# Right ventricular thrombus with pulmonary embolism. A rare presentation of Behçet's disease

# Thrombus du ventricule droit avec embolie pulmonaire. Présentation rare de la maladie de Behçet.

Mariem Jeber, Leila Abid, Rania Hammami, Salma Charfeddine, Amine Bahloul, Tarek Ellouz, Dorra Abid, Faten Triki, Souad Mallek, Mourad Hentati, Samir Kammoun

*Service de cardiologie, Hôpital Hédi Chaker, sfax-Tunisie*
*Correspondance: Mariem Jeber, Service de cardiologie, Hôpital Hédi Chaker, sfax-Tunisie, email: mariemjabeur2@gmail.com*

**Résumé**

Nous rapportons l'observation d'un patient se présentant pour douleur thoracique associée à une dyspnée. Les explorations retrouvaient un thrombus ventriculaire droit compliqué d'embolie pulmonaire. Le diagnostic étiologique concluait à une maladie de Behçet. Un traitement à base de cyclophosphamide et de prednisone durant 3 mois a permis la résorption quasi complete du thrombus.

Mots-clés

Maladie de Behçet, Thrombus intracardiaque, Embolie pulmonaire

**Summary**

We report the case of an adult patient presenting with chest pain and dyspnea. Detailed clinical work up of the patient showed right ventricular thrombus and pulmonary embolism along with the prescribed criteria for the diagnosis of Behcet's disease. Three months of therapy with cyclophosphamide and prednisolone resulted in near complete clinicoradiological response.

Keywords

Behcet's disease, intracardiac thrombus, pulmonary embolism

*Correspondance*
*Mariem Jeber, Service de cardiologie, Hôpital Hédi Chaker, sfax-Tunisie.*
*mariemjabeur2@gmail.com*

## INTRODUCTION

Behcet's disease (BD) is a multisystem, chronic inflammatory disorder of unknown etiology. First described by Behcet Hulusi in 1937 in a patient with oral and genital ulcer and iridocyclitis [1]. Other organ involvement includes skin, joint, central nervous system, gastrointestinal tract, lung and cardiovascular system. This disease is frequent among the Mediterranean, Middle east and Far eastern populations [2].

Cardiopulmonary involvement include wide spectrum of abnormalities. Cardiac manifestations in Behcet's disease occur in about 1-5% of cases [3]. It includes coronary artery disease, recurrent pericarditis, myocardiopathy and endocardiac abnormalities. Intracardiac thrombus formation is a rare and serious complication. It often occurs in association with pulmonary thromboembolism, in about 52% of cases [4]. Young males seem to be most at risk and the right heart is the most frequent site of involvement [4].

Pulmonary involvement includes abnormalities of the vessel lumen and its wall, lung parenchyma, pleura and mediastinal structures [5]. Pulmonary arteries are the second most common site of arterial involvement preceded by the aorta [6]. Aneurysms are more common than thrombosis [6]. Thrombosis of the pulmonary arteries is usually in situ [7].

Considering the rarity of this disease and its distinct presentation with intracardiac thrombus and pulmonary embolism, we report this case.

## CASE REPORT

A 29-year-old male presented with chest pain and dyspnea NYHA stage IV of two days duration. The patient complained of low grade fever, decreased appetite and weight loss since one month. On active questioning, the patient revealed recurrent oral and genital ulcers during last 6 months. The patient denied any history of similar illness in his family.

General physical examination showed respiratory rate 25 breaths/minute, blood pressure 120/70 mm Hg, and pulse rate 115 beats/minute. Pallor and lymphadenopathy were absent. Examination of oral cavity showed multiple oral ulcers. Scars of old ulcers were present on the scrotum.

Routine laboratory tests revealed hemoglobin 12.7 g/dL, total leukocyte counts 10860/mm3 with neutrophils at 75% and ESR at 80 mm in first hour. His fasting blood sugar, renal and hepatic functions were within normal limits. His serum immunological profile showed no anomalies (Negative cANCA, pANCA, rheumatoid factor and antinuclear antibody).

