Available online at www.rajournals.in

RA JOURNAL OF APPLIED RESEARCH
ISSN: 2394-6709
DOI:10.47191/rajar/v9i4.11
Volume: 09 Issue: 04 April-2023

International Open Access

Impact Factor- 8.174

Page no.- 221-225

A Rare Case of Behçet Disease Presenting with sepsis, Pulmonary Embolism and Right Ventricular Thrombus

M.A. Skandaji[1], M. Ibenchekroun[1], O. Elkhabouty[1], A. Sefiani[1], H. Leghlimi[2], H. Hanani[3], Z. Mehsseni[1], N. Fellat[1]

[1] Cardiology A department IBN SINA Hospital, Rabat, Morocco
[2] Cardiovascular reanimation department IBN SINA Hospital, Rabat, Morocco
[3] Internal medicine department IBN SINA Hospital, Rabat, Morocco

ARTICLE INFO

Published Online:
17 April 2023

Corresponding Author:
M.A. Skandaji

ABSTRACT

Behçet Disease is a chronic systemic inflammatory disorder that manifests as recurring mouth ulcers and a number of systemic symptoms, such as genital ulcers, eye disease, skin lesions, arthritis, genital warts and gastrointestinal, neurologic, or vascular disease. Both men and women are equally affected by it, and it is most frequent along the historic Silk Road, which runs from eastern Asia to the Mediterranean region. The majority of people begin to exhibit symptoms between the ages of 20 and 40. (1)

We describe a rare case of a 21-year-old man with two right ventricular masses and pulmonary embolism due to a not yet diagnosed Behçet's disease.

KEYWORDS: Behçet disease, thrombus, embolism, heart

CASE REPORT

A 21 year-old man was admitted in our hospital with a 5-month history of recurrent fever, chest discomfort and dry cough the patient had recurrent oral ulcers (at least 2 times a year) and genital ulcers (at least once a year).He also had hemoptysis. There was no history of eye, joint, or neurological involvement. He did not have any other systemic manifestations. On taking a detailed history, he had recurrent fever for 1 month before hospitalization, with temperature spikes of around 38.5°C. During this period, a practitioner treated him with amoxicillin plus clavulanic acid and paracetamol. The fever persisted, although less frequent over the next few days. There was a significant loss of weight. He was not on any long-term medications and had no history of tobacco or alcohol abuse. His family history was unremarkable. On the day of admission, he was afebrile at 37°C. His blood pressure was 120/70 mm Hg and his heart rate was 90/minute. His systemic examination was normal apart from a genital lesion. Laboratory findings showed the following: total white blood cell count at 14.3 x 10^9 /L (neutrophils 81%) , hemoglobin at 9.7 g/dL; platelets at 260 x 10^9 /L ; aspartate transaminase at47 U/L; alkaline phosphatase, 148 U/L, C-reactive protein (CRP) at 279 mg/ L, procalcitonin at 1.3 and blood cultures were negative. His temperature spiked to 39°C on the night of admission. After blood cultures was taken, intravenous Triaxon, Bactrim and ciprofloxacin and oral paracetamol was empirically started to treat any possible underlying infection. A transthoracic echocardiography (TTE)(Figure 1) was performed to rule out predisposing cardiac conditions. The TTE showed a left ventricular ejection fraction at 65%, normal left ventricular systolic function, no right ventricular systolic impairment but lightly dilated RV/LV >1) and tow masses in the right ventricle (RV) of 30 and 20 mm as longest axis; knobbly, like thrombus. The first one sits on surface of septum but appears separate from endocardium. It has not a typical aspect of an invasive tumor. The other one on the RV free wall and no atrioventricular septal defect was seen.

