


An adolescent with Behçet's aortitis mimicking infective endocarditis: a case report

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Background

Aortic valve involvement is rare in patients with Behçet's disease (BD); however, recurrent prosthetic valve detachment after valve surgery has frequently been reported. We report a rare case of Behçet's aortitis involving the aortic valve, mimicking active infective endocarditis (IE) with perivalvular abscess.

Case summary

A 16-year-old boy, with an unknown case of BD, presented with pyrexia of unknown origin, severe aortic valve regurgitation, vegetation, and perivalvular abscess in the aortic valve. All cultures tested negative for microorganisms. As we suspected IE, aortic valve replacement was performed. After the initial surgery, recurrent prosthetic valve detachment and pseudoaneurysm formation occurred, which resulted in the diagnosis of BD. The patient underwent a modified Bentall procedure, in which the valve conduit was proximally sutured to the left ventricular outflow tract instead of the aortic annulus. Immunosuppressive therapy was initiated on the 10th postoperative day. His condition became stable, and additional surgery was not required.

Discussion

The echocardiographic findings of Behçet's aortitis involving the aortic valve resemble those of aortic valve IE. Modified Bentall procedure, combined with effective immunosuppressive therapy, may be useful in preventing prosthetic valve detachment.

Keywords

Case report • Behçet's disease • Aortitis • Infective endocarditis • Modified Bentall procedure

Learning points

- Echocardiographic findings of Behçet's aortitis involving the aortic root (vegetation or perivalvular abscess) resemble those of infective endocarditis of the aortic valve.
- Persistent increase in C-reactive protein level after aortic valve surgery despite antibiotic use would suggest the possibility of non-infectious aortitis.
- The modified Bentall procedure combined with immunosuppressive therapy may be useful in preventing prosthetic valve detachment in patients with Behçet's disease.

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Introduction

Aortitis is a rare and life-threatening complication of Behçet's disease (BD), sometimes involving the aortic root, leading to aortic valve regurgitation (AR).^{1,2} Standard aortic valve surgery often results in recurrent prosthetic valve detachment due to the aortic annulus being fragile.³ We report an unusual case of Behçet's aortitis involving the aortic valve with vegetation and perivalvular abscess as well as severe AR, mimicking active infective endocarditis (IE) of the aortic valve.

Timeline

Time point	Medical events
15 days before admission	Antibiotics therapy started for pyrexia.
Day 1	Admission with the possibility of infective endocarditis. All cultures were negative.
Day 20	Aortic valve replacement (AVR) performed. No microorganisms cultured from valve tissue.
Day 32	Re-AVR performed for prosthetic valve detachment.
Day 59	Repair for an aortic pseudoaneurysm performed.
Day 85	Diagnosis of Behçet's disease confirmed.
Day 88	Modified Bentall procedure performed for the recurrence of valve detachment.
Day 98–104	Methylprednisolone (200 mg/day) administrated.
Day 105–118	High-dose prednisone (60 mg/day) administrated. Antibiotics gradually discontinued.
Day 119, 133, 147	The dosage of prednisone reduced to 50, 40, and 35 mg/day.
Day 160	The patient discharged on prednisone 30 mg/day.
Day 250	No recurrence of valve detachment detected.

Case presentation

A 16-year-old previously healthy Japanese boy was admitted with pyrexia of unknown origin. Fifteen days prior, oral antibiotic therapy had been initiated. His blood pressure was 120/70 mmHg, heart rate was 100 beats/min, and body temperature was 38.5°C. A grade 3/6 diastolic murmur was auscultated. Laboratory tests showed a leucocyte count of 12.2×10^3 , haemoglobin level of 12.0 mg/dL, and C-reactive protein (CRP) level of 5.6 mg/L. Transoesophageal echocardiography demonstrated severe AR due to right coronary cusp (RCC) prolapse, vegetation-like mass on RCC, and perivalvular abscess (*Figure 1A* and *Video S1*). Multi-slice computed tomography revealed an aortic root pseudoaneurysm, suggestive of a perivalvular abscess cavity (*Figure 1B*). Initial and follow-up blood cultures were negative for microorganisms.

