Utility of Serum IgG4 Levels in a Multi-Ethnic Population

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Background:
(1) Autoimmune pancreatitis (AIP) is significantly complicated with malignancy, however life prognosis remains unknown. (2) Approximately 20% of AIP shows no elevation of IgG4, which represents a low disease activity state, however long-term outcome of this group remains unknown. This study clarified life prognosis of AIP and long-term outcome of IgG4-negative AIP.

Materials & Methods:
(1) We enrolled 96 AIP patients who were followed for more than 3 years, and the standardized incidence ratio (SIR) for malignancy and the standardized mortality ratio (SMR) were calculated. (2) We compared the clinical and laboratory findings of 85 IgG4-positive AIP patients with those of 13 IgG4-negative AIP patients with regard to long-term clinical course.

Results:
(1) The overall SIR for malignancies was 2.11 (95% CI 1.38-2.85), whereas SMR was 0.61 (95% CI: 0.33-0.90), representing that AIP patients developed malignancy significantly more frequently than did the general population, whereas the mortality rate of AIP patients was significantly lower than that of the general population. (2) IgG4-positive patients exhibited a higher rate of diabetes and steroid treatment (p=0.013 and p=0.023, respectively) as well as higher values of disease activity markers, such as soluble IL2 receptor (p=0.003) and immune complex (p=0.004). The prevalence of malignancies and mortality rate were comparable between the groups.

Conclusions:
(1) Patients with AIP have a higher rate of malignancy but lower mortality rate, likely owing to regular follow-up testing and resultant early diagnosis. (2) IgG4-negative AIP represents a low disease activity state in terms of less frequent steroid treatment, however, the prevalence of malignancy and mortality rate are similar to those of IgG4-positive AIP.