

Kidney involvement by IgG4-related sclerosing disease

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A 54-year-old Caucasian woman was admitted due to persistent febrile syndrome, sweating and abdominal pain. Fever marked the onset of the patient's medical history 10 years earlier. The physical examination revealed normal findings.

Laboratory data showed normal white blood cell count, mild microcytic anaemia with a normal reticulocyte count, important inflammatory syndrome (C-reactive protein 40 mg/L, IL-6 23 pg/mL), normal renal function and no urinary abnormalities. Autoimmune serology was negative. An abdominal computed tomography scan revealed multiple nodular masses in the right kidney associated with a large lumboaortic adenopathic mass (Figure 1). A renal biopsy showed severe interstitial fibrosis and tubular atrophy. The borders of the sclerotic lesion were heavily infiltrated by lymphocytes and plasma cells (Figure 2). The morphology and anatomic distribution of the lesions raised the differential diagnosis of tubulointerstitial nephritis versus a lymphoma or plasma cell dyscrasia (Figure 3). Immunohistochemistry and *in situ* hybridization studies (including CD3, CD20, CD68, CD138, lambda LC, kappa LC, PAX5, desmin,

ALK-1, AE1/AE3 cytokeratin, HMB-45) did not support a lymphoproliferative process. CD20 and PAX5 highlighted well-formed follicles and occasional germinal centres, while CD3 stained primarily small, mature-appearing T-cells. CD138+ plasma cells were abundant and kappa/lambda ISH showed polytypic distribution. Reed-Sternberg and/or Hodgkin cells were not identified, although focally, eosinophils were prominent. However, IgG4 immunostaining showed a marked increase in IgG4-positive plasma cells at the borders of the sclerotic lesions. We made a diagnosis of IgG4-related sclerosing autoimmune disease. Evolution was favourable following immunosuppressive therapy (prednisone and cyclophosphamide), with an important decrease in the size of the lumboaortic adenopathic mass and right kidney nodular masses, and remission of fever and of the inflammatory syndrome.

IgG4-related disease is a systemic immune-mediated disease that typically manifests as fibroinflammatory masses that can affect almost any organ system. Frequently, imaging studies of the kidney involvement

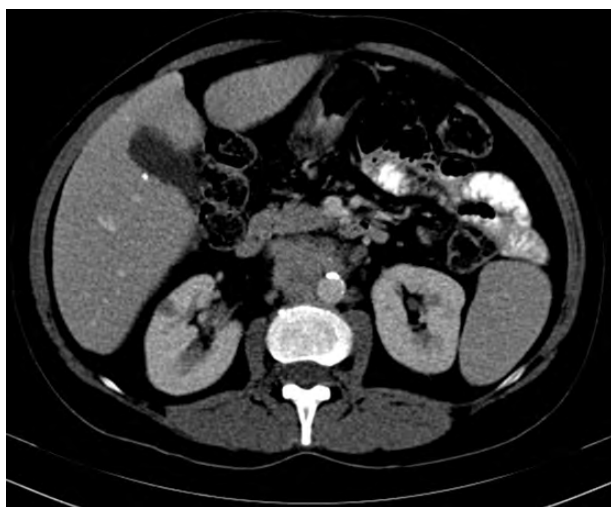


Fig. 1. Computed tomography scan of the abdomen: nodular masses in the right kidney associated with a large lumboaortic adenopathic mass.