Culture-negative IE in the aortic valve was suspected; hence, empirical antibiotic therapy consisting of ceftriaxone and sulbactam-ampicillin was initiated. Twenty days after the initiation of antibiotics, aortic valve replacement (AVR; INSPIRIS, Edwards Lifesciences, Irvine, USA) was performed. A 3-mm mobile vegetation-like mass in the RCC and a perivalvular abscess at the junction of the right and left coronary cusps were revealed (*Figure 2*). The abscess cavity was

debrided and repaired with glutaraldehyde-preserved equine pericardium. No microorganisms were cultured from the abscess or the valve tissue. However, 12 days later, the patient required a repeat AVR (ATS-AP, ATS Medical, Minneapolis, USA) to fix prosthetic valve detachment. Seventeen days later, further operation was required due to formation of a pseudoaneurysm at the aortic cannulation site. The levels of CRP increased further after the initial surgery and did not normalise despite antibiotic use (*Figure 3*). Serum procalcitonin level was between 0.01 ng/mL and 0.09 ng/mL during the hospital stay.

Due to the unusual postoperative course, the possibility of non-infective aortitis was considered. On day 85 of hospitaliza-

tion, the diagnosis of BD was confirmed on the basis of previous episodes of recurrent oral, genital, and skin ulcers, coupled with human leucocyte antigen B51 (HLA-B51) positivity. Histopathological examination of the aortic valve showed myxomatous valvulopathy without significant leucocyte infiltration, suggestive of Behçet's valvulitis (*Figure 4*).¹

Twenty-nine days after the third operation, the modified Bentall procedure (MBP) proposed by Chen *et al.* was performed to treat the recurring valve detachment.⁷ Modified Bentall procedure was performed as follows: the proximal end of a straight graft was sutured directly to the left ventricular outflow tract (LVOT) (leaving the aortic annulus) with a double continuous 4-0 polypropylene suture. The prosthetic valve (SJM Regent, St Jude Medical, St Paul, USA) was sutured into the graft 10 mm from its end, with a continuous 4-0 polypropylene suture. The coronary artery buttons were anastomosed to the composite graft through a 7-mm graft to prevent coronary artery kinking (*Figure 5*). The cardiopulmonary bypass and aortic cross-clamp times were 292 min and 183 min, respectively. Immunosuppressive therapy was initiated on the 10th postoperative day. The patient was administered methylprednisolone (200 mg/day) for 7 days, followed by 14 days of high-dose oral prednisone therapy (60 mg/day). The levels of CRP quickly

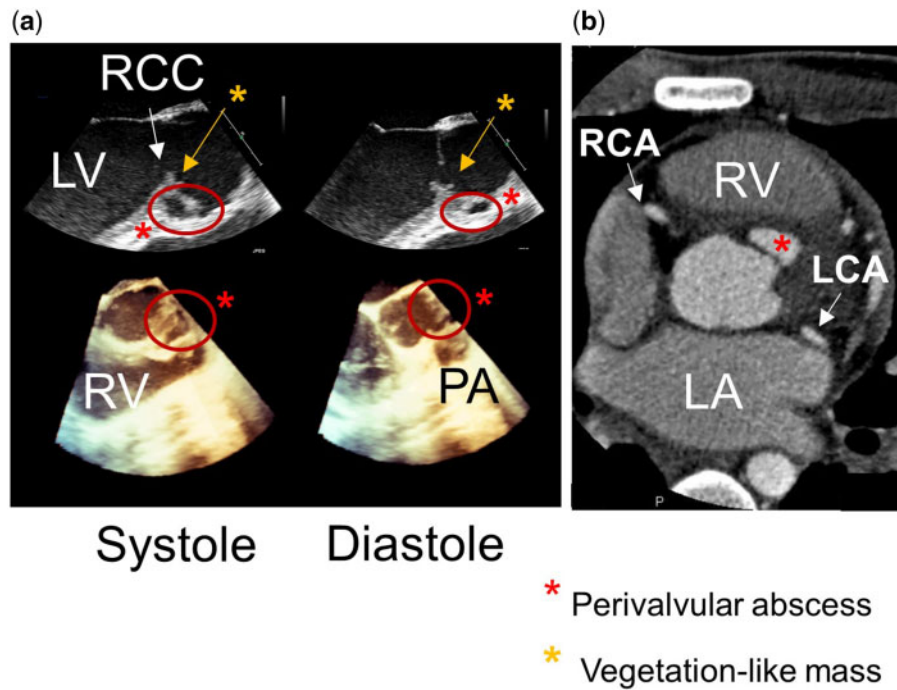
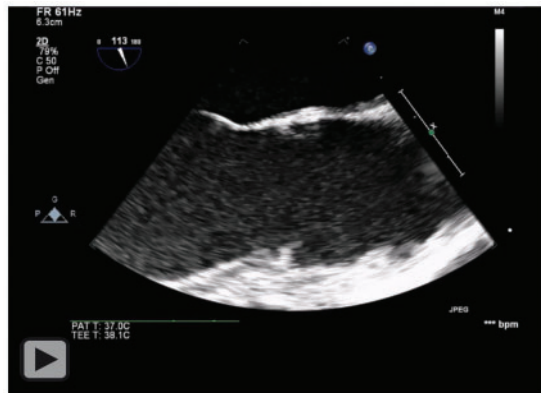