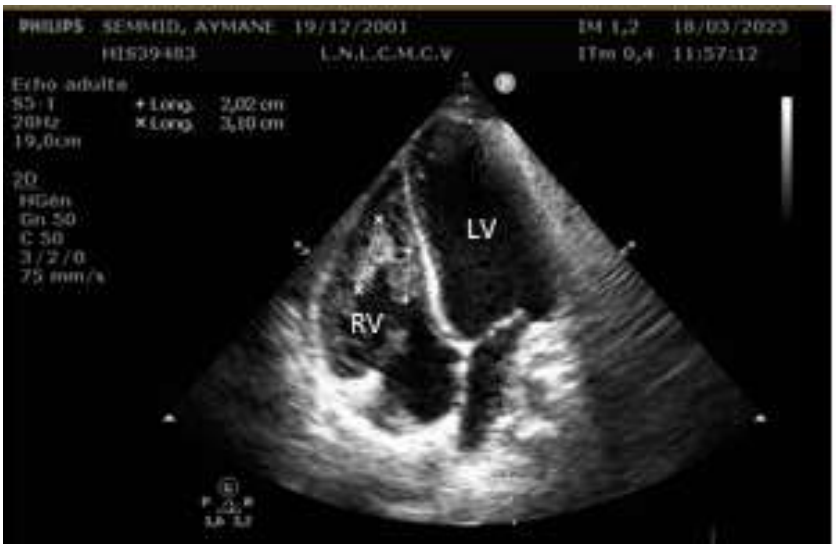

Figure 1 : A 4 chambre TTE showing the two RV embolism

A Chest CT (figure 2) was performed to search eventual thrombus in the pulmonary system and reveals an apical segmental pulmonary embolism.

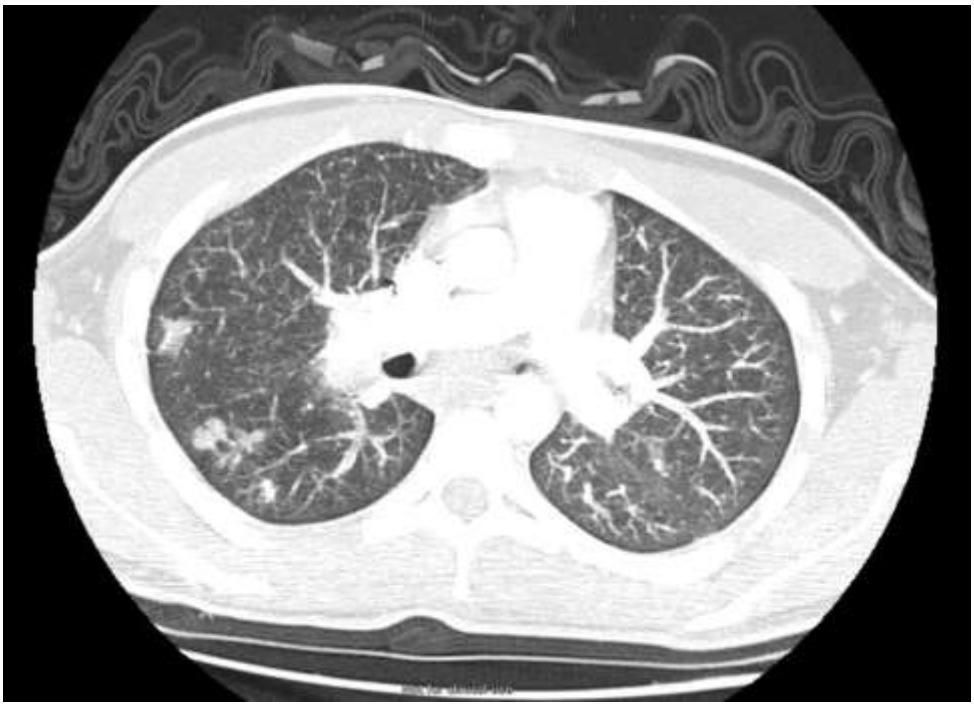

Figure 2 : CT showing an apical segmental pulmonary embolism

As per the recommendations, a cardiac magnetic resonance imaging (MRI) was performed to further evaluate the nature of the intrathoracic masses. The MRI revealed a right ventricular lesion suggestive of two thrombus (Figure 3). He was started on anticoagulation subsequently and remained relatively afebrile for the next few days.

