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CASE REPORT
Cardiovascular surgery

Flanged Bentall procedure for paravalvular leakage and pseudoaneurysm after root replacement in Behcet's disease and infective endocarditis: a case report

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Background Behcet's disease is a multi-systemic inflammatory disorder. Paravalvular leakage and aortic pseudoaneurysm are rare in patients with Behcet's disease after aortic root replacement. Complicated post-operative infective endocarditis can make the treatment more difficult. We applied a flanged Bentall procedure to treat one such case.

Case summary A 27-year-old man with aortic regurgitation and Behcet's disease underwent aortic root replacement. Post-operative electrocardiogram showed a complete atrioventricular block. One year after the operation, he underwent percutaneous temporary pacemaker implantation and endovascular stent graft exclusion because of pseudoaneurysm of the ascending aorta. Post-operative fever and blood culture confirmed infective endocarditis. Examination showed paravalvular leakage and pseudoaneurysm recurrence. Then, the patient underwent a third operation in our hospital. Aortic root replacement with a flanged composite valved conduit was performed. Immunosuppressants and antibiotic treatment were given after surgery. After 3 months, the cardiovascular examination was normal, and the patient was in good condition.

Discussion Surgical treatment of aortic regurgitation caused by Behcet's disease was characterized by a high rate of paravalvular leakage, which led to reoperation and high mortality. Combined infective endocarditis would further increase the difficulty and risk of treatment. It is important to maintain effective immunosuppressive therapy while monitoring serum biomarkers and inflammation indicators. The potential hazards of immunosuppressants are increased risk of infection and poor tissue healing. In our case, targeted antibiotic treatment and appropriate immunosuppressive therapy were well balanced. The flanged Bentall procedure was also the key to success, which could increase aortic effective orifice area and reduce the risk of dehiscence.

Keywords Behcet's disease • Infective endocarditis • Paravalvular leakage • Pseudoaneurysm • Flanged Bentall • Case report

ESC curriculum 4.1 Aortic regurgitation • 7.5 Cardiac surgery • 9.1 Aortic disease

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Learning points

- To understand the importance of effective immunosuppressive therapy and targeted antibiotic treatment of paravalvular leakage due to Behcet's disease and infective endocarditis.
- To recognize that the flanged Bentall procedure could increase aortic effective orifice area, reduce the risk of dehiscence, and be helpful for exposure and haemostasis.
- To understand the importance of education for patients with continuous immunosuppressive therapy and monitoring of Behcet's disease.
- To recognize that open surgery rather than endovascular stent therapy should be the first choice for pseudoaneurysm of ascending aorta.

Introduction

Behcet's disease is a multi-systemic inflammatory disorder of unknown aetiology with high incidences in Asian, Middle Eastern, and Mediterranean populations.[1] Paravalvular leakage and aortic pseudoaneurysm are rare in patients with Behcet's disease after aortic root replacement.[2,3] Post-operative paravalvular leakage of patients, complicated with infective endocarditis, can make the treatment more difficult. Sub-annular endomyocardial implantation of a flanged composite valved conduit is beneficial to treat these patients.[4] We applied flanged Bentall procedure to treat one such case.

Summary figure

Time	*Events*
Before first presentation	Exertional dyspnoea for 6 months. Recurrent oral and genital ulceration for 5 years.
The first operation in local hospital	Aortic root replacement with a 20 mm Medtronic ATS mechanical valve and a 26 mm graft. Post-operative electrocardiogram showed complete atrioventricular block.
Six months after the first operation	The cardiovascular examination was normal and the patient stopped prednisone by himself.
One year after the first operation	Sudden chest pain. Computed tomography revealed a pseudoaneurysm.
The second operation in local hospital	Endovascular stent graft exclusion and percutaneous temporary pacemaker implantation.
Four days after the second operation	Fever. Blood cultures were positive for *Streptococcus viridis* and *Streptococcus oralis*.
Four months after the second operation	Transoesophageal echocardiography showed paravalvular leakage. Computed tomography showed pseudoaneurysm recurrence and type I endoleak.
The third operation in our hospital	Aortic root replacement with a flanged composite valved conduit (flanged Bentall). Immunosuppressants and antibiotic treatment lasted after surgery.
Three months follow-up	The cardiovascular reexamination was normal. No heart failure hospitalizations nor major cardiovascular events.

