent course. Crescentic forms are rare and remain poorly characterized.

Case Description: We report three kidney transplant recipients with crescentic IgAN and unfavorable graft outcomes.

A 41-year-old man with biopsy-proven IgAN underwent a second deceased donor kidney transplant in 2019 after loss of a previous graft due to recurrent disease. Induction immunosuppression included antithymocyte globulin, followed by tacrolimus, mycophenolate mofetil and prednisone. Progressive albuminuria developed early after transplantation, reaching a urinary albumin-to-creatinine ratio (uACR) of 4.8 g/g. At 22 months post-transplant, serum creatinine increased to 1.8 mg/dL from a baseline of 1.3 mg/dL. Kidney biopsy demonstrated recurrent IgAN (M1E1S1T0C2) with diffuse mesangioproliferative and focal membranoproliferative features and crescents involving 40% of glomeruli, without evidence of rejection. Corticosteroid therapy following the Pozzi regimen was initiated, but subsequent

cytomegalovirus infection required valganciclovir therapy and reduction of immunosuppression. Five years after the biopsy, graft dysfunction and persistent proteinuria remained evident.

A 35-year-old man with end-stage kidney disease secondary to IgA vasculitis underwent deceased donor transplantation in 2018. Eighteen months later, he developed nephrotic-range albuminuria (uACR 4.4 g/g), hematuria and graft dysfunction, with serum creatinine of 2.2 mg/dL. Kidney biopsy revealed crescentic IgAN (M0E1S1T1C2) with necrotizing lesions, arteritis and dominant C3 deposition, raising suspicion of overlap with C3 glomerulopathy. Electron microscopy demonstrated subendothelial and intramembranous electron-dense deposits, supporting complement-mediated injury. Therapeutic options including corticosteroids, cyclophosphamide, rituximab, plasma exchange and eculizumab were discussed but declined by the patient. Progressive graft dysfunction led to graft loss 2 years after biopsy.

A 70-year-old man underwent kidney transplantation in 2018 for end-stage kidney disease of unknown etiology. Approximately 3 years post-transplant, he developed nephrotic-range albuminuria and progressive graft dysfunction, with serum creatinine of 2.5 mg/dL. Kidney biopsy demonstrated de novo crescentic IgAN (M0E1S1T0C1) with crescents involving 20% of glomeruli and associated fibrinoid necrosis. Supportive treatment with optimization of renin-angiotensin system blockade and diuretics was implemented. Escalation of immunosuppression was not pursued because of advanced age and significant comorbidities. Renal function progressively deteriorated, and hemodialysis was initiated 2 months after biopsy.

Across all cases, presentation was characterized by albuminuria, hematuria and rapid graft dysfunction. Histopathology consistently demonstrated crescentic IgA deposition without evidence of antibody-mediated rejection or C4d positivity.

Discussion and Learning Points: Crescentic IgAN after kidney transplantation appears to represent a rare but aggressive phenotype associated with rapid graft dysfunction and poor outcomes. These cases highlight the limited efficacy or feasibility of current therapeutic strategies and support the hypothesis that complement-mediated mechanisms may contribute to disease severity in selected patients.

PO 91
ACUTE TUBULAR NECROSIS ASSOCIATED WITH THE USE OF ENFORTUMAB VEDOTIN

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[2] LAP-UNILABS

Category: Nephrotoxicity, Acute Kidney Injury

Background: Enfortumab vedotin has emerged as a therapeutic option for treating metastatic urothelial carcinoma unresponsive to previous lines of therapy. Despite its demonstrated efficacy in terms of overall survival and tumor response rate, its safety remains under investigation. The most common adverse effects are peripheral neuropathy, cutaneous toxicity, hyperglycemia, and asthenia. However, cases of renal toxicity have also been described. The underlying mechanism is not yet fully understood and may involve drug-induced tubular toxicity.

Case description: A 49-year-old man with high cardiovascular risk, with a past medical history of hypertension with target-organ damage (left ventricular hypertrophy), type 2 diabetes mellitus and grade I obesity, was diagnosed with high-grade urothelial carcinoma in 2022. He started neoadjuvant chemotherapy with cisplatin and gemcitabine in August 2022, which was discontinued in November 2022 because of hepatic toxicity. In January 2023, the patient underwent curative surgery. The patient remained under outpatient surveillance, and by the end of 2024, disease progression with lymph node involvement was detected and confirmed by biopsy of a left external iliac lymph node conglomerate. In April 2025, he resumed treatment with cisplatin

and gemcitabine for 3 months, followed by maintenance therapy with avelumab. In January 2026, new evidence of nodal recurrence was identified, and treatment with enfortumab vedotin was initiated in March 2026. Three weeks after starting the treatment, the patient presented to the hospital with an erythematous and pruritic skin rash involving the upper and lower limbs symmetrically, as well as hyperglycemia that was difficult to control at home. Laboratory evaluation revealed acute kidney injury (creatinine 1.8 mg/dL from a baseline of 1.2 mg/dL), mild hypokalemia (potassium 3.2 mmol/L), metabolic acidosis (bicarbonate 15.5 mmol/L), and hypophosphatemia (0.7 mg/dL). Urinalysis showed leucocyturia (10–25 leukocytes per high-power field), with a protein-to-creatinine ratio of 1.1 mg/mg and an albumin-to-creatinine ratio of 59 mg/g. Renal ultrasonography revealed no abnormalities. The patient was admitted to the hospital, and during hospitalization, renal function remained stable, although oral bicarbonate and potassium supplementation was required, with a good response. The diagnostic hypotheses included acute interstitial nephritis or acute tubular necrosis associated with enfortumab vedotin, with associated Fanconi syndrome secondary to tubular injury. To clarify the etiology of acute kidney injury, a renal biopsy was performed, revealing findings compatible with acute tubular necrosis and tubulointerstitial nephritis lesions with signs of chronicity.
The patient was discharged with outpatient follow-up and was maintained on oral bicarbonate and potassium chloride supplementation. Following a multidisciplinary discussion between the nephrology and oncology departments, enfortumab vedotin was discontinued. The patient remains under follow-up, with serum creatinine levels returning to baseline values and resolution of hypophosphatemia and hypokalemia, although metabolic acidosis persists. At the oncology multidisciplinary meeting, a definitive decision was made to discontinue enfortumab vedotin and initiate trastuzumab.