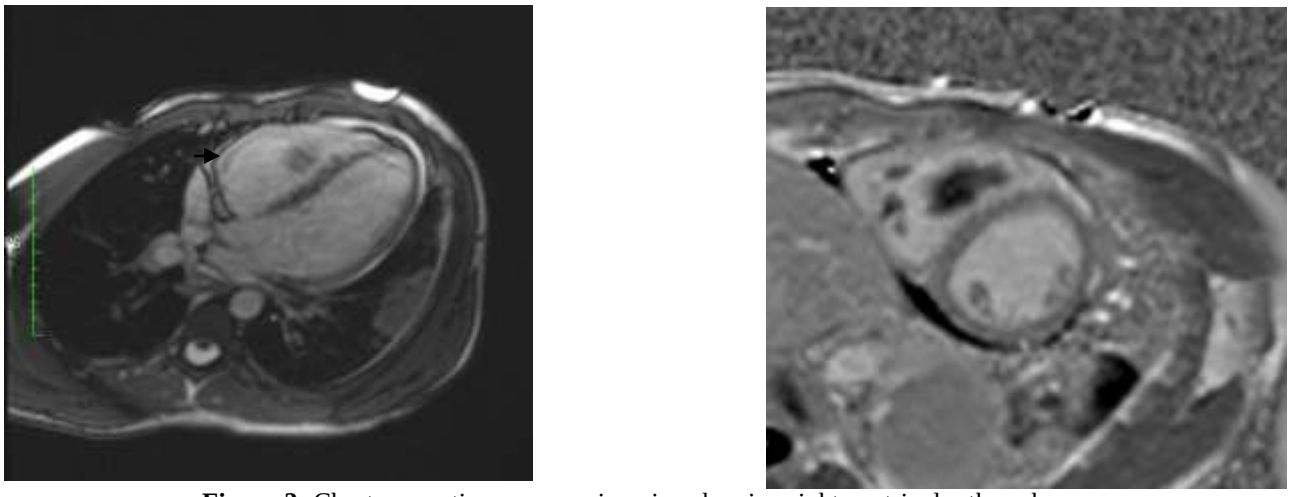

Figure 3: Chest magnetic resonance imaging showing right ventricular thrombus

A CTAP scan was performed to search for any masses in the rest of the body. It was negative.

The patient remained afebrile under the same antibiotherapy. Anticoagulation remained until the infection was irradicated. Then corticotherapy was started.

DISCUSSION

The ICBD criteria requires at least three points for the diagnosis of Behçet Disease: genital aphthosis (two points), ocular lesions (two points), oral aphthosis (one point), skin lesions as pseudofolliculitis or erythema nodosum (one point), vascular lesions (superficial phlebitis, DVT, large vein thrombosis, arterial thrombosis, or aneurysm) (1 point), and pathergy (1 point). According to history, this patient fits both the diagnostic criteria of Behçet Disease.(2-3)

Vascular involvement has been considered to result from systemic vasculitis, and it occurs in 5 to 10% of these patients. (4)

Although rare, Behçet Disease complicated by right ventricular thrombosis and/or pulmonary embolism has been reported (5-6). And the right heart is the most common site of involvement (7-8). 1.9% of Behcet's disease patients have been documented to have the potentially fatal complication known as cardiac thrombus. It generally affects the right heart chambers and typically affects men under the age of 40.

Behcet's disease cardiovascular signs have been documented in 7%–46% of people, with a high death rate of patients with apparent cardiovascular involvement surpassing 20%. Although thrombosis is the most typical symptom of cardiovascular Behcet, occasional cases of pericarditis, myocarditis, coronary disease, congestive heart failure, ventricular thrombosis, aortic aneurysm, and valvular disease have also been reported(7-8).

