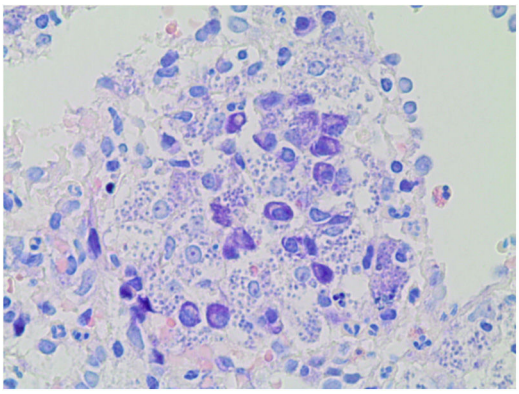

Fig. 2 – Splenic biopsy, Giemsa staining. Microorganisms corresponding to *Leishmania* with rounded or oval morphology inside the cytoplasm of macrophages/histiocytes.

and renal biopsy was performed at the same time, with the splenic biopsy providing the diagnosis of leishmaniasis and the renal biopsy the diagnosis of immune-mediated glomerulonephritis associated with this infectious disease. In a patient with constitutional symptoms, pancytopenia and renal lesion, clinician's should consider leishmaniasis. The rapid initiation of specific therapy with amphotericin B for opportunistic infection by *Leishmania* led to a good evolution of the patient.

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The need for renal biopsy in oncology patients on check-point inhibitors check-point inhibitors: New triggers for extracapillary glomerulonephritis extracapillary glomerulonephritis

La necesidad de la biopsia renal en paciente oncológico con inhibidores de check-point: nuevos trigger para glomerulonefritis extracapilar

Dear Editor,

Checkpoint inhibitors (CPI) have revolutionised cancer treatment. They act by inhibiting T-lymphocyte (TL) receptors, cytotoxic T-lymphocyte antigen 4 (CTLA4) or programmed cell death 1 (PD1) receptors or their ligands, triggering TL dysregulation and hyperactivation, which causes immune-related adverse events (irAE). In the kidneys, the most common is immune-mediated interstitial nephritis, but there is also

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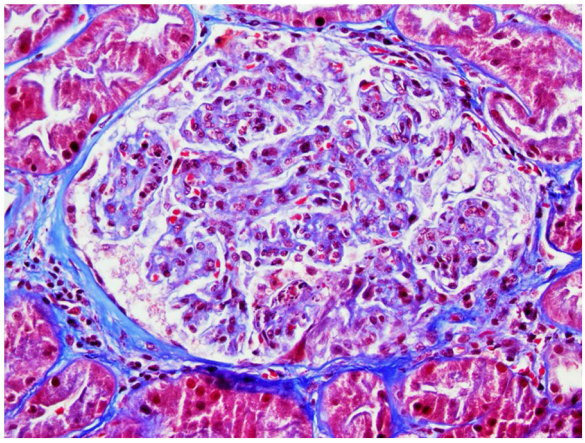

Fig. 1 – Masson's trichrome. Glomerulus with global endocapillary hypercellularity, leucocytoclasia and red cell fragmentation in the capillary tuft close to the vascular pole.

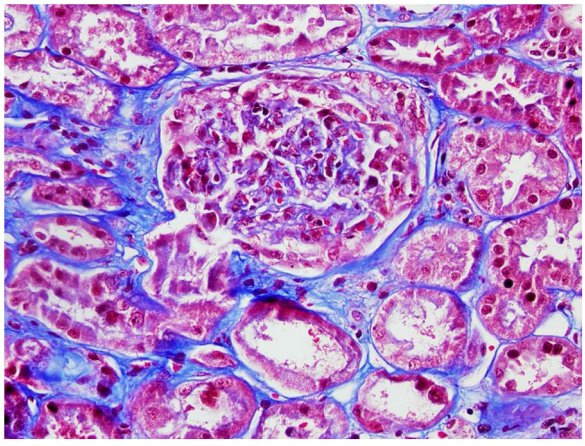

Fig. 2 – Masson's trichrome. Glomerulus with epithelial crescent.

pauci-immune glomerulonephritis, podocytopathies and C3 glomerulopathy, which worsen the prognosis.[1–6] We present a case of pauci-immune glomerulonephritis (PIGN) during treatment with durvalumab, antiPD-L1, in a patient with squamous cell carcinoma of the lung.

This was a 71-year-old male with a history of hypertension, type 2 diabetes, dyslipidaemia and squamous cell carcinoma of the lung (G2 pT3 pN1, PD-L1 < 1%) diagnosed in 2018; surgical treatment and chemotherapy with four cycles of cisplatin and vinorelbine. Maintenance treatment with durvalumab, 12 cycles, until 14/11/2019, remaining disease free (Figs. 1 and 2).

