

**Abstract Submission No. : 2460**

## **Clinical features and outcomes of Immunoglobulin G4-related Disease including Immunoglobulin G4-related kidney disease**

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**Objectives:** Immunoglobulin G4-related disease (IgG4-RD) is a newly recognized disease, and a few epidemiologic studies about this disorder have been published. This research aimed to describe the clinical, laboratory, and histopathological features and outcomes of IgG4-RD.

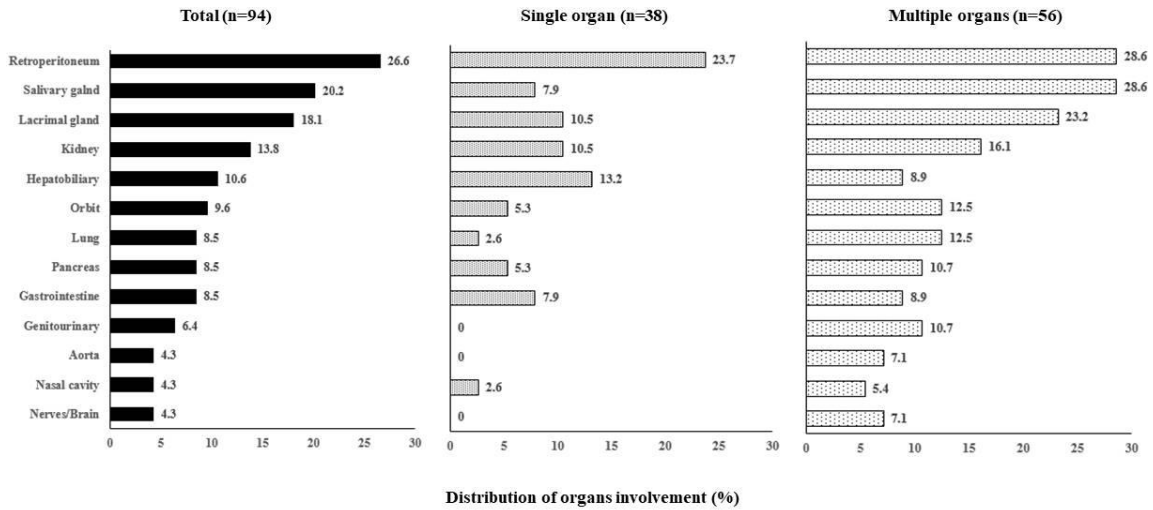
**Methods:** Ninety-four patients who satisfied the comprehensive diagnostic criteria on IgG4-RD were included in this study. Fifty-eight (61.7%) were men. The mean age was 54.8 years, and the median follow-up duration was 32.9 months. The clinical feature between single and multiple organ involvement and with or without kidney involvement groups were evaluated based on symptoms and laboratory findings. The clinical outcome was assessed according to treatment strategies and response.

**Results:** Of 94 patients, 56 (59.6%) had multiple organs involvement. It showed a variety of symptoms and organs involved. Patients with multiple organ involvement had higher serum IgG and IgG4 levels than those with single organ involvement. Those with IgG4-related kidney disease (IgG4-RKD) had worse renal function. The incidence of peripheral blood eosinophilia and hypocomplementemia was higher in patients with renal involvement than in those without. Glucocorticoids-based therapy was most commonly used. (79.8%). Thirty-nine (41.5%) achieved complete remission. Eighteen (19.1%) relapsed after response to treatment. Eight (61.5%) of 13 patients with IgG4-RKD experienced improvement in renal function after treatment. None of the patients died during the follow-up period.

**Conclusions:** Kidney or other organ involvement is not significantly associated with clinical outcomes. Since IgG4-RD has different clinical features, it should be accurately diagnosed. Therefore, all physicians must actively diagnose and treat the condition.

Figure 1. Distribution of organ involvement

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**Figure 2. Treatment response**

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