Clinical Rheumatology
https://doi.org/10.1007/s10067-021-05720-9

CLINICAL IMAGE

A rarely seen manifestation in Behcet's disease: intracardiac thrombosis

Reşit Yıldırım [1] · Mustafa Dinler [1] · Nazife Şule Yaşar Bilge [1] · Timuçin Kaşifoğlu [1]

Received: 20 January 2021 / Revised: 19 March 2021 / Accepted: 24 March 2021
© International League of Associations for Rheumatology (ILAR) 2021

Case presentation

A 27-year-old male presented with acute-onset dyspnea, pain, and diffuse swelling in both lower extremities. Past medical history was noncontributory except for recurrent oral ulcers. Physical examination revealed tachycardia, papulopustular lesions on the upper back of the body, and positive Homans sign in the lower extremities bilaterally. Lung examination was clear, but mild sinus tachycardia was present on electrocardiogram (ECG). Laboratory investigations showed high acute phase reactants and elevated D-dimer levels. Doppler imaging of the lower extremities exhibited bilateral widespread deep vein thrombosis throughout the femoral veins. No evidence of pulmonary embolism, vasculitis, and pulmonary aneurysm was found by computed tomography (CT) pulmonary angiography. However, a hypoechoic lesion consistent with thrombus formation was detected incidentally in the right ventricle of the heart (Fig. 1). Intracardiac thrombus formation was confirmed by transesophageal echocardiogram (TEE) (Fig. 2). Results of pathergy test and HLAB51 gene mutation were unrevealing. Lupus anticoagulant, anticardiolipin, and anti-β2 glycoprotein I antibodies were negative. Tests for factor V Leiden mutation, homocysteine, antithrombin III, prothrombin G20210A mutation, and proteins C and S were inconclusive. The patient was diagnosed with Behcet's disease (BD) based on International Criteria for Behcet's Disease (ICBD) [1]. High-dose intravenous steroid and cyclophosphamide in combination with warfarin were initiated. After completing the third dose of cyclophosphamide, control TEE demonstrated no evidence of intra-cardiac thrombosis. Six cycles of monthly cyclophosphamide were planned. The patient currently is receiving 6 mg of methylprednisolone and warfarin.

Discussion

Behcet's disease (BD) is primarily known by its mucocutaneous and ocular manifestations [2]. However, vascular involvement in BD, which may be seen in about 40% of patients, constitutes the hidden face of the disease leading to high morbidity and mortality rates. The most common site of thrombosis is the deep veins of the lower extremities, but atypical thrombosis such as portal venous thrombosis, pulmonary artery thrombosis, sagittal sinus thrombosis, and intracardiac

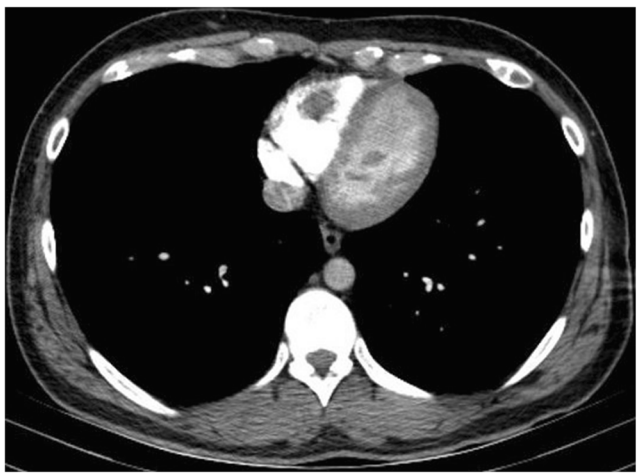


Fig. 1 Hypoechogenic lesion within the right ventricle of the heart seen by pulmonary CT angiography

✉ Reşit Yıldırım
celeng18@gmail.com

Mustafa Dinler
drmdinler@gmail.com

Nazife Şule Yaşar Bilge
suleyasar@yahoo.com

Timuçin Kaşifoğlu
timucinkasifoglu@hotmail.com

[1] Department of Internal Medicine, Division of Rheumatology, Osmangazi University School of Medicine, Meselik Kampusu, 26480 Eskisehir, Turkey

