Morbus Behçet associated aortitis mimicking infective endocarditis

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Summary

A patient with Morbus Behçet was admitted with severe symptomatic aortic regurgitation caused by aortitis that mimicked infective endocarditis. After aortic valve replacement and antibiotic treatment, regurgitation and systemic inflammation recurred. Infection could be ruled out and the patient was finally treated with immunosuppressive therapy and repeat aortic valve replacement using a mechanical composite graft. Morbus Behçet associated aortitis is rare but may mimic infective endocarditis. This is an important fact for clinicians and surgeons involved in the treatment of suspected endocarditis and valve replacements.

Key words: Morbus Behçet; aortitis; infective endocarditis; aortic valve replacement; aortic regurgitation

Case report

A 44-year-old patient of Serbian origin was admitted to the emergency department with severe dyspnoea, which had begun three days earlier. The patient was suffering from Behçet's disease treated with methotrexate, prednisone and colchicine. Clinical examination revealed signs of decompensated heart failure and chest X-ray confirmed pulmonary oedema. The patient was intubated, mechanically ventilated and urgently transferred to our hospital. Transoesophageal echocardiography showed severe aortic regurgitation due to a destroyed non-coronary cusp and a large mobile vegetation (fig. 1A). Laboratory tests showed leucocytosis of 19.5 G/L, elevated C-reactive protein (53.7 mg/L) and an accelerated blood sedimentation rate (84 mm/h). The body temperature was 39 degree Celsius. Infective endocarditis was suspected, blood cultures were drawn and empiric antibiotic therapy was initiated. Due to refractory cardiogenic shock, urgent surgical replacement of the aortic valve (bioprosthetic Edwards Perimount Magna Ease valve) was performed. Intraoperatively the non-coronary cusp was found to be destroyed with a big vegetation attached to it. Adjacent to the destroyed cusp the surgeon found a small cavity, presumably an abscess of the aortic root. Therapy for Behçet's disease was continued as described above.

Figure 1
A: Mobile vegetation in the left ventricular outflow tract attached to the non-coronary cusp (transoesophageal echocardiography).
B: Severe paraprosthetic aortic regurgitation (yellow-blue flow) seen on transthoracic echocardiography in the parasternal long axis view.