On 17/11/2019 he was admitted for acute renal failure; creatinine 4.5 mg/dl, microhaematuria (300 red blood cells/μl), clinical and analytical nephrotic syndrome (proteinuria 6 g/day, albumin 2.2 mg/dl). Immunological study normal/negative; renal biopsy: 21 glomeruli, 6 globally sclerotic. Inflammatory hypercellularity in capillary lumens, mononucleated, focally neutrophils, fragmented red blood cells and leucocytoclastic phenomenon. Four glomeruli with epithelial crescents. Interstitial fibrosis, tubular atrophy and mild chronic inflammation. The arteries and arterioles appeared normal. Direct immunofluorescence no IgA, IgG, IgM, C3, C1q, kappa, lambda deposits. Diagnosis: pauci-immune glomerulonephritis with extracapillary proliferation in 27% of glomeruli. He was started on intravenous steroids (500 mg × 3 days) orally at a dose of 1 mg/kg/day and adjusted intravenous cyclophosphamide (500 mg/m^2).

After one month, outpatient follow-up with oral steroids and cyclophosphamide; creatinine 3–3.5 mg/dl, CKD-EPI 16.5–18 ml/min, nephrotic proteinuria and microhaematuria.

Readmission after second bolus of cyclophosphamide; creatinine 7.14 mg/dl, urea 319 mg/dl and proteinuria 4.18 g/24 h without nephrotic syndrome, severe haematuria. Renal replacement therapy (RRT) was started; intermittent haemodialysis, maintaining a diuresis of 1500–2000 cc/day. Subsequently, he continued intravenous cyclophosphamide and a regular haemodialysis programme, with close follow-up. We have noted a decrease in proteinuria to <1.5 g/day, diuresis 2 l and a CrCl 12–18 ml/min with no need for RRT until now and oncological disease in complete remission.

Checkpoints are regulators of the TL immune response. Their blockade with immunotherapy promotes a state of lymphocyte dysregulation, leading to hyperstimulation of TL and better control over tumour cells. However, their main drawback is irAE, exacerbating autoimmune diseases such as PIGN. PIGN is characterised by positive serum antineutrophil cytoplasmic antibodies (ANCA), sometimes, as in our patient, they are negative, speculating that they are ANCA against another epitope or other undetected autoantibodies.[3]

PIGN has been associated with aberrant PD1 expression in some subjects and increased TL hyperactivity, with the risk of developing PIGN increased by immunotherapy treatment. In our case we could postulate that there was this aberrant expression which triggered PIGN,[7] and stopping the immunotherapy combined with immunosuppressive treatment was able to control the TL hyperactivity, which then led to remission of the PIGN.

Polymorphisms in PDCD1 (the gene that encodes PD1) have also been described, which increase susceptibility for developing PIGN; therefore, a genetic analysis could be useful to prevent the development of PIGN in patients requiring treatment with CPI and to consider other treatments.[8,9]

The close relationship between oncology and nephrology leads to early renal biopsy and assessment by nephrology, with early diagnosis increasing renal and overall survival.

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Pseudoxanthoma elasticum and hereditary renal hypouricemia: Complication of systemic disorders or different entities? Presentation of a case

Pseudoxantoma elasticum e hipouricemia renal hereditaria: ¿complicación de afección sistémica o entidades diferentes? Presentación de un caso

Dear Editor,

Pseudoxanthoma elasticum (PXE) is a rare genetic disorder characterised by fragmentation and calcification of elastic fibres in the skin and tunica media of arteries. Extracutaneous clinical manifestations are rare, the most common being hypertension, angina pectoris, stroke, intermittent claudication, upper gastrointestinal bleeding, angioid streaks in the retina and thickened skin.

Cases have been reported of possible association with other autoimmune diseases, such as systemic lupus erythematosus (SLE),[1] ankylosing spondylitis[2] and rheumatoid arthritis. Urologically, cases have been reported of ruptured ureters after ureteroscopy and there would seem to be a greater predisposition to urinary tract infections.[3]

There are no case reports linking PXE and uric acid metabolism disorders.

We present the case of a patient with PXE and hereditary renal hypouricaemia (HRH).

This was a 63-year-old woman under follow-up by Nephrology with a history of lupus nephritis class IV and mixed cryoglobulinaemia type III. No renal lithiasis. No visual problems. She first developed asymptomatic yellowish papular lesions on her neck. Suspected PXE was confirmed by skin biopsy and genetic study, by the presence of heterozygous mutation c.3662G>A (p.R1221H) in the ABCC6 gene.

In addition, analytical tests showed alterations in uric acid metabolism. The most recent test showed uricaemia 2.3 mg/dl, fractional excretion of uric acid (FEUa) 12.95%, proteinuria 0.14 g/24 h and normal urine sediment. No glycosuria or hypercalciuria. Normal acid–base balance.

After obtaining informed consent, a genetic study was requested which showed a heterozygous (+) mutation of the pathogenic variant c.1400C>T (p.T467M) in the SLC22A12 (URAT1) gene.

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