Figure 1 Preoperative findings. (A) The transoesophageal echocardiogram showed severe right coronary cusp prolapse, an oscillating mass of 3 mm × 4 mm on the right coronary cusp (yellow asterisk), and perivalvular abscess with a cavity (red asterisk). View [Supplementary material online, Video S1](#). (B) Multi-slice computed tomography revealed a pseudoaneurysm in the aortic root, suggestive of the abscess cavity. LA, left atrium; LV, left ventricle; LCA, left coronary artery; PA, pulmonary artery; RCA, right coronary artery; RCC, right coronary cusp; RV, right ventricle.



Video S1 Preoperative transoesophageal echocardiographic findings mimicking those of aortic valve infective endocarditis.

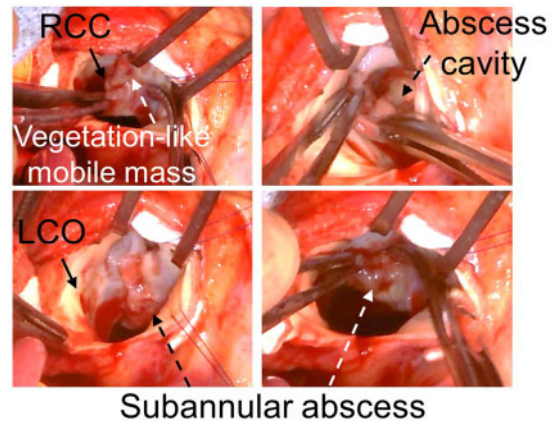


Figure 2 Intraoperative findings of the initial aortic valve replacement. A vegetation-like mobile mass was found on the right coronary cusp. Aortic peri- and subannular abscess was formed at the junction of the right and left coronary cusp, with an abscess cavity. LCO, left coronary orifice; RCC, right coronary cusp.

decreased and continued to remain within the normal range, even when the antibiotics were discontinued (*Figure 3*). Repeated echocardiography revealed no signs of prosthetic valve detachment. The patient was discharged with instructions to take medium-dose prednisone (30 mg/day) for 14 days. Seven months after MBP (to date), he has had no postoperative complications and continues to be on low-dose prednisone.

Discussion

To the best of our knowledge, this case involves the youngest patient with BD who required aortic valve surgery. Male sex and younger

