

IgG4-related respiratory disease and its mimickers

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Objective:

IgG4-related disease (IgG4-RD) is defined as a multi-organ disease including pulmonary disorders. Its dual characteristics are elevated serum IgG4 concentrations and infiltration of abundant IgG4-positive plasma cells in the lesions. Both findings are essential for diagnosis, but IgG4-positive plasma cells may be present in the tissue of respiratory diseases including lung cancer and Castleman's disease. To clarify the clinical features of IgG4-related respiratory disease (IgG4-RRD) and delineate differential points from mimickers, we studied the clinical details of IgG4-RRD and its mimickers.

Method:

The study group of Intractable Diseases from the Ministry of Health, Labour and Welfare, Japan (MHLW) conducted a multicenter retrospective study of IgG4-RRD in Japan and collected the data of non-IgG4-related respiratory diseases (non-IgG4-RRD) presenting high serum IgG4 concentrations and abnormal findings in chest images suggesting IgG4-RRD. We evaluated these patients based on the comprehensive diagnostic criteria for IgG4-RD (CDC) 2011 and the proposed diagnostic criteria for IgG4-RRD.

Results:

Data of 46 patients (Male 26, Female 20) were collected. The median age was 65 (range 43-85) years old. Final diagnoses of mimickers were as follows: eosinophilic pneumonia (with bronchial asthma), eight cases; anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis, 15 cases; lymphoproliferative disorders/lymphoma, seven cases; interstitial pneumonia, four cases; sarcoidosis, two cases; and others, 10 cases. In laboratory data, the median serum levels of IgG and IgG4 were 1950mg/dl (range 937-7537) and 327mg/dl (range 120-2040), respectively. The median serum level of CRP was 1.23 mg/dl. When evaluating non-IgG4-RRD cases using diagnostic criteria, seven cases were diagnosed as "definite" IgG4-RD by CDC 2011, and one case of lung cancer with ANCA-positive interstitial pneumonia led to the "definite" by the proposed diagnostic criteria for IgG4-RRD.

Conclusion:

Some patients with non-IgG4-RRD fulfilled the criteria for IgG4-RD. These mimickers were difficult to differentiate from IgG4-RRD. The organ-specific diagnostic criteria are helpful when clinical manifestations are similar.
