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Case Report

Cardiovascular Manifestations in Behcet's Disease : Unusual Mode of Revelation

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Abstract

Behcet's disease is a multisystemic, chronic, inflammatory disorder of unknown etiology. It is characterized by recurrent buccal aphthosis, genital ulcers, and uveitis. It is more frequent in young adults, with a predominantly male population from the Mediterranean and Middle East.

Additional manifestations in other locations (gastrointestinal tract, genitourinary tract, central nervous system, cardiovascular system, and lungs) can occur and can impact the clinical progress and prognosis. Although cardiac involvement during Behçet's disease is uncommon and intracardiac thrombosis is exceptional.

We report the case of a 32-years-old man admitted for a 3-month history of fever, loss of weight with an intracardiac thrombus in the context of Behçet's disease.

Introduction

Behcet's disease is a systemic inflammatory disease primarily involving the oral and genital mucosa, skin, and eyes [1]. It can also affect several other sites (pulmonary, gastrointestinal, genitourinary, central nervous system, and cardiovascular) which could influence the prognosis of the disease [2].

The onset is insidious with peak age in young adulthood (25-30 years), but also occasionally in children before the age of 16 years [3].

Cardiac involvement is rare, with serious consequences: intracardiac thrombosis, cardiomyopathy, pericarditis, valvulopathy, acute coronary syndrome, and conduction disorders [4] . Despite its sporadic occurrence, it is strongly correlated with mortality.

We report the case of a young man admitted for management of fever with loss of body weight, a condition that was present for three months.

Case Presentation

A 32-years- old man with no medical history was hospitalized for an isolated prolonged fever progressing for more than 3 months with marked inflammatory state (C-reactive protein at 158 mg/l and high and erythrocyte sedimentation rate at 83 mm). No infectious symptoms were identified.

The initial physical examination revealed a temperature of 38.5°C and found multiple scrotal ulcerations.

The electrocardiogram showed a Sinus tachycardia.

The laboratory tests indicated anemia, hemoglobin at 9.2 g/dL, neutrophil leukocytosis at 13120/L, and C-reactive protein at 158 mg/l. Haemocultures were negative, as was the bronchial aspirate culture for mycobacteria.

Body computed tomography (CT) has been performed and showed no anomaly. There was no evidence of pulmonary embolism, vasculitis, and pulmonary aneurysm in CT pulmonary angiography.

We realized transthoracic echocardiography (TTE) that detected a right atrial echogenic mass of 13×15 mm consistent with thrombus **(Figure 1)**.

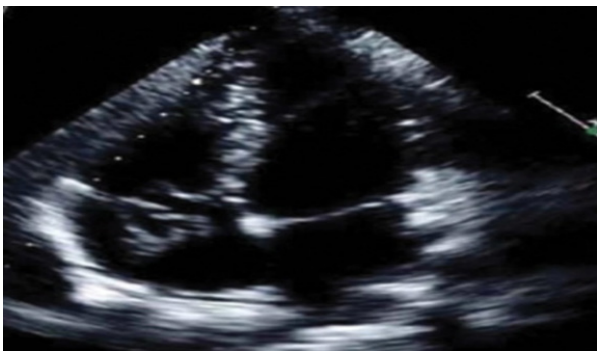

Figure 1: Transthoracic echocardiography, in apical four-chamber view, showing a large right atrium thrombus 13×15 mm.

With the exclusion of infection, it was assumed that the intracardiac masse was secondary to heart involvement by Behçet's disease, considering the patient's history of recurrent genital ulcers, the presence of the scrotal ulceration scars, and a pathergy test that came out positive.The patient received intravenous

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high-dose methylprednisolone followed by oral prednisone and anticoagulation therapy.The thrombus in the right atrium decreased substantially in size. At 4 months' follow-up, there was complete resolution of the thrombus in the right atrium.

Discussion

Cardiac involvement can occur as intracardiac thrombus, endocarditis, myocarditis, pericarditis, endomyocardial fibrosis, coronary arteritis, myocardial infarction, and valve disease [5]. Pericardial involvement has been reported as the most common manifestation of cardiac involvement in some series [6, 7]. The clinical presentation may be acute pericarditis, hemorrhagic pericardial tamponade, constrictive pericarditis, recurrent pericarditis, or even just a small asymptomatic pericardial effusion [8,9]. Endomyocardial involvement usually manifests as endomyocardial fibrosis on the right and / or left side of the heart [10].

