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Case Report

Pulmonary Embolism in Behçet's Disease: When Vasculitis Drives Thrombosis: A Case Report

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Abstract

Behçet's disease is a chronic multisystem inflammatory disorder in which vascular involvement represents a major cause of morbidity and mortality. Although venous thrombosis is common, pulmonary embolism remains a rare and potentially life-threatening manifestation related to an inflammation-driven thrombotic mechanism. We report the case of a 70-year-old man with long-standing Behçet's disease complicated by previous superior vena cava thrombosis, who was diagnosed with pulmonary embolism. Clinical presentation was nonspecific. Clinical probability assessment using the Wells and revised Geneva scores guided the diagnostic approach. Electrocardiographic findings were non-diagnostic, while transthoracic echocardiography allowed hemodynamic assessment and risk stratification. Computed tomography pulmonary angiography confirmed the diagnosis. This case underscores the diagnostic and therapeutic challenges of pulmonary embolism in Behçet's disease and highlights the need for individualized management balancing immunosuppressive therapy and anticoagulation

Keywords: Behçet's disease, vascular involvement, pulmonary embolism.

INTRODUCTION

Behçet's disease (BD) is a chronic, relapsing, multisystem inflammatory disorder of unknown etiology, characterized by recurrent mucocutaneous, ocular, and systemic manifestations. Although mucocutaneous involvement is the most common feature, systemic complications—particularly vascular involvement—are the primary determinants of morbidity and mortality [1,2].

Vascular involvement affects approximately 25–40% of patients with BD and may involve both the venous and arterial systems. Venous thrombosis is the most frequent vascular manifestation, commonly affecting the deep veins of the lower limbs, the inferior and superior vena cava, and cerebral venous sinuses. Arterial involvement, although less frequent, includes aneurysmal and occlusive lesions and is associated with a poorer prognosis [2,3].

Pulmonary vascular involvement is relatively uncommon but potentially life-threatening. Pulmonary embolism is a rare complication of Behçet's disease, with a reported prevalence below 5% among patients with vascular involvement [3,4]. This apparent rarity is explained by the distinctive pathophysiology of thrombosis in BD, which is primarily driven by inflammatory vasculitis rather than classical embolic migration from peripheral veins. Thrombi formed in this context are typically firmly adherent to the inflamed vessel wall, limiting their embolic potential [4,5]. Despite its low frequency, pulmonary embolism remains a major diagnostic and therapeutic challenge in patients with Behçet's disease.

CASE REPORT

1. Patient information

A 70-year-old man with no cardiovascular risk factors had been followed for Behçet's disease since 1980 and was treated with colchicine, with irregular follow-up. His medical history was marked by the occurrence of superior vena cava thrombosis in 1989, for which long-term anticoagulant therapy was initiated. The patient presented to the emergency department with acute pleuritic chest pain associated with sudden-onset dyspnea, prompting urgent medical evaluation.

2. Clinical findings

On admission to the cardiac intensive care unit, the patient was tachycardic (heart rate: 121 bpm) with borderline blood pressure (102/56 mmHg). Physical

examination revealed bilateral basal crackles on lung auscultation. Cardiac auscultation showed no murmurs, and abdominal examination was unremarkable.

3. Diagnostic assessment

The initial electrocardiogram (ECG) showed left axis deviation with signs of left ventricular hypertrophy. **Fig 1**