Published online: 30 March 2021

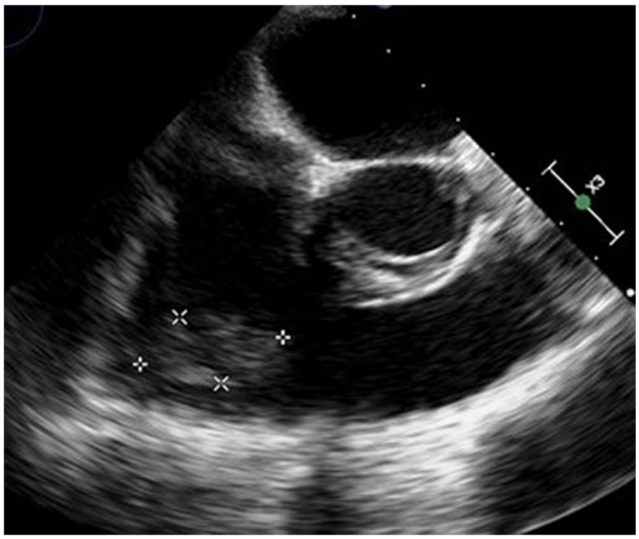


Fig. 2 Intracardiac thrombus in the right ventricle seen by transesophageal echocardiography

thrombosis may also be seen [3]. Cardiac thrombus in BD has been reported rarely (1.9%) in which nearly 40% of those may be the presenting manifestation [4]. It usually occurs in men younger than 40 years and mostly involves the right chambers of the heart [4, 5]. Intracardiac thrombus in BD is a life-threatening condition. Although no proven treatment is currently available, aggressive immunosuppression with combination of cyclophosphamide and high-dose steroid is recommended based on limited case-control studies and experiences from the treatment of other vascular involvement in BD. Of note, many experts also suggest anticoagulation with warfarin [4, 5]. In summary, the presence of intracardiac thrombus formation in the right-sided cardiac chambers even in the absence of the characteristic clinical features of BD should raise suspicion for diagnosis of BD, especially in a young male originating from the Mediterranean Basin or the Middle East.

Declarations

Disclosures None

Consent for publication We obtained written informed consent from the patient for publication of this case report together with any accompanying images.

References

1. Davatchi F, Assaad-Khalil S, Calamia KT, Crook JE, Sadeghi-Abdollahi B, Schirmer M, Tzellos T, Zouboulis CC, Akhlagi A, Al-Dalaan A, Alekberova ZS, Ali AA, Altenburg A, Arromdee E, Baltaci M, Bastos M, Benamour S, Ben Ghorbel I, Boyvat A, Carvalho L, Chen W, Ben-Chetrit E, Chams-Davatchi C, Correia JA, Crespo J, Dias C, Dong Y, Paixão-Duarte F, Elmuntaser K, Elonakov AV, Graña Gil J, Haghdoost A-A, Hayani RM, Houman H, Isayeva AR, Jamshidi, Kaklamanis P, Kumar A, Kyrgidis A, Madanat W, Nadji K, Namba S, Ohno I, Olivieri J, Vaz Patto N, Pipitone MV, de Queiroz F, Ramos C, Resende CM, Rosa C, Salvarani MJ, Serra F, Shahram H, Shams KE, Sharquie M, Sliti-Khanfir T, Tribolet dA, Vasconcelos C, Vedes J, Wechsler BY, Cheng K, Zhang Z, Ziaei N (2014) The International Criteria for Behçet's Disease (ICBD): a collaborative study of 27 countries on the sensitivity and specificity of the new criteria. Journal of the European Academy of Dermatology and Venereology 28 (3):338–347
2. Kural-Seyahi E, Fresko I, Seyahi N, Ozyazgan Y, Mat C, Hamuryudan V, Yurdakul S, Yazici H (2003) The long-term mortality and morbidity of Behçet syndrome: a 2-decade outcome survey of 387 patients followed at a dedicated center. Medicine (Baltimore) 82(1):60–76
3. Saadoun D, Wechsler B, Desseaux K, Le Thi Huong D, Amoura Z, Resche-Rigon M, Cacoub P (2010) Mortality in Behçet's disease. Arthritis Rheum 62(9):2806–12
4. Wang H, Guo X, Tian Z, Liu Y, Wang Q, Li M, Zeng X, Fang Q (2016) Intracardiac thrombus in patients with Behcet's disease: clinical correlates, imaging features, and outcome: a retrospective, single-center experience. Clinical Rheumatology 35(10):2501–2507
5. Emmungil H, Yaşar Bilge NŞ, Küçükşahin O, Kılıç L, Okutucu S, Gücenmez S, Kalyoncu U, Kaşifoğlu T, Turgay M, Aksu K (2014) A rare but serious manifestation of Behçet's disease: intracardiac thrombus in 22 patients. Clin Exp Rheumatol 32(4 Suppl 84):S87–92

Publisher's note Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.