Cases of cardiomyopathy have been described which may be ischemic, non-ischemic or inflammatory in nature. Clinically, it can manifest as systolic or diastolic heart failure, or more subtly, asymptomatic systolic or diastolic dysfunction [11,12]. Cardiovascular involvement is estimated between 7% and 46%. Vasculitis lesions that can affect veins and arteries of all sizes are present. The typical form of arterial involvement is manifested by a real or false aneurysm. The most common complication is superficial vein thrombosis, followed by deep vein thrombosis [13]. In arterial involvement, which is less common than venous involvement, the carotid, pulmonary, aorta, iliac, femoral and popliteal arteries are more frequently involved.

Coronary aneurysms can be seen during angiography procedures. Some of these aneurysms are asymptomatic, while others manifest as acute coronary syndrome [14,15]. These aneurysms are isolable, and most are evident with coronary stenosis and are sometimes seen with multiple arterial aneurysms [16,17]. Valsalva sinus aneurysms and aortitis are the most frequently reported cardiac complications, affecting the root of the aorta [18]. Valsalva sinus aneurysms can be seen alone or with other sinus aneurysms and can lead to acute or chronic aortic failure.

In our case, the cardiac involvement was manifested by an intracavitary thrombus. It can be one of the first signs of disease with pulmonary embolism, or can cause cerebral embolism passing through the patent foramen ovale. Often, the right ventricle is involved, but the left ventricle has been shown to be involved as well [19, 20].

The pathophysiology of thrombotic predisposition is still unknown. Various mechanisms have been proposed [2]:

- Endothelial cell ischemia or disruption, which leads to enhancement of platelet aggregation.
- The presence of antiphospholipid antibodies, which is reported to occur in 18% of cases.
- The presence of prothrombotic factors, such as deficiencies of protein S, protein C, and antithrombin.
- Elevated von Willebrand factor antigen levels, which have recently been observed in association with Behçet syndrome.
- Fibrinolysis anomaly due to endothelial cell damage after the deposition of antigen-antibody (immune complex disease).
- Homozygosis for the factor V Leiden mutation or the prothrombin gene, which increases the risk of thrombosis by a factor of 6 or 3, respectively.

Transesophageal and transthoracic echocardiography are generally sufficient to enable a diagnosis of cardiac thrombosis. The mass is usually heterogeneous and echogenic, involving the ventricles rather than the atria [21] and the right heart more often than the left [20].

Ultrasonic imaging might suggest a differential diagnosis a large vegetation, for example, or an intracardiac tumor such as a myxoma, or endomyocardial fibrosis but the clinical context helps to clarify the diagnosis [22]. In any event, the resolution of the mass under anticoagulant or antiplatelet therapy establishes a retrospective diagnosis, because it distinguishes thrombus from tumor [20].

MRI can differentiate between a tumor and an intracardiac thrombus, and in the case of endomyocardial fibrosis, it represents the gold standard for diagnosis showing a 3-layer appearance: a healthy outer myocardial layer, an intermediate layer of fibrosis, and finally internal thrombotic structures [23].

There is no consensus regarding the treatment of cardiac thrombosis in association with Behçet syndrome. The object of treatment is to control the underlying disease and to resolve the cardiac thrombus. Evidence for treatment is based on case series reported in the literature, the majority of which were treated with a combination of anticoagulants and immunosuppressants (azathioprine or cyclophosphamide) with superior results [4].

However, an associated aneurysm of the pulmonary artery especially the presence of large or bilateral aneurysms makes the use of these therapies difficult because of the risk of a devastating hemoptysis.

Surgery might become necessary in cases of cardiac thrombosis that is massive, is recurrent after medical treatment, or is associated with cardiac congestion [19].

Conclusion

Cardiac involvement in Behçet's disease is an unusual manifestation that conditions the prognosis. The detection of an intracardiac mass in a young subject should lead to the diagnosis of intracardiac thrombus in the context of Behçet's disease. Echocardiography is important, and treatment is essentially based on immunosuppressants and anticoagulation.

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