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Valve prolapse in Behçet's disease

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SUMMARY Valve prolapse was diagnosed solely by echocardiography in three consecutive patients with Behçet's disease. Two patients had prolapse of the posterior mitral valve leaflet, but no clinical manifestations of valve prolapse. In the third patient aortic valve prolapse was associated with physical signs of aortic regurgitation and left ventricular failure.

Valve prolapse in these cases may have resulted from structural and functional derangement caused by the underlying non-specific vasculitis that occurs in Behçet's disease.

Behçet's disease (or syndrome) is now recognised to be an autoimmune disorder showing evidence of multisystem involvement and widespread vasculitis as its basic pathohistological features.[1–4] Associated cardiovascular lesions have been well documented in recent years and have actually been considered as the major cause leading to fatal outcome.[5 6] Prolapse of cardiac valves is one of the complications not previously described. This report presents the brief histories, related clinical data, and echocardiographic features indicating mitral or aortic prolapse in three consecutive cases and suggests the frequent development of such pathological changes in the course of Behçet's disease.

Patients and methods

CASE REPORTS

Case 1—A 36 year old woman presented to our clinic in 1975 suffering from an intermittent low grade fever associated with oral and genital ulcers, conjunctivitis, erythema nodosum on both legs, and arthralgia. Physical examination of the cardiovascular system was normal. The erythrocyte sedimentation rate was 34 mm in the first hour. The electrocardiogram showed frequent ventricular extrasystoles. She was treated with oral steroids with good symptomatic relief. In December 1979 segmental colitis was diagnosed at laparotomy. The cardiovascular manifestations appeared in April 1982 when she had attacks of palpitation and pronounced dyspnoea, and typical echocardiographic signs of mitral posterior leaflet prolapse were found. Hypertension was diagnosed in February 1983.

Case 2—A 63 year old woman was admitted in August 1978 with a history of fever (up to 39°C) a month before and painful orogenital ulcers three days after the onset of fever. Physical examination showed severe anaemia, bilateral conjunctival hyperaemia, small ulcers on the buccal mucosa and tip of the tongue, and dark red nodules on both legs. Her erythrocyte sedimentation rate was 120 mm in the first hour. A skin biopsy specimen from the left forearm showed thickening and mild inflammatory cell infiltration of the wall of some small veins. After admission her symptoms were controlled with antipyretic and antibiotic drugs. She was discharged in September 1978. Follow up examinations in 1983 detected no abnormal findings, but a cross sectional echocardiogram showed typical signs of mitral valve prolapse (Figure).

Case 3—A 58 year old man was seen in 1969 with low grade fever, ulcers in the mouth, chronic conjunctivitis, and erythema nodosum on the dorsum of the feet. In August 1982 he began to experience frequent attacks of precordial tightness without any obvious precipitating causes. The electrocardiogram showed left anterior fascicular block and left ventricular hypertrophy. He was admitted in January 1984 with shortness of breath and orthopnoea, mild oedema of both legs, and multiple small ulcers on the buccal mucosa, tongue, and external genitalia. His blood pressure was 140/80 mm Hg and the cardiac rhythm regular. There was a blowing diastolic murmur of moderate intensity at the left sternal border (second and third intercostal spaces). The erythrocyte sedimentation rate was 54 mm in the first hour. Immune complexes were present in the blood, and he

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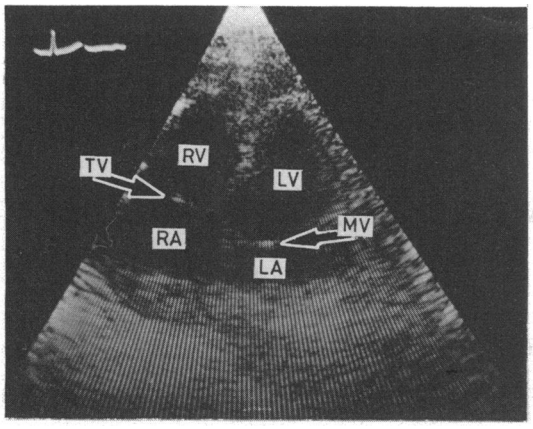

Figure *Case 2: Cross sectional echocardiogram in the apical view of four cardiac chambers showing the mitral valve (MV) bulging beyond the atrioventricular annulus into the left atrium (LA) during mid and late systole. RV, right ventricle; LV, left ventricle; RA, right atrium; TV, tricuspid valve.*

had raised concentrations of serum IgA and IgG. The cross sectional echocardiogram showed prolapse of the right coronary cusp of the aortic valve and associated aortic incompetence. After admission, oral steroids, cardiotonics, and diuretics were given to improve cardiac function. but cardiac insufficiency and pulmonary congestion were refractory to these measures. Oral ulceration could be controlled only with cyclophosphamide.

ECHOCARDIOGRAPHIC FINDINGS

In cases 1 and 2 the mid and late systolic hammocking of the mitral valve was shown on the M mode echocardiogram and in the cross sectional apical four chamber view (Figure). The mitral leaflets bulged beyond the atrioventricular valve annulus into the left atrium during systole. In case 3 prolapse of the aortic valve cusp during diastole was shown on the cross sectional image of the heart in the long parasternal axis view.

Discussion

The clinical features of all three cases fulfil the diagnostic criteria of the complete type of Behçet's disease. The cardiovascular manifestations in Behçet's disease have attracted attention in recent years. The incidence as reported by James was around 30%, and the vascular lesions appeared to be the major cause of death in those with such complications.[3 5 6] Valve prolapse occurred in three consecutive patients, who make up 50% of our total series of six patients with Behçet's disease. This frequent association of the two conditions in the absence of other obvious aetiological factors suggests that a causal relation may exist. Recently, it has been suggested that mitral valve prolapse may be an autoimmune disease.[7] It occurs in collagen vascular diseases such as polyarteritis nodosa, scleroderma, and systemic lupus erythematosus and in disorders of collagen formation such as Marfan's syndrome.[8 9] We consider that the valve prolapse in the present three cases may have been the result of structural damage and functional derangement caused by the underlying non-specific vasculitis of Behçet's disease, but histological examination would be necessary to confirm or refute such a hypothesis.

The aortic regurgitation detected in case 3 caused cardiovascular deterioration, and hence such prolapsing valve lesions may have important prognostic consequences. The situation was less serious in the two patients with mitral valve prolapse. In both there were no physical signs of prolapse, and the diagnosis was made purely on echocardiographic grounds. Such an absence of physical signs is common in patients with mitral valve prolapse due to any cause. The clinical significance of echocardiographically documented prolapse in Behçet's disease is at present unknown, but follow up will establish whether such patients develop overt mitral regurgitation or the other complications of mitral valve prolapse such as arrhythmias, emboli, and bacterial endocarditis. In the meantime it seems sensible, in a multisystem disorder like Behçet's disease, to document the presence or absence of cardiac valve prolapse by routine echocardiographic screening.

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