

Answering Reviewers (Written in blue)

Reviewer #1:

1. The etiology of this disease has not been determined to be autoimmunity. The authors are required to correct the description as appropriate.

We have correct it as following: IgG4-RD is a chronic fibrotic disease mediated by immunity. At present, it is considered that immunologic derangement and infection act as risk factors that activate a large number of lymphocytes to participate in the immune response. Lymphocytes release cytokines, such as interleukin 4 (IL-4), IL-5, IL-10, IL-13 and transforming growth factor β (TGF- β), leading to eosinophilia and elevated serum IgG4 and IgE, which ultimately cause characteristic fibrosis of IgG4-RD.

2. Radiological findings of the renal parenchyma are also important in diagnosing IgG4-related kidney disease (IgG4-RKD). The authors should describe whether the patient showed typical radiological findings of the renal parenchyma on enhanced CT or not.

There was no obvious abnormal density shadow in the parenchyma of either kidney on the CT, while the enhanced CT scan showed that the parenchymal enhancement in both kidneys was weakened.

3. Findings of IgG4, IgG, CD38, or CD138 immunostaining should be demonstrated as figures.

We have demonstrated as Figure 2.

4. The initial dose of glucocorticoids (GC) seemed to be relatively high. The body weight of the patient and the reason to choose such dose should be described.

Considering that there were no contraindications for glucocorticoid treatment, meanwhile although the patient weighed 60 kg approximately, due to the serious condition of him and multiple organ injuries, the patient was treated with enough glucocorticoids at 60 mg/d as initial dosage, and the dosage was gradually reduced as renal function getting better and finally maintained at 10 mg/d.

5. In Figure 2, the authors should explain the finding in each figure (A-F) in more detail.

We have explained the finding in each figure (A-F) in more detail in Figure 2.

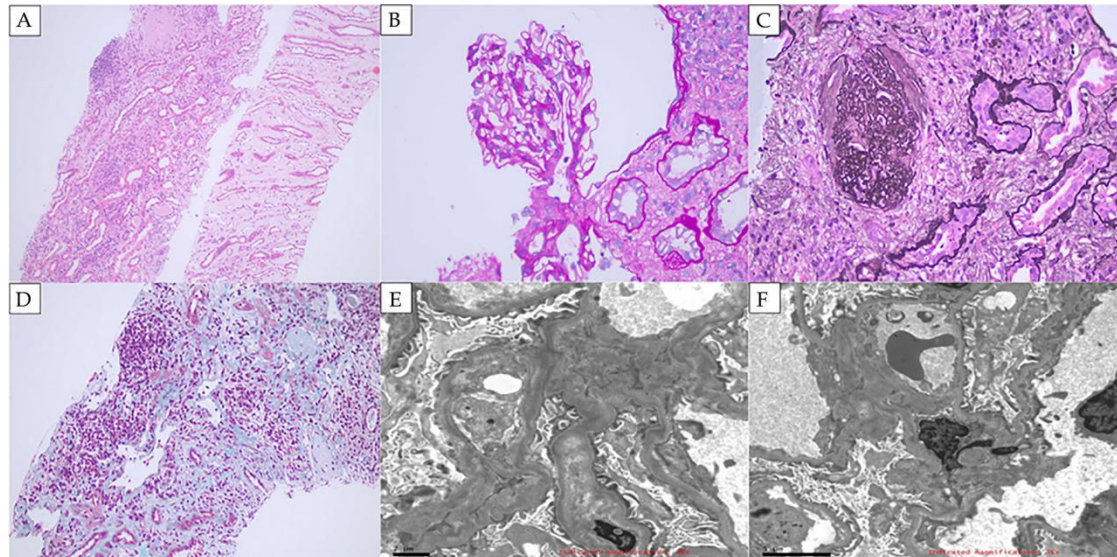


Figure 2 Histopathological findings on the kidney biopsy. A: The renal interstitium was infiltrated by plasma cells and lymphocytes predominantly with fibrosis like car width (light microscopy: hematoxylin and eosin (HE), $\times 100$); B: As shown in the picture B, the injury of the glomerular lesion was mild (light microscopy: periodic acid-Schiff (PAS) staining, $\times 400$); C: In the figure C, we could find one glomerular sclerosis and striate fibrosis in the renal interstitium (light microscopy: Masson and periodic acid-sliver methenamine (PASM), $\times 400$); D: The figure was in the Masson 200 staining, and the lesion was the same as Figure A. In addition, IgG4 plasma cells >10 cells/high powered field and IgG4 plasma cells / IgG plasma cells $>40\%$ were observed by immunohistochemistry. At the same time, there were CD 38-positive plasma cells and CD 138-positive plasma cells; E and F: Most fusion of glomerular podocytes was observed by electron microscopy, while no electron dense granules were observed in the subepithelium, mesangial

area, or subendothelium.

Reviewer #2: 建议

1. Writing in the english language can be further reviewed: on page 2 line 3, "an autoimmune disease recognized by" would better convey the meaning of the sentence; on page 7 line 15 "the middle part of the ureter"; on page 8 line 25 "biopsy confirmed that the patient had"; on page 11 lines 11 and 28 ureterostenosis and hydronephrosis.

We have polished the english language according to suggestion of the reviewer.

2. On the history of past illness section, it is mentioned that the patient had a thyroidectomy - it would be interesting to know what was the indication, and if it was possibly related to the condition described (IgG4-related thyroid disease). On the outcome and follow-up section, it would be interesting to know for how long did the patient receive hemodialysis.

The patient had a thyroidectomy and according to the question of the reviewer, we followed up the patient and the patient said that the pathological findings after the thyroidectomy were benign. At the same time, the patient's thyroid function examination showed no significant abnormalities. So it was not related to IgG4-related thyroid disease. The patient receive hemodialysis for four days. And As the kidney biopsy confirmed that the patient had IgG4-RKD, he was treated with glucocorticoids.