

## KSN 2017 Abstract

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### Clinical aspects of IgG4-related retroperitoneal fibrosis in single center study

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**Objectives :** Retroperitoneal fibrosis (RF) is a rare condition characterized by the proliferation of fibrous tissue in the periaortic and periliac retroperitoneum. Along with increasing awareness of IgG4-related disease, we try to find the prevalence of IgG4-related RF and compare its clinical feature with non IgG4-related RF.

**Methods :** The material of this retrospective single center study included 21 patients with retroperitoneal fibrosis between January 2006 to December 2016. We entered all clinical and laboratory information for statistical analysis to determine the prevalence, clinical feature and outcomes of IgG4-related retroperitoneal fibrosis.

**Results :** In total 21 patients with retroperitoneal fibrosis, 10 patients (47.6%) were diagnosed with IgG4-related RF (2 with 'definite', 2 with 'probable' and 6 with 'possible' IgG4-related RF). The average age was 62.2 years (range, 41–79) and 6 (60%) were male. All lesions were detected around the infrarenal portion of the abdominal aorta and 7 cases simultaneously affected the iliac arteries. Hydronephrosis was present in 8 cases (80%) and acute kidney injury was observed in 6 patients.

The median serum IgG4 at diagnosis was 641 (range, 24.8–3660). The elevation of serum IgG4 level was not observed in 2 patients. Eight patients were initially treated with steroid therapy. Seven of them (87%) showed radiological response, which showed poor correlation with serum IgG4 level. Comparing with 11 patients of non IgG4-related retroperitoneal fibrosis (52.4%), there were no statistically significant difference between the groups in age, gender distribution, incidence of hypertension, diabetes mellitus, heart failure, malignancy, acute kidney injury, proteinuria, or hematuria, concentration of hemoglobin, white cell counts, inflammatory markers, and complements.

**Conclusions :** This is a pilot study to evaluate the clinical and laboratory features of IgG4-related retroperitoneal fibrosis. Larger studies are needed for prevalence, prognosis of IgG4 related or non related RF.

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**Keywords :** IgG4-related retroperitoneal fibrosis, Retroperitoneal fibrosis, IgG4-related disease