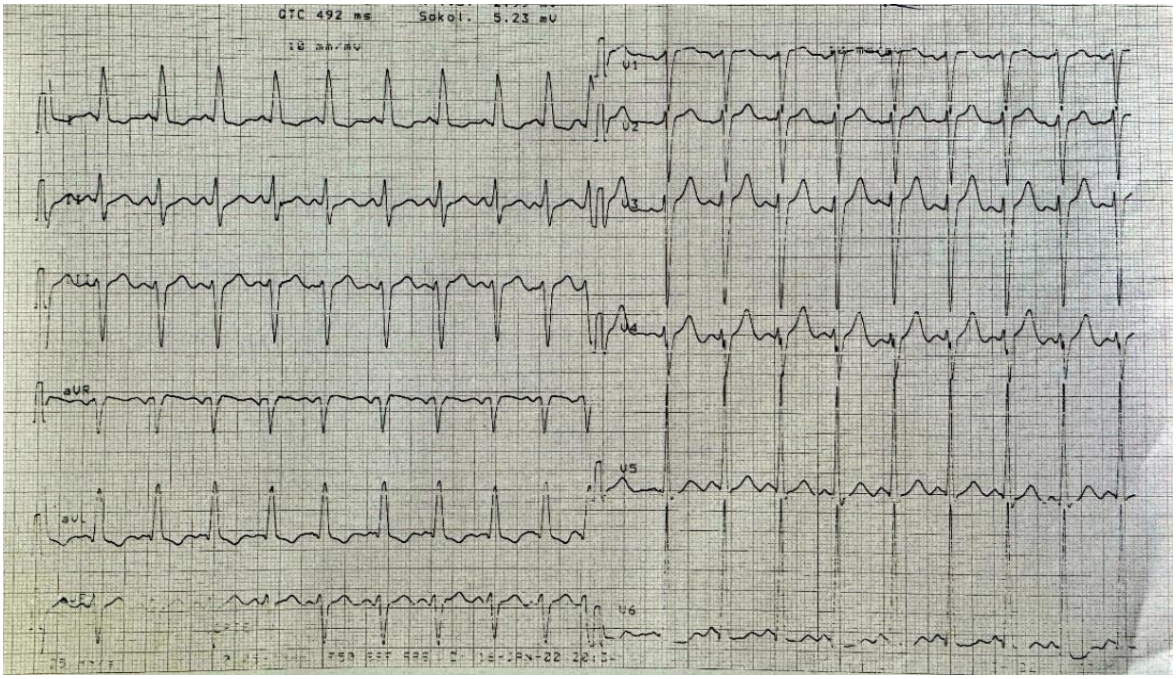

Figure 1: Twelve-lead electrocardiogram showing sinus rhythm with left ventricular hypertrophy according to the Sokolow–Lyon criterion (Sokolow index = 50 mm)

Bedside transthoracic echocardiography revealed global left ventricular hypokinesia with a reduced left ventricular ejection fraction of 34%, associated with elevated left ventricular filling pressures, while right ventricular size and systolic function were preserved. Fig 2