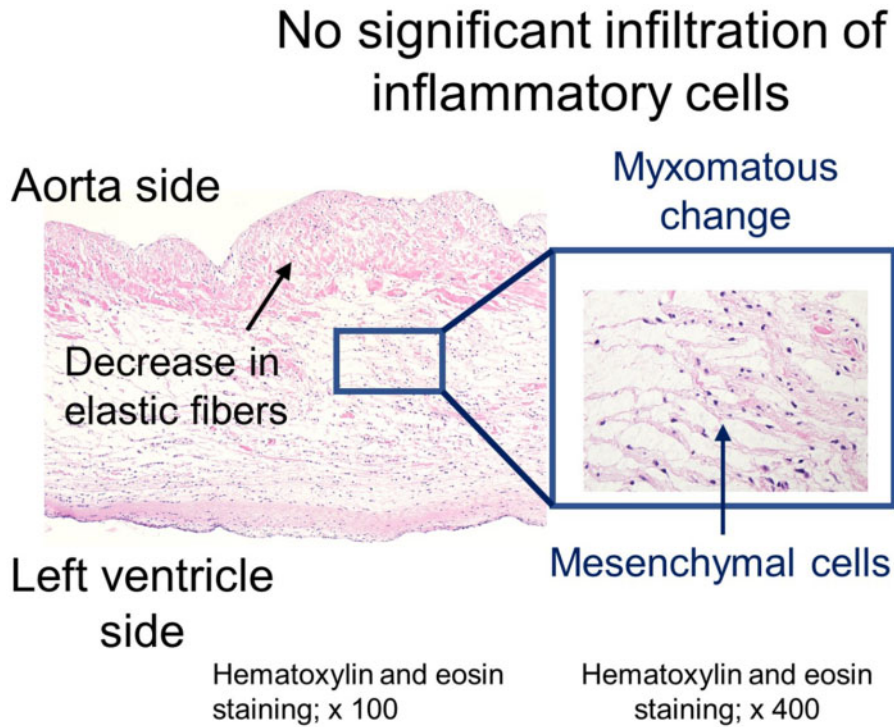


Figure 3 Clinical course during hospitalization. AVR, aortic valve replacement; BD, Behçet's disease; MBP, modified Bentall procedure; m-PSL, methyl-prednisolone; PSL, prednisolone; PVL, perivalvular leak.

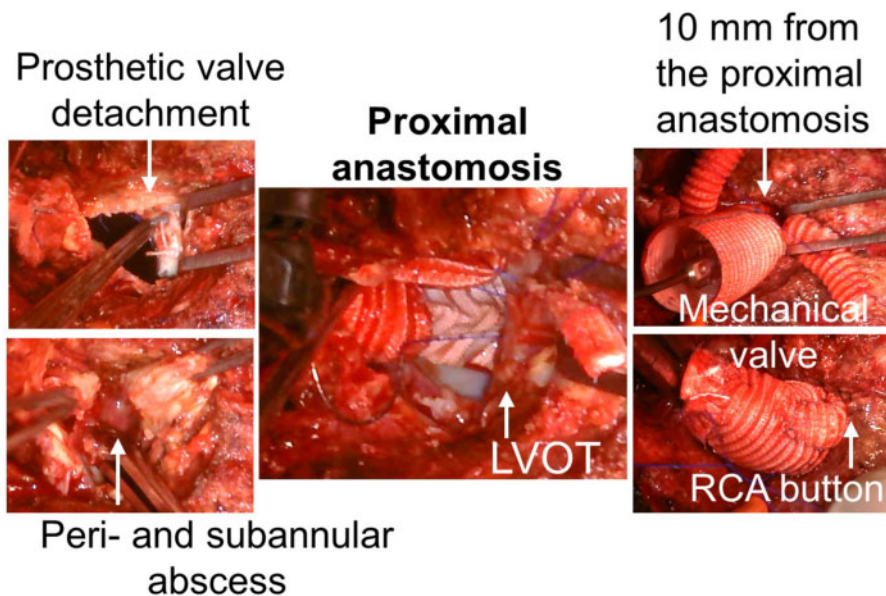


Figure 4 Histopathological findings of the native aortic valve (haematoxylin and eosin staining). Myxomatous change of the aortic valve without significant infiltration of the inflammatory cells was seen. LVOT, left ventricular outflow tract; RCA, right coronary artery.