Discussion and learning points: This case describes acute kidney injury in the context of acute tubular necrosis secondary to enfortumab vedotin use, associated with Fanconi syndrome due to tubular toxicity. The increasing incidence of oncological diseases and the growing use of chemotherapy make this topic highly relevant. The approach to and management of chemotherapy related toxicity may represent a significant challenge in patient care, showing the importance of reporting and characterizing cases like this.

PO 92
FROM PROLIFERATIVE TO MEMBRANOUS DOMINANCE IN REFRACTORY LUPUS NEPHRITIS: SUCCESSFUL TREATMENT WITH VOCLOSPORIN

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Background
Systemic lupus erythematosus (SLE) is an autoimmune disease in which lupus nephritis (LN) remains a major cause of morbidity and organ damage. Histological transition from proliferative (class III/IV) to membranous (class V) LN may reflect different immunopathological mechanisms and treatment responses. Persistent nephrotic-range proteinuria despite immunologic improvement remains challenging in refractory LN and highlights the value of repeat kidney biopsy. Voclosporin, a novel calcineurin inhibitor approved for active LN, reduces proteinuria and improves renal outcomes, although evidence in refractory membranous-predominant disease remains limited.

Case description

A 38-year-old Asian woman was diagnosed with SLE in 2011 based on malar rash, leukopenia, positive ANA, anti-SSA and anti-dsDNA antibodies, and compatible skin biopsy findings. Sicca symptoms suggestive of secondary Sjögren syndrome and childhood psoriasis were also present. Initial azathioprine and leflunomide were discontinued because of intolerance and leukopenia.

In February 2018, lupus nephritis developed with hematuria, proteinuria (UPCR 2 g/g), hypocomplementemia, anemia, and elevated anti-dsDNA titers. Kidney biopsy showed class IV LN (activity index 8; chronicity index 0). Mycophenolate mofetil (MMF), glucocorticoids, and

tacrolimus induced complete renal and immunologic remission by May 2018 (Figure 1 and 2), although MMF dose reduction was required because of stomatitis and leukopenia.

Renal relapse occurred in November 2019 with progressive proteinuria, recurrent hematuria, and rising anti-dsDNA titers. Between 2020 and 2022, nephrotic-range proteinuria persisted despite MMF, corticosteroids, and tacrolimus, exceeding 8 g/24h by late 2021, while renal function remained preserved (Figure 1). A second biopsy in August 2022 revealed class IV LN with class V features and predominantly active lesions (activity index 7; chronicity index 1). MMF optimization and tacrolimus adjustment failed to induce remission.

Rituximab initiated in January 2023 achieved CD19 depletion, complement normalization, and reduced serologic activity, but only partial renal response (Figures 1 and 2). Belimumab was added in August 2024, stopped in April 2026 after respiratory infection. Despite transient improvement, nephrotic-range proteinuria persisted, reaching 12.3 g/day by late 2025 (Figure 1).

A third biopsy in August 2025 showed mixed class IV/V LN with class V-dominant features, low inflammatory activity, and early chronic lesions, supporting transition to membranous-predominant disease.

Because of unstable tacrolimus exposure and worsening renal function (eGFR 67 mL/min/1.73 m²), treatment was switched to voclosporin in March 2026. At follow-up in May 2026, proteinuria decreased to 3.1 g/g with improved renal function (eGFR 82 mL/min/1.73 m²) (Figure 1).

Discussion and learning points

This case highlights dissociation between immunologic and renal activity in refractory LN. Persistent nephrotic-range proteinuria despite serologic improvement suggested transition to membranous-predominant disease. Failure of MMF, tacrolimus, rituximab, and belimumab underscores the complexity of refractory LN and the value of repeat kidney biopsy. Favorable response to voclosporin suggests it may represent an effective option in refractory class IV/V LN with membranous-dominant features.

PO 93
MEMBRANOUS NEPHROPATHY STRIKES BACK: RECURRENCE AFTER KIDNEY TRANSPLANTATION IN A PATIENT WITH BK VIRURIA

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Background:
Recurrence of primary membranous nephropathy (MN) after kidney transplantation is a well-recognized cause of post-transplant proteinuria and may negatively impact graft survival. Circulating anti-PLA2R antibodies are strongly associated with disease recurrence and may facilitate early diagnosis. In transplant recipients, concomitant BK viral replication may complicate the diagnostic approach and therapeutic strategy, particularly when immunosuppression reduction is required.

Case description:
A 58-year-old man presented in 2014 with nephrotic syndrome (proteinuria 7.3 g/day). Immunologic and viral studies were negative, and malignancy screening was unremarkable. Native kidney biopsy established the diagnosis of primary MN. He was treated with cyclosporine and renin–angiotensin system blockade but evolved with recurrent nephrotic syndrome and progressive kidney dysfunction, reaching ESKD in 2016.

In 2020, he underwent deceased-donor kidney transplantation from a 21-year-old donor. The transplant was considered immunologically high risk because of preformed non–donor-specific class I anti-HLA antibodies. Induction therapy consisted of thymoglobulin (cumulative dose 7.5 mg/kg), followed by maintenance immunosuppression with prednisone, tacrolimus, and mycophenolate mofetil. Immediate graft function was achieved, with serum creatinine of 0.95 mg/dL at discharge.

During the first post-transplant year, BK viruria without viremia was detected, prompting immunosuppression minimization, with subsequent virologic improvement.

In 2026, the patient developed progressive proteinuria and hypoalbuminemia, with albuminuria of 1.7 g/day and serum albumin of 27 g/L, while graft function remained stable. Investigation revealed hypogammaglobulinemia with preserved IgA and IgM, absence of complement consumption or monoclonal gammopathy, persistently negative donor-specific anti-HLA antibodies, and rising BK viruria without viremia. Given the differential diagnosis between recurrent MN and BK virus–associated nephropathy, an allograft biopsy was performed.

Light microscopy (figure 1) showed thickened glomerular basement membranes with spikes on silver stain, without significant mesangial proliferation, tubulitis, capillaritis, or chronic interstitial damage. Immunofluorescence (figure 2) revealed granular IgG and C3c deposits along glomerular capillary walls, with both kappa and lambda light chains, while C1q and C4d were negative. Immunohistochemistry demonstrated weak focal IgG4 staining and no evidence of polyomavirus infection. Serum anti-PLA2R antibodies were markedly elevated (587 U/mL).

