Aortic regurgitation and false aortic aneurysm formation in Behçet's disease

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Summary

Behçet's disease is associated with vascular complications. Aortic regurgitation associated with this condition has never previously been described. A case is now reported in which this occurred and which was followed postoperatively by a false aneurysm of the ascending aorta.

Case report

A 26-year-old man was referred to out-patients with severe leg ulceration in December 1974. He had had mouth and scrotal ulceration since the neonatal period and had observed that minor skin trauma was invariably followed by sepsis. He had no symptoms referable to the cardiovascular system and there was no past history of rheumatic fever.

There was a positive family history in that his mother, brother and nephew all had episodes of mucocutaneous ulceration.

He was thin and pale. There were no skeletal features of Marfan's syndrome. He had a single aphthous ulcer on his tongue and a symmetrical eruption over the extensor aspect of his lower limbs which comprised bluish papules and nodules; some hyperkeratotic and some pustular. Annular lichen planus was present at the base of the glans penis. In addition he had aortic regurgitation. BP was 140/50 mmHg.

Investigations were: Hb, 14.7 g/dl; WBC, $9.9 \times 10^9/l$; ESR, 10 mm/hr; urea and electrolytes, normal; serum proteins and electrophoresis, normal; serological tests for syphilis, negative; chest X-ray, normal; metacarpo-phalangeal index, normal; sacro-iliac joints, normal. Blood group, AB Rh-negative; tissue type: HLA A2.XB15.27/CW1. Skin biopsy showed non-specific epidermal reaction, dermal downgrowth and some degree of scarring. Immunofluorescence was negative.

From January 1975 to January 1978 he developed progressive cardiac failure which became unresponsive to digoxin and diuretic therapy. Cardiac catheterization and angiography confirmed severe aortic regurgitation, and in February 1978 he underwent aortic valve replacement with an aortic homograft. At operation it was noted that the aortic valve was scarred and perforated and did not resemble a valve damaged by rheumatic disease. Histology of the valve showed non-specific changes of irregular fibrosis, without an inflammatory infiltrate. The postoperative course was complicated by thoracotomy wound sepsis and several days of complete dysphagia due to oral ulceration.

He improved over the next 3 months but within 5 months marked deterioration had occurred. He was readmitted in July 1978 and was anaemic, pyrexial and in left ventricular failure. Two small pustules were present on the thoracotomy wound and there was a pulsatile mass to the right of the sternum. Signs of aortic regurgitation had recurred. The Hb had fallen to 10.2 g/dl, ESR had risen to 60 mm/hr and WBC was $14.4 \times 10^9/l$. Five blood cultures and 2 wound swabs were negative. Chest X-ray showed cardiomegaly and prominent dilatation of the ascending aorta.

Aortography established the diagnosis of a large false aneurysm of the ascending aorta. In August 1978 this was resected and the aorta was repaired. The false aneurysm communicated with the aortic lumen at the site of the previous aortic cannulation.

Histology of the resected aneurysm showed mainly fibrous tissue with focal calcification, proliferation of capillaries and lymphatic infiltration. Again, the postoperative course was complicated by mouth ulceration and superficial chest wound dehiscence.

He has remained well for the past 6 months (1979) since the second operation.

FIG. 1. Photomicrograph of section through aortic valve (Elastic Van Gieson).

Discussion

Behçet's disease was first described in 1937 as the triad of relapsing iritis and oral and genital ulceration. The disease is now well recognized as affecting other systems and Mason and Barnes enlarged the diagnostic features accordingly in 1969.

In the present patient, the diagnosis was made on the basis of 3 major criteria; relapsing oral and genital ulceration and pustular skin lesions and one minor criterion, the positive family history.

Reports of cardiovascular abnormalities have been limited to venous thrombosis (Enoch *et al.*, 1968), carditis (Lewis, 1964), and pericarditis (Sigel and Larson, 1965). Two cases of aortic aneurysms have been described (Hills, 1967; Mounsey, 1965), one case developed a false aneurysm of the lumbar aorta following osteomyelitis of the lumbar spine.

The authors believe the aortic regurgitation in their patient was secondary to his Behçet's disease. The macroscopic appearance of a scarred, perforated valve and the non-specific histology (Fig. 1) are rarely seen in valve disease due to congenital abnormalities, rheumatic fever or syphilis.

The development of a false aneurysm of the ascending aorta through the previous aortic cannulation site could also be secondary to the Behçet's disease. Poor tissue healing and sepsis are common in this condition. Both were present in this patient and both are well recognized precedents to false

aneurysm formation. In addition the histology of the resected aneurysm is similar to that reported by Enoch *et al.* (1968) and Hills (1967).

The patient is HLA B27 positive. This haplotype is found with increased frequency in Behçet's disease and in ankylosing spondylitis and Reiter's disease, when there is also an association with aortic regurgitation. No common pathogenic mechanism has yet been identified.

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