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Abdominal aortic aneurysm and IgG4-associated disease

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Introduction: Until recently, aortic abdominal aneurysm (AAA) has been exclusively presented as a consequence of the atherosclerotic process affecting the aorta. Subsequent transcriptomic study has shown that it is not always the case. AAA may result from aortitis or periaortitis that are syndromes of the so-called IgG4-related disease (IgG4-RD). IgG4-related disease represents a relatively newly defined condition comprised of a collection of disorders characterized by IgG4 hypergammaglobulinemia and the presence of IgG4-positive plasma cells in the affected organs with fibrotic or sclerotizing changes. IgG4-RD was identified as a possible cause for vasculitis of large vessels.

Experimental Aim: In our study, we performed an examination of bioptic AAA samples in order to identify patients with the findings characteristic of IgG4-RD.

Patients & Methodology: We examined a total of 114 patients with AAA requiring surgery. In each patient, a biopsy sample was taken from the aneurysm pouch at the place of the largest dilation, established macroscopically, during the surgery. Subsequently, histopathological examination was performed, as well as the examination of the presence of IgG4-producing plasma cells. IgG, and IgG1-IgG4 was determined in serum of the patients.

Results: In the AAA group of patients, an increased IgG4 level of more than 1.35 g/L was present in four patients. In two of these patients, immunohistochemistry showed 30 to 50 IgG4+ plasma cells/hpf; however, histopathological findings were negative with regard to IgG4-RD. In a further two patients, IgG4 levels exceeded 1.35 g/L and more than 50/IgG4+ plasma cells/hpf were found in the histological preparation. In addition, there was positive histopathological feature (storiform fibrosis, dense lymphoplasmacytic infiltrate and obliterative phlebitis) that confirmed IgG4-RD. In further five patients, more than 50 IgG4+ plasma cells/hpf were found in the aneurysm samples, as well as histopathological findings indicative of IgG4-RD. However, serum IgG4 levels were within a normal range in these five patients. In another 13 patients an increased number of IgG4+ plasma cells were present, with more than 30 IgG4+ and less than 50 IgG+ plasma cells/hpf; however, IgG4 serum levels were less than 1.35 g/L and no IgG4-RD histopathological features were found.

Conclusion: Some AAAs is considered to be a disease associated with IgG4-RD. This finding makes it possible to provide a targeted anti-inflammatory or Immuno-suppressive treatment to these patients.

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