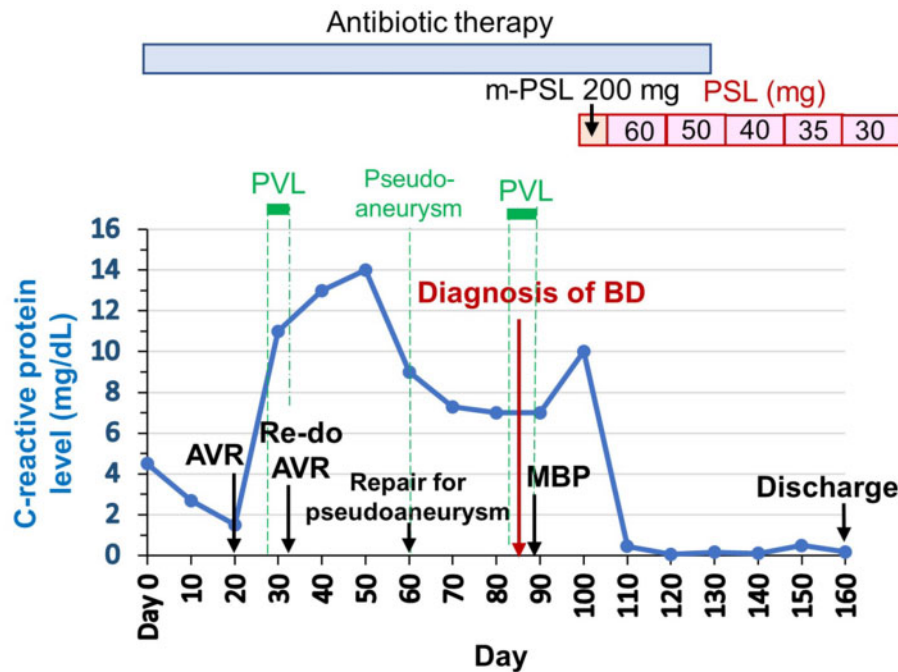


Figure 5 Intraoperative findings of the modified Bentall procedure. The prosthetic mechanical valve was detached due to extensive abscess involving the annulus tissue. The proximal end of a straight graft was sutured to the left ventricular outflow tract myocardium instead of the annulus tissue. MBP, modified Bentall procedure; m-PSL, methyl-prednisolone; PVL, perivalvular leak.

age of disease onset (<25 years) are major risk factors for clinical severity in BD.⁴

Behçet's aortitis involving the aortic root is extremely rare, and sometimes results in the formation of a sterile vegetation or perivalvular abscess as well as significant AR, mimicking aortic valve IE.^{1,2,5-7} Due to the fragility of the aortic annulus, which is further driven by refractory inflammation by surgical invasion, recurrent prosthetic valve detachment is a common complication of surgery for AR with BD, especially in the absence of immunosuppressive therapy (Supplementary material online, Figure S1).⁸ Therefore, early and confirmed diagnosis of aortic valve involvement in BD is necessary to determine the appropriate treatment strategy (Supplementary material online, Figure S2). In this case, persistent increase in CRP level after the initial surgery despite antibiotic use, combined with the preoperative echocardiographic findings, strongly suggested the possibility of non-infectious aortitis involving the aortic valve.

Modified Bentall procedure, in which the valved conduit is proximally sutured to the LVOT instead of the fragile aortic annulus, aids in preventing prosthetic valve detachment and improving surgical outcomes in patients with Behçet's aortitis because the LVOT myocardium is rarely affected by BD.⁹ This technique may also contribute to decreased tension on button coronary anastomosis, resulting in a low incidence of coronary artery kinking or anastomotic pseudoaneurysm.¹⁰ Immunosuppressive therapy is essential for controlling inflammation and improving surgical outcomes in patients with BD. However, postoperative steroid administration is controversial because of its characteristic of increasing susceptibility to deep infection.¹¹ In our case, immunosuppressive therapy was immediately

initiated after the removal of the mediastinal drains, resulting in an uneventful postoperative course.

In conclusion, the echocardiographic findings of Behçet's aortitis involving the aortic root can resemble those of IE of the aortic valve. Physicians should be aware of this to avoid delays in diagnosis and improper treatment. Modified Bentall procedure combined with effective immunosuppressive therapy may be useful in preventing prosthetic valve detachment.

Lead author biography



Kaoru Hattori, MD received the PhD degree from Graduate School of Medicine, Hirosaki University in 2017. She is currently working as a Clinical Fellow in Cardiovascular Surgery department at Yamato Seiwa Hospital, Kanagawa, Japan. Her research interest is mainly focused on aortic valve disease and aortic valve-related aortopathy, particularly associated with bicuspid aortic valve.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

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