

## **Analysis of 84 patients with IgG4-related disease and malignancy**

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### **Objective:**

Recently, some reports have described the relationship between IgG4-RD and malignancy. However, patient background factors such as affected organs and ethnicity in these reports were different. Furthermore, only a few reports analyzed IgG4-RD patients with malignancy who were treated by rheumatologists. This prompted us to analyze the clinical features of IgG4-RD patients with malignancy.

### **Methods:**

Between November 2004 and September 2015, we retrospectively evaluated 84 patients with IgG4-RD in our hospital. We analyzed the prevalence of malignancy, relationship between the appearance of malignancy and diagnosis of IgG4-RD, type of cancer, and related factors. We compared mean age, gender, laboratory data, affected organs and therapy between the malignancy and non-malignancy groups. We also analyzed the standardized incidence ratio (SIR) of newly recognized malignancy after the diagnosis of IgG4-RD.

### **Results:**

There were 53 and 31 male and female patients, respectively with a mean age of 64.9 (range 41-81) years. Average observation period was 4.6 years. Mean serum levels of IgG and IgG4 were 2324±1040 mg/dL, 704±644 mg/dL. The mean number of affected organs was 2.8. The affected organs were as follows: salivary glands (53.6%), lacrimal glands (48.8%), lung (36.9%), retroperitoneum/ periaorta (29.8%), kidney (23.8%) and pancreas (21.4%). Twenty-one malignancies developed in 18 of 84 patients (21.4%), before the diagnosis of IgG4-RD in 11 malignancies in 11 patients (mean 4.3 years earlier, range 0.6-14 years), and after 10 malignancies in 9 patients (mean 1.7 years later, range 0-9 years). Colon cancer, prostate cancer and malignant lymphoma (ML) were seen in three patients each, and gastric, lung and renal cancers and in two patients each. ML developed after the onset of IgG4-RD in all patients. Only two patients developed malignancy in the same organ as affected by IgG4-RD. No significant differences were seen in mean age, gender, laboratory data, affected organs and the prevalence of corticosteroid therapy or the mean dose of prednisolone. We analyzed SIR of newly recognized malignancy after the diagnosis of IgG4-RD in 83 patients who were observed for more than one year. We found that SIR within one year of diagnosis of IgG4-RD was 4.62 and that of the total period of observation was 1.92.

### **Conclusions:**

The present study clarified the incidence, timing, type of malignancy in patients with IgG4-RD treated by rheumatologists. Malignancies developed at a high frequency. However, no specific characteristics were identified, making periodic screening for malignancy of particular importance.