Case presentation

A 27-year-old man went to the local hospital because of exertional dyspnoea for the past 6 months. Oral diuretics were given. He had a history of recurrent oral and genital ulceration for 5 years. Imaging examination showed aortic regurgitation and aortic root aneurysm. He underwent aortic root replacement with a 20 mm Medtronic ATS mechanical valve and a 26 mm graft. During the operation, the thickened aortic cusps were perforated and changes of arteritis were obvious. There was no obvious annulus at the attachment of non-coronary cusp, so the suture was placed on the left ventricular outflow tract. Post-operative electrocardiogram showed a complete atrioventricular block. His heart rate was about 40 b.p.m., without dizziness or syncope, so a permanent pacemaker was not implanted. The histopathology of resected tissue showed fibromyxoid valvulopathy with leucocyte infiltration. Behcet's disease was diagnosed, and oral prednisone (10 mg/day) was administrated after surgery. After 6 months, the cardiovascular examination was normal, and the patient stopped prednisone by himself.

One year after the operation, the patient was admitted to the local hospital due to sudden chest pain. Computed tomography revealed a pseudoaneurysm at the distal anastomosis of the ascending aorta. He underwent percutaneous temporary pacemaker implantation and endovascular stent graft exclusion. Two 36 mm tube grafts were deployed. The patient had fever 4 days after endovascular intervention. Blood cultures were positive for *Streptococcus viridis* and *Streptococcus oralis*. The temporary pacemaker was removed. Targeted antibiotic therapy according to drug sensitivity results was initiated. During the next 4 months, the patient's temperature fluctuated between 36°C and 38°C. Laboratory examination showed a white blood cell count of 1.06–8.58 $*$ 10^9/L (reference range: 4–10 $*$ 10^9/L), a haemoglobin of 90–111 g/L (reference range: 120–160 g/L), a B-type natriuretic peptide of 1400–1900 pg/mL (reference range: 0–150 pg/mL), a C-reactive protein (CRP) of 47–63 mg/L (reference range: 0–8 mg/L), and erythrocyte sedimentation rate (ESR) of 12–36 mm/h (reference range: 0–15 mm/h). Subsequent blood cultures were negative. Transoesophageal echocardiography (*Figure 1*) showed paravalvular leakage and dysfunction of one mechanical valve leaflet. Computed tomography showed pseudoaneurysm recurrence and type I endoleak. Then, the patient was referred to our hospital.

For cardiopulmonary bypass, femoral arterial cannulation and atrial cannulation were used. The pseudoaneurysm was 6 cm $*$ 4 cm, and the adhesion around the aorta was severe. After cross-clamping of the aorta, the aneurysm was incised, and the malfunctional valved conduit was removed. The aortic root replacement with a flanged composite valved conduit (flanged Bentall; *Figure 2*) was performed.[4] A 22 mm supra-annular mechanical valve Medtronic AP360 and a 26 mm vascular graft Maquet were used. The flange of the conduit (5 mm in length) was implanted to the aortic annulus with interrupted pledgeted sutures. The Cabrol technique with an 8 mm graft was preferred due to tension on the left coronary anastomosis. The right coronary button was anastomosed end-to-side to the composite graft. The distal anastomosis of the graft to the transected aorta was performed using a continuous 4–0 polypropylene suture. After surgery, a permanent pacemaker was implanted. Immunosuppressants included oral prednisone (1 mg/kg/day) and cyclophosphamide (100 mg/day). Cultures of aortic tissues and grafts were negative.