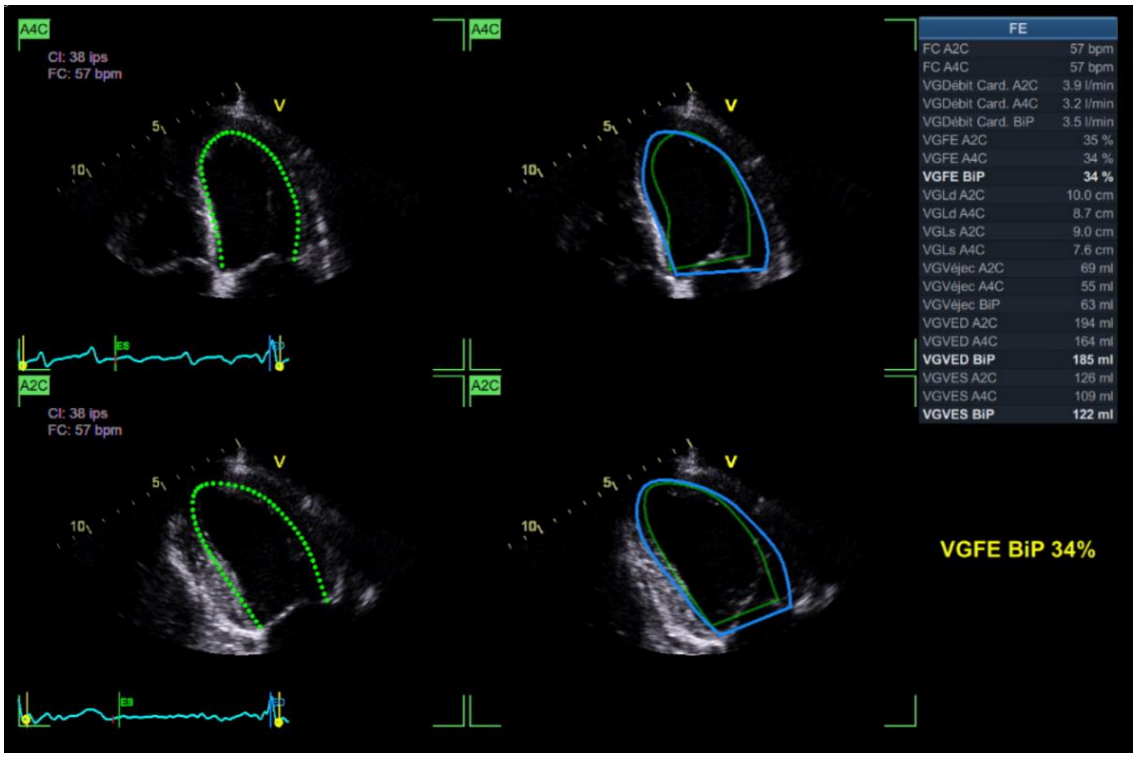

Figure 2: Transthoracic echocardiographic image showing a reduced left ventricular ejection fraction (LVEF)

Given the clinical suspicion of pulmonary embolism, clinical probability scores were calculated: the Wells score was 3 and the revised Geneva score was 9, indicating an intermediate clinical probability.

Laboratory investigations showed markedly elevated D-dimer levels (1000 ng/mL), elevated C-reactive protein (128 mg/L), and positive cardiac troponin levels (116 ng/L). Renal function was mildly impaired, while hematologic parameters were within normal ranges.

Urgent computed tomography pulmonary angiography demonstrated bilateral subsegmental pulmonary emboli. In addition, imaging findings were consistent with chronic thrombosis of the superior vena cava, involving the brachiocephalic trunk and jugular veins. Fig 3

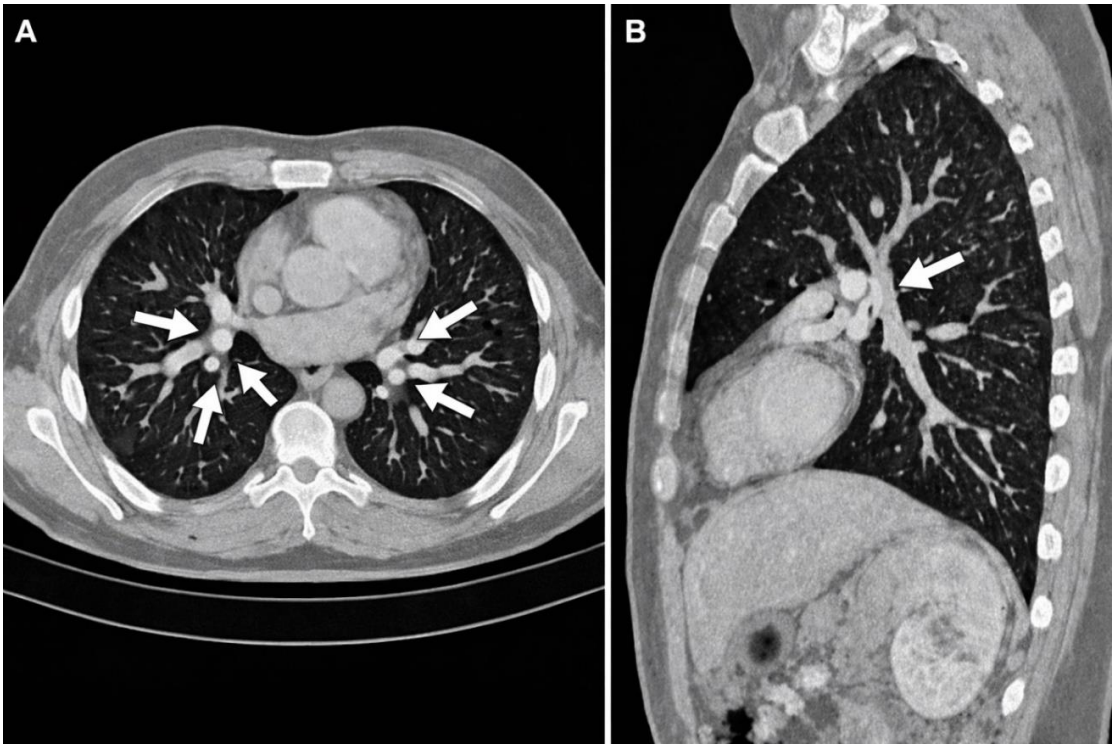

Figure 3: Images of computed tomography pulmonary angiography; A. Axial computed tomography pulmonary angiography (CTPA) demonstrating bilateral subsegmental pulmonary emboli (white arrows). B. Sagittal reconstruction confirming a left subsegmental pulmonary embolus (white arrow).”