Pulmonary angiography was performed and showed pulmonary embolism interesting lobar arterial branch and a soft tissue density focus in the right ventricle suggestive of thrombus.

Echocardiography showed a 16 × 14 mm mass in right ventricular cavity with no evidence of any structural heart disease (Figure 1).

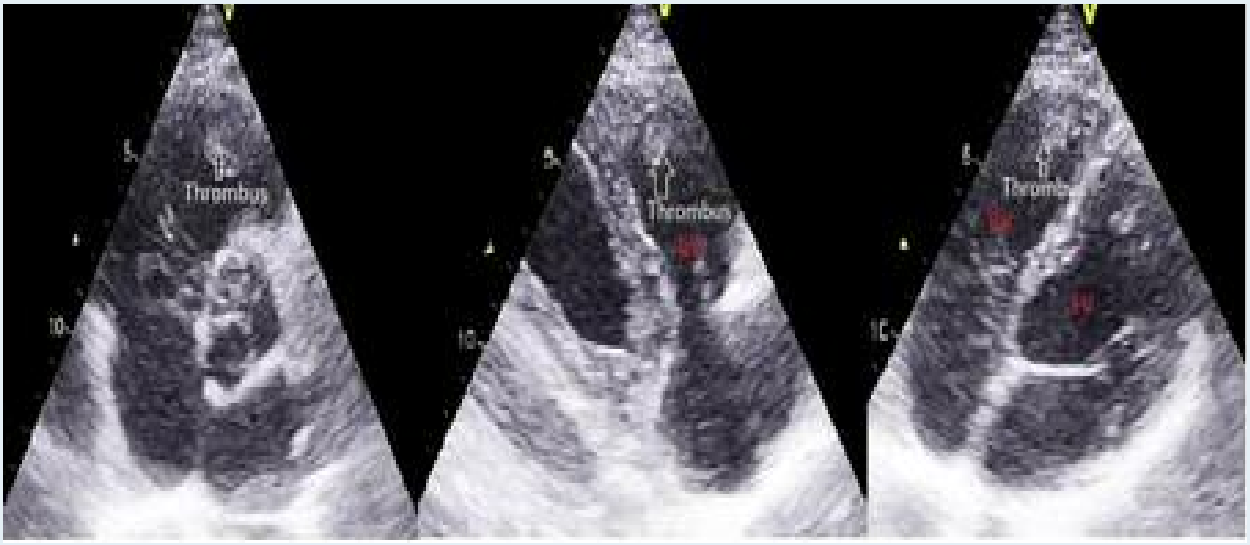


**Figure 1:** : Echocardiography showed an organized thrombus 16x14mm adherent to the anterior wall of the right ventricle. RV : Right ventricle ; LV : Left ventricle

Cardiac Magnetic resonance imagering (MRI) confirmed the existence of a 25mm thrombus hanging on the anterior wall of the right ventricle (Figure 2).

