Aortitis With Severe Aortic Regurgitation in Behcet’s Disease: A Case Report

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Behcet’s aortitis is a rare, but one of the most severe complications of Behcet’s disease. We report a 24-year-old woman who was noted initially to have aortitis and severe aortic regurgitation caused by Behcet’s disease. After receiving aortic valve replacement, aortoplasty and immunosuppressant therapy, her condition became stationary. As far as we are aware, she is the youngest case that has undergone surgery. The early onset of hemodynamic decompensation is considered to be related to delay in diagnosis and lack of steroid treatment. [J Formos Med Assoc 2010;109(1):82–84]

Key Words: aortic regurgitation, aortitis, Behcet’s disease

Behcet’s disease is a chronic, recurrent, multisystemic inflammatory disorder without definite etiology. It occurs worldwide, but has the highest incidence in Japan and Eastern Mediterranean countries.1 Currently, the most commonly used diagnostic criteria are those of the International Study Group for Behcet’s disease or the revised diagnostic criteria of the Behcet’s Disease Research Committee of Japan.1 Behcet’s disease can be divided into three subtypes: neuro-, entero-, and vasculo-Behcet’s disease.2 Cardiovascular complications are uncommon (7–29%), especially those that involve the heart or aorta, but frequently they are associated with mortality and morbidity.3

Here, we report a patient without a diagnosis of Behcet’s disease, until aortitis with severe aortic regurgitation was demonstrated.

Case Report

A 24-year-old woman who denied any systemic disease was admitted because of intermittent chest tightness and progressive exertional dyspnea for 6 months. Auscultation revealed a grade 2/6 systolic sea-gull murmur and a grade 3/6 diastolic murmur at the left middle sternal border. Her blood pressure was 96/40 mmHg and her heart rate was 80 beats/min. Upon detailed review, she had no significant known medical history, but had recurrent aphthous and genital ulcers, and skin lesions for 2 years. Hence, Behcet’s disease, incomplete form, was suspected and a work-up was performed. C-reactive protein was 2.06 mg/dL (normal range, <0.6 mg/dL). The erythrocyte sedimentation rate was 66 mm/hour and >120 mm/2 hours. The complement C3 and C4 levels were within normal limits and the autoantibodies (including antinuclear antibody, anti-extractable nuclear antigen, and rheumatoid factor) were negative. The Venereal Disease Research Laboratory test was non-reactive.

Electrocardiography showed left ventricular (LV) hypertrophy. Echocardiography revealed moderate-to-severe eccentric aortic regurgitation and marked LV enlargement with suboptimal...
contractility (LV end-diastolic dimension, 6.4 cm; ejection fraction, 58%). Cardiac catheterization showed that the LV end-diastolic pressure was 8 mmHg, the systemic blood pressure was 90/36 mmHg, and the pulmonary artery pressure was 22/8 mmHg. Aortography at the aortic root revealed severe aortic regurgitation without aortic valve prolapse (Figure 1). There were irregularities in the ascending aortic wall, without dilatation. Hence, Behcet’s disease with aortitis was confirmed.

The patient underwent aortic valve replacement with a 19-mm St. Jude Regent™ mechanical valve (St. Jude Medical, Inc., St. Paul, MN, USA) and aortoplasty. Thickened aortic valve with impaired coaptation, irregular and thickened aortic wall with supravalvular aortic stenosis, dilated LV chamber, and a small amount of pericardial effusion were found during surgery (Figure 2). The histological examination revealed myxomatous degeneration with mild lymphocytic infiltration. The postoperative course was uneventful. Immunosuppressant, anticoagulant, and anti-failure treatment was given. Follow-up echocardiography showed normalized LV dimensions with improved contractility, and C reactive protein level returned to normal. After 2.7 years follow-up her current condition is stationary in New York Heart Association functional class I.

Figure 1. Ascending aortogram with (A) right anterior oblique view and (B) lateral view showing irregular ascending aortic wall with severe aortic regurgitation.

Figure 2. (A) Irregular and thickened aortic wall (arrow) with supravalvular aortic stenosis, (B) thickened aortic valve with impaired coaptation (arrow), dilated left ventricular chamber, and a small amount of pericardial effusion, were found during surgery.
Discussion

Vasculo-Behcet’s disease is a rare systemic vasculitis that involves large vascular lesions. Three major manifestations in order of frequency are: venous occlusion, arterial occlusion, and arterial aneurysm formation. Treatment of Behcet’s disease is symptomatic and empirical but remains unsatisfactory because of variable manifestations with uncertain etiology and pathogenesis. However, most presentations are responsive to corticosteroids and possibly additional immunosuppressant therapy. In our patient, the diagnosis of Behcet’s disease was delayed until aortitis with severe aortic regurgitation was noted. No steroid or immunosuppressant therapy was administered preoperatively. This might be the reason that, to the best of our knowledge, our patient is the youngest case of Behcet’s aortitis to undergo surgical management. This emphasizes the importance of steroid treatment to delay the onset of aortitis symptoms.

Aortitis with or without valvulitis complicating Behcet’s disease is extremely rare and frequently results in significant aortic regurgitation with hemodynamic decompensation, for which surgical management is usually required, but difficult because of the inflammatory and fragile tissue. The most serious complications after surgical treatment are prosthetic valve detachment, bypass graft occlusion, and pseudoaneurysm formation. Ando et al reported a high prosthetic valve detachment rate of 40% and a low 5-year freedom from reoperation (64%) in patients with Behcet’s disease, in contrast to 4.6% and 96%, respectively, of those with Takayasu’s arteritis. The cause of the differences between the two diseases is still unknown. In a study by Okada et al, the reoperation rate was 7.4% per patient-year and the mortality rate was 3.7% per patient-year.

Several surgical techniques have been developed to lower the complication rate, including the translocated Bentall procedure, Cabrol’s method, and valved conduit reconstruction. Preoperative steroid administration is recommended to reduce inflammation. However, postoperative steroid therapy is controversial. In a study by Isomura et al, early postoperative steroid therapy increased the susceptibility to infection, disturbance of wound healing, and valvular detachment. However, Ando et al and Suzuki et al suggested that continuous postoperative steroid administration has the benefit of inflammation control, and reduces the incidence of complications, especially those in the active inflammatory stage.

In patients with Behcet’s disease or other autoimmune diseases, aortitis or other cardiovascular complications should be evaluated carefully in those with chest discomfort. Steroid administration is important, especially preoperatively, which not only decreases inflammatory reactions, but also reduces the postoperative steroid dosage and diminishes the associated side effects. Moreover, because of high reoperation and mortality rates, long-term follow-up is mandatory after surgery.

References