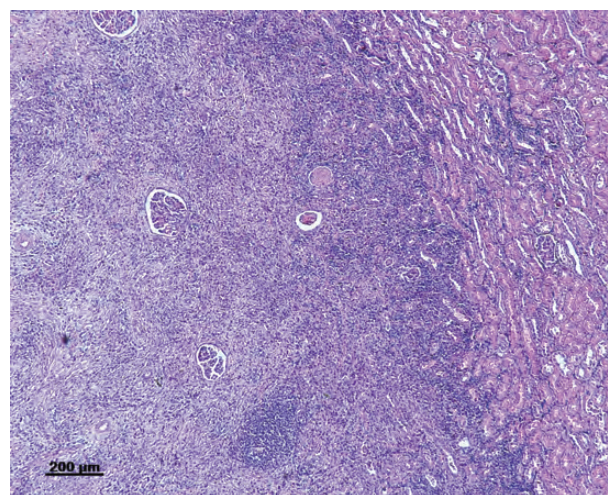


Fig. 2. Renal biopsy: a fibroinflammatory process extensively replaces normal renal interstitium. The borders of the sclerotic lesion are heavily invaded by a dense lymphoplasmacytic infiltrate. (haematoxylin–eosin stain; original magnification, $\times 20$)

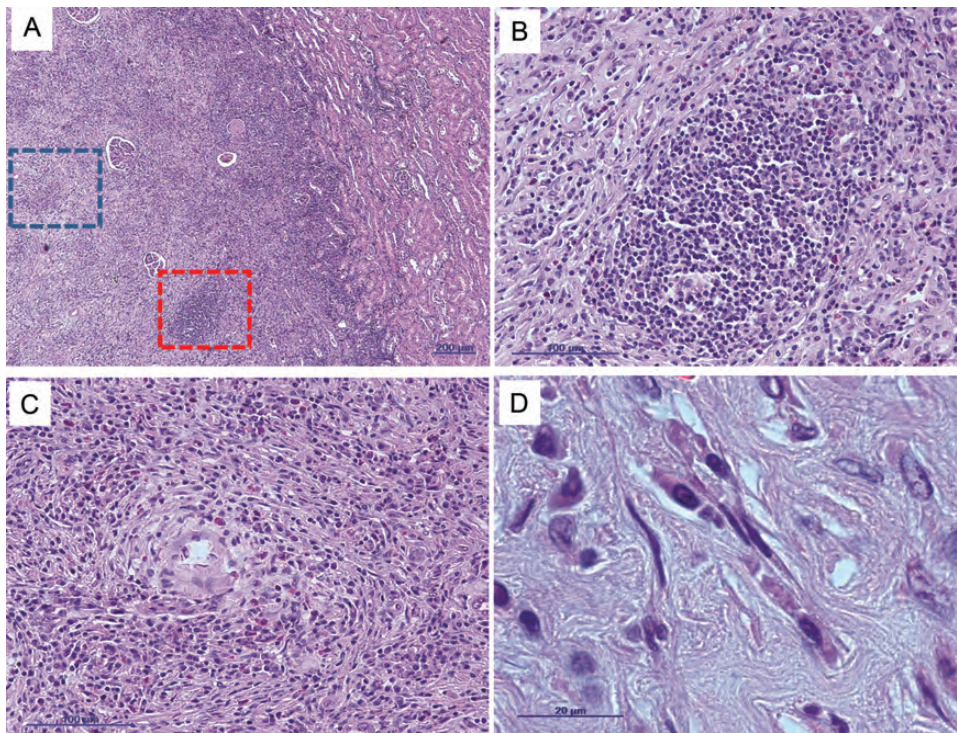


Fig. 3. (A) Normal kidney architecture (right) is replaced by severe inflammatory cell infiltration and fibrosis (left). (B) Well-formed follicles are visible in the inflammatory cell infiltration (high magnification of the red square marked area in A). (C) The prominent lymphoplasmacytic infiltrate presents a high number of eosinophils (high magnification of the blue square marked area in A). (D) Collagen fibres form large, irregular bundles between inflammatory cells, showing a whorled pattern of fibrosis (storiform fibrosis).

by IgG4-related sclerosing disease reveals singular or multiple nodular masses affecting one or both kidneys, sometimes leading to nephrectomy to exclude renal malignancy. Moreover, histopathological diagnosis is challenging because differential diagnosis with lymphoma or plasma cell dyscrasia is difficult.

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Conflict of interest statement. None declared.

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