The histologic findings, together with positive anti-PLA2R antibodies and nephrotic-range proteinuria, supported the diagnosis of recurrent primary MN in the renal allograft.

Discussion:
This case highlights the diagnostic challenge of post-transplant proteinuria in patients with concurrent BK viruria. Although recurrent MN was strongly suggested by the clinical presentation and elevated anti-PLA2R antibodies, kidney allograft biopsy remained essential to exclude BK virus–associated nephropathy and other transplant-specific pathologies that could significantly influence therapeutic decisions and graft prognosis.

Importantly, the biopsy not only confirmed recurrent MN but also excluded active polyomavirus nephropathy and significant chronic injury, supporting a different immunosuppressive approach. The case further emphasizes the value of integrating serologic biomarkers with histopathology in transplant recipients and illustrates that recurrent MN may initially present with preserved graft function despite clinically significant proteinuria. Early recognition and timely intervention are crucial to optimize long-term graft outcomes.

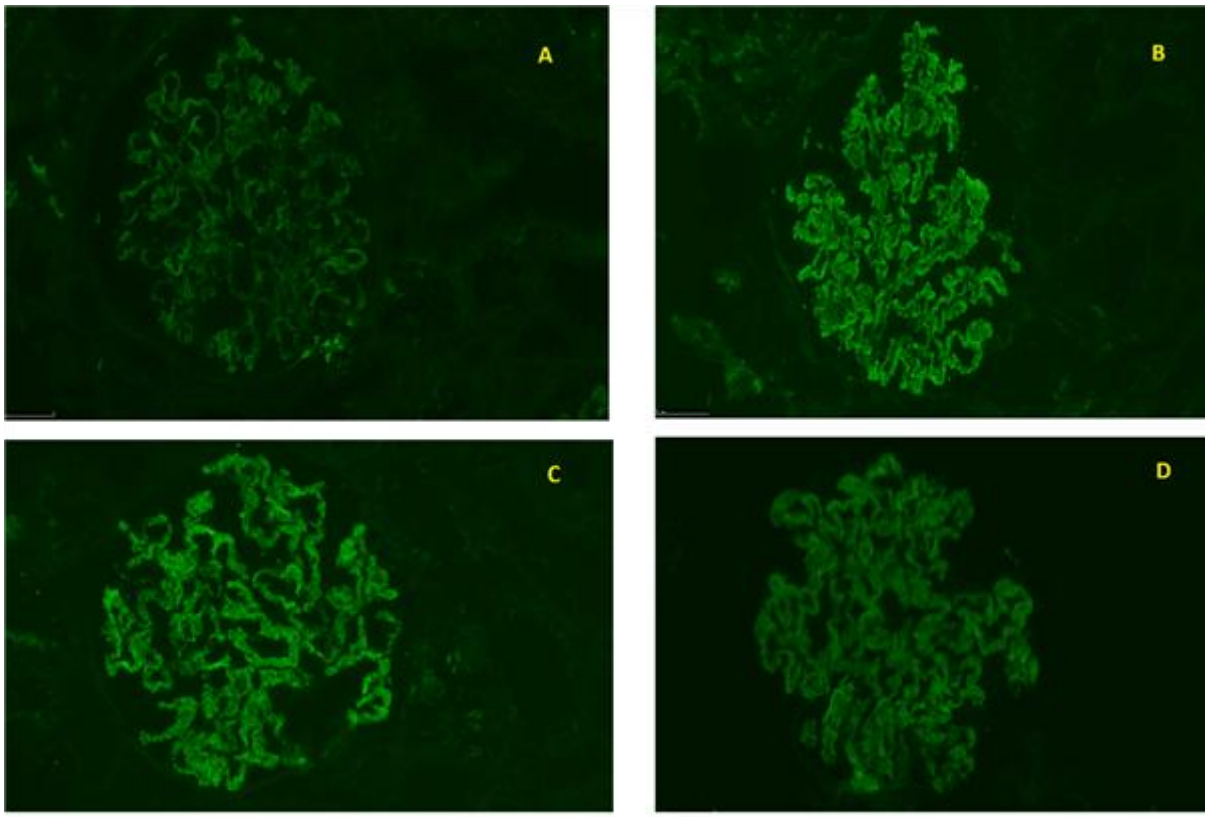

Figure 2. Immunofluorescence demonstrated granular membranous IgG (B) staining with C3 (A) and both kappa (C) and lambda (D) light chains.

