CASE REPORT

Behçet's disease with a large intracardiac thrombus: a case report

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Abstract

Behçet's disease is recognised as a chronic multisystem disorder with vasculitis as its underlying pathological process. Cardiac involvement is rare and often associated with poor prognosis. A case of a 33 year old man with Behçet's disease, presenting with a large right ventricle and right atrial thrombus, is reported. Two dimensional (cross sectional), colour Doppler, and transoesophageal echocardiography, angiography, computed tomography, and magnetic resonance imaging were used to diagnose the disease. With cyclophosphamide and dexamethasone treatment, the cardiac lesions progressively resolved.

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suffered from both genital and oral ulcers over an eight month period and deep peripheral vein thrombosis in the lower extremities.

On physical examination the patient complained of breathlessness, and looked ill and thin. Blood pressure was 110/80 mm Hg, respiration rate was 29 per minute, and pulse rate was 90 per minute, and regular. There was no murmur on cardiac examination, and ECG was normal.

Abnormal biochemical and haematological parameters included a haemoglobin concentration of 9 g/dl (5.6 mmol/l), and an erythrocyte sedimentation rate of 96 mm/hour. Liver and kidney function tests were normal. C reactive protein concentration was raised, antinuclear antibody and anti-DNA tests were negative, and HLA-B5 and pathergy tests were positive. Haptoglobin and α acid glycoprotein concentrations were raised.

The patient's chest x ray was normal. Two dimensional (cross sectional) and colour Doppler echocardiographic examination (HP Sonos 5500) revealed multiple cardiac masses in the right atrium and a very large thrombus (approximately 4×4 cm) in the right ventricle. Transthoracic and transoesophageal echocardiography showed that there was an abnormal structure attached to the right ventricular apex, which was protruding into the cavity (Fig 1A, 2A, 3A). Echocardiography showed the left ventricle was normal in size, and no vegetations were found on the heart valves. Transthoracic and transoesophageal echocardiography were repeated frequently during treatment.

Coronary arteries, left ventriculography, and aortic root angiography were normal. Computed tomography and magnetic resonance imaging of the patient's chest showed multiple pulmonary infarcts in the lower lobes of the lungs.

Repeated blood and urine cultures, immunoelectrophoresis, serological investigations, skin tests, bone marrow aspiration, and biopsy did not provide any evidence of bacterial endocarditis, malaria, fungal infections, tuberculosis, or malignancy.

The diagnosis of Behçet's disease was based on the criteria of the international study group.⁹ The patient had a positive pathergy test, and recurrent oral and genital ulceration consistent with Behçet's disease.

Behçet's disease is a chronic, inflammatory, and multisystemic condition of unknown aetiology, clinically characterised by recurrent oral and genital ulceration and skin eruptions, inflammatory ocular involvement, and neurological manifestations.¹⁻⁴ Involvement of skin, joints, and gastrointestinal, nervous, respiratory, and cardiovascular systems is also recognised.⁵⁻⁷ Although superficial and deep peripheral vein thrombosis is seen in a quarter of these patients, cardiac involvement is extremely rare.⁸

The disease has a worldwide distribution, but it is mainly observed in Turkey, other Mediterranean areas, and Japan.⁵ It affects mainly young adults, with male patients experiencing a more severe clinical progress than female. Adult males are primarily affected, most often in their 30s or 40s. Viral, genetic, several immunological abnormalities, and environmental factors have been described in the pathogenesis of the disease.^{2,3}

We report a case of a man with Behçet's disease with unusual manifestations and rare complications, which posed several diagnostic and therapeutic challenges.

Case report

A 33 year old man was admitted to our hospital with a 25 day history of dyspnoea, paroxysmal nocturnal dyspnoea, orthopnoea, cough, and haemoptysis of unknown origin. He had

Figure 1 Transthoracic echocardiography on apical four chamber. (A) Before treatment. (B) After treatment. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

Figure 2 Transthoracic echocardiography on parasternal short axes. (A) Before treatment. (B) After treatment. AO, aorta.