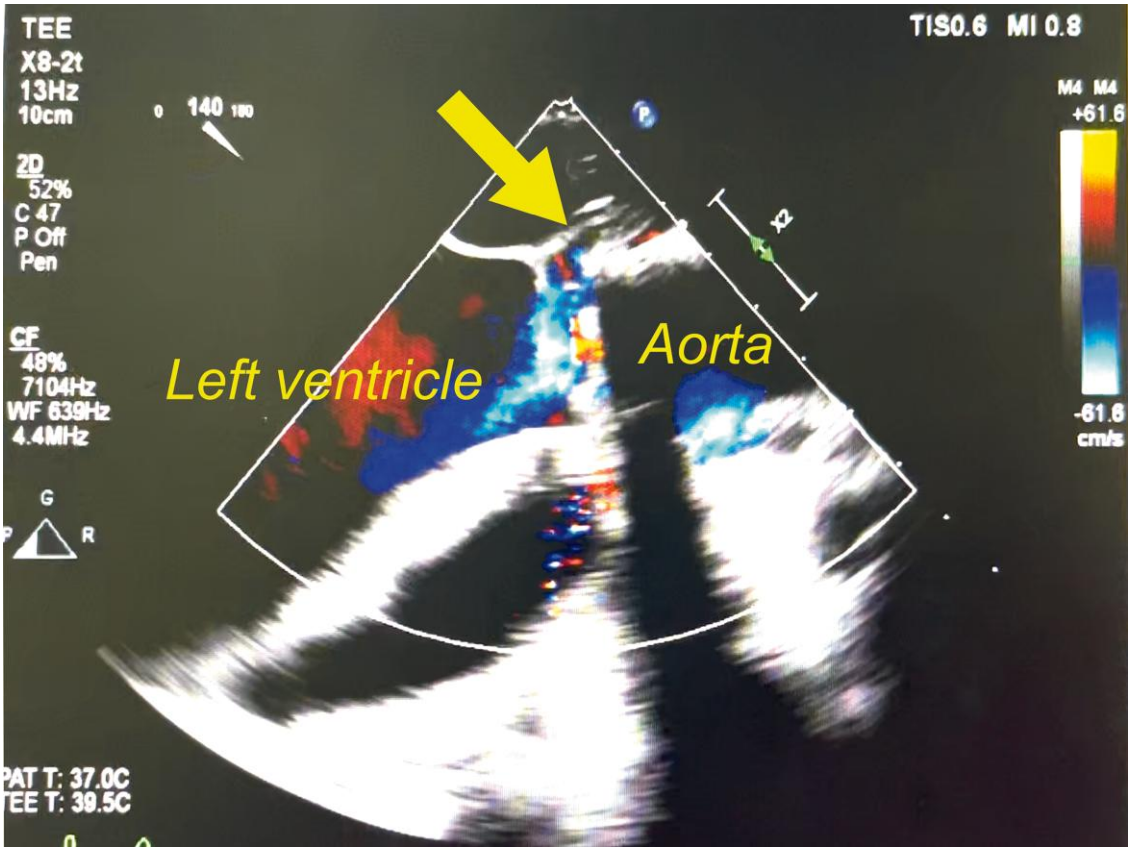

Figure 1 Transoesophageal echocardiography before the third operation. The arrow shows the leakage area around the valve.

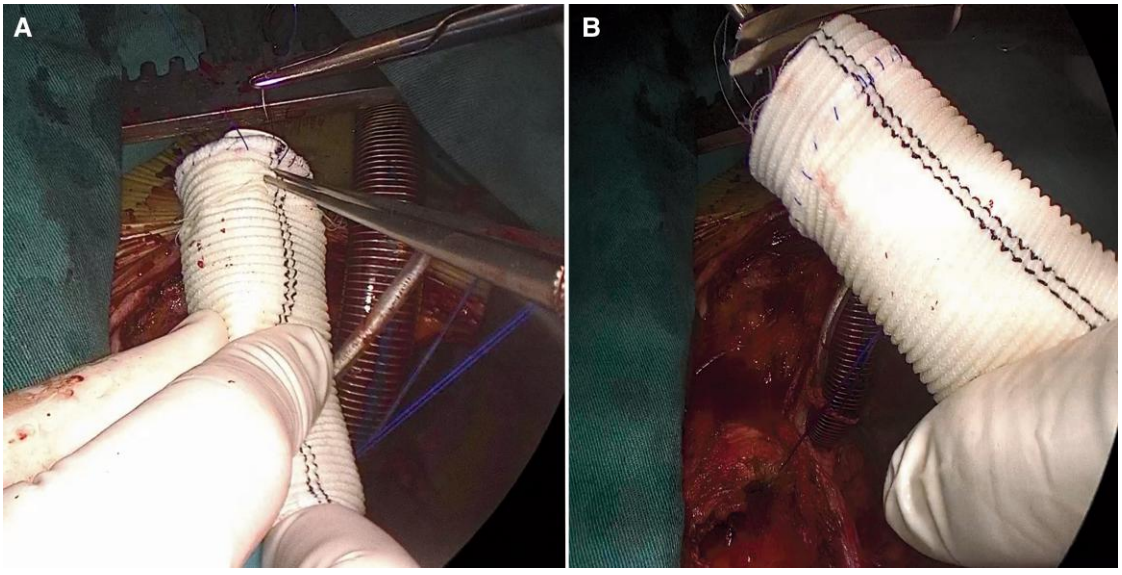

Figure 2 Preparation of a flanged composite graft. (*A*) A segment of the vascular graft is everted outward to form the flange. A mechanical valve is inserted and sutured to the graft with a continuous 4–0 polypropylene. (*B*) The flange of the conduit is returned to its original position, and the flanged composite graft is ready.