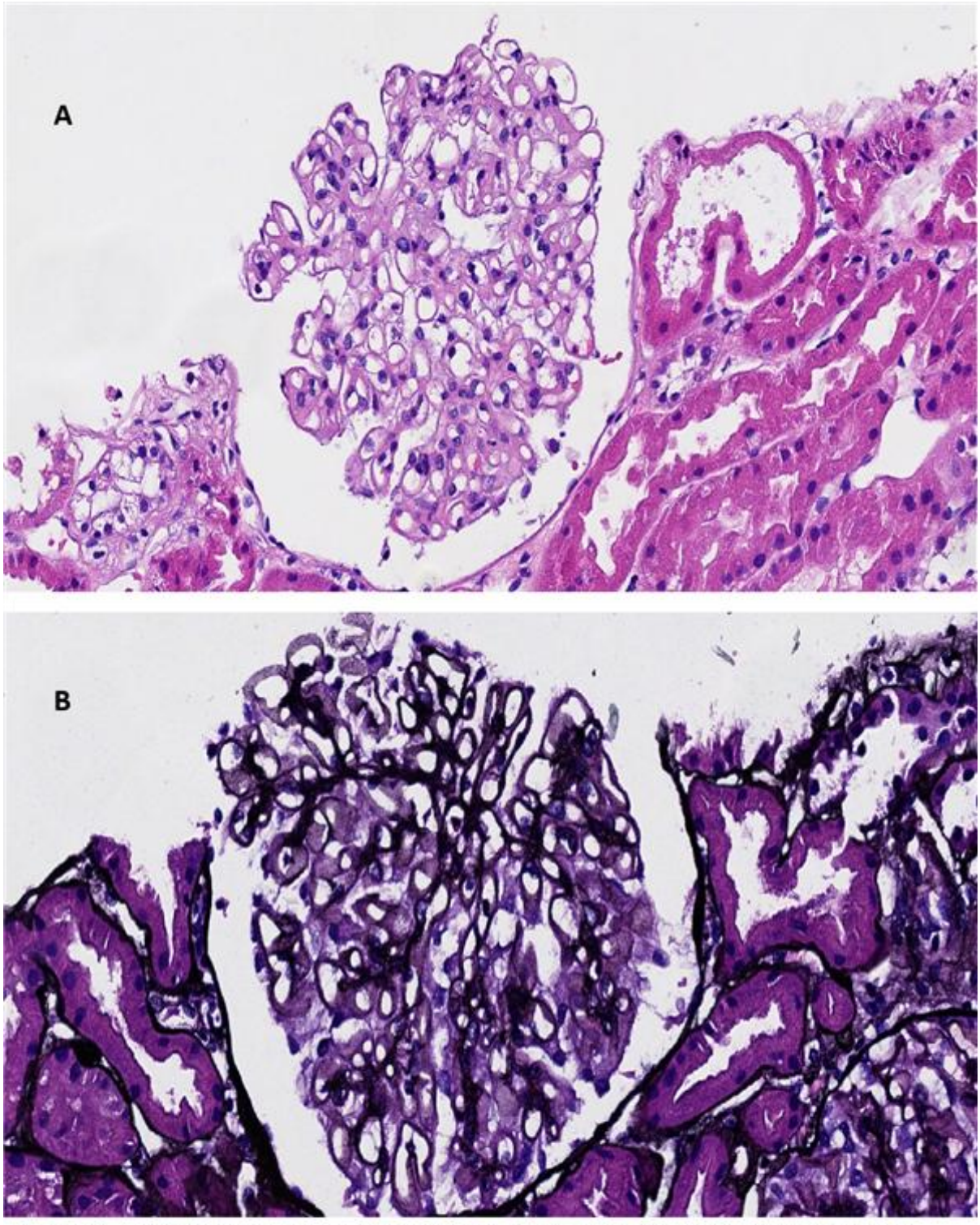

Figure 1. (A) Hematoxylin and eosin (H&E) stain showing diffuse thickening of the glomerular capillary walls without significant hypercellularity. (B) Silver stain (Jones methenamine silver) demonstrating characteristic basement membrane "spikes," consistent with Membranous nephropathy.

PO 94
AN UNEXPECTED CAUSE OF NEPHROTIC SYNDROME IN A KIDNEY-PANCREAS TRANSPLANT RECIPIENT

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Background

Post-transplant nephrotic syndrome may arise from diverse causes and poses a diagnostic challenge, particularly in the late post-transplant period. The differential diagnosis includes recurrent primary disease, transplant-specific complications, and de novo glomerulopathy. We report a case of nephrotic syndrome developing after kidney-pancreas transplantation.

Case description

A 38-year-old woman presented with progressive edema and weight gain, seven years after simultaneous kidney-pancreas transplantation.

Her medical history included type 1 diabetes complicated by retinopathy, neuropathy, peripheral artery disease, and presumed diabetic nephropathy leading to end-stage kidney disease (ESKD). She had started dialysis eight years earlier due to ESKD with nephrotic syndrome, without hematuria. Native kidney biopsy was not performed. She had been treated with hemodialysis for two months followed by peritoneal dialysis for thirteen months before successful transplantation. Post-transplant follow-up was uneventful, with preserved kidney and pancreatic graft function, under maintenance immunosuppression with tacrolimus, mycophenolic acid, and prednisolone.

At presentation, laboratory evaluation revealed new-onset nephrotic range proteinuria (urinary protein-to-creatinine ratio (UPCR) 5.1 g/g), hypoalbuminemia (3.1 g/dL), microscopic hematuria, and impaired kidney graft function (serum creatinine 2.1 mg/dL from a baseline of 1.0 mg/dL). Pancreatic graft function remained preserved.

Kidney graft biopsy revealed glomeruli with lobular appearance, endocapillary proliferation, glomerular basement membrane thickening, and capillary pseudothrombi. No peritubular capillaritis was noted. Immunofluorescence studies showed glomerular deposition of IgM, C1q, C4c, C4d, and both kappa and lambda light chains. These findings supported the diagnosis of immune complex (IC)-mediated membranoproliferative glomerulonephritis (MPGN).

Extensive evaluation for secondary causes, including autoimmune disease, infection, monoclonal gammopathy, and neoplasia, was negative. Importantly, both donor-specific antibodies (DSA) and cryoglobulins were absent. No underlying etiology was identified, and a diagnosis of idiopathic IC-mediated MPGN was made.

Given the development of nephrotic syndrome despite standard maintenance immunosuppression, rituximab was initiated (1 g administered twice, two weeks apart).

At one-month follow-up, the patient is asymptomatic with improved proteinuria (UPCR 2.3 g/g) and stable kidney function (serum creatinine 1.9 mg/dL).

Discussion

Nephrotic syndrome developing years after transplantation raises a broad differential diagnosis, and kidney graft biopsy plays a pivotal role.

Recurrent diabetic nephropathy was unlikely due to preserved pancreatic graft function. Although the native kidney disease was not biopsy-proven, post-transplant recurrence of MPGN typically occurs earlier.

Chronic antibody-mediated rejection can present as late-onset post-transplant nephrotic syndrome, even in the absence of prior acute rejection episodes or detectable DSA. Other transplant-associated causes of proteinuria include viral infection and drug-related toxicity.

De novo glomerulopathies, such as podocytopathies, membranous nephropathy, or MPGN, are rare but recognizable entities in kidney grafts. MPGN warrants a comprehensive evaluation for secondary causes, but this case illustrates that despite extensive work-up, an idiopathic form may be diagnosed.

Importantly, de novo glomerulopathy can occur late after transplantation, even under adequate immunosuppression, and may require escalation of therapy. Early recognition and accurate histopathological characterization are crucial for guiding management and preserving graft function

PO 95
EDEMA AND HYPERTENSION: THE IMPORTANCE OF DIFFERENTIAL DIAGNOSIS

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Category: Glomerular diseases

Background: Leukodermic male, 71-year-old, autonomous in normal daily living activities. He has a history of essential hypertension, obesity, and is an active smoker (83 pack-years). He has no medical follow-up, is non-adherent to medication and his vaccination schedule is outdated.

