

Title: *Tubulointerstitial nephritis associated with IgG4-Related Disease*

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Introduction: IgG4-Related Disease (IgG4-RD) is a multi-systemic fibroinflammatory condition that mimics malignancy. This report describes the case of a patient with a history of Diffuse large B-cell lymphoma (DLBCL) and a late diagnosis of IgG4-RD with important kidney involvement.

Case presentation: Male patient, 75 years old, diagnosed with DLBCL, achieving complete remission in 2009 after treatment with R-CHOP (Rituximab, Cyclophosphamide, Doxorubicin, Vincristine). Between 2012 and 2024, he developed recurrent lymphadenopathies with increased uptake on PET-computed tomography (CT). Five lymph node biopsies demonstrating only reactive alterations. In January 2025, PET-CT showed diffuse uptake in the pancreas corresponding to a 2.8 cm solid pancreatic lesion, and a pancreatic biopsy demonstrated non-neoplastic tissue. In April 2025, the patient developed prostatic symptoms, an increase in serum creatinine (Scr) (2.1 mg/dL, from a baseline of 0.8 mg/dl), and left hydronephrosis with periaortic tissue on CT. Following transurethral resection and double-J stenting, Scr improved to 1.3 mg/dL, and prostatic pathology revealed chronic prostatitis with plasmacytosis. In December 2025, the patient developed proteinuria (1.25 g/day) and a recurrent increase in Scr (2.6 mg/dL). The kidney biopsy revealed chronic interstitial nephritis with 80% storiform fibrosis and numerous IgG4-positive plasma cells (IgG4:IgG ratio 2/3, >70 cells/HPF), supporting a diagnosis of IgG4-RD (Figure), further corroborated by highly elevated serum IgG (>12,056 mg/dL). Corticosteroid therapy (prednisone 40 mg/day) was initiated in February 2026, resulting in progressive improvement of Scr (1.3 mg/dL) after 2 weeks of therapy.

Discussion: Tubulointerstitial nephritis can be observed in up to 15% of IgG4-RD cases and should be included in the differential diagnosis in the context of mass-forming lesions and hypermetabolic lymphadenopathies.