Patients frequently arrive at the hospital with fever hemoptysis, hemorrhagic rash, cough, dyspnea and chest discomfort. When the pulmonary artery is affected, fever is more prevalent (9-10).

Therefore, obtaining a complete history is crucial for further workup and management, particularly with patients who have not yet received a Behçet Disease diagnosis. Early identification in these situations can be difficult because the initial symptoms, such as fever, cough, dyspnea, chest pain, and hemoptysis, are frequently nonspecific(11).

However, even in the lack of the defining clinical features of the condition, Behçet Disease diagnosis may still be given if a patient has a mass in the right-sided cardiac chambers(12)

This is particularly applicable if the patient is a young male from the Eastern Asia, Mediterranean basin, or the Middle East (13) though such conditions can also occurs in patients without predisposing ethnic or geographic factor(14).

When a chest CT scan was performed on this patient to evaluate pyrexia of unknown origin, pulmonary embolism and a right ventricle thrombus were unintentionally discovered. His clinical manifestations were conclusively linked to the underlying Behçet Disease especially after a review of the patient's prior medical history.

The management of Behçet Disease, particularly with large-vessel manifestations (15), depends on an early diagnosis and treatment. For the detection of lung and intracardiac thromboembolism, chest CT or MRI are frequently used (6) and echocardiography is performed to evaluate the efficacy of therapy and follow-up

Before being admitted to the hospital, the patient received treatment with oral amoxicillin and clavulanic acid but his temperature and other symptoms persisted. His fever decreased while he was hospitalized after starting anticoagulant medication in addition to the empirical antibiotic therapy. Although it is uncommon, pulmonary embolism with thrombosis has been linked to reports of high fever and fever of this cause responds to anticoagulant therapy (19).In addition, fever is another frequent Behçet Disease appearance(20).

Medical treatment is the cornerstone of the treatment (21). The most favored and highly effective method is anticoagulation with/without immunosuppression therapy. Cyclophosphamide and corticosteroids are known to be effective(22-5) . However, surgery to remove intracardiac thrombus has rarely been performed owing to hemodynamic

compromises (23). An extensive examination should be conducted to rule out any potential infections before the start of immunosuppression treatment.

Because of lack of data, the comparison of efficacy between anticoagulant therapy alone and anticoagulant therapy together with immunosuppression therapy is not well known.

CONCLUSION

Because of lack of specificity, early diagnosis may be challenging. Therefore, accurate imaging tests, a comprehensive physical exam, and through history are all necessary for diagnosis. Anticoagulation with or without immunosuppression therapy is used as the method of management, and echocardiography is used to closely monitor the size of the thrombus.

An integrated management strategy involving the vascular physician, rheumatologist, and cardiologist will eventually be beneficial for these patients.

