

Inflammatory Abdominal Aortic Aneurysm with Immunoglobulin Heavy Chain Gene Rearrangement

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Abstract: Inflammatory abdominal aortic aneurysm (IAA) is an aortic manifestation of IgG4-related disease. However, the pathogenesis of IgG4-related disease is obscure. Herein, we report the case of a 74-year-old Japanese man with IAA, associated with IgG4-positive plasma cell infiltration and oligoclonal bands of immunoglobulin heavy chain (IgH) gene rearrangement. The patient presented with a growth of pulsating abdominal mass. Computed tomography revealed an infrarenal type of abdominal aortic aneurysm and right common iliac arterial aneurysm. Histologically, the aneurysmal wall of the aorta showed marked fibrous thickening of tunica adventitia with severe lymphoplasmacytic infiltration, including lymphoid follicles, perineural inflammatory cell infiltration, and obstructive phlebitis. Immunohistochemical analysis showed the infiltration of many IgG4-positive plasma cells and formation of B-cell rich lymphoid follicles in the aneurysmal wall. The mean percentage value of IgG4-positive plasma cells was found to be $64.9 \pm 14\%$ of all IgG-positive plasma cells. Light chain immunoglobulin restriction was absent. Polymerase chain reaction analysis showed oligoclonal bands of IgH gene rearrangement in all samples obtained from the aneurysmal wall. Thus, the present paper describes for the first time an IAA case with extensive infiltration of numerous IgG4-positive plasma cells, as well as oligoclonal bands of IgH gene rearrangement.

Key words : Inflammatory abdominal aortic aneurysm, IgG4-positive plasma cell, IgH gene rearrangement, Polymerase chain reaction, Immunohistochemistry