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Intracardiac Thrombus in the Right Heart: An Atypical Presentation of Behçet's Disease

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Intracardiac thrombosis is a rare complication of Behçet's disease (BD), which can present as an intracardiac tumor. Its discovery precedes the diagnosis of Behçet's disease in half of the cases.
We report the observation of a 64-year-old patient with a history of bipolar aphthosis, admitted to our department for etiological work-up of a prolonged fever disorder. Transthoracic echocardiography revealed a right ventricular mass appended to the tricuspid valve. Cardiac MRI was consistent with thrombus. The case was discussed in a multidisciplinary team including cardiologists, radiologists and internists, and the diagnosis of Behçet's disease with cardiac complications was established. The patient was treated with immunosuppressive therapy, with a favorable clinical and echocardiographic evolution.
The discovery of an intracardiac mass should prompt the diagnosis of cardiac thrombus and Behçet's disease, even in the absence of predisposing ethnic or geographical factors.

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Keywords: Right ventricular mass; cardiac mass; Behçet's disease; intracardiac thrombus; echocardiography.

1. INTRODUCTION

Behçet's disease (BD) is an inflammatory, multisystemic vasculitis characterized by the frequency and benignity of cutaneous-mucosal and articular manifestations, and the severity of ocular, central neurological, vascular, and especially arterial and digestive manifestations [1].

This condition predominantly affects men (twice as often as women) between the ages of 20 and 40 years. It is common in the Far East and around the Mediterranean basin. Diagnosis relies on clinical presentation and international criteria [1,2]. Behçet's disease progresses in episodes, sometimes spontaneously regressive, and its treatment is mainly symptomatic due to numerous unknowns regarding its etiology and pathophysiology [1].

The frequency of cardiac involvement varies between less than 1% and 6% in clinical series and 16.5% in an autopsy series [3]. All three layers of the heart can be affected, including pericarditis, myocardial involvement, valvular abnormalities, coronary artery, and conduction tissue involvement. Intracardiac thrombosis is very rare, with a recent literature review reporting 25 cases. This complication typically occurs in young men from the Mediterranean basin and the Middle East, predominantly affecting the right heart chambers [3].

2. CLINICAL CASE

We report the clinical case of a 64-year-old chronic smoker who was admitted to the emergency department with a febrile altered mental status associated with respiratory distress. The patient had a history of weight loss, unquantified fever for over two months, recurrent oral and genital ulcers, and a history of pseudofolliculitis.

At admission, the general examination revealed a confused patient with a Glasgow Coma Scale score of 13/15, fever of 39°C, tachycardia at 120 bpm, respiratory rate of 30 breaths per minute with oxygen saturation at 92%, and signs of respiratory distress. Pulmonary examination revealed a focus of consolidation in the right lung with crackling rales. Cardiovascular examination was unremarkable, and neurological examination did not reveal any motor or sensory deficits.

An emergency thoracic CT scan revealed a likely infectious origin focal consolidation in the right lower lobe without signs suggestive of pulmonary embolism. Given the febrile altered mental status, a cerebral MRI was requested, which showed nonspecific signal abnormalities in the white matter.

The biological assessment showed an inflammatory syndrome with a CRP level of 445 mg/l and a white blood cell count of 14,000/mm3. Additionally, there was an evidence of functional renal insufficiency, indicated by an estimated glomerular filtration rate (eGFR) of 25 ml/min/1.73 m². Despite the fever, three sets of blood cultures yielded no growth.

The patient was initially managed in the intensive care unit and started on antibiotic therapy with a third-generation cephalosporin combined with gentamicin. There was a marked clinical improvement with resolution of fever and dyspnea, and initial improvement in laboratory parameters, including a follow-up CRP of 180 mg/l, white blood cell count of 8600/mm3, and eGFR of 60 ml/min/1.73 m². However, there was a subsequent rise in CRP to 254 mg/l and recurrence of fever.

Given the prolonged fever and deterioration in general condition, a transthoracic echocardiogram was performed, revealing the presence of a pedunculated, mobile mass measuring 28 x 15mm attached to the tricuspid valve without evidence of stenosis or regurgitation. Differential diagnoses considered based on the location of the mass included a right heart myxoma, vegetation, thrombus, or intracardiac metastatic lesion.

The patient stayed in the intensive care unit for 10 days before being transferred to the cardiology department for further diagnostic and therapeutic management of the intra-cardiac mass, following stabilization.

For diagnostic confirmation and better tissue characterization of the mass, a cardiac MRI was performed, revealing a tricuspid peri-valvular mass. It appeared inflammatory, avascular, with a significant thrombotic component. No detectable contrast uptake was observed within the mass, suggesting a non-obstructive thrombus.