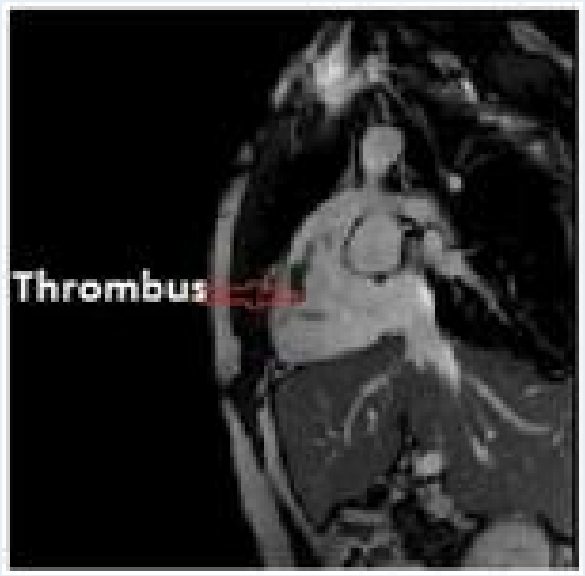


**Figure2 :** MRI showed a 25mm right ventricular thrombus

The diagnosis of Behcet's disease with intracardiac thrombus and pulmonary embolism was made on the clinical and radiological findings.
Cyclophosphamide at 0,6 g/m2 monthly was prescribed, along with 30 mg of prednisolone /daily tapered over 1 month to 10 mg daily associated with anticoagulation. After 3 months of therapy, repeat echocardiography showed resolution of right ventricular thrombus (Figure 3).

## DISCUSSION

The diagnosis of BD is considered on the basis of newer international criteria for Behçet's disease (ICBD)[8]. This new criteria has additionally incorporated vascular manifestations (VMs) to the earlier five criteria of international study group [1].
Intracardiac thrombi in BD may result from endomyocardial fibrosis, which may be a sequele of vasculitis involving endocardium, myocardium or both [9]. As intracardiac thrombus is tightly attached to the endocardium, embolism from the cardiac cavity seems to be relatively uncommon [10]. Young males seem to be most at risk than female and the right heart is the most frequent site of involvement for intracardiac thrombus formation [4,11].
The diagnosis of intracardiac thrombi in BD 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 tumor and myxoma [10,12].
In a series of 137 patients with BD only one patient was found to have right ventricular thrombus [13]. Similarly, only one patient was observed to have intracardiac thrombus out of 56 (1.78%) patients by Uçan et al [14].

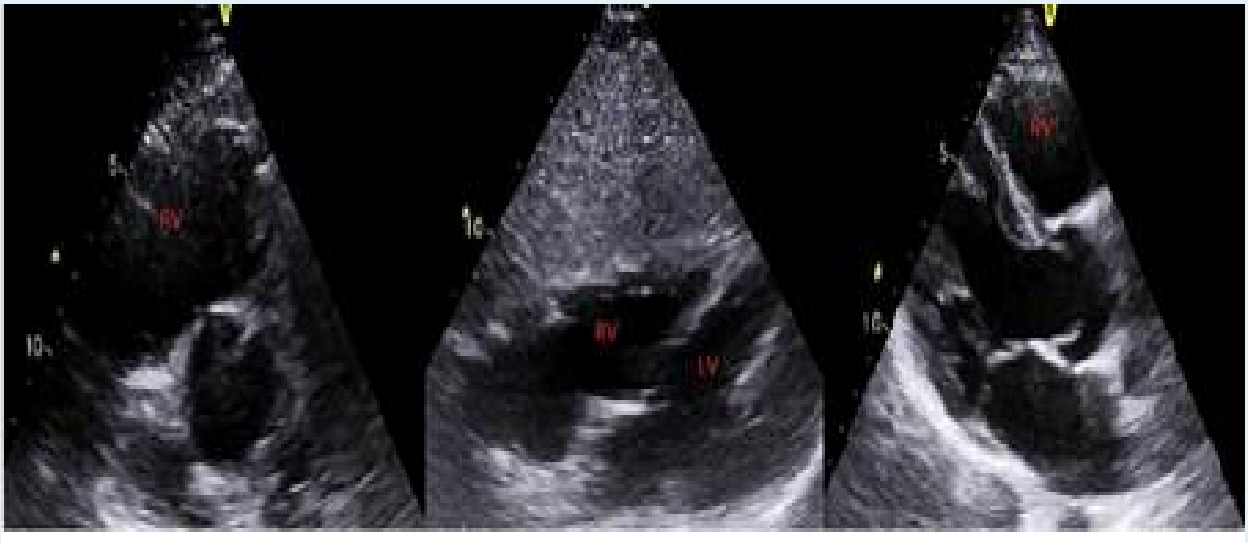

**Figure 3:** :: Echocardiography showed resolution of the right ventricular thrombus

It is well established that Behcet's disease predisposes strongly to venous and arterial thrombosis [4]. Recurrent superficial and deep thrombophlebitis of the lower extremities are the most common abnormalities, followed by vena cava thrombosis [4]. Fifty per cent of patients with right ventricular thrombus had pulmonary arterial thrombosis [4].