Case description: The patient presented with hypertension, headache, progressively worsening bilateral and symmetrical lower limb edema, orthopnea, and fatigue with minimal exertion, with a 5-day history. The patient also reported almost daily use of nonsteroidal anti-inflammatory drugs (NSAIDs). Laboratory tests revealed acute kidney injury with creatinine of 3.93 mg/dL, NTproBNP 3155 pg/mL, albumin 2.1mg/dL and a slight increase in inflammatory parameters (such as C-reactive protein 3.79 mg/dL). Urinalysis showed +++ proteinuria (200-400 mg/dL), + hematuria, + leukocyturia, a protein/creatinine ratio of 13,693 mg/g and an albumin/creatinine ratio of 10,180 mg/g. Imaging studies (electrocardiogram, transthoracic echocardiogram and chest X-ray) had no alterations. Also, serum protein electrophoresis and immunological studies (in particular anti human immunodeficiency virus (HIV) antibodies) were performed with no abnormal results. Renal and bladder ultrasound showed aspects suggestive of medical nephropathy, namely slight attenuation of parenchymal sinus differentiation. During hospitalization, there was progressive worsening of peripheral edema, dyspnea, which requiring oxygen therapy, and creatinine levels (maximum of 7.7 mg/dL), leading to the start of renal replacement therapy and a request for collaboration with Nephrology. A biopsy was performed, revealing focal segmental glomerulosclerosis (FSGS) with a collapsing pattern and acute interstitial nephritis. Corticosteroid therapy was initiated, resulting in improved renal function and inflammatory parameters.

Discussion and Learning points: FSGS is a nephropathy that primarily affects podocytes, characterized by glomerular sclerosis and severe nephrotic syndrome. The collapsing variant of this disease is the most aggressive and severe form, and the one with the worst prognosis. It is characterized by the collapse of glomerular vessels. And is associated with viral infections, namely SARS-CoV-2 and HIV, may have a genetic cause (mutation in the APOL1 allele) or an idiopathic one. The most common clinical presentation is massive nephrotic syndrome and rapidly developing renal failure. The prognosis depends essentially on rapid diagnosis and rapid commence of therapy, although it remains guarded due to corticosteroid resistance. This clinical case highlights the importance of differential diagnosis and management of patients with

peripheral edema and hypertension, which may indicate a chronic disease that progressively leads to renal failure.

Keywords: Focal segmental glomerulosclerosis, Collapsing focal segmental glomerulosclerosis

PO 97
DIAGNOSTIC CHALLENGES IN RECURRENT FSGS: FROM NATIVE KIDNEY DISEASE TO POST-TRANSPLANT RECURRENCE

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Category: Glomerular diseases

Background: Focal segmental glomerulosclerosis (FSGS) is a histopathologic pattern of injury that manifests as nephrotic syndrome and can be primary, genetic, secondary or of unknown cause. Etiology determines treatment, prognosis, and recurrence risk after kidney transplantation, that can reach 55% in primary FSGS, making it a major cause of graft loss.

Case description: We report the case of a white male patient who presented with nephrotic syndrome at age 35, with no family history of kidney disease. Kidney biopsy showed diffuse foot process effacement, supporting a diagnosis of primary FSGS, and treatment with high dose corticosteroids was initiated. The disease was resistant to steroids and calcineurin inhibitors, raising doubts about its primary etiology. Secondary causes were excluded and genetic testing revealed a heterozygous ACTN4 variant (c.1154A>G), also present in his asymptomatic mother and classified as non-pathogenic after expert review.

The patient maintained nephrotic range proteinuria and progressed to end-stage kidney disease 7 years after diagnosis. At 45 years of age, he underwent deceased donor kidney transplantation, with 3 human leucocyte antigen mismatches (A1, B1, DR1) and without donor specific HLA antibodies. Induction immunosuppression included basiliximab, and maintenance therapy tacrolimus, mycophenolate mofetil and prednisolone. At four months post-transplant, due to discordant graft function and nephrotic-range proteinuria, a graft biopsy was performed revealing acute antibody-mediated rejection (AMR) with no FSGS lesions on light microscopy, but >80% of foot process effacement in electron [LS1] microscopy. Anti-nephrin antibodies were negative. Treatment with high dose corticosteroids, plasmapheresis, rituximab and intravenous immunoglobulin was promptly initiated, but plasmapheresis had to be interrupted due to severe hemorrhagic complications. Partial response was achieved, with a minimal proteinuria of 1.8 g/g creatinine and a stable serum creatinine around 2 mg/dl.

At one-year post-transplant, the patient presents stable graft function but persistent nephrotic range proteinuria, despite treatment with enalapril, dapagliflozin and finerenone. Due to persistent severe leucopenia, rituximab was withdrawn and micofenolate mofetil was transiently replaced with everolimus, without worsening proteinuria.

Discussion and learning points: The diagnosis of primary FSGS remains difficult to establish with certainty, especially when resistance to treatment is observed. In this case, only after early post-transplant recurrence was the etiology confirmed. In this patient with poor native kidney disease control, post-transplant recurrence treatment was further limited by immunosuppression related adverse reactions, resulting in early chronic graft dysfunction despite a favorable histocompatibility profile. Other highlights from this case include:

1. Genetic variants in FSGS require careful interpretation, as they may not influence clinical course.
2. High risk of recurrence of primary FSGS post-transplant, requiring early and frequent monitoring of proteinuria.

3. Recurrent FSGS and AMR may coexist and should both be considered in post-transplant proteinuria.
4. Plasmapheresis may be a key component of both AMR and recurrent FSGS treatment.

PO 98
Immunoglobulin A nephropathy secondary to cirrhosis presenting as nephrotic syndrome with acute kidney injury

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Background: Secondary immunoglobulin A (IgA) nephropathy associated with chronic liver disease may present with glomerular haematuria, nephrotic-range proteinuria and acute kidney injury (AKI), mimicking other causes of AKI.

