Immunoglobulin G4-related Kidney Disease as a Cause of Acute Renal Insufficiency

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Immunoglobulin G4 (IgG4)-related kidney disease is a systemic autoimmune disease which characterized by elevated serum IgG4 and dense infiltration of IgG4-positive plasma cells into tubular interstitium. It can be a mimicker of acute renal insufficiency. We herein report a rare case of IgG4-related kidney disease as a cause of acute renal insufficiency.

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INTRODUCTION

Immunoglobulin G4 (IgG4)-related disease is an immune-mediated condition characterized by infiltration of abundant IgG4-positive plasma cells and lymphocyte into various organs, such as the pancreas, gallbladder, liver, lacrimal gland, and lymph nodes.[1] Renal involvement is referred as IgG4-related kidney disease. We report a rare case of IgG4-related kidney disease as a cause of acute renal insufficiency.

CASE REPORT

A 69-year-old man was admitted for fatigue lasting for 2 weeks. Laboratory data showed acute renal insufficiency with serum creatinine gradually increased from 407 μmol/L to 711 μmol/L within 1 month. His hemoglobin was 96 g/L and proteinuria was 1+. Anti-antinuclear antibody was positive with a titer of 1:320, but anti-antineutrophil cytoplasmic antibody was negative. Serum IgG level was 26.9 g/L (reference range, 7.00 g/L to 16.00 g/L); IgG4, 19.4 g/L (reference range, 0.03 g/L to 2.01g/L); complement C3, 0.63 g/L; and complement C4, 0.15 g/L. Urinary κ chain level was 0.29 g/L and λ chain level was 0.30 g/L. No dysmorphic plasmocyte was found in bone marrow aspiration (Figure 1). Except the irregular outline of the two kidneys, no abnormality was found in the lung, liver, or pancreas when examined by computed tomographic scan (Figure 2). No bone destruction was found by flat bone radiography.

Kidney biopsy indicated infiltration of numerous

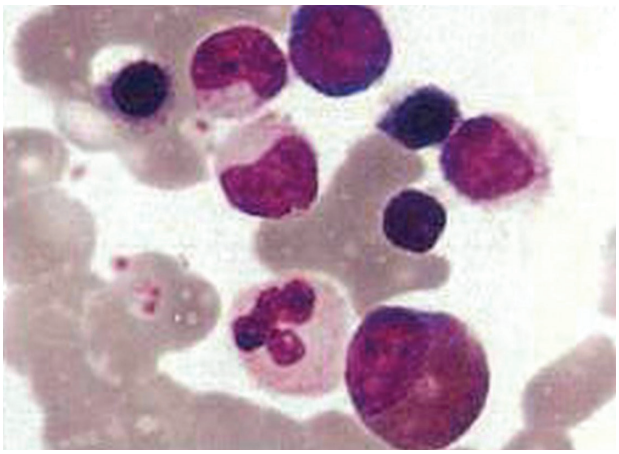

Figure 1. Bone marrow characteristics revealed by oil immersion lens. Romanowsky-Giemsa staining of bone marrow showing the plasmocyte percentage is approximately 2% to 4% without immature cells (× 100).

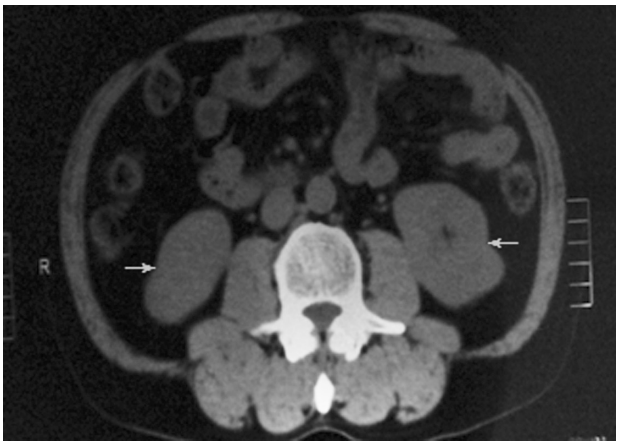

Figure 2. Imaging features of the kidneys. Abdomen computed tomography at the level of kidney exhibiting bilateral irregular outline (white arrows) of the kidneys.

