中文題目：以腎病症候群表現的 IgG4 相關性腎病

英文題目：IgG4-related lung disease presenting as nephrotic syndrome

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Introduction:

IgG4-related disease (IgG4-RD) is a rare recognized immune-mediated fibroinflammatory disease that can involve various organs, especially pancreas, aorta, biliary tract, salivary glands, thyroid, orbit, lymph nodes, and kidney. The IgG4-related kidney disease (IgG4RKD) was first described in 2004 by Japan author. Since then, IgG4 related kidney disease (IgG4RKD) has gradually been recognized worldwide. However, due to the rare case in the real world. The rates of misdiagnosis and missed diagnosis are often high We present a pathologic proven IgG4 related kidney disease, nephrotic syndrome as its initial manifestation.

Case presentation:

A 47-year-old male patient admits for a decline in renal function accidentally founded, accompanied by foamy urine, fatigue, and poor appetite noted for several months, and a diagnosis of nephrotic syndrome was made.

Laboratory test results describe in Table1. For the suspicion of IgG4-related disease, a renal biopsy was been performed. Light microscopy results were chronic interstitial nephritis with increase IgG4+ plasma cells (Figure1). The immunofluorescence assay and electron microscopy are described in figure 2 and figure 3. Based on the immunohistochemical findings, clinical manifestations, and exclusion of other secondary factors, a diagnosis of IgG4-TIN was made. Her renal function and protein urine ameliorated after systemic steroid use.

Discussion:

IgG4-related disease was recognized internationally in 2011, Hamano et al reported that patient with autoimmune pancreatitis, has high serum IgG4 concentrations. Later, multiple studies showed this new disease frequently involves multiple organs, including the Kidney. The disease has been known as IgG4-related kidney disease (IgG4RKD). TIN with multiple organ damage is a typical manifestation. The typical renal pathological features of IgG4-RKD include renal interstitial fibrosis and focal lymphoplasmacytic infiltration. Typical immunohistochemistry shows interstitial IgG4+ plasma cell infiltration (> 10/HPF), and more than 40% of IgG+ plasma cells.

Our current patient had an initial disease manifestation of nephrotic syndrome, a pathological diagnosis of IgG4-TIN was made after renal biopsy. The disease was alleviated after systemic steroid treatment. In our view, IgG4RKD should be a differential diagnosis in patients presenting with nephrotic syndrome to avoid a missed diagnosis and delayed treatment owing to delayed treatment will increase the risk of renal failure.

Conclusion:

This case reminds us that the differential diagnosis of nephrotic syndrome and Renal function deterioration should not neglect the possibility of IgG4 related disease,
treatable disease easily resulting in treatment delay.