Case description: A 67-year-old man with essential hypertension, type 2 diabetes mellitus, atrial fibrillation, heart failure with preserved ejection fraction of unclear aetiology, Child–Pugh B alcoholic liver cirrhosis and multifocal hepatocellular carcinoma, on first-line palliative sorafenib 200 mg twice daily since January 2025, presented to the emergency department on 1 April 2026 with progressive peripheral oedema, scrotal oedema and abdominal distension. Clinical examination revealed anasarca, ascites and non-oliguric AKI, with a serum creatinine of 2.3 mg/dL and metabolic acidosis, compared with a baseline creatinine of 1.2 mg/dL in March 2026. He was afebrile, normotensive and had a normal heart rate, with no neurological, respiratory or cardiovascular dysfunction. Inflammatory markers were not elevated. Renal and bladder ultrasound showed preserved kidneys, with no hydronephrosis. Urinalysis revealed proteinuria of 300 mg/dL, approximately 200 erythrocytes/µL and 15 leucocytes/µL; urinary sediment examination showed numerous non-squamous epithelial cells, with no other elements, and dysmorphic erythrocytes were present. The initial urine culture was negative. Ascitic fluid analysis showed a serum–ascites albumin gradient greater than 1.1, with no evidence of spontaneous bacterial peritonitis.

Liver assessment demonstrated progressive hypoalbuminaemia, from 3.2 g/dL in 2025 to 2.8 g/dL in March 2026 and 2.4 g/dL during admission, mild cholestasis, thrombocytopenia of 86 × 10?/L and a normal INR. Despite withdrawal of diuretics and administration of albumin, renal function deteriorated. Spot urine quantification revealed a protein-to-creatinine ratio of 31 g/g and an albumin-to-creatinine ratio of 18 g/g. Further investigation showed elevated serum IgA, 718 mg/dL, mildly reduced C3, 88.9 mg/dL, and normal C4. Immunological testing was negative, including rheumatoid factor, ANA, ANCA and anti-GBM antibodies. No monoclonal peak, abnormal free light-chain ratio, haptoglobin consumption or LDH elevation was detected. HBV, HCV and HIV markers, blood cultures and ascitic fluid culture were negative.

In view of a possible sorafenib-associated glomerulopathy, sorafenib was discontinued, without renal recovery. Computed tomography showed no progression of hepatocellular carcinoma. Renal biopsy revealed IgA nephropathy with a membranoproliferative pattern, mesangial and endocapillary proliferation, double contours, three cellular crescents in 18 glomeruli, mild interstitial fibrosis/tubular atrophy, and mesangial and capillary wall deposits of IgA and C3c. The presentation was interpreted as IgA nephropathy secondary to cirrhosis, with no apparent relationship to the drug or to tumour progression. Owing to progressive azotaemia and anasarca refractory to maximised diuretic therapy, the patient was started on a regular haemodialysis programme.

Discussion: This case illustrates the multiplicity of causes of AKI in patients with cirrhosis. Nephrotic-range proteinuria with albumin predominance and dysmorphic microhaematuria point towards glomerular involvement, making renal biopsy crucial to distinguish cirrhosis-associated secondary IgA nephropathy from sorafenib toxicity, paraneoplastic glomerulopathy or thrombotic microangiopathy. This entity may display a membranoproliferative pattern, follow a progressive course and culminate in chronic haemodialysis, adversely affecting vital prognosis. No specific

treatment has been established, and immunosuppression remains particularly controversial owing to the infectious risk in this population.

PO 19
Predictors of Renal Replacement Therapy, Infections, and Mortality in ANCA-Associated Vasculitis: A Single-Center Cohort Study

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Background
Antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis comprises a heterogeneous group of disorders frequently involving the kidneys. Outcomes may be influenced not only by treatment strategies but also by patient characteristics and disease severity. This study aimed to evaluate clinical predictor factors associated to renal replacement therapy (RRT), infectious complications, and mortality.

Materials and Methods
This retrospective cohort study included patients admitted to the Nephrology Department of ULS Médio Tejo with ANCA-associated vasculitis between January 2019 and December 2025. Data collected included demographics and laboratory parameters; comorbidities; immunosuppression (IS) therapy (cyclophosphamide, rituximab, or Rituxvas), RRT requirement during hospitalization, infectious complications within the first year after diagnosis, and mortality at 12 and 24 months.

Continuous variables showed non-normal distribution. Categorical variables were compared using the Chi-square or Fisher's exact test (n<5), and continuous variables using the Mann–Whitney U test. Binary logistic regression was performed to identify independent predictors of RRT, infections and mortality.

Results
Twenty-seven patients were included (mean age 69.8±12.2 years; 52% female), with predominance of MPO-ANCA (89%, n=24). At admission, 22% (n=6) had pre-existing CKD, with a mean baseline creatinine of 1.72 mg/dL.
IS included cyclophosphamide (59%; n=16), Rituxvas (19%, n=5), rituximab (7%; n=2), and no IS (15%, n=4).
During hospitalization, RRT was required in 41% (n=11) of patients, with partial renal recovery (PRR) in 45% (n=5) of these. RRT requirement was associated with DM (p=0.048), and cyclophosphamide use with PRR (p=0.040); however these association did not remain significant in multivariable logistic regression.
Infections occurred in 41% (n=11) of patients within the first year, 91% (n=10) requiring hospitalization. Cyclophosphamide was associated with infections in univariable analysis (p=0.045), but not after multivariable analysis. In contrast, Rituxvas regimen emerged as a predictor of infectious complications (OR 27.9; 95% CI 1.34–580; p=0.003).
Mortality was 3% (n=1) at 12 months and 11% (n=3) at 24 months. Age ≥85 years was associated with mortality at 24 months (p=0.0049); however, no independent predictors of mortality were identified in multivariable analysis.

Discussion and conclusion

In this cohort, ANCA-associated vasculitis was characterized by high rates of RRT requirement and infectious complications. Notably, the Rituxvas regimen was independently associated with infections, suggesting a potential impact of treatment intensity on infection risk. Although these findings should be interpreted with caution due to the small sample size, they highlight the importance of careful patient selection and close monitoring when using immunosuppressive therapy. Larger studies are warranted to further clarify the impact of treatment strategies on clinical outcomes.