Case Report

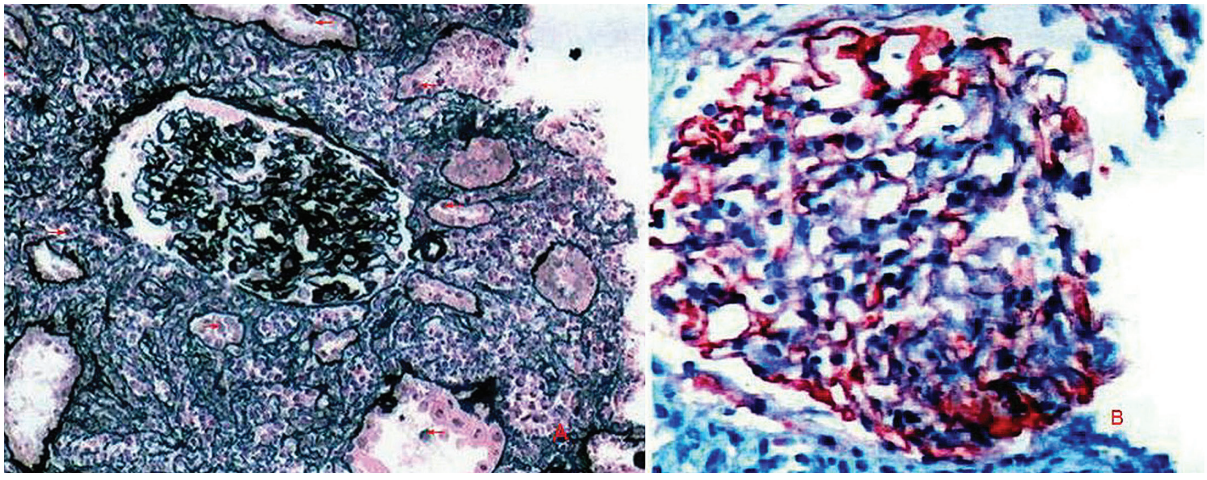

Figure 3. Histological findings of renal biopsy specimen. Left, IgG4 stain indicating significant IgG4-positive lymphoplasmacyte infiltration (arrows) with fibrosis in the renal interstitium, and the maximum IgG4-positive plasma cells are more than 50 per high-power field (× 10). Right, Immunohistochemical examination of glomerulus showing minor abnormalities. Complement C3d (one of the cleavage segments of complement 3) stain demonstrates only vascular loop is C3d positive. No κ chain and λ chain deposit is observed in kidney tissue (× 40).

IgG-positive and IgG4-positive plasma cells in the tubulointerstitium (Figure 3). The patient was diagnosed with IgG4-related kidney disease and received methylprednisolone without any side effects and recovered promptly with the serum creatinine dropped to 320 μmol/L in 2 weeks and IgG4 decreased to 3.2g/L at the 2-month follow-up visit.

DISCUSSION

Known as organ enlargement and nodular lesions, IgG4-related kidney disease consists of abundant infiltration of lymphocytes and IgG4-positive plasma cells in various organs simultaneously or consecutively.[1] The kidney may be the first or the only affected organ. The following clues may imply diagnosis of IgG4-related kidney disease according to the recent documents[2]: (1) acute or chronic kidney dysfunction with abnormal urinalysis; (2) kidney histopathology indicating an IgG4-positive plasmacyte count greater than 10 per high-power field or a ratio of IgG4 to IgG-positive plasma cells greater than 40% or characteristic "storiform" fibrosis surrounding the nests of lymphocyte or plasmacyte; (3) renal imaging demonstrating multiple low-density lesions, diffuse renal enlargement, solitary hypovascular mass in the kidney, and hypertrophic lesion of the kidney pelvic wall with irregularity of the kidney pelvic surface by contrast-enhanced computed tomography; (4) high polyclone serum immune globulin levels especially an IgG4 level higher than 1.35 g/L, or with high levels of IgE and hypocomplementemia; and (5) eosinophilic granulocyte increases.

Glomerulonephritis-associated IgG4-related disease is comparatively rare; however, cases of membranous glomerulonephritis or membranoproliferative glomerular disease have already been described.[3,4] Immunoglobulin G4-related disease often shows an effective responsiveness to glucocorticoid, while there are relatively high rates of recurrence in 20% to 40% patients after stopping corticosteroid therapy or during the steroid tapering period.[5] A repeat course of corticosteroids with an immunomodulator such as azathioprine is often administered.[6] Immunosuppressive agents can be used in the case of corticosteroid refractory.[7] Yet there is no standardized regimens for these agents; thus, further randomized trials are needed.

CONFLICT OF INTEREST

None declared.

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