While being investigated, the patient was treated with high dose intravenous heparin for 10 days, during which time the thrombi did not resolve. After the diagnosis was made, treatment was initiated with dexamethasone, cyclophos-

Figure 3 Transoesophageal echocardiographic image of the patient. (A) Before treatment. (B, C) After treatment.

phamide, and colchicine. The patient was placed on 1 g intravenous cyclophosphamide administered over 30 minutes, and 40 mg intravenous dexamethasone, warfarin 0.5 mg/kg per day, and colchicine (50 mg) three times daily. The patient's clinical condition improved dramatically. The huge thrombus in the right ventricle decreased substantially in size and was barely noticeable on an echocardiogram before discharge. After treatment, near complete lysis of the thrombus was observed with minimal residual thrombus detectable at one, two, and three months post-discharge on two dimensional echocardiography (figs 1B, 2B, 3B,C). No episodes of pulmonary emboli were detected following medical treatment.

Discussion

Behçet's disease is recognised as a chronic multisystem disorder affecting systemic organs and characterised by oral and genital ulceration, dermatitis, and recurrent uveitis. Recently, cardiovascular involvement has been reported to occur in about 1–7 % of patients

with Behçet's disease. The reported complications include endomyocardial fibrosis in both the right and left ventricles, thrombus in the pulmonary artery or right ventricle, pericarditis, myocarditis, coronary arteritis, and valvar disease.¹⁰⁻¹³

Our patient had a highly mobile mass in the right ventricle and multiple small masses in the right atrium. Endocarditis has been described recently as being associated with an organised thrombus in the right ventricle.¹⁴ However, the patient did not show the clinical manifestations of infective endocarditis.

Histological examination of biopsies or surgical specimens may be helpful in determining the pathological features of structures affected by Behçet's disease. Cardiovascular histology typically reveals infiltration by inflammatory cells, neovascularisation, fibrosis, and thrombosis.¹⁰⁻¹² However, we did not perform a right ventricular biopsy in our patient as this was deemed to involve excessive risk, and management would not have been affected by the results.

The pathologic mechanism of microvascular thrombus formation in vasculitis is believed to be caused by endothelial cell ischaemia or disruption that leads to enhanced platelet aggregation.¹⁵ Decreased release of vascular tissue plasminogen activator has been reported in systemic and cutaneous vasculitis.¹⁶ Impaired fibrinolysis as a result of endothelial cell injury from deposited immune complexes is another possible mechanism. Prolonged euglobulin lysis times and abnormal fibrin concentrations were found in several types of vasculitis, including Behçet's disease.¹⁷ Another possible pathologic mechanism of thrombosis in Behçet's disease is attributed to the presence of antiphospholipid antibodies, which are reported to be present in 18% of cases.¹⁸

Although Mendes and colleagues² surgically treated a right ventricular thrombus in an unusual case of Behçet's disease, two weeks later their patient returned with recurrent right ventricular thrombus. After medical treatment complete lysis was achieved in that case. In our patient medical treatment proved extremely successful.

This case provides a good example of Behçet's disease, involving an interesting occurrence in a young man who had clinical findings of pulmonary emboli secondary to large right ventricular thrombus and right atrial

multiple small thrombi. According to our experience, apical four chamber appearance was thought to be necessary to obtain the best images on two dimensional echocardiographic examination. On the other hand, transoesophageal echocardiographic images can be necessary in order to view the ventricle or right atrium clearly.

The present findings suggest that in patients with features compatible with Behçet's disease, thrombi in both the right atrium and right ventricle can be present. Early echocardiography seems advisable in patients with Behçet's disease in order to determine any cardiac involvement, in spite of its rarity. First line treatment is medical, but a thrombus can become massive and extensive, so congestive heart failure, for example, may demand surgical treatment.

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