PO 22
PERFORMANCE OF THE MAYO CLINIC MGRS PREDICTION TOOL IN A REAL-WORLD MULTICENTRE PORTUGUESE COHORT

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Background

A substantial proportion of patients with monoclonal gammopathy (MG) and chronic kidney disease (CKD) have renal impairment unrelated to monoclonal gammopathy of renal significance (MGRS). Although abnormal serum free light chain ratio, higher proteinuria, and hematuria are independent predictors of MGRS, kidney biopsy (KB) remains the gold standard for diagnosis. Identifying patients most likely to benefit from this invasive procedure is challenging, particularly when relative contraindications such as obesity or the need for interruption of anticoagulation are present.

The Mayo Clinic MGRS prediction tool was developed to estimate the probability of MGRS using clinical and laboratory parameters. In this study, we aimed to assess its performance in a real-world multicentre Portuguese cohort. To our knowledge, this is the first evaluation of this tool in a European population.

Materials and Methods

We conducted a retrospective multicentre study including adults who underwent native KB in the context of MG and kidney failure or proteinuria across three Portuguese centres between January 2015 and June 2025. MG was defined by a positive serum protein electrophoresis, serum immunofixation, or an abnormal free light chain ratio. Patients with haematological malignancies requiring treatment or with missing data necessary to apply the prediction tool were excluded.

The Mayo Clinic MGRS prediction tool was applied to each patient to estimate the probability of biopsy-proven MGRS.

Continuous variables were assessed for normality using the Shapiro–Wilk test and are presented as mean ± standard deviation or median and interquartile range (IQR). Comparisons between groups were performed using the independent samples t-test for normally distributed variables and the Mann–Whitney U test for non-normally distributed variables. Categorical variables are presented as counts and percentages and were compared using the chi-square test or Fisher's exact test. Statistical analyses were performed using jamovi (version 2.6.44). A p-value < 0.05 was considered statistically significant.

Model discrimination was assessed using the area under the receiver operating characteristic curve (AUC). Diagnostic performance was evaluated at predefined probability thresholds of 0.10 and 0.25, by calculating sensitivity, specificity, positive predictive value (PPV), and negative predictive value (NPV).

Results

We identified 37 patients, 13 (35.1%) had MGRS and 24 (64.9%) did not. AL amyloidosis (n= 8; [61.5%]) was the most common lesion in the MGRS group and diabetic nephropathy (n= 7; [29.1%]) in the non-MGRS group. Histological diagnoses are presented in **Table 1**. There were no significant differences in age, sex, blood pressure, or kidney function between groups. However, diabetes was significantly more frequent in the non-MGRS group (p= 0.003).

The model demonstrated good discriminative ability, with an AUC of 0.82. The prediction tool was strongly associated with MGRS (OR 83.6, 95% CI 4.8–1458, p = 0.002). At a probability threshold of 0.10, sensitivity was 92.3% and specificity was 54.2%, with a PPV of 52.2% and a NPV of 92.9%. At a threshold of 0.25, sensitivity was 69.2% and specificity was 75.0%, with a PPV of 60.0% and an NPV of 81.8%.

Discussion and Conclusion

In this multicentre Portuguese cohort, the Mayo Clinic MGRS prediction tool demonstrated good discriminative performance, consistent with the results from the external validation cohort from the University of Minnesota. Although sensitivity was lower in our population, specificity was similar or slightly higher.

At the 0.10 threshold, the model showed high sensitivity and NPV, supporting its potential role in identifying patients at low risk of MGRS, enabling a rule-out strategy.

Our results suggest the tool may be useful for clinical risk stratification and biopsy prioritization. Results should be interpreted in light of the retrospective design and small sample size.

PO 25
REAL-WORLD EXPERIENCE WITH TOLVAPTAN IN ADPKD AT A TERTIARY CENTER: TRANSLATING EVIDENCE INTO PRACTICE

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Background

Autosomal dominant polycystic kidney disease (ADPKD) is the most prevalent genetic kidney disease leading to kidney failure. Tolvaptan, a vasopressin V2 receptor antagonist, slows kidney function decline and kidney growth in patients with rapidly progressive disease. However, its implementation in clinical practice is challenging, requiring careful selection, dose titration and monitoring for aquaretic effects and hepatotoxicity. Real-world data on early treatment experience remain limited, particularly regarding tolerability, dose optimization and short-term persistence.

Materials and Methods

We conducted a retrospective, observational study of the initial experience with tolvaptan in 10 patients with ADPKD at a tertiary referral center. Baseline data at treatment initiation included demographic, clinical, laboratory and imaging characteristics (Mayo Imaging Classification). Safety monitoring followed a strict protocol: twice-monthly kidney and liver function and electrolyte tests during the first month, followed by monthly follow-up. Treatment variables included starting dose, titration speed, maximum dose achieved and reasons for dose limitation. All adverse events

were systematically recorded, with a focus on aquaretic symptoms and their impact on daily functioning.

Results

A total of 10 patients with ADPKD receiving tolvaptan were analyzed with a mean age of 43.3 years and 60% were female. Mean treatment duration was 7.1 weeks (range: 4-16 weeks). All patients met established criteria for rapidly disease progression, based on both Mayo Imaging Classification and a historical estimated glomerular filtration rate (eGFR) decline >5 mL/min/1.73 m²/year. Mean baseline eGFR was 40.9 mL/min/1.73 m² and mean total kidney volume was 2534.2 mL. All patients initiated tolvaptan at a dose of 45/15 mg, followed by stepwise titration. Two patients (20%) achieved the maximum dose (90/30 mg) within 4-6 weeks, while the remaining patients (80%) were maintained on an intermediate dose (60/30 mg), with further up-titration limited by non-aquaretic symptoms, mainly asthenia and gastrointestinal intolerance. Aquaretic symptoms were universal but did not limit dose escalation. No cases of hepatotoxicity, electrolyte disturbances or significant hyperuricemia were observed. Treatment persistence was 100%, with no dose reductions or interruptions.

Discussion and conclusion

In this real-world cohort, early experience suggests that initiation of tolvaptan is feasible and associated with high treatment persistence, despite expected aquaretic symptoms. While these effects were consistently reported, they did not represent a major barrier to treatment continuation or dose escalation. Instead, non-aquaretic symptoms, particularly asthenia and gastrointestinal intolerance, were the main factors influencing dose optimization, leading most patients to stabilize at intermediate doses (60/30 mg). These findings highlight the importance of individualized titration strategies and careful clinical monitoring in routine practice.

