

Case Report

Management of Behçet Disease with Multiple Complications

TARIK EL HOUARI¹, LATIFA OUKERRAJ¹, LAMIAE GHZAIEL¹, IBTISSAM FELLAT¹,
MOUNIA AZEROUAL², KHALID SERRAJ², ZOUBIDA TAZI MEZALEK², MOHAMMED CHERTI¹,
JAMILA ZARZUR¹, MOHAMMED ARHARBI¹

¹Division of Cardiology, ²Division of Internal Medicine, Ibn Sina Hospital, Rabat, Morocco

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We present the case of an 18-year-old male patient with Behçet's disease who presented with dyspnoea, fever and haemoptysis. A diagnosis of right ventricular thrombosis associated with endomyocardial fibrosis, a pulmonary artery aneurysm, and cerebral vein thrombosis was made. Considering the absence of haemodynamic compromise and the risk of recurrence after surgical treatment for cardiac thrombus, we preferred medical management, which consisted of immunosuppression and anticoagulation. During a follow-up period of 9 months we observed complete dissolution of the thrombus and dramatic improvement of the patient's clinical status.

Behçet's disease is a multi-systemic disease and may present with vascular, cutaneous, pulmonary, neurological, rheumatological, gastrointestinal and genito-urinary manifestations.¹ Cardiac abnormalities have been described in only 1-5% of cases of Behçet's disease,² with intracardiac thrombosis and endomyocardial fibrosis being rarely observed. We report a new case of right intracardiac thrombosis associated with endomyocardial fibrosis, cerebral vein thrombosis and bilateral pulmonary aneurysm.

Case presentation

An 18-year-old man was admitted to our hospital with a 45-day history of dyspnoea, cough, haemoptysis of unknown origin, and recurrent episodes of chest pain and fever. He had suffered from both genital and oral ulcers over a 3-month period.

On physical examination the patient complained of breathlessness, and looked ill and thin. Blood pressure was 100/60 mmHg, respiration rate was 26 per minute, and pulse rate was 90 per minute and regu-

lar. There was no murmur on cardiac examination, and the ECG was normal. The patient's chest X-ray was also normal. Abnormal biochemical and haematological parameters included a haemoglobin concentration of 8 g/dl, and an erythrocyte sedimentation rate of 42 mm/hour. Liver and kidney function tests were normal. Antinuclear antibody and anti-DNA tests were negative, while the pathergy test was positive.

Two-dimensional and colour Doppler echocardiographic examinations revealed a cardiac mass in the right ventricle. Transthoracic echocardiography revealed an abnormal structure attached to the right ventricular apex, protruding into the cavity (Figure 1A). Transthoracic echocardiography showed that the right ventricle was normal in size; no vegetations were found on the heart valves.

Thoracic computed tomography angiogram revealed a giant aneurysm in the right pulmonary artery with a peripheral thrombus and a small aneurysm in the left pulmonary artery (Figure 2). Cerebral computed tomography showed a cerebral vein thrombosis in the right lateral sinus.

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Address:
Tarik El Houari

Division of Cardiology
Ibn Sina Hospital
Rabat, Morocco
e-mail:
htarik-78@yahoo.fr

A regimen of 6-monthly boluses of intravenous cyclophosphamide, anticoagulant and prednisone 1 mg/kg/day was started. The biological assessment during follow up showed a clear regression of the markers of the inflammation. Since haemoptysis persisted, the patient benefited from an embolisation of the aneurysm of the right pulmonary artery. Transthoracic echo examinations three weeks and nine months after the beginning of the treatment showed that the thrombus size was diminished, as were the bright echoes of the right ventricular endocardium (Figure 1 B).

Discussion

Cardiac involvement in Behçet's disease is very rare, but is usually associated with a poor prognosis. Myocarditis, pancarditis, acute myocardial infarction, conduction system abnormalities, and valvular disease have been reported.³ Intracardiac thrombus formation is very uncommon.⁴ It is seen more often in male than in female patients, while younger patients tend to be affected more frequently.⁵ In a series of 137 patients with Behçet's disease only one patient developed right ventricular thrombus.⁶ Intracardiac thrombi in Behçet's disease may result from endomyocardial fibrosis, which may be a sequela of vasculitis involving endocardium, myocardium, or both.⁷ The diagnosis of intracardiac thrombi in Behçet's disease may be made using cardiac magnetic resonance imaging, computed tomography, and transthoracic echocardiography, which may show a mass in the heart chambers, sometimes indistinguishable from infective vegetations or from a tumour.

