

Plasma Cell-Rich Tubulointerstitial Nephritis (TIN) without Increased IgG4 Positive Plasma Cells Resembling IgG4-TIN: a Report of 4 Cases

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Objectives:

IgG4-related tubulointerstitial nephritis (TIN) is characterized by a plasma cell-rich TIN, expansile storiform interstitial fibrosis, tubular basement membrane immune complex deposits, and an increase in IgG4 positive plasma cells. We describe a series of four patients with a typical histological appearance of IgG4-TIN but without a significant increase in IgG4 positive plasma cells.

Method and Materials:

Four plasma cell-rich TIN cases are identified that showed morphological features resembling IgG4-TIN but did not show a moderate or marked increase in IgG4 positive plasma cells. Clinical history was obtained. Pathology reports were reviewed and pathological features recorded.

Results:

The specimens consisted of 3 biopsies and 1 nephrectomy. The average patient age was 70 years (range 62 to 79) and the male to female ratio was 3:1. Three patients presented with elevated creatinine (average: 5.2 mg/dL, range: 1.9-10.2 mg/dL); one presented with kidney masses. Extra-renal involvement was identified in 3 cases: one with lesions in pancreas, retroperitoneum, retroperitoneal lymph nodes, mediastinum, and mediastinal lymph nodes; one with suspected pancreatic involvement; and one with salivary gland involvement. Histologically, three cases revealed expansile storiform fibrosis pattern; one biopsy showed acute interstitial nephritis. Immunofluorescence staining was performed on the three biopsies. All of these showed granular tubular basement staining for IgG and kappa and lambda light chains (1-3+, scale 0-3). All four cases showed <10 or no IgG4 positive plasma cells per high power field with IgG4 to IgG ratio of <40%.

Conclusion:

An increased number of IgG4-positive plasma cells may not be essential in the diagnosis of IgG4-TIN. Further understanding of the pathogenesis of IgG4-related disease may bring light to refine the current diagnostic criteria of IgG4-related disease.