Cardiac involvement and superior vena caval obstruction in Behçet's disease

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Roguin, N., Haim, S., Reshef, R., Peleg, E., and Riss, E. (1977). Thorax, 33, 375-377.
Cardiac involvement and superior vena caval obstruction in Behçet's disease. A patient with Behçet's disease developed the rare complications of pericarditis and, two months later, superior vena caval obstruction. The latter complication was investigated by angiography to exclude other causes. As a result of this investigation the caval obstruction was managed conservatively, the symptoms and signs improving spontaneously.

Recent studies of Behçet's syndrome have identified a variety of symptoms besides the original triad of recurrent aphthous stomatitis, uveitis, and genital ulceration (Berlin, 1960; Haim, 1970; Haim *et al.*, 1974). However, cardiac involvement in this disorder has rarely been reported (Godeau *et al.*, 1972).

We report a haemodynamic and angiographic study of a patient with Behçet's disease who developed both pericarditis and superior vena caval syndrome.

Case report

A 35-year-old man was referred with a few days' history of fever, weakness, and chest pain accentuated by thoracic movements.

He was in good general health, with a temperature of 38°C, blood pressure 120/80 mmHg, and pulse regular, 110 per minute. There was no paradoxical pulse or Kussmaul sign and the lungs were clear. The first and second heart sounds were normal but a third heart sound and grade 2/6 systolic murmur were heard at the apex, and a pericardial friction rub was present over the precordium maximal at the lower left sternal border. The liver was palpable 6 cm below the costal margin. The electrocardiogram showed sinus rhythm, ST elevation in frontal and precordial leads, and, a few days later, diffusely negative T waves. Chest radiography revealed moderate cardiomegaly without signs of pulmonary venous hypertension.

The blood sedimentation rate was 42 mm in the first hour, C-reactive protein positive; the remaining routine laboratory investigations were normal.

Corticosteroid therapy was followed by a gradual improvement of symptoms. The fever and chest pain disappeared, as did the systolic murmur and the pericardial friction rub. A chest film 10 days after admission showed diminution of the cardiac shadow.

During his stay in hospital the patient developed painful ulceration in the mouth and on the genitals and also a follicular rash. These symptoms proved later to be of a recurrent nature. At the site of intracutaneous injection of normal saline solution an erythematous area with a pustule at its centre developed.

The diagnosis of Behçet's disease associated with pericarditis and suspected myocardial involvement was, therefore, established. The patient was discharged and readmitted two months later because of swelling of the neck and face, worsening after rest in bed. Physical examination at this time revealed marked jugular engorgement, mild cyanosis of the face, collateral venous circulation on the upper part of the chest, superficial thrombophlebitis at the site of venepuncture in the right arm, and a third heart sound at the apex. No murmurs or pericardial friction rub were present. The liver was palpable 1 cm below the costal margin, no oedema was detected in the legs, and the blood pressure was 120/80 mmHg. The blood sedimentation rate was 75 mm in the first hour; chest radiography revealed widening of the superior

mediastinum and a normal cardiac shadow. The electrocardiogram showed diffuse ST-T changes.

The results of cardiac catheterisation are shown in the Table. A catheter introduced via the left antecubital vein failed to pass to the lower part of the superior vena cava (SVC). A second catheter was, therefore, introduced via the left saphenous vein, and a heart catheterisation was performed. However, the second catheter failed to pass to the upper portion of the SVC. A 26 mmHg gradient was found at the site of the obstruction. Angiography revealed total obstruction of the SVC below the outlet of the azygos vein (Fig. 1) and recanalisation of the left innominate vein with rich collateral circulation (Fig. 2). Injection of contrast medium showed an intraluminal filling defect below the obstruction.

Table Haemodynamic data

Haemodynamic data	Pressure (mmHg)	Oxygen Sat. %
Left innominate vein	30 mean	
Right innominate vein	30 mean	
Superior vena cava (below obstruction)	4 mean	
Inferior vena cava	4 mean	
Right atrium	5 mean	76.5
	a:8 x:4 v:6 y:2	
Pulmonary artery	25/17	77
Right ventricle	30/10	75.5
Left ventricle	120/10	94
Left atrium	13 mean	94
Pulmonary vein	13 mean	94

Fig. 1 Superior venacavagram showing obstruction of the superior vena cava below the outlet of the azygos vein.

Fig. 2 Injection of contrast medium in the left subclavian vein showing recanalisation of the left innominate vein and rich collateral circulation.

The condition of the patient gradually improved without surgical intervention and he was finally discharged. During a follow-up period of 18 months no worsening of the cardiac state was noted; however, the patient continued to suffer recurrent orogenital ulceration and a follicular rash.

Discussion

The diagnosis of Behçet's disease in our case is based upon the presence of recurrent orogenital ulceration, folliculitis, and the positive cutaneous hypersensitivity reaction to intradermal injection of saline, which is a unique feature of this malady (Haim *et al.*, 1976).

Two findings lend this case a special interest: the pericardial involvement and the contribution of the haemodynamic and angiographic study in evaluating the prognosis and the management of the superior vena caval obstruction. The latter is an accepted complication of Behçet's disease (Haim *et al.*, 1971; Kansu *et al.*, 1972; Chajek and Fainaru, 1975), and, in the absence of other apparent aetiology, we feel justified in relating it to this disorder.

The literature reveals very few reports on cardiac involvement in Behçet's disease. Oshima *et al.* (1963) reported a patient with Behçet's disease associated with incomplete right bundle branch block; McMenemey and Lawrence (1957) diagnosed at necropsy a myocardial degeneration

in a patient with this disorder; and Lewis (1964) described a Behçet's patient with signs of pericarditis and myocarditis. Two additional cases were also reported by Godeau *et al.* (1972) and by Sigel and Larson (1965). The cardiac affection in our case presented clinical and electrocardiographic findings typical of an acute pericarditis. Besides, there were also indications of a possible myocardial damage as suggested by the third heart sound with systolic murmur at the apex together with the incomplete right bundle-branch block and the increased cardiac shadow.

The cardiac catheterisation and the angiographic demonstration of the exact site of obstruction enabled us to assume a favourable prognosis without the surgical intervention which might have been considered had the obstruction involved the outlet of the azygos or both venae cavae.

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