Abdominal aortic aneurysm and IgG4-related disease

Miroslav Prucha[1], Petr Sedivy[2], Petr Stadler[2], Pavel Zdrahal[2]

[1] Department of Clinical Biochemistry, Hematology and Immunology, Na Homolce Hospital, Prague, Czech Republic.

[2] Department of Vascular Surgery, Na Homolce Hospital, Prague, Czech Republic.

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 affected organs with fibrotic or sclerotizing changes. IgG4-RD was identified as a possible cause of vasculitis of large vessels.

Experimental aim

In our study, we performed an examination of bioptic AAA sample in order to identify patients with findings which are characteristic for IgG4-RD.

Patients and methodology

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

Table 2. Characteristics and parameters of patients fulfilling criteria of IgG4-RD

Patient	Sex	Age	IgG4 - serum (G/L)	IgG4+/HPF	Histopat-hology
1	male	69	0.55	86	positive
2	male	59	0.45	81	positive
3	male	68	0.77	51	positive
4	male	66	1.87	169	positive
5	male	61	2.41	72	positive
6	male	64	0.11	84	positive
7	male	60	0.47	53	positive

Figure 1. Typical histopathological feature of lymphoplasmatic infiltrate and storiform fibrosis

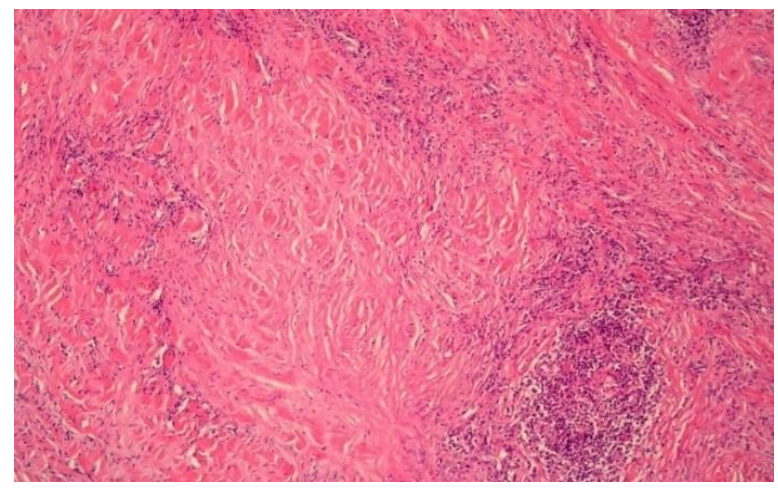
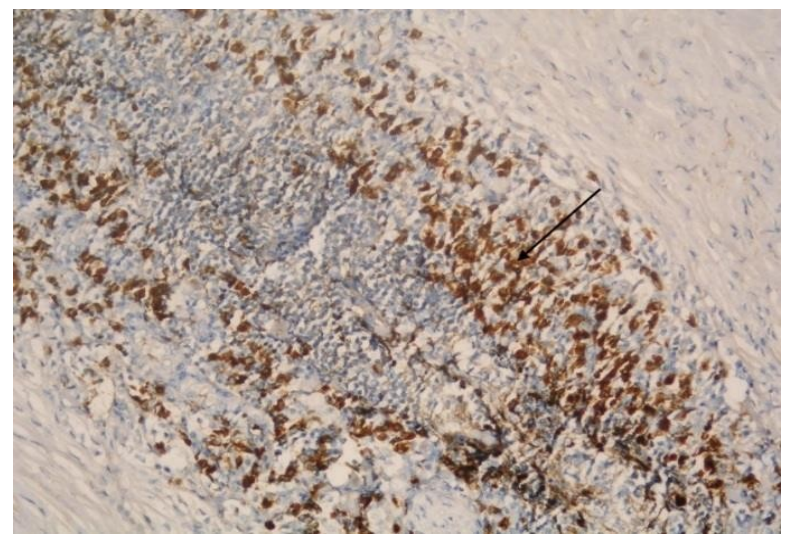


Figure 2. By immunohistochemistry for IgG4, the majority of the IgG+ plasma cells expressed IgG4



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, but 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 a 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. Table 2 shows an overall summary of the results. 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 are considered to be a disease associated with IgG4 - RD. This finding provides a possibility to use a targeted anti-inflammatory or immunosuppressive treatment in these patients.

This work was supported by an MH CZ - DRO (Hospital Na Homolce – NNH, 00023884), IG144101.