4. Diagnosis

Acute pulmonary embolism with intermediate–low risk, according to the Pulmonary Embolism Severity Index (PESI class II), occurring in the context of Behçet’s disease with previous major venous thrombosis.

5. Therapeutic intervention

The patient was initially treated with low-molecular-weight heparin, followed by a rapid switch to direct oral anticoagulant therapy after clinical stabilization, in addition to standard heart failure treatment.

6. Follow-up and outcomes

The clinical course was favorable, with progressive improvement of dyspnea and chest pain. Hemodynamic parameters stabilized, and no signs of clinical deterioration were observed. The patient was discharged on oral anticoagulation, with close outpatient follow-up.

7. Patient perspective:

The patient reported that the sudden onset of chest pain and shortness of breath was frightening and significantly affected his sense of well-being. Being informed of the possible complication of Behçet’s disease, particularly given his previous history of venous thrombosis, caused considerable anxiety. After initiation of anticoagulant therapy and appropriate medical management, the patient experienced progressive symptom relief. The patient expressed satisfaction with the care received and agreed to the publication of this case for educational purposes.

8. Informed Consent: Written informed consent was obtained from the patient for publication of this case report and accompanying images.

DISCUSSION

Pulmonary embolism (PE) represents a rare but clinically significant expression of vascular involvement in Behçet’s disease (BD). In contrast to the high prevalence of venous thrombosis observed in BD, PE

remains uncommon, with a reported frequency below 5% among patients with vascular involvement [3,4]. This discrepancy highlights the unique thrombotic mechanisms underlying vascular disease in BD.

The relative rarity of PE in BD, despite the high frequency of venous thrombosis, can be explained by the distinctive pathophysiological mechanisms of thrombosis in this disease. Unlike classical thromboembolic disease, in which emboli typically originate from deep veins of the lower limbs, thrombosis in BD is primarily driven by inflammation of the vessel wall. Immune-mediated vasculitis leads to endothelial activation and injury, resulting in local thrombus formation. These thrombi are characteristically firmly adherent to the inflamed endothelium, which significantly limits their tendency to detach and embolize [5]. Consequently, in situ pulmonary artery thrombosis is more common than true embolic migration from peripheral veins.

The pathophysiology of pulmonary vascular involvement in BD reflects a complex interplay between inflammation, endothelial dysfunction, and coagulation abnormalities. Pro-inflammatory cytokines, such as tumor necrosis factor-α and interleukin-6, promote endothelial damage, increase tissue factor expression, and suppress fibrinolysis, thereby creating a prothrombotic environment [6]. In addition, neutrophil hyperactivation and oxidative stress further contribute to vascular injury. Pulmonary embolism may therefore result either from in situ thrombosis of the pulmonary arteries or, less frequently, from embolization of peripheral venous thrombi. Importantly, pulmonary artery thrombosis may coexist with pulmonary artery aneurysms, a hallmark and particularly severe manifestation of BD-associated pulmonary vasculitis [7].

Clinically, PE in BD often presents with nonspecific symptoms, including dyspnea, pleuritic chest pain, cough, and occasionally hemoptysis. Hemoptysis is a key clinical feature, as it may suggest aneurysmal involvement rather than isolated thrombotic disease. Fever is frequently observed and is usually related to systemic inflammation rather than infection. Physical findings range from mild hypoxemia and tachycardia to signs of acute right ventricular strain in more severe cases.

Electrocardiographic findings are variable and nonspecific in pulmonary embolism. Sinus tachycardia is the most commonly reported abnormality, while signs suggestive of right ventricular overload—such as right axis deviation, right bundle branch block, or anterior T-wave inversions—may be observed in cases of significant pulmonary vascular involvement. Overall, ECG abnormalities mainly reflect hemodynamic consequences and should be interpreted in conjunction with clinical evaluation and imaging findings [8].

In the diagnostic approach to suspected pulmonary embolism, clinical probability scores, including the Wells score and the revised Geneva score, are essential tools for estimating pre-test probability and guiding subsequent investigations. Their use helps rationalize D-dimer testing and imaging strategies, particularly in Behçet's disease, where clinical presentation is often atypical and inflammatory manifestations may confound diagnosis.