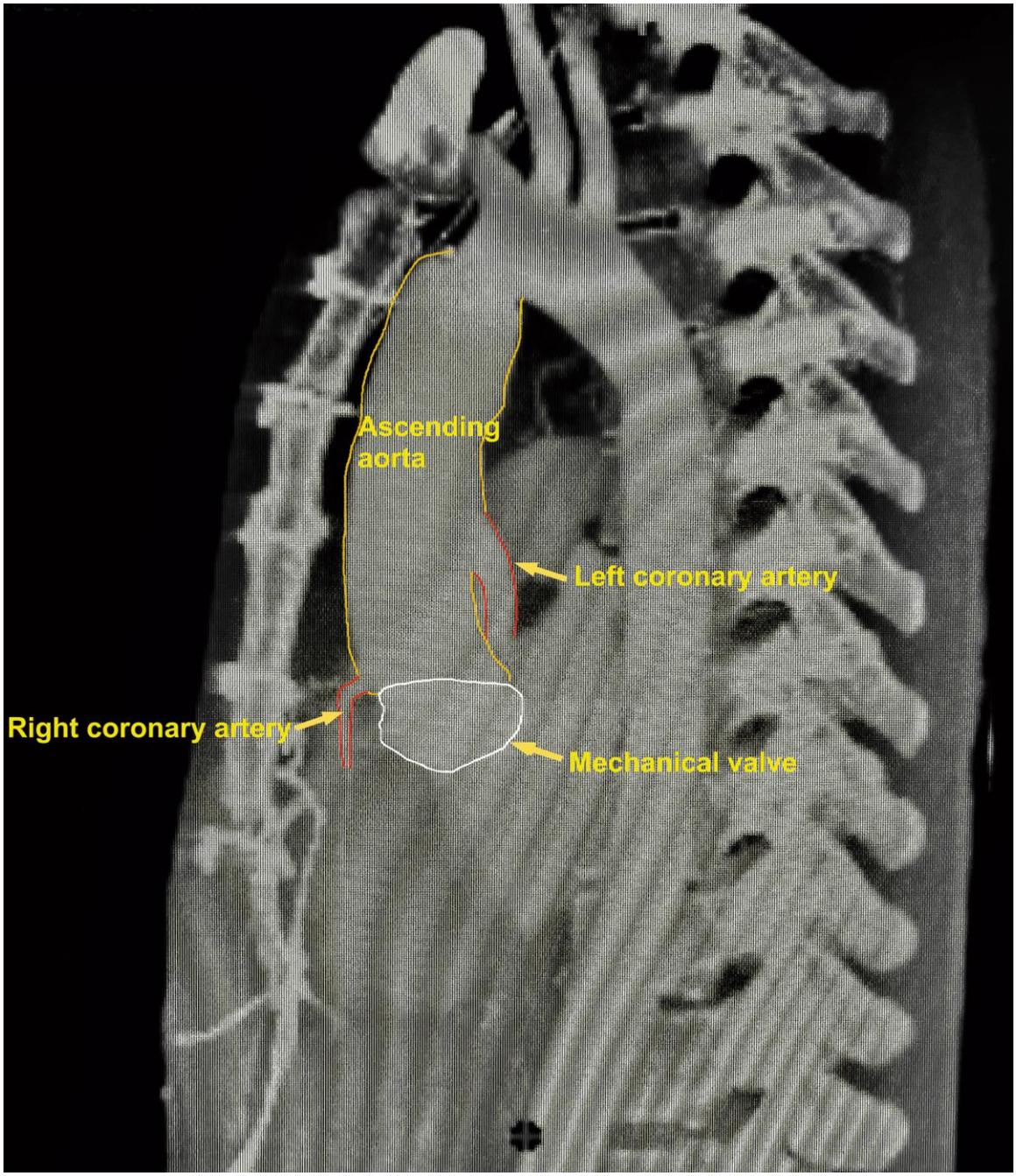

Figure 3 Computed tomography during follow-up after the third operation.

