

Right Ventricular Thrombus: An Unusual Manifestation of Behcet's Disease

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Abstract: Behçet syndrome is a multisystemic, chronic, inflammatory disorder of unknown cause. It is characterized by recurrent buccal aphthosis, genital ulcers, and uveitis with hypopyon. The mean age at which it occurs is 20 to 30 years; men are 2 to 5 times more often affected than women. Additional manifestations in other locations (skin, joints, gastrointestinal tract, genitourinary tract, central nervous system, cardiovascular system, and lungs) can appear and can influence the clinical course and prognosis. Although cardiac involvement during Behçet syndrome is uncommon and intracardiac thrombosis is exceptional. In this unusual case a young man had symptoms that primarily related to recurrent right ventricular thrombi and pulmonary thromboemboli.

Keywords: Behcet's disease; right ventricular thrombi; pulmonary thromboemboli.

1. Introduction

Behcet's disease (BD) is a multisystemic inflammatory disease with a clinical spectrum that has greatly expanded since it was first described in 1937 as a triple complex of recurrent oral genital ulcers and uveitis.

Behçet's disease is recognized as a systemic vasculitis involving both arteries and veins of any size. Vascular involvement has occurred in one-third of patients. Most vascular events consist of recurrent superficial or deep vein thrombosis. Arterial thrombosis is less frequent.

Cardiac manifestations of Behcet's disease include endocarditis, myocarditis, pericarditis, endomyocardial fibrosis, coronary arteritis with or without myocardial infarction, aneurysms of the coronary arteries, valve dysfunction, conduction system disturbances, and intracardiac thrombosis (ICT). ICT is a rare manifestation of the disease. [1]

We report a case of a young man who had symptoms that primarily related to recurrent right ventricular thrombi and pulmonary thromboemboli.

2. Case Report

A 27-year-old man with a three weeks history of dyspnoea, cough and haemoptysis was admitted. At the age of 25, he had suffered from painful oral and genital ulcerations and polyarthralgias. At that time, examination revealed bilateral papillary oedema.

Laboratory tests on admission revealed: haemoglobin of 11 g/dl, erythrocyte sedimentation of 100 mm/hr and C reactive protein concentration of 70 mg/l.

In our case, this complication of Behcet's disease was diagnosed by echocardiography who objectified the presence

of thrombus in the right atrium measuring 15x18 mm, a thrombus in the right ventricle measuring 18x29 mm. (Figure 1 and 2)

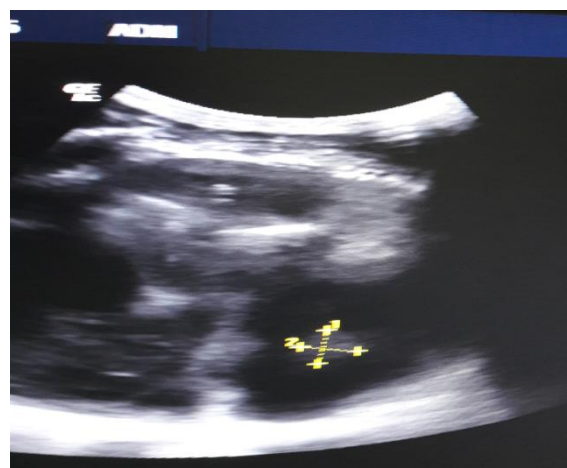


Figure 1: Image showing thrombus in the right atrium measuring 15x18 mm



Figure 2: Image showing thrombus in the right ventricle measuring 18x29 mm

The angioscanner found the presence of hypodense images in the lower lobar artery, bilaterally, compatible with emboli. Further identification of a hypodense structure occupying the right ventricle extended 27x16 mm in favour of thrombus. Analysis of the remainder of the pulmonary parenchyma finds triangular lesions with a basic peripheral external base and with lesions of pulmonary infarction. (Figure 3 and 4)

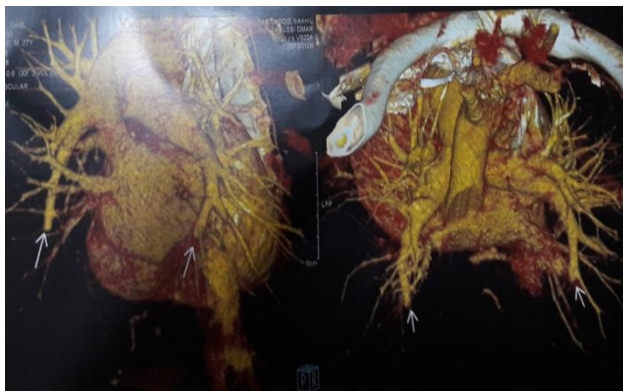


Figure 3: Images showing bilateral emboli of the lower lobar artery



Figure 4: Images showing the lesions of pulmonary infarction

The patient was given immunosuppressive and anticoagulant therapy.

We conclude that thrombi especially in the right heart cavities can be present in Behcet's disease without causing specific symptoms but can lead to pulmonary embolism. Early echocardiography seems advisable to detect the presence of cardiac involvement and medical therapy can be effective in the resolution of intracardiac thrombosis in the setting of Behcet's disease.

3. Discussion

Behcet's Disease is now recognized as a multisystem illness in which the initial manifestations may not be confined to the classic symptom complex of oral and genital ulcerations and recurrent uveitis.

Vascular involvement is frequent and has been reported to occur in up to one third of patients [2-3].

Venous lesions are the most common abnormality and usually consist of recurrent superficial and deep thrombophlebitis, most often involving the lower extremities

[2-3]. Thromboses of the vena cavae are the second most common vascular lesion and often result in obstruction of the vessel. Despite the high frequency of venous thrombosis, pulmonary emboli are a rare complication. Arterial thrombosis and spontaneous aneurysm formation, especially of the aorta and pulmonary artery, are less common in Behcet's disease but responsible for the majority of vascular deaths. [2-3]

Cardiac involvement in Behcet's disease is uncommon but such abnormalities as myocarditis marantic endocarditis, and coronary artery aneurysms have been described. [4] Intracardiac thrombi are extremely rare. [5-6] In a series of 137 patients with Behcet's disease, Koc et al. described only one case of intracardiac thrombus within the right ventricle that was not associated with clinical embolic events.

These authors also performed an extensive review of the literature on Behcet's disease and found only two cases of intracardiac thrombi. In one case the patient had symptoms consistent with recurrent pulmonary emboli but no evidence of venous thrombosis.

Transthoracic echocardiography revealed a mass in the right ventricle, which was found to be thrombus at the time of surgical excision. In a recent report Vanhaleweyk et al. described a patient in whom thrombus in the right atrium and both ventricles was visualized by transthoracic echocardiography and nuclear magnetic resonance imaging. Heparin and immunosuppressive therapy resulted in disappearance of the thrombi without clinical evidence of systemic embolization. [7]. Transesophageal echocardiography to document the presence of both right ventricular and superior vena cava thrombi has not been previously described.

This case represents another unusual manifestation of Behcet's disease in which the clinical presentation was likely related to recurrent pulmonary emboli originating from the right ventricular thrombus, although in situ thrombosis and emboli from the pulmonary artery cannot be excluded. Transesophageal echocardiography documented the thrombi, which helped support the diagnosis of Behcet's disease, and was useful in monitoring the patient's clinical course.

4. Conclusion

We conclude that thrombi, especially in the right-heart cavities, are possible complications of Behcet syndrome that can lead to pulmonary embolism. Early echocardiography is advisable for the detection of cardiac involvement, and medical treatment should be the first choice of therapy.

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