A case with both biopsy-proven IgG4-RD and ANCA-associated vasculitis

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Objective:

] IgG4-RD and ANCA-associated vasculitis (AAV) have common features, such as affecting lesions and pathological findings. Serological and histopathological findings, such as elevated serum IgG4 concentrations and infiltration of IgG4-positive plasma cells can be also found in AAV cases. The two diseases need careful differential diagnosis. However, we herein present a rare case of IgG4-RD concomitant with AAV, both of which were biopsy-proven.

Methods:

We referred to previous reports of cases with both IgG4-RD and AAV, and compared our case with them.

Results:

A 63-year-old male was introduced to the Department of Respiratory Medicine with suspicion of lung cancer, because he was found to have nodules in his bilateral lung apexes. The left one was resected. However, histopathology revealed storiform fibrosis, obliterating phlebitis and remarkable infiltration of IgG4-positive plasma cells (IgG4/IgG ratio = 0.6), all of which were diagnostic for IgG4-RD. Ground glass opacities (GGOs), which were considered to be signs of IgG4-related lung disease (IgG4-RLD), appeared and disappeared in his both lungs. He was not treated because he did not complain of any symptoms. When he was 66 years old, he was diagnosed as having scleritis. Laboratory tests revealed elevated CRP levels, positive MPO-ANCA (179 U/mL), proteinuria and microhematuria. He was referred to the Department of Rheumatology and renal biopsy was performed. Light microscopic examination showed cellular crescents and necrotizing lesions with exudative changes in glomerulus, whereas immunofluorescence microscopy showed slight or no deposits of IgG, IgG4 and complements in glomerulus and interstitia, leading to the diagnosis of necrotizing crescentic glomerulonephritis due to AAV. After high-dose of PSL was started, CRP was quickly normalized and MPO-ANCA titers were decreased. Scleritis, proteinuria and microhematuria were also disappeared. Azathioprine was added as a maintenance therapy. Six months later, chest CT revealed improvement of GGOs.

Conclusion:

There have been 3 reported cases with both biopsy-proven IgG4-RD and AAV in the literature (Tosovsky, 2012; Alexandraki, 2015; Della-Torre, 2016). There have not been cases with IgG4-RLD and ANCA-associated glomerulonephritis as our case. In all the concomitant cases, PSL and immunosuppressants were used to control AAV. Whereas IgG4-RD responds well to middle-dose PSL, AAV usually needs high-dose PSL and immunosuppressants. In terms of treatment plan, the differential diagnosis is important.