CASE REPORT

Intracardiac Thrombus in a Young Man: Don’t Forget Behçet’s Disease!

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Abstract

Intracardiac thrombosis is an exceptional complication of Behçet’s disease. The management of this involvement is difficult due to the risk of recurrence. We present the case of a young man admitted to our hospital for intermittent fever. The microbiologic investigations did not show any causative germ. We discovered a right ventricle thrombus on echocardiography. We confirmed the diagnosis of pulmonary embolism on CT angiogram. The patient developed oral and genital ulcerations which were consistent with Behçet’s syndrome. The thrombus had disappeared after treatment with anticoagulant, corticosteroid and immunosuppressors. Intracardiac thrombosis can reveal Behçet’s disease. An exhaustive examination and close monitoring should be performed in order to reveal pathognomonic signs as soon as possible and to promptly start the appropriate treatment.

Key words: Behçet’s disease, intracardiac thrombus, pulmonary embolism

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Introduction

Behçet’s disease (BD) is a chronic, multisystemic vasculitis that affects mainly young adults in Mediterranean, Middle Eastern and Far Eastern countries (1). It is characterized by recurrent oral and genital aphthous ulceration, skin lesions and ocular inflammation. This disease frequently affects other systems including neurological, locomotor or vascular systems. Cardiac thrombosis is a rare and serious complication, which can reveal the syndrome in half of the cases (2). The management of this complication is difficult because of the risk of thrombosis recurrence even after surgical resection of the thrombus. We report a case of Behçet’s syndrome in an adolescent, discovered by an intracardiac thrombus associated with a pulmonary embolism.

Case Report

A previously healthy 20-year-old man was admitted to our hospital for the evaluation of intermittent fever over a 2-month period. On physical examination, the patient appeared weak and thin; his body temperature was 38°, blood pressure 100/60 mmHg and heart rate=110 bpm. The heart sounds were normal without any murmurs. The lung fields were clear to auscultation. The remainder of the physical examination was unremarkable. The electrocardiogram showed a sinus tachycardia with a right bundle branch block. The X ray chest showed hilar enlargement.

We noted an inflammatory syndrome in the laboratory results. Microbiologic studies identified no causative organisms. Hemocultures and serology tests for Q fever, Legionella, Bartonella, Tropheryma whipplei, Chlamydia, Mycoplasma, and Brucella were negative.

The echocardiography showed a left ventricle ejection fraction of about 50%, a mild right ventricle enlargement with a 1.5 cm × 2 cm mass on the apex. This mass appeared hyperechoic and well circumscribed (Fig. 1); on color Doppler, it seemed to be vascularized. Pulmonary arterial hypertension (54 mmHg) was noted.

A thoracic spiral computed tomographic confirmed the diagnosis of pulmonary embolism as it showed thrombi in
Figure 1. Apical 4-chamber view on transthoracic view: the right ventricular thrombus.

Figure 2. CT angiogram: thrombi in the bifurcation and in the main pulmonary arteries.

both pulmonary arteries reaching the bifurcation (Fig. 2). Thus, we started treatment with heparin.

The thrombophilia study did not show any abnormalities, and anti-nuclear, anti-DNA, antiENA, and anti-neutrophil cytoplasm antibody tests were all negative. Tumor markers were also negative.

Two weeks later, the patient complained of painful oral ulcers and acneiform lesions on the torso and scar lesions of a few millimeters on the scrotum. At the same time we noted a positive HLA-B51 serologic typing. Taking these findings together, the patient was diagnosed with Behçet’s disease (BD) as he fulfilled The International Study Group of Behçet’s Disease criteria (3). Corticosteroid treatment was started with intravenous bolus of methylprednisolone (1 g per day for 3 days) followed by oral prednisone 1 mg/kg per day, associated to colchicine (1 mg per day). An immunosuppressive therapy with intravenous cyclophosphamide (1 gram monthly) was added to the treatment, yielding immediate improvement in clinical and laboratory parameters.

In addition, warfarin was administered to maintain the international normalized ratio (INR) at 2.5 or above. Two months later, the right ventricular thrombus and pulmonary hypertension had disappeared on transthoracic. The patient was still being monitored, without recurrence, one year later.