REFERENCES

1. Aksu T, Tufekcioglu O.: Intracardiac thrombus in Behçet's disease: four new cases and a comprehensive literature review. Rheumatol Int 2015; 35: pp. 1269-1279.
2. Kobayashi T, Kishimoto M, Swearingen CJ, et al. Differences in clinical manifestations, treatment, and concordance rates with two major sets of criteria for Behçet's syndrome for patients in the US and Japan: data from a large, three-center cohort study. Mod Rheumatol 2013 May; 23(3):547–553
3. Davatchi F, Sadeghi Abdollahi B, Shahram F, et al. Validation of the International Criteria for Behçet's disease (ICBD) in Iran. Int J Rheum Dis 2010;13(1):55–60
4. Kale A, Akyildiz L, Akdeniz N, Kale E. Pregnancy complicated by superior vena cava thrombosis and pulmonary embolism in a patient with Behcet disease and the use of heparin for treatment. Saudi Med J 2006;27(1):95–97
5. Hammami R, Abid L, Frikha F, et al. Intracardiac thrombus in a young man: don't forget Behçet's disease!. Intern Med 2012; 51(14):1865–1867
6. Aksu T, Tufekcioglu O.: Intracardiac thrombus in Behçet's disease: four new cases and a comprehensive literature review. Rheumatol Int 2015; 35: pp. 1269-1279.
7. Khammar Z, Berrady R, Boukhrissa A, et al. [Intracardiac thrombosis in Behçet disease: clinical presentation and outcome of three cases]. J Mal Vasc 2011;36(4):270–273
8. Yıldırım R, Dinler M, Yaşar Bilge NŞ, Kaşifoğlu T: A rarely seen manifestation in Behcet's disease: intracardiac thrombosis. Clin Rheumatol 2021; 40: pp. 4355-4356.
9. 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(4, Suppl 60):S76–S78
10. Ozer N, Ciftçi O, Demirci M, et al. Right ventricular outflow tract thrombus in a case of Behcet's disease. J Am Soc Echocardiogr 2005;18(4):380–382.
11. Houman M, Ksontini I, Ben Ghorbel I, et al. Association of right heart thrombosis, endomyocardial fibrosis, and pulmonary artery aneurysm in Behçet's disease. Eur J Intern Med 2002; 13(7):455.
12. Gopathi S, Hurt RT, Guardiola J. Intracardiac thrombus in Behcet's disease: a rare case in the United States. Respir Med CME 2011;4 (4):154–156.
13. Fekih M, Fennira S, Ghodbane L, Zaouali RM. [Intracardiac thrombosis: unusual complication of Behcet's disease]. Tunis Med 2004; 82(8):785–790
14. Darie C, Knezinsky M, Demolombe-Rague S, et al. [Cardiac pseudotumor revealing Behçet's disease]. Rev Med Interne 2005;26 (5):420–424
15. Kajiya T, Anan R, Kameko M, et al. Intracardiac thrombus, superior vena cava syndrome, and pulmonary embolism in a patient with Behçet's disease: a case report and literature review. Heart Vessels 2007;22(4):278–283.
16. San Luis Miranda R, Lázaro Castillo JL, Enciso Gómez R, et al. [Right ventricular thrombus and pulmonary artery aneurysms in Behçet's disease. Report of one case]. Arch Cardiol Mex 2007;77 (2):130–136
17. Emad Y, Abdel-Razek N, Gheita T, el-Wakd M, el-Gohary T, Samadoni A. Multislice CT pulmonary findings in Behçet's disease (report of 16 cases). Clin Rheumatol 2007;26(6):879–884.
18. Sayin AG, Vural FS, Bozkurt AK, Oz B, Uygun N. Right atrial thrombus mimicking myxoma and bilateral pulmonary artery aneurysms in a patient with Behçet's disease—a case report. Angiology 1993;44(11):915–918
19. Calvo-Romero JM, Lima-Rodríguez EM, Pérez-Miranda M, BureoDacal P. Low-grade and high-grade fever at presentation of acute pulmonary embolism. Blood Coagul Fibrinolysis 2004;15(4): 331–333.
20. Granel B, Berbis P, Serratrice J, Pelletier J, Disdier P, Weiller PJ. [An unusual cause of long-term fever: Behçet disease]. Presse Med 1999;28(40):2221–2222.
21. Alibaz-Oner F., et. al.: Behcet disease with vascular involvement: effects of different therapeutic regimens on the incidence of new relapses. Medicine 2015; 94: pp. e494.

22. Noureddine M, Charei N, Drighil A, Chraibi N. [Right intracardiac thrombus in Behçet's disease]. Arch Mal Coeur Vaiss 2004;97 (9):925–928.
23. Espinosa G, Blanco I, Antón JM, Sánchez M, Macchiarini P, Barberà JA. Chronic thromboembolic pulmonary hypertension in Behçet's disease: effectiveness of endarterectomy. Clin Exp Rheumatol 2010;28(4, Suppl 60):S79–S81.