Computed tomography pulmonary angiography remains the diagnostic modality of choice, enabling accurate identification of pulmonary artery thrombosis and detection of associated aneurysmal lesions, which are critical for therapeutic decision-making. Echocardiography provides complementary information by assessing right ventricular function and estimating pulmonary pressures.

Risk stratification using the Pulmonary Embolism Severity Index (PESI) is essential for estimating early mortality risk and directly informing management decisions. In patients with Behçet's disease, PESI classification helps identify those who may benefit from a conservative treatment strategy while ensuring appropriate monitoring and individualized therapeutic planning [8].

Management of PE in BD differs substantially from that of conventional PE. Immunosuppressive therapy represents the cornerstone of treatment, targeting the underlying vasculitic process. High-dose systemic corticosteroids are usually initiated, often combined with immunosuppressive agents such as cyclophosphamide or azathioprine. Biologic therapies targeting tumor necrosis factor-α have demonstrated efficacy in refractory or severe cases [9]. The role of anticoagulation remains controversial. While anticoagulants may be considered in carefully selected patients without pulmonary artery aneurysms, they are generally avoided in the presence of aneurysmal disease because of the high risk of life-threatening hemoptysis [10].

The prognosis of pulmonary embolism in BD largely depends on early diagnosis, careful assessment of pulmonary vascular involvement, and prompt initiation of appropriate immunosuppressive therapy. With adequate treatment, outcomes may be favorable; however, pulmonary vascular involvement remains a major cause of mortality, underscoring the need for long-term follow-up and multidisciplinary management.

CONCLUSION

Pulmonary embolism is a rare but potentially severe manifestation of vascular involvement in Behçet's disease. Its occurrence reflects the complex interplay between inflammation, endothelial dysfunction, and thrombosis rather than classical thromboembolic mechanisms. Early recognition of pulmonary vascular involvement and careful imaging assessment are

essential to guide appropriate management. This case highlights the importance of considering pulmonary embolism in patients with Behçet's disease presenting with acute respiratory symptoms and underscores the need for individualized therapeutic strategies, balancing immunosuppressive therapy and anticoagulation. Long-term follow-up and multidisciplinary management remain crucial to improving outcomes in this high-risk population.

CONSENT FOR PUBLICATION

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Conflicts of Interest: The authors declare no conflicts of interest.

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BIBLIOGRAPHY

1. Yazici H, Seyahi E, Hatemi G, Yazici Y. Behçet syndrome: a contemporary view. *Nat Rev Rheumatol.* 2018;14(2):107–119.
2. Maldini C, Lavalley MP, Cheminant M, de Menthon M, Mahr A. Relationships of HLA-B51 with Behçet's disease clinical characteristics. *Rheumatology (Oxford).* 2012;51(5):887–900.
3. Kural-Seyahi E, Fresko I, Seyahi N, *et al.,* The long-term mortality and morbidity of Behçet syndrome. *Medicine (Baltimore).* 2003;82(1):60–76.
4. Tascilar K, Melikoglu M, Ugurlu S, *et al.,*Vascular involvement in Behçet's syndrome. *Rheumatology (Oxford).* 2014;53(11):2018–2022.
5. Seyahi E. Behçet's disease: vascular involvement and therapeutic challenges. *Best Pract Res Clin Rheumatol.* 2016;30(2):279–295.
6. Butta NV, Fernández-Bello I, López-Longo FJ, Jiménez-Yuste V. Endothelial dysfunction and altered coagulation in Behçet disease. *Semin Thromb Hemost.* 2015;41(6):621–628.
7. Erkan F, Gul A, Tasali E. Pulmonary manifestations of Behçet's disease. *Thorax.* 2001;56(7):572–578.
8. Konstantinides SV, Meyer G, Becattini C, *et al.,*2019 ESC Guidelines on acute pulmonary embolism. *Eur Heart J.* 2020;41(4):543–603.
9. Hatemi G, Christensen R, Bang D, et al EULAR recommendations for the management of Behçet's syndrome. *Ann Rheum Dis.* 2018;77(6):808–818.
10. Seyahi E, Melikoglu M, Akman C, *et al.,* Pulmonary artery involvement in Behçet disease. *Medicine (Baltimore).* 2012;91(1):35–48.

ABBREVIATIONS (ALPHABETICAL ORDER)

1. **BD** – Behçet's disease
2. **ECG** – Electrocardiogram
3. **PE** – Pulmonary embolism
4. **PESI** – Pulmonary Embolism Severity Index