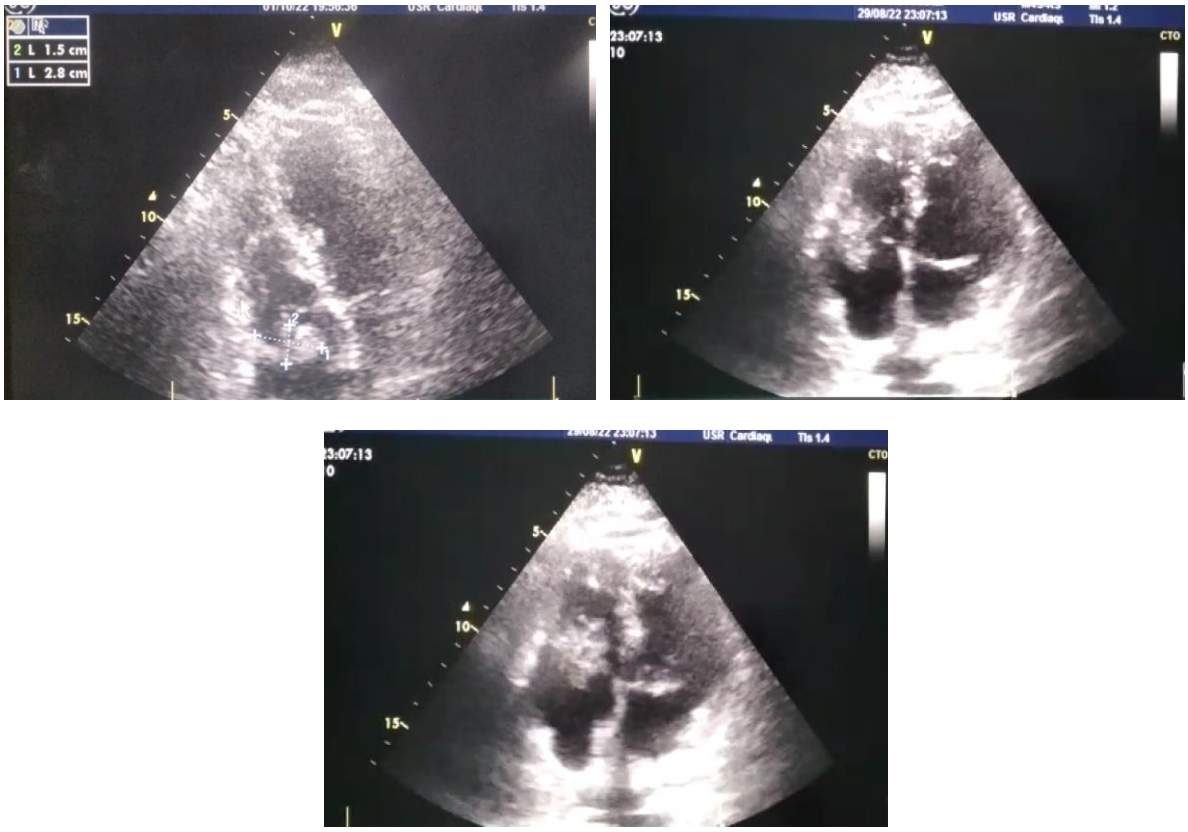

Fig. 1, 2, 3. Echocardiography demonstrates an intra-cardiac, pedunculated, mobile mass attached to the tricuspid valve

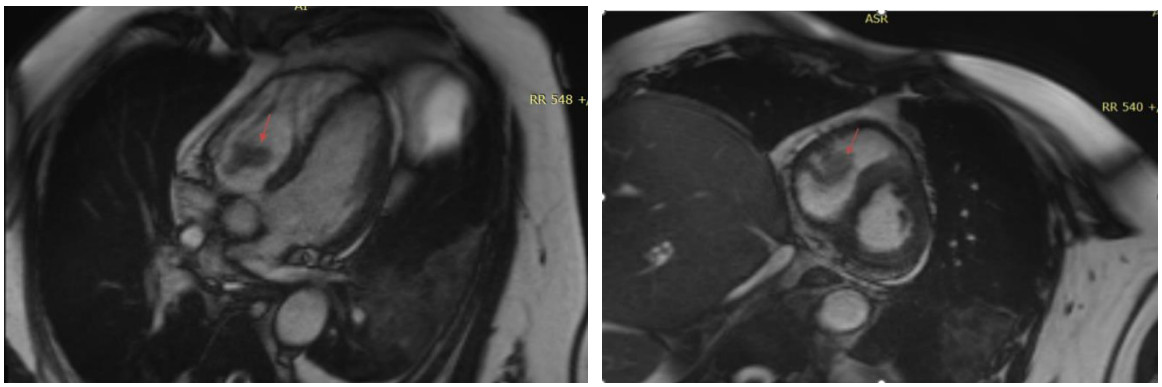

Fig. 4, 5. Cardiac MRI showing a peri-tricuspid valvular mass suggestive of a thrombus

The atypical location of the thrombus led us to consider the diagnosis of Behçet's disease. Given the patient's history of recurrent oral and genital ulcers and pseudofolliculitis, the diagnosis of Behçet's disease was confirmed with 4 points according to the International Criteria for Behçet's Disease (ICBD) score.