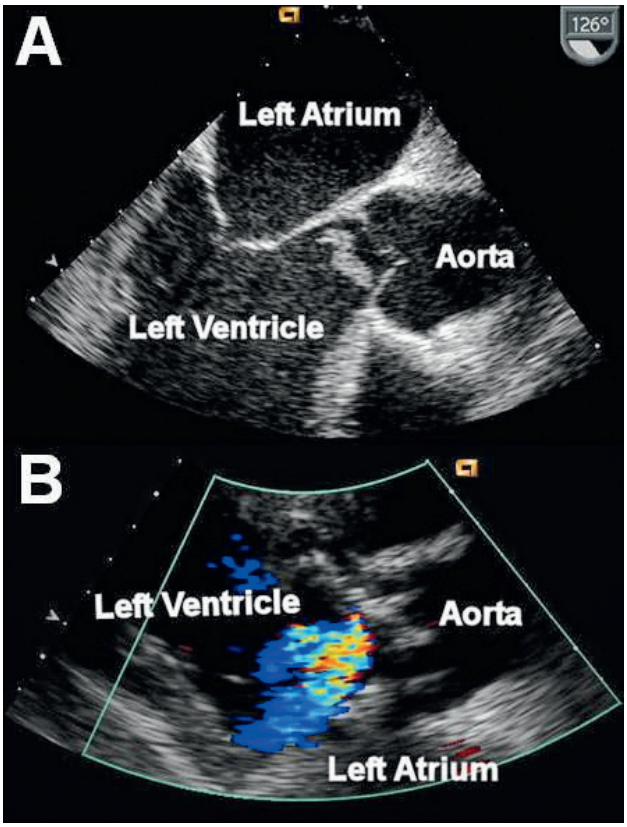

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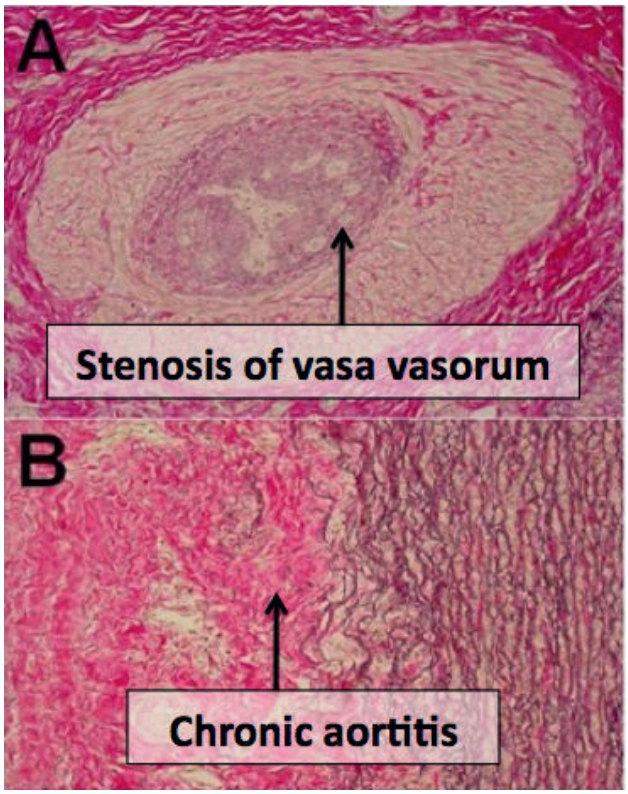
Figure 2
A: Severely stenosed vasa vasorum in the adventitia of the ascending aorta (Elastica-Van-Gieson-stain, 200 x). B: Chronic aortitis with disintegration of elastic fibres in the media of the ascending aorta (Elastica-Van-Gieson-stain, 200 x).



Blood cultures and serological tests for microorganisms associated with infective endocarditis remained negative. The retrieved vegetation was of a non-specific inflammatory nature and eubacterial polymerase chain reaction of the specimen was negative. Antibiotic therapy with amoxicillin, clavulanic acid, rifampicine and gentamicin was administered for four weeks while immunotherapy for Behçet's disease remained unchanged. Two months after replacement of the aortic valve, while the patient had more or less recovered, he presented to the emergency department again because of progressive dyspnoea over a few days. Transthoracic echocardiography showed a well functioning aortic valve prosthesis with trivial transvalvular but severe paraprosthetic regurgitation (fig. 1B). Inflammatory markers, including C-reactive protein (44 mg/L) and leukocyte count (18.1 G/L), were again markedly elevated. Six blood cultures were drawn, but remained sterile. A new working hypothesis assuming Behçet-associated aortitis mimicking infective endocarditis was proposed [1–3]. Hence, immunosuppressive therapy with high doses of prednisone, azathioprine and infliximab was begun without any additional antibiotics. Inflammatory markers normalised within one week of treatment. Due to the severity of the paraprosthetic regurgitation a conservative approach, as previously described [1], could not be pursued. After two weeks of intensified immunosuppressive therapy, elective surgical aortic root and valve replacement was performed using a mechanical composite graft (ATS Medical). Sutures were reinforced with a xenopericardial layer to prevent recurrent leakage. Histopathological examination of the excised aorta (fig. 2A/B) showed chronic aortitis characterised by dense fibrosis, small collections of lymphocytes and plasma cells, disintegration of elastic lamellas and stenotic vasa vasorum. Furthermore fragmented and thinned elastic fibres in the tunica media were documented [4]. These findings are compatible with Morbus Behçet associated aortitis, although overlapping morphological features are found in different aortic diseases (e.g. Takayasu, syphilis).

Two months later the patient was doing well and transthoracic echocardiography showed an unremarkable composite graft.

Discussion

Morbus Behçet is a systemic autoimmune disease characterized by inflammation of small blood vessels. As there is no specific diagnostic test, the diagnosis is based on clinical signs [5] and exclusion of other inflammatory vascular aetiologies (e.g. lupus, rheumatoid arthritis). Common features include painful oral and genital ulcers, inflammatory eye disease, erythema nodosum and neurological involvement. Cardiovascular manifestations of Behçet's disease are rare but manifold and may include pericarditis, coronary artery vasculitis with myocardial infarction, cardiomyopathy, thromboembolism, aortic aneurysm and aortitis [6]. Morbus Behçet's associated aortitis may involve the aortic valve with features mimicking infective endocarditis [1–3]. It is important to consider this possibility in patients with known or suspected Behçet's disease [1–3]. In these cases, the therapeutic goal is suppression of vascular inflammation by intensive immunosuppressive therapy. If aortic valve replacement is indicated, a composite graft or Bentall-type operation [4, 7], instead of solely bioprosthetic valve replacement, should be considered to eliminate the possibility of recurrent aortitis.

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