Antibiotic treatment (ceftriaxone) lasted for 8 weeks. After 3 months, the cardiovascular examination was normal, and the patient was in New York Heart Association functional class I. Computed tomography during follow-up was normal (*Figure 3*).

Discussion

Behcet's disease complicated with infective endocarditis is rare. Surgical treatment of aortic regurgitation caused by Behcet's disease was characterized by a high rate of paravalvular leakage, which led to reoperation and high mortality.[2,5] Combined infective endocarditis would further increase the difficulty and risk of treatment. Ghang *et al.*[5] reported their surgical experience of 23 patients with Behcet's disease. One case had post-operative bacterial mediastinitis and underwent a re-operative bioprosthetic root replacement. Guo *et al.*[6] reported their experience of 644 Behcet's disease patients. Only one patient developed infective endocarditis and paravalvular leakage in the third month after the first operation and died suddenly before reoperation. Jeong *et al.*[2] reported their

results of 19 patients with aortic regurgitation attributable to Behcet's disease. 36.8% (7 of 19) of their patients were suspected to have infective endocarditis based on echocardiographic findings and elevated inflammatory markers, but no microorganisms were cultured. Only one of them survived. It is important to maintain effective immunosuppressive therapy while monitoring serum ESR and CRP levels. However, the potential disadvantage is an increased risk of infection and poor tissue healing. In our case, targeted antibiotic treatment and appropriate immunosuppressive therapy were well balanced. The pseudoaneurysm occurred after the patient stopped the medication on his own and was caused by the recurrence of Behcet's disease. The occurrence of paravalvular leakage may be related to both the recurrence of Behcet's disease and infective endocarditis. Drug treatment of prosthetic valve endocarditis should last ≥6 weeks and the duration of treatment is based on the first day of effective antibiotic therapy, as recommended by the ESC guidelines.[7] After the third operation of our patient, targeted antibiotic treatment lasted for 8 weeks, and blood cultures were negative during follow-up.

We used the flanged Bentall procedure in this case. The artificial graft has better elasticity, which is conducive to implantation of a larger valve. The diameter of the aortic annulus in this patient was only 20 mm. We implanted a 22 mm Medtronic AP360 supra-annular mechanical valve. The aortic effective orifice area of this patient increased from 1.3 to 2.1 cm^2. In Behcet's disease, the recurrent inflammatory changes lead to fragility in both the aortic annulus and wall. Sub-annular endomyocardial implantation of a flanged composite valved conduit is beneficial to prevent paravalvular leakage. The flange of the composite graft could decrease tension and vibration of the suture ring in the whole cardiac cycle, thereby reducing the risk of dehiscence. It is also beneficial to the exposure and haemostasis of aortic root.[8]

There are two lessons to learn. First, Behcet's disease is characterized by relapses and remissions. Education for patients is essential. Immunosuppressive therapy must not be stopped at will. It is important for patients to pay enough attention to the disease and comply with the treatment. Second, open surgery rather than endovascular stent therapy should be the first choice for pseudoaneurysm of ascending aorta.[9] If this patient had not received endovascular stent therapy, he may have not suffered from infective endocarditis.

This is a case of the treatment of paravalvular leakage and pseudoaneurysm due to Behcet's disease and infective endocarditis. The flanged Bentall procedure was used. The short-term results are good, although long-term follow-up is necessary.

Patient perspective

Behcet's disease is characterized by relapses and remissions. Immunosuppressive therapy must not be stopped at will. It is important for patients to pay enough attention to the disease and comply with the treatment.

Lead author biography

Dr Xiangyang Qian has worked for 26 years in Fuwai Hospital, National Center for Cardiovascular Diseases, Beijing, China. He has been a chief physician of cardiovascular surgery since 2012. His main areas of interest include aortic surgery and cardiac valvular surgery, especially in the treatment of complex diseases such as acute aortic type A dissection, aortic root aneurysm, complex aortic arch lesions, and total thoracoabdominal aortic aneurysm.

Supplementary material

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

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: None declared.

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Data availability

The data underlying this article are available in the article.

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