Cyclophosphamide and corticosteroids are the main stay of medical treatment of this life-threatening disease carrying poor prognosis. However, early diagnosis and aggressive therapy result in remission with resolution of right ventricular thrombus. There is no controlled trial of anticoagulant and thrombolytic agents in BD [13]. Recently, Pigaet al.[15] successfully used thrombolytic therapy for recurrent right ventricular thrombosis in a patient with BD.

## REFERENCES


1. Criteria for diagnosis of Behçet's disease. International study group for Behçet's disease. Lancet.1990;335:1078–80.
2. Düzgün N, Küçükşahin O, Atasoy KÇ, Togay Işıkay C, Gerede DM, Erden A, et al. Behçet's disease and intracardiac thrombosis: A report of three cases. Case Rep Rheumatol. 2013;2013:637015.
3. Wechsler B, Du LT, Kieffer E. Cardiovascular manifestations of Behçet's disease. Ann Med Interne (Paris) 1999;150:542–54.
4. Mogulkoc N, Burgess MI, Bishop PW. Intracardiac thrombus in Behçet's disease: A systematic review.Chest. 2000;118:479–87.
5. O'Duffy JD, Carney JA, Deodhar S. Behçet's disease. Report of 10 cases, 3 with new manifestations.Ann Intern Med. 1971;75:561–70.
6. Erkan F, Gül A, Tasali E. Pulmonary manifestations of Behçet's disease. Thorax. 2001;56:572–8.
7. Yılmaz S, Cimen KA. Pulmonary artery aneurysms in Behçet's disease. Rheumatol Int. 2010;30:1401–3.
8. International Team for the Revision of the International Criteria for Behçet's Disease. "Evaluation of the International Criteria for Behçet's disease (ICBD)" Clin Exp Rheumatol. 2006;24(Suppl 42):S13.
9. Houman M, Ksontini I, Ben Ghorbel I, Lamloum M, Braham A, Mnif E, et al. Association of right heart thrombosis, endomyocardial fibrosis, and pulmonary artery aneurysm in Behçet's disease. Eur J Intern Med.2002;13:455.
10. Erkan F, Esen K, Tunaci A. Pulmonary complications of Behçet's disease. Clin Chest Med.2002;23:493–503.
11. Vanhaleweyk G, el-Ramahi KM, Hazmi M, Sieck JO, Zaman L, Fawzy M. Right atrial, right ventricular and left ventricular thrombi in (incomplete) Behçet's disease. Eur Heart J. 1990;11:957–9.
12. El Houari T, Oukerraj L, Ghzaiel L, Fellat I, Azeroual M, Serraj K, et al. Management of Behçet disease with multiple complications. Hellenic J Cardiol. 2009;50:420–2.
13. Kiraz S, Ertenli I, Oztürk MA, Haznedaroğlu IC, Celik I, Calgüneri M. Pathological haemostasis and "prothrombotic state" in Behçet's disease. Thromb Res. 2002;105:125–33.
14. Uçan ES, Kıter G, Abadoğlu Ö, Karlikaya C, Akoğlu S, Bayindir Ű. Thoracic manifestations of Behçet's disease: Reports of the Turkish authors. Turkish Respiratory Journal. 2001;2:39–44.
15. Piga M, Puchades F, Mayo I, D'Cruz D. Successful thrombolytic therapy for recurrent right ventricular thrombosis in Behçet's disease. Clin Exp Rheumatol. 2010;28(Suppl 60):S76–8.