Case report

Mitral and aortic regurgitation in Behçet's syndrome

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SUMMARY A 35-year-old man with definite Behçet's disease developed acute mitral and aortic regurgitation. Valvular disease, we believed, was another manifestation of this disease. Cardiac involvement in Behçet's disease and the role of prednisone therapy are discussed.

Behçet's syndrome is now recognised as a multisystem disease characterised by a relapsing inflammatory process of uncertain aetiology.¹ Whereas there is abundant literature on vascular disease, only recently has cardiac involvement been recognised as a feature.²

Case report

A 35-year-old man was found to have Behçet's syndrome in July 1981. The diagnosis was made on the basis of recurrent painful oral and genital aphthous ulcers, asymmetrical polyarthritis, subcutaneous thrombophlebitis, hyper-reactivity of the skin (positive 'pathergy' test), and iridocyclitis. Prednisone was started and then slowly decreased.

Seven months later the patient developed a new outbreak of ulcers and arthritis. He denied fever, dyspnoea, or chest pain. On admission the physical findings included: blood pressure 130/70 mmHg, temperature 36.5°C, pulse of 120/min, and several painful oral and genital aphthous ulcers. There was tenderness over the ankles, more prominent on the right side. Cardiac auscultation and phonocardiography showed a grade 3/6 pansystolic murmur with an S₃ at the apex, a grade 3/6 systolic ejection murmur, and a grade 3/6 diastolic murmur along the left sternal border (Fig. 1). The chest radiograph showed mild cardiomegaly without venous pulmonary congestion. The electrocardiogram showed no abnormalities except sinus tachycardia. Severe valvular mitral and aortic dysfunction, fluttering of the mitral valve, and mild left ventricular dilatation were identified by M mode and 2-D

echocardiography. No vegetations were found. Nine blood cultures (without prior antibiotic therapy) and tests for antinuclear antibodies, rheumatoid factor, brucella, and rickettsia were negative. The streptozyme test, Venereal Disease Research Laboratory test, and fluorescent treponeme antibody absorption test also proved negative. Prednisone 120 mg daily was started, and four weeks later signs of aortic regurgitation disappeared, though mitral insufficiency continued without S₃ (Fig. 1). Cardiac catheterisation carried out at this time showed mild mitral regurgitation but no aortic regurgitation. Endomiocardial biopsy disclosed non-specific fibrosis. Prednisone was gradually reduced and finally discontinued. Three years later the mitral insufficiency murmur still persisted together with mild dilatation of the left ventricle, but clinically the patient was free of symptoms.

Discussion

Our patient fulfilled the criteria proposed for Behçet's syndrome (BS) by various authors.¹⁻³ Seven months after the diagnosis he developed an acute mitral and aortic insufficiency. The absence of findings of other collagen vascular diseases and the exclusion of infective endocarditis, the relationship with a flare of the systemic disease process, and the outcome with immunosuppressive therapy alone (without antibiotics) led us to conclude that valvular disease was secondary to BS itself.

Fainaru and Chajek stated 'Cardiac involvement in BS is so rare that it may be coincidental'.⁴ However, myocarditis has been reported in a few patients,⁵ also pericarditis.⁵ Davies found chronic inflammation and thrombosis on the right ventricle,⁶ and Buge reported endocardial fibrosis.⁷ Two cases

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Fig. 1 The upper tracing shows a holosystolic murmur of mitral regurgitation in the mitral area and an early systolic murmur followed by an early diastolic murmur in the aortic area. The carotid pulse (CG) synchronous with the phonocardiogram is normal. The lower tracing recorded one month later shows the disappearance of the early diastolic murmur. The holosystolic mitral murmur remains unchanged.

of endocarditis, one of chronic aortic regurgitation and one of severe, acute aortic regurgitation and sinus of Valsalva's aneurysm,⁸⁻¹⁰ have been reported. Our patient presents further evidence that valvular disease should be added to the spectrum of BS, and therefore we suggest that patients with BS should be evaluated carefully for signs of valvular disease.

The role of corticosteroids in the management of valvular involvement of BS is uncertain owing to the small number of reported cases. Although we cannot exclude the possibility that the aortic regurgitation may have resolved spontaneously, high dose prednisone therapy did lead to a partial resolution of the valvular disease. Therefore we

recommend a trial of this drug for those patients with BS and presenting symptoms of related valvular disease, with the aim of postponing or even avoiding valve replacement.

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