Differentiating the thrombus from a myxoma can also be challenging; thus, transoesophageal echocardiographic imaging⁸ and cardiac magnetic resonance

imaging⁹ may be necessary. The mass is usually immobile with a broad-based attachment.¹⁰ Treatment of intracardiac thrombi associated with Behçet's disease is mainly through medication, namely antithrombotic and immunosuppressive therapy, although surgical intervention may be necessary in cases refractory to medical treatment, cases with haemodynamic compromise, or when recurrent episodes of pulmonary embolism ensue. The association of endomyocardial fibrosis and Behçet's disease was first described in 1977 at necropsy.¹¹ Endomyocardial fibrosis in Behçet's disease may be a sequela of endocarditis or myocarditis, or both, complicated by mural thrombus and predominantly involving the right ventricle. Corticosteroids and anticoagulants or anti-aggregants may prevent the development of endomyocardial fibrosis. If endomyocardial fibrosis is complicated by cardiac failure, surgical excision is successful in the short term.¹² In our patient, intracardiac thrombosis was associated with endomyocardial fibrosis, cerebral vein thrombosis and pulmonary aneurysms. This association has not been previously reported.

Pulmonary artery aneurysms are very rare (1.1%),¹³ have a poor prognosis and are one of the leading causes of patient death in Behçet's disease. The pulmonary manifestations appear, on average, 3.6 years after the first extrapulmonary manifestation. Haemoptysis is the major pulmonary manifestation and occurred in 77% of reported cases. Chest X-ray films showed abnormal findings in 90% of cases. Helical computed tomography is currently the method of choice for confirming the diagnosis, because it provides an excellent vascular image, requiring only a small quantity of contrast material. Magnetic resonance imaging is also helpful in the diagnosis of pulmonary artery aneurysms and pseudoaneurysms. Cur-

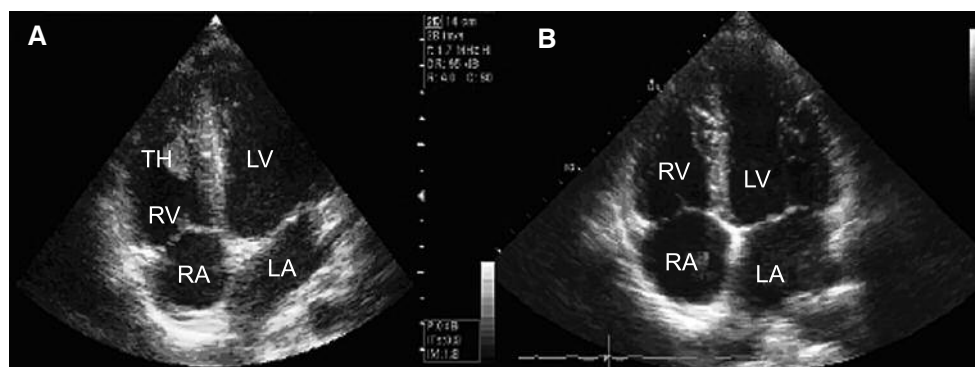


Figure 1. Transthoracic echocardiogram (apical 4-chamber view) showing the right ventricular thrombosis. A: before treatment, B: after treatment. LA – left atrium; LV – left ventricle; RA – right atrium; RV – right ventricle; TH – thrombus.

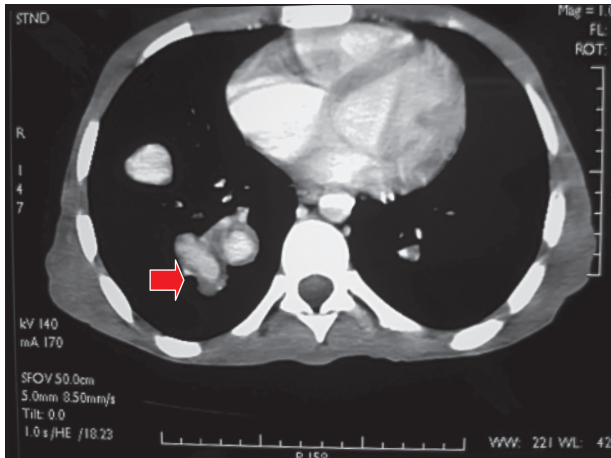


Figure 2. Computed tomography of the chest showing an aneurysm in the right pulmonary artery, with a peripheral thrombus, and a small aneurysm in the left pulmonary artery.

rently, the mainstay of treatment is immunosuppression, especially when introduced in the early stages before irreversible damage to the arterial wall develops. Anticoagulant therapy holds potential hazards for patients with aneurismal dilatation of the pulmonary blood vessels. When the disease is localised to one area of the lung, surgical excision, followed by medication, is indicated to prevent fatal pulmonary haemorrhage.¹⁴ Endovascular embolisation may be of value in treatment-resistant haemoptysis, as in our case.^{15,16}

Neuro-Behçet's disease is usually observed after a long duration of the disease. In this case all of the complications were inaugural. Occurrence of neurological involvement as a first presentation is quite exceptional in paediatric Behçet's disease.¹⁷ Intracranial hypertension, with or without cerebral vein thrombosis, appears in 11-35% of all patients with neuro-Behçet's disease.¹⁸ Treatment of cerebral vein thrombosis in Behçet's disease is not standardised. Long-term follow up has not been available in most reports.¹⁹ The use of anticoagulants in the treatment of cerebral vein thrombosis remains controversial.

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