IgG4-related respiratory disease and its mimickers

Shoko Matsui, Seijiro Minamoto, Hiroshi Yamamoto, Tomohiro Handa, Yuko Waseda, Michiaki Mishima, Keishi Kubo
Health Administration Center, University of Toyama, Osaka Prefectural Medical Center for Respiratory and Allergic Diseases Shinshu University School of Medicine, Department of Respiratory Medicine, Kyoto University Graduate School of Medicine, JCHO Kanazawa Hospital, Saiseikai Noe Hospital, Nagano Prefectural Hospital Organization

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-RD) and delineate differential points from mimickers, we studied the clinical details of IgG4-RD 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-RD in Japan and collected the data of non-IgG4-related respiratory diseases (non-IgG4-RD) presenting high serum IgG4 concentrations and abnormal findings in chest images suggesting IgG4-RD. We evaluated these patients based on the comprehensive diagnostic criteria for IgG4-RD (CDC) 2011 and the proposed diagnostic criteria for IgG4-RD.

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-RD 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-RD.

Conclusion:

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