**Discussion**

The prevalence of cardiovascular involvement in Behçet’s disease varies from 7.7% to 43%, according to the different ethnic group (1, 4). Venous lesions are the most common abnormality and usually consist of recurrent deep vein thromboses, most often involving the lower extremities. Thrombosis of the vena cava is also a common vascular lesion in BD, but intracardiac thrombosis is rare. In a series of 137 BD patients only one case of intracardiac thrombus was described (5). Ozkan et al. (6) demonstrated that there is no difference in the prevalence of echocardiographic abnormalities between patients with BD and control subjects. Since the first description by Buge in 1977 (7) up to date, only about 50 cases have been reported (1, 2, 8-17).

The clinical manifestations of the disease start with fever (50.9%), hemoptysis (55.4%) and dyspnea (36.2%). It can be associated with a venous thrombosis of the superior and/or inferior vena cava (45.3%), pulmonary embolism (41.5%), and aneurysms of the pulmonary arteries (38.3%) (17). The discovery of Behçet’s disease by intracardiac thrombosis, as in the current case, is little described. Such a complication occurs in males (81.25%) with an average age of 28 years.

Intracardiac thrombosis is often associated with deep vein thrombosis and with thrombosis of the vena cava, respectively, in 50% and 22% of cases (18), with pulmonary complications (aneurysm of the pulmonary artery (9, 12) or pulmonary embolism) or with endomyocardial fibrosis (2, 19). It usually involves the right side of the heart (right chambers in 78% of cases: the right atrial chamber (8) and the right ventricle (9, 10, 13, 16)), more often than the left ventricle (11, 20). In the patient, the thrombus was found in both right and left ventricles. Involvement of the left heart has rarely been reported or discussed (11, 18, 20). Intracardiac thrombus often precedes other manifestations of Behçet’s disease as in the current patient.

Transesophageal and transthoracic echocardiography are generally sufficient to reveal this complication. The mass is usually heterogeneous and echogenic, involving the ventricles rather than the atria and the right heart more often than the left (right ventricle in 78%) (18). The main differential diagnoses are primary heart tumour and infectious endocarditis. The differentiation from atrial myxoma is sometimes difficult based on the fact that the appearances of the myxoma may be atypical and the surface may be covered by thrombus (12, 17). Imaging tests as CT chest, and MRN could be helpful in the assessment of thoracic manifestations of BD including thrombus of the systemic veins, heart and pulmonary arteries. Scintigraphy can be used to determine whether a thrombus is acute or chronic.

In the present observation, the diagnosis of the thrombus
was obvious on echocardiography. Nevertheless, the etiologic investigations were not contributory. The lack of pathognomonic symptoms or definitive laboratory tests makes BD a difficult and sometimes overlooked diagnosis. We should also consider epidemiologic data like the Mediterranean origin of the patient, young age and male sex.

The pathogenic mechanism underlying the thrombotic tendency among patients with Behçet’s disease is not well known. Some authors (17) have incriminated the presence of antiphospholipid antibodies or prothrombotic factors (deficiencies of protein S, protein C, and antithrombin III), but in the present patient all of the thrombophilia tests were negative. Others have suggested fibrinolysis anomaly due to endothelial cell damage after the deposition of antigen-antibody (like α enolase antibodies) (21).

The association between hyperhomocysteinemia and thrombosis has been investigated in some studies in BD patients. Recently, La Regina and colleagues (22) showed via a large metaanalysis that hyperhomocysteinemia may be considered to be associated with thrombosis in BD patients and that the treatment with folic acid supplementation could improve the ophthalmic and vascular lesions.

Regarding the treatment of cardiac thrombosis associated with Behçet’s disease, there is no consensus (13). The aim is to resolve the cardiac thrombus and to prevent the recurrence of this complication. Anticoagulants alone do not seem to be effective; systemic glucocorticoid and immunosuppressive agents are highly recommended before using anticoagulants (21). Surgery might be limited to the cases with massive thrombosis, recurrence of the complication despite an optimal medical treatment, and when there is a cardiac congestion (22).

The prognosis of cardiac thrombosis during BD is poor. A high mortality was noted due to massive hemoptysis, recurrence and post-surgical complication.

Conclusion

It is critical to keep in mind Behçet’s syndrome in patients with intracardiac thrombus in order to institute the appropriate medical treatment as soon as possible. The combination of methylprednisolone, cyclophosphamide, and warfarin seems to be a good option to treat such a complication.

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References