Baseline clinical and laboratory features of IgG4-related disease: retrospective Japanese multicenter study of 334 cases

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Objective:
IgG4-related disease (IgG4-RD) shows an extremely diverse clinical spectrum. Each clinician encounters a clinically distinct patient population depending on his/her specialty, and it is difficult to avoid institutional bias in a single center. This prompted us to conduct a relatively large-scale multicenter study with well-experienced physicians of IgG4-RD including rheumatologists, gastroenterologists, pulmonologists and nephrologists to better clarify the baseline clinical and laboratory features of IgG4-RD.

Methods:
We retrospectively evaluated 334 patients with IgG4-RD diagnosed between 1996 and 2015 in five institutions. The diagnosis of IgG4-RD was made based on the comprehensive diagnostic criteria or criteria of each organ. We analyzed the serum levels of IgG, IgG4, C3, C4, CH50 and CRP, affected organs, and target organs for biopsy and treatment. We also analyzed the prevalence of diabetes mellitus (DM) and malignancy.

Results:
Two hundred and five patients were male, and 129 were female. The mean age was 63.8 years (range 25-91). At diagnosis, mean serum IgG4 was 755 mg/dL, and elevated in 95.5%. Patients with low C3 and C4 were seen in 34.7% and 33.7%, respectively. Logistic regression analysis showed
that kidney, lung and pancreas lesion independently influenced hypocomplementemia. Mean and median serum levels of CRP were 0.42 and 0.10 mg/dL, respectively, and 90.2% of patients had CRP less than 1.0 mg/dL. Mean number of organs involved was 3.2 (range 1-11); salivary gland (SG) was the most frequently involved organ, followed by lacrimal gland (LG), pancreas, retroperitoneum (RP)/periaorta, kidney, and lung. Single organ involvement was seen in 11.4%. RP/periaorta, lung and kidney were more frequently affected in males, and LG and SG were more frequently affected in females. We analyzed the prevalence of biopsy of the affected organs, and found that the major SG was biopsied in 49.0% of patients, followed by kidney (37.5%), LG (35.1%), lung (33.8%), pancreas (12.8%) and RP/periaorta (1.2%). Corticosteroid was administered to 78.0%, and recurrence was noted in 22.0% of patients. The prevalence of DM was 34.4% and was significantly higher in those with than without AIP (46.2% vs. 30.0%, p=0.005). The prevalence of malignancy was 17.1%.

Conclusions:
The present study clarified the baseline clinical features of IgG4-RD by a retrospective multicenter study. We found that serum level of CRP was less than 1.0 mg/dL in 90% of patients. Moreover, in more than 95% of patients, elevated serum IgG4 levels were useful for the diagnosis of IgG4-RD.