Key-words: Autosomal dominant polycystic kidney disease (ADPKD), Tolvaptan, Real-world evidence.

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Long-Term Outcomes and Predictors of Kidney Disease Progression of a Pre-Pandemic Biopsy-Proven Lupus Nephritis Cohort (2013–2019) from Northern Portugal

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Background: Lupus nephritis (LN) is one of the most frequent and severe manifestations of systemic lupus erythematosus and remains a major predictor of poor prognosis. Despite advances in LN management, a substantial proportion of patients remain at risk of disease progression and adverse outcomes. This study aimed to characterize the clinical and laboratory features of a cohort of LN patients from a major Portuguese university hospital and to gain insights into long-term outcomes and risk factors for kidney disease progression.

Methods: We conducted a retrospective observational cohort study of patients with biopsy-proven LN diagnosed between 2013 and 2019, with follow-up through May 2025. Cases were identified from the institutional Renal Biopsy Registry. Demographic, clinical, and laboratory data were extracted from electronic medical records. Kidney outcomes included evolution of proteinuria, rate of kidney function decline assessed by estimated glomerular filtration rate (eGFR), incidence of end-stage kidney disease (ESKD), and all-cause mortality. Kidney disease progression was defined as an annualized decline in eGFR ≥3 mL/min/1.73 m². Multivariable logistic regression analysis was performed to identify predictors of eGFR decline.

Results: A total of 52 patients were included (84.6% female; mean age at kidney biopsy of 35.6 ± 13.5 years). Class IV was the predominant histopathological pattern (63.5%), followed by class V (11.5%) and class III (9.6%). Thirty-four patients (65.4%) were newly diagnosed with LN, while 18 (34.6%) underwent repeat biopsy during the study period, most commonly due to lupus flare (83.3%). At baseline, the mean eGFR was 92.1 ± 40.5 mL/min/1.73m², and proteinuria exceeded 1 g/day in 78.8% of patients, of whom 36.6% presented with nephrotic-range proteinuria. One year after treatment initiation, 57.7% of patients achieved a complete renal response according to EULAR/ERA-EDTA criteria, while the remainder achieved partial response. Over a median follow-up of 7.8 years, proteinuria progressively declined: 69.2% achieved levels <0.5 g/day, 21.1% had proteinuria between 0.5–1.0 g/day, and 9.6% maintained nephrotic-range proteinuria at last follow-up. Kidney disease progression occurred in 15.4% (n=8), three patients (5.8%) developed ESKD, and two patients (3.8%) died. In multivariable analysis, global glomerular sclerosis, kidney impairment at presentation, and changes in proteinuria over time were independent predictors of kidney disease progression.

Conclusion: In this biopsy-proven LN cohort from a Portuguese tertiary center, the predominance of class IV disease and disease relapse suggest a selection toward more severe cases. Despite this, the majority achieved sustained proteinuria reduction and stable renal function, supporting the effectiveness of local treatment strategies. However, a clinically meaningful subset experienced kidney disease progression, underscoring that long-term renal risk remains significant even under contemporary management. Global glomerular sclerosis, baseline kidney impairment, and proteinuria trajectory were independent predictors of progression. Further studies are warranted to clarify these outcomes and inform optimized management strategies.

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Minimal change disease in adults: predictors of AKI and long-term outcomes; A 24-year bicentric cohort study

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Background Minimal Change Disease (MCD) is a rare cause of nephrotic syndrome in adults. While well described in children, **adult-onset MCD remains poorly defined**, with limited data on its presentation, prognostic markers, and therapeutic response.

Materials and Methods This retrospective bicentric cohort study was conducted in two national tertiary nephrology centers and included 45 adults with biopsy-confirmed MCD diagnosed between January 2000 and January 2025. Demographic, clinical, laboratory, and histological data were collected, along with immunosuppressive treatments and outcomes. Group comparisons were performed using t-tests with unequal variances for continuous variables and chi-square tests for categorical variables. Predictors of AKI were assessed using multivariable logistic regression. Statistical analyses were performed using Stata 19, with significance set at $p<0.05$. Continuous variables were summarized as median and IQR.

Results Median age at diagnosis was 38 years (IQR 30), with male predominance (64.4%). Most patients were of European origin (73%), followed by African origin (15.5%). Secondary causes were identified in 13.3%. At presentation, median creatinine was 0.92 mg/dL (IQR 0.78), albumin 1.7 g/dL (IQR 1.33), and proteinuria 6930 mg/day (IQR 7475). Erythrocyturia was present in 65.1%. Histology revealed chronic structural changes, including global glomerulosclerosis in 32%, tubular atrophy in 23%, interstitial fibrosis in 25%, and chronic vascular lesions in 69%. IgM deposition was present in 35% of biopsies.

AKI occurred in 44.4% of patients; 47.4% met KDIGO stage 3 and 11% required dialysis. Renal recovery occurred in 85%, although two remained dialysis-dependent. Older age (OR 1.09; 95%

CI 1.03–1.15; p=0.002) and higher proteinuria (OR 1.3676; 95% CI 1.1064-1.6904; p=0.004) independently predicted AKI.

Steroids were used in 93% of patients, with complete remission in 77.5%, partial in 12.5%, and steroid-resistance in 10%. Relapses occurred in 65%, including 15% frequently relapsing and 35% steroid-dependent patients. Steroid-related complications occurred in 58.5%. Rituximab was used in 31%, achieving complete remission in all, though 58% relapsed. Calcineurin inhibitors were used in 38% and mycophenolate in 11%. **Median follow-up was 6.08 years (IQR 8.70).** Renal function remained stable in most patients and only two progressed to CKD KDIGO ≥3. Four patients died.

Discussion and Conclusion In this bicentric national cohort, adult MCD frequently presented with severe nephrotic syndrome and a high incidence of AKI. Age and proteinuria were independent predictors of AKI, underscoring the relevance of early risk stratification. Although remission rates with steroids and rituximab were high, relapses were common and steroid toxicity substantial, highlighting the limitations of a steroid-centric approach. Despite recurrent disease activity, long-term renal outcomes were generally favorable, with only a minority progressing to advanced CKD. These findings refine the characterization of adult MCD and support more individualized therapeutic strategies.