Left Ventricle to Pulmonary Arterial Fistula due to Behçet’s Disease

Surgical treatment of aortic regurgitation due to Behçet’s disease is difficult. A 57-year-old male with Behçet’s disease underwent aortic valve replacement with a mechanical valve for aortic regurgitation in 1995. Due to prosthetic valve detachment, 5 months thereafter he underwent a Bentall type operation with a composite graft. Due to complication of the left ventricle to pulmonary arterial fistula, 6 months later a third operation was performed for closure of the fistula. He is doing well at present 5 years after the third operation. Left ventricle to pulmonary arterial fistula is an exceedingly rare complication and has not been reported in the literature. (Jpn J Thorac Cardiovasc Surg 2004; 52: 135–138)

Key words: aortic regurgitation, Behçet’s disease, aortic root replacement, fistula

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Aortic regurgitation (AR) caused by Behçet’s disease is very rare.⁴ Surgical treatment of AR due to Behçet’s disease is difficult, because the incidence of postoperative complications such as prosthetic valve detachment and pseudoaneurysm is very high.⁵ We report a case of ruptured aneurysm of the left ventricle (LV) outflow into the pulmonary artery (PA) after a Bentall type operation with Behçet’s disease. This is an exceedingly rare complication and has not been reported in the literature.

Case

The case is a 57-year-old male with a history of recurrent oral ulcers, pustulosis, colon ulcer and arthralgia. He underwent aortic valve replacement (AVR) with SJM23A for AR at another hospital on June 28, 1995. He did not have a history of dental diseases before AVR. Steroids were not administered. He began to complain of anterior chest pain on effort from September 1995. Aortogram revealed massive AR due to prosthetic valve detachment on October 12, 1995 and on November 7, 1995 he was admitted to our hospital. His white blood cell (WBC) count was 9,900/mm³ and C reactive protein (CRP) was 4.5 mg/dl. He underwent a Bentall type operation using a composite graft (Gelseal graft 28 mm with SJM27A) on November 15, 1995. The dehiscence was located around the right and non-coronary cusp. The tissue around the dehiscence was friable and edematous. Root abscess and several vegetations were observed beneath the posterior commissure. This abscess cavity was closed using mattress valve sutures with Teflon felt after debridement. Aortic cross-clamp time, cardiopulmonary bypass time and operation time were 140 minutes, 240 minutes and 9 hours, respectively. There was complication with low cardiac output syndrome and he was supported by intra-aortic balloon pumping for 2 days. All cultures were negative before and after the second operation and tissue sections stained for bacteria did not demonstrate any organisms, but he was diagnosed to have prosthetic valve endocarditis according to operative findings, that is, presence of an abscess cavity and several vegetations. Antibiotics were administered intravenously. Laboratory data showed a normalized WBC count and CRP on the 40th day after surgery, but the WBC count and CRP increased from the 60th day with a WBC count of 18,700/mm³ and CRP of 15.8 mg/dl (Fig. 1). Steroid therapy with prednisolone at 10 mg/day was commenced on the 92nd day and CRP reached the normal range on the 96th day. Computed tomography (CT) scan and echocardiogram showed an aneurysmal
change of the LV outflow and pulmonary stenosis by aneurysm of the LV outflow on the 114th day (Fig. 2A, B). Color mapped Doppler showed a new LV to PA shunt on March 20, 1996. Cardiac catheterization revealed O₂ step up between the main PA and right ventricle (RV) outflow. Qp/Qs was 2.42. The pressure gradient between the main PA and RV outflow was 30 mmHg. The pressure of the right atrium and main PA were 11 mmHg and 44/16 (22) mmHg, respectively. Right ventriculogram showed main PA stenosis. He was diagnosed to have a LV to PA fistula due to a ruptured aneurysm of the LV outflow and pulmonary stenosis due to compression by an aneurysm.

The third operation was conducted on May 8, 1996. The aneurysm ruptured to the PA and the defect was in the posterior wall of the main PA with the diameter of the defect being 15 mm (Fig. 3). The aneurysmal wall was very thin and the diameter of the aneurysm was 25 mm. There was no vegetation around the fistula and the surface of the aneurysm was very smooth. Communication between the PA and LV outflow was just beneath the prosthetic valve of aortic position and just above the pulmonary valve. The fistula was closed by mattress sutures with Teflon felt via PA tomy on cardiopulmonary bypass. Aortic cross-clamp time, cardiopulmonary bypass time and operation time were 69 minutes, 98 minutes and 5 hours 10 minutes, respectively. He satisfactorily recovered from the third operation. Echocardiogram confirmed no residual shunt. He was discharged on June 15, 1996. Systolic murmur was newly audible at 2 months postoperatively. A small LV to PA shunt was detected on echocardiogram, but the volume of shunt is not increasing. He is doing well and working at present 5 years after the third operation. Prednisolone (5 mg/day) is administered at present for normalized CRP.

**Discussion**

This patient was diagnosed to have an incomplete type of Behçet’s disease based on his clinical history. He underwent AVR for AR at another hospital and was complicated with prosthetic valve detachment. The incidence of prosthetic valve detachment is higher in patients with AVR than a valve conduit operation for AR due to Behçet’s disease. We selected a Bentall type operation using a composite graft as the second procedure in this case.

Pseudoaneurysmal formation is sometimes complicated in patients with Behçet’s disease. In this case, aneurysmal change appeared in the LV outflow and the aneurysm ruptured into the main PA. An LV outflow tract PA fistula has been reported in only one case due to infective endocarditis. This complication has not been reported in Behçet’s disease. Suzuki et al. have recommended postoperative steroid therapy in the active stage of Behçet’s disease. It was difficult for us to decide whether to use steroids just after the Bentall