The patient was started on therapeutic doses of heparin, corticosteroids, and immunosuppressants after ruling out an ongoing infection and latent tuberculosis. One week later, there was significant improvement in symptoms, particularly with the achievement of afebrile status after the initial bolus, and improvement in inflammatory markers, with a follow-up CRP of 12 mg/l. The patient was hospitalized in the cardiology department for 15 days. A follow-up transthoracic echocardiogram (TTE) performed before discharge showed a reduction in the size of the mass, measuring 24 x 10 mm.

Although the patient showed clinical and biological improvement with treatment, monthly follow-up echocardiograms revealed persistent thrombus before confirming its complete resolution after 6 months of treatment.

3. DISCUSSION

Although Behçet's disease is found across a wide geographical area extending from the Mediterranean basin to the Far East, Behçet's disease complicated by intracardiac thrombus appears to predominantly affect patients from the Mediterranean basin and the Middle East [1,2].

In the literature, the most common cardiac involvement during Behçet's disease (BD) is acute benign pericarditis occurring contemporaneously with an inflammatory flare of the disease. Myocarditis, occurring in the absence of lesions in the major coronary arteries, has also been reported. Additionally, rare cases of spontaneous ventricular aneurysms or post-myocardial infarction aneurysms have been described [4]. Intracardiac thromboses are exceptionally rare in BD, often preceding the diagnosis of BD in half of the cases, as in our observation. Since the first autopsy case reported by Buge et al. in 1977 [5], around forty cases of intracardiac thromboses have been reported in BD. They are frequently associated with endomyocardial fibrosis and may also be associated with specific pulmonary arterial involvement, such as pulmonary aneurysms or parenchymal involvement. However, our patient did not have endomyocardial fibrosis or aneurysms. The etiology of these thromboses in BD remains unclear, and the role of antiphospholipid antibodies in this disease is not established [5]. The majority of cardiac thromboses in BD affect the right heart chambers, with involvement of the left heart being less common.

Echocardiography can sometimes raise suspicion of a thrombus, although the appearance may also suggest a myxoma or another tumor. Cardiac MRI and scintigraphy can contribute to the diagnosis of cardiac thrombus.

In our case, the diagnosis of Behçet's disease was made based on clinical criteria, as the patient had recurrent cutaneous-mucosal lesions (≥4 points according to the ICBD score). However, the clinical presentation with fever and altered sensorium, along with the presence of a large intra-cardiac right- ventricular mass, delayed the final diagnosis. The etiological diagnosis of a cardiac mass is always challenging, especially when it involves the right heart chambers. In these cases, it is crucial to differentiate between thrombus, neoplasms, or vegetations. In our patient, cardiac MRI was essential for the final diagnosis, revealing the presence of an inflammatory mass with a significant thrombotic component in a patient suspected of having Behçet's disease.

Currently, there are no guidelines or randomized controlled trials regarding the treatment of cardiac involvement in Behçet's disease. Therefore, we rely on data from published case reports and the experience of high-volume centers for guidance.

Several authors have reported the resolution of intracardiac thrombus following medical treatment of Behçet's disease. Corticosteroids have been used alone or in combination with colchicine and/or immunosuppressants (such as azathioprine, cyclophosphamide, or cyclosporine). Treatments with vitamin K antagonists or aspirin have also been employed, although their use is challenging in cases associated with hemoptysis [3]. In fact, some cases in the literature have shown that anticoagulation alone did not promote regression or resolution of these masses, and resolution was only achieved after introducing immunosuppressive therapy [6,7].

Dincer et al. successfully treated a post-surgical recurrent cardiac thrombosis with fibrinolytics [8]. However, cardiac surgery is generally not recommended due to high rates of recurrence and embolization [9].

In our case, we initially pursued a conservative therapeutic approach, including therapeutic anticoagulation, which proved unsuccessful. Regression was achieved after adding an immunosuppressive treatment. The failure of anticoagulant therapy alone could potentially be explained by the inflammatory component of the mass.

4. CONCLUSION

Intracardiac thrombosis is a rare complication of Behçet's disease, which can present as an intracardiac tumor. Its discovery precedes the diagnosis of Behçet's disease in half of the cases. This case presented increased complexity

in terms of diagnostic management because the diagnosis of Behçet's disease was uncertain at the time of the cardiac complication.

A multidisciplinary approach, including clinical discussion among clinical cardiology, cardiac imaging, cardiac surgery, and internal medicine, was crucial to recognize the final diagnosis and its complications and to formulate effective treatment.

CONSENT

As per international standards or university standards, patient(s) written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standards or university standards written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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