KSN 2018
Seoul, Korea · May 17 - 20

**Abstract Type : Poster**
**Presentation No. : POT 019**

# A case of IgG4-related disease presented as a renal pelvis mass

**Cheolgu Hwang**, Sang Heon Song, In Seong Park, Harin Rhee, Mi Yeun Han, Ihm Soo Kwak, Youhyun Jeon, Dong Eon Kim, Yeo Jin An
Department of Internal Medicine-Nephrology, Busan National University Hospital, Korea, Republic of

**Case Study:** IgG4-related disease (IgG4-RD) is one of the newly recognized systemic inflammatory disorders, characterized by massive IgG4+ plasma cell infiltration, causing enlargement, nodules of the various organs, simultaneously or metachronously. The disease usually affects the pancreas, liver, salivary glands, and lymph nodes. The kidney is a frequently affected organ, and a variety of renal manifestations of IgG4-RD is glomerular lesions, pyelitis, and multiple renal nodules.
A 36-year-old woman was presented with right kidney mass accidentally found during a health screening. Computed tomography showed a 3.5cm, enhancing mass at the arterial phase in pelvis of right kidney and right ureter (Figure 1). This mass presumed to represent an IgG4 related sclerosing disease and kidney biopsy was performed. Microscopic examination showed a dense lymphoplasmacytic infiltration with fibrosis. Immunohistochemical study was performed and abundance of positive IgG4 plasma cells was proved (Figure 2). Serum IgG4 level measured after kidney biopsy was 1960 mg/dL (reference values 3-201mg/dL). She complained of eyelid edema and suspected inflammation of the lacrimal gland. She was diagnosed with IgG4-RD and started steroid treatment. Eyelid edema improved within 10 days after starting steroid treatment. It is currently taking 40mg of prednisolone.
IgG4-RD was firstly described in patients with sclerosing cholangitis, called autoimmune pancreatitis, and later observed in other organs. Numerous case series have expanded the spectrum of this disorder and shown involvement of multiple organ systems in addition to the kidney. Kidney involvement present with a kidney nodule that may misinterpret as a renal cell carcinoma. Unnecessary surgery can be avoided if the IgG4-RD of the kidney is considered in the differential diagnosis.

Figure 1. CT image

