Common Variable Immunodeficiency and Takayasu’s Arteritis

Sir,

Common variable immunodeficiency (CVI) is frequently associated with the development of neoplastic and/or autoimmune disorders [1]. Well-known associated autoimmune diseases are systemic lupus erythematosus, dermatomyositis, idiopathic thrombocytopenic purpura, autoimmune hemolytic anemia, primary biliary cirrhosis, Graves disease, and sarcoidosis [2]. We did not find any report on the association of CVI and Takayasu arteritis.

Case Report

A 42-year-old woman with a history of recurrent herpes zoster infections and bilateral bronchiectasias was diagnosed as having CVI 4 years prior to the present admission. She had recently been diagnosed as having severe hypertension and on admission she had a blood pressure of 200/120 mm Hg and an intense periumbilical systolic murmur. The ECG and chest x-ray were normal and blood analyses showed a low total protein value (43 g/l) due to a hypogammaglobulinemia of 2 g/l with deficiency of IgG, IgM, and IgA. The serum complement level was normal and the antitissue antibodies (antinuclear, antismooth muscle, and anti-DNA) were negative. An abdominal ultrasound examination showed a large aneurysm of the abdominal aorta and a left kidney diminished in size. A CT confirmed the presence of the aortic aneurysm, a finding that was not present on a previous CT scan performed 4 years earlier. A digital intravenous angiography subtraction examination of this region revealed a large dilated abdominal aorta with bilateral stenosis of the renal arteries, multiple stenosis in the upper mesenteric artery and a complete abolished function of the left kidney (fig. 1). The thoracic aorta and supraaortic vessels appeared normal in this examination.

The pathological analysis of the portion of the aortic wall taken at surgery revealed the presence of intimal fibrosis, a chronic inflammatory infiltrate (lymphocytes, plasma cells and histiocytes) and giant cells. These findings were considered consistent with Takayasu arteritis with severe aortic involvement.

Fig. 1. Large dilated abdominal aorta with bilateral stenosis of the renal arteries, multiple stenosis in the upper mesenteric artery and a complete abolished function of the left kidney.

Comment
Circulating antibodies against the vessel wall have been detected in patients with Takayasu arteritis [3]. Another experimental study has described inflammatory lesions of the aortic wall as the result of immunological reactions of both cellular and humoral types [4]. Immunologically the patients show defective T lymphocyte function, increase in the serum level of IgG and a reduction in serum complement constituents C3 and C4, indicating the possibility of formation of a complement-binding immune complex [5].

The presence of Takayasu arteritis in a patient with CVI raises the possibility of a new type of associated autoimmune condition.


References

Announcements
Third ‘Berliner Dialyseseminar’
November 23–24, 1990, Berlin (West)
The main purpose of the meeting is to provide basic knowledge on the entire field of renal replacement therapy. There will be only invited lectures.
For further information please write to:
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Charles E. Culpeper Foundation Scholarships in Medical Science
The Charles E. Culpeper Foundation is currently accepting applications for its 1991 Scholarships in Medical Science Program designed to support the career development of academic physicians. Up to 3 awards of $100,000 per year for 3 years will be made to United States medical schools on behalf of candidates who are US citizens, who have received their MD degree from an US medical school in 1982 or later, and who are judged worthy of support by virtue of the quality of their research proposals. All scientific research relevant to human health is eligible for consideration. No institution may nominate more than one candidate.
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