Acute Kidney Injury from Isolated IgG4-related Infiltrative Kidney Disease, A Case Report

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Abstract

Background: IgG4-related disease (IgG4-RD) is a systemic disease which can involve nearly all organ. Renal involvement includes acute kidney injury from interstitial nephritis, membranous nephropathy and renal infiltrative lesion. Symptomatic IgG4-related kidney disease (IgG4-RKD) without extrarenal organ involvement is extremely rare condition.
Case report: We reported a case of 48-year-old male with isolated renal IgG4-RD presented with abdominal pain. His laboratory investigation showed acute kidney injury, proteinuria and hypoalbuminemia. Whole abdominal CT scan showed infiltrative lesions at both kidneys and ureters with hydronephrosis. His kidney biopsy revealed lymphocytic and plasma cell infiltration, 82 plasma cell/HPF, storiform fibrosis without obliterative phlebitis. After treatment with corticosteroid, he had improvement of renal function and had remission of the infiltrative lesion.
Conclusion: Isolated renal IgG4-RD is rare and can be difficult to diagnosed. The disease can be present as acute interstitial nephritis, glomerular disease, retroperitoneal fibrosis, and rarely mass or infiltrative lesions. The disease likely to response well to corticosteroid; thus, the diagnosis shall not be delayed. The delayed diagnosis may lead to organ fibrosis and dysfunction.

History

A 48-year-old Thai male presented with right side abdominal pain for 3 months. His pain was dull aching in nature and radiated to the right groin. He reported experiencing nocturia twice per night and noted an increase in foamy urine. He also had weight loss of 13 kilograms (92 Kg to 79 Kg) within 3 months**.**

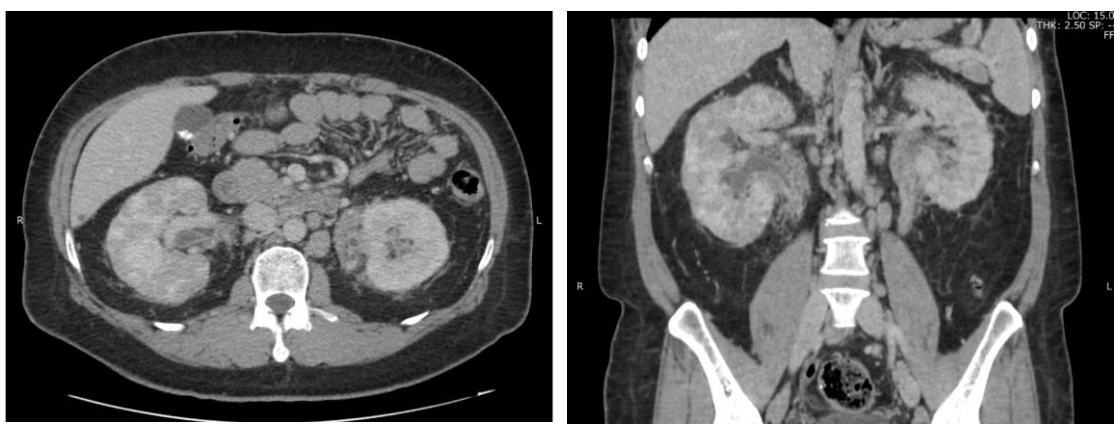

Figure 1: Post venous contrast CT whole abdomen in axial view (left) and coronal view (right) showed bilateral kidneys infiltrated with heterogenous infiltrative lesions.

Investigation

-CT abdomen was performed and found bilateral kidney infiltration (Figure 1)
-His kidney biopsy was performed and revealed interstitial infiltration with lymphocytes, IgG-4 staining plasma cells, eosinophils and some neutrophils (Figure 2)

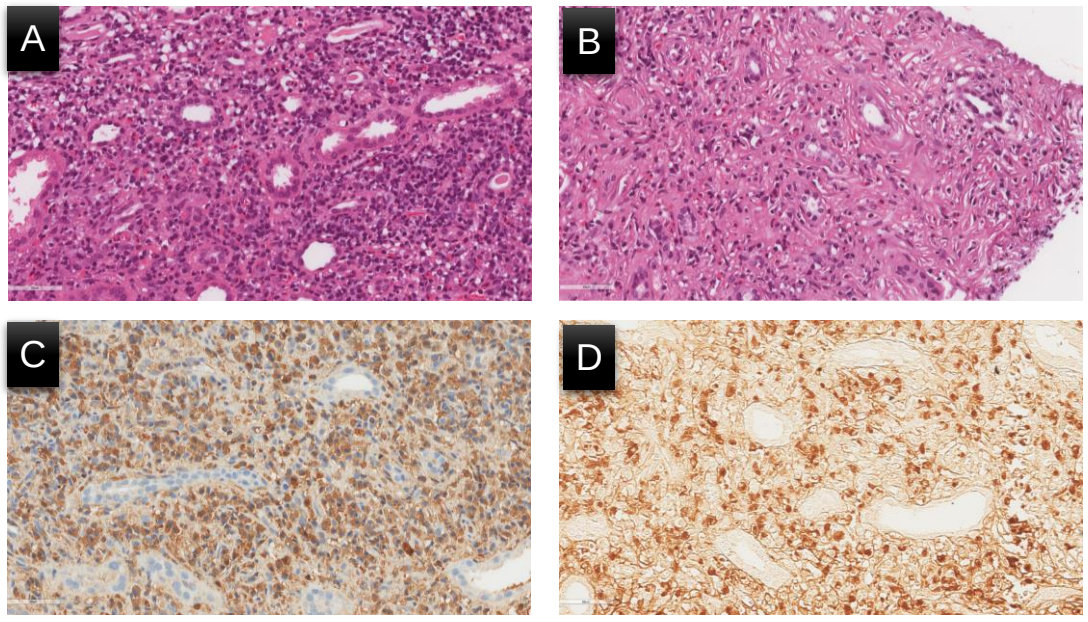

Figure 3: The kidney biopsy demonstrates interstitial infiltration with lymphocytes, plasma cells, eosinophils and some neutrophils (A, H&E 400x). Focus of storiform fibrosis is shown (B, H&E 400x). Immunoperoxidase staining of renal interstitial shows IgG staining plasma cells (C, 400x) and IgG-4 staining plasma cells (D, 400x).

Treatment

He was treated with oral prednisolone at a dosage of 40 mg/day for three months. The prednisolone dosage was gradually tapered, and azathioprine at 50 mg/day was introduced to address steroid-related side effects. His symptoms showed improvement throughout the treatment course. Within one month after initiation of treatment, his serum creatinine decreased from 1.76 mg/dL to 1.22 mg/dL, further reducing to 1.0 mg/dL within four months. Concurrently, his serum albumin increased from 3.2 mg/dL to 4.0 mg/dL three months after treatment. The serum IgG-4 levels decreased from 4,480 mg/dL to 493 mg/dL within six months, eventually normalizing at one year. A repeat CT scan of the entire abdomen at six months post-treatment revealed complete remission of kidney and ureter infiltration, as well as bilateral hydronephrosis

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