CASE REPORT / OLGU SUNUMU

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Segmental Pulmonary Embolism Secondary to Iliac Vein Thrombosis in a Young Male with Behçet's Disease: A Multimodal Diagnostic Case Report

Behçet Hastalığı Olan Genç Bir Erkekte İliak Ven Trombozuna Sekonder Segmenter Pulmoner Emboli: Multimodal Tanısal Olgu Sunumu

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Abstract

Behçet's disease (BD) is a chronic multisystem vasculitis characterized by recurrent mucocutaneous lesions and the potential to affect major organs. Among its most serious complications is vascular involvement, particularly venous thrombosis. We report the case of a 29-year-old man with established BD and a history of deep vein thrombosis who presented with sudden-onset dyspnea. Imaging revealed complete occlusion of the left iliac vein together with bilateral segmental pulmonary emboli. Transthoracic echocardiography demonstrated mild right ventricular enlargement and an elevated systolic pulmonary artery pressure, consistent with right ventricular strain. This case underscores the significant thromboembolic potential of BD and highlights the importance of comprehensive cardiovascular assessment using multimodal imaging in affected individuals.

Keywords: Behçet's disease, pulmonary embolism, iliac vein thrombosis, echocardiography, vasculitis

Öz

Behçet hastalığı (BH), tekrarlayan mukokutanöz lezyonlarla karakterize, çoklu organ sistemlerini tutabilen kronik ve sistemik bir vaskülittir. Hastalığın en ciddi komplikasyonlarından biri vasküler tutulum olup, özellikle venöz tromboz sık gözlenmektedir. Bu olgu sunumunda, daha önce derin ven trombozu öyküsü bulunan 29 yaşındaki erkek hasta, ani başlangıçlı dispne ile başvurmuştur. Yapılan görüntüleme incelemelerinde sol iliak vende tam obstrüksiyon ile birlikte segmenter pulmoner emboliler saptanmıştır. Transtorasik ekokardiyografi, sağ ventrikülde basınç artışı ile uyumlu gerilme bulguları ortaya koymuştur. Bu olgu, BH'nin tromboembolik komplikasyonlara yatkınlığını vurgulamakta ve bu hasta grubunda kapsamlı kardiyovasküler değerlendirmenin önemine dikkat çekmektedir.

Anahtar Kelimeler: Behçet hastalığı, pulmoner emboli, iliak ven trombozu, ekokardiyografi, vaskülit

INTRODUCTION

Behçet's disease (BD) is a chronic, relapsing inflammatory disorder involving multiple organ systems, typically presenting with recurrent oral and genital ulcers, uveitis, and characteristic skin lesions. A particularly severe manifestation of BD—especially in young male patients—is vascular involvement. Venous thrombosis occurs in up to 40% of patients and is a major contributor to morbidity and mortality (1). Thrombotic events frequently affect large-caliber veins, such as the femoral and iliac veins, and may extend into the inferior vena cava (IVC) (2).

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BD-related thrombosis differs from traditional hypercoagulable states in that it involves intense perivascular inflammation and endothelial injury, thereby causing thrombi to adhere firmly to the vessel wall. This characteristic has traditionally been thought to reduce the risk of embolization. However, pulmonary embolism (PE), though historically considered rare in BD, is increasingly recognized, particularly in patients with extensive or recurrent venous thrombosis (3).

Unexplained dyspnea in BD patients may lead to underdiagnosis of PE, partly due to assumptions of low embolic risk. Transthoracic echocardiography (TTE) is a valuable tool in such settings, assessing right ventricular function, estimating pulmonary pressures, and detecting indirect evidence of PE or chronic thromboembolic pulmonary hypertension (CTEPH) (4).

We present the case of a young male with BD and a history of deep vein thrombosis (DVT), who developed segmental pulmonary emboli secondary to complete occlusion of the iliac vein. This report highlights the substantial thrombotic potential of BD and reinforces the role of multimodal cardiovascular imaging in the evaluation of these patients, even when they appear hemodynamically stable.

CASE PRESENTATION

A 29-year-old man with a four-year history of BD—diagnosed based on recurrent oral and genital ulcerations, a positive pathergy reaction, and episodic uveitis—presented with acute shortness of breath and swelling of the left lower extremity. Two years earlier, he had experienced a DVT in the left leg, which was treated successfully with anticoagulation. He had no known hereditary thrombophilia and no family history of thrombotic disorders.

On physical examination, engorged superficial collateral veins were visible over the left lower abdomen, extending across the midline, suggesting chronic venous outflow obstruction (Figure 1). The left calf was enlarged, firm, and exhibited non-pitting edema with prominent varicosities (Figures 2 and 3), consistent with long-standing venous insufficiency.

Computed tomography (CT) angiography demonstrated complete thrombotic occlusion of the left common iliac and external iliac veins. Additionally, bilateral segmental pulmonary arterial filling defects were noted, consistent with acute pulmonary emboli. TTE revealed mild right ventricular dilation and an estimated systolic pulmonary artery pressure of approximately 40 mmHg (normal <35 mmHg). Figure 4 shows that the IVC was mildly dilated with reduced inspiratory collapse, consistent with elevated central venous pressure. (Normal IVC diameter <21 mm with >50% collapsibility).

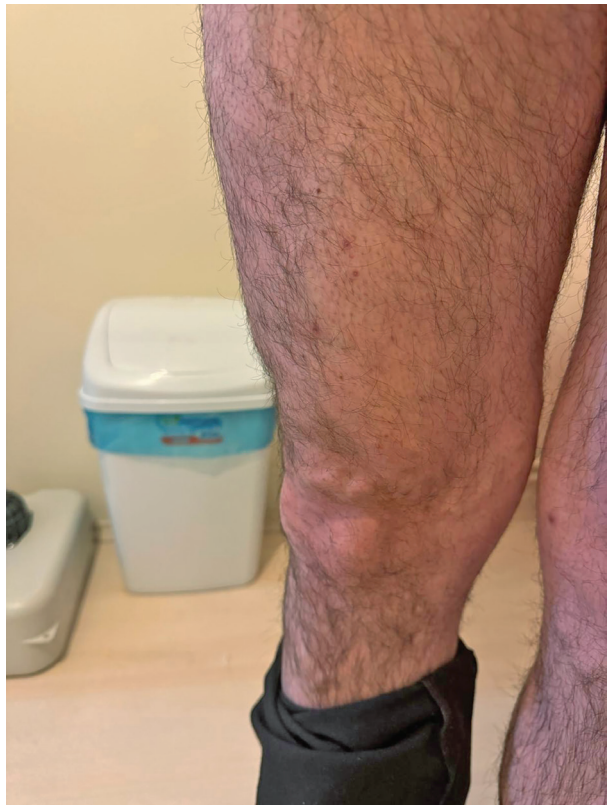

Figure 1. Visible varicose veins of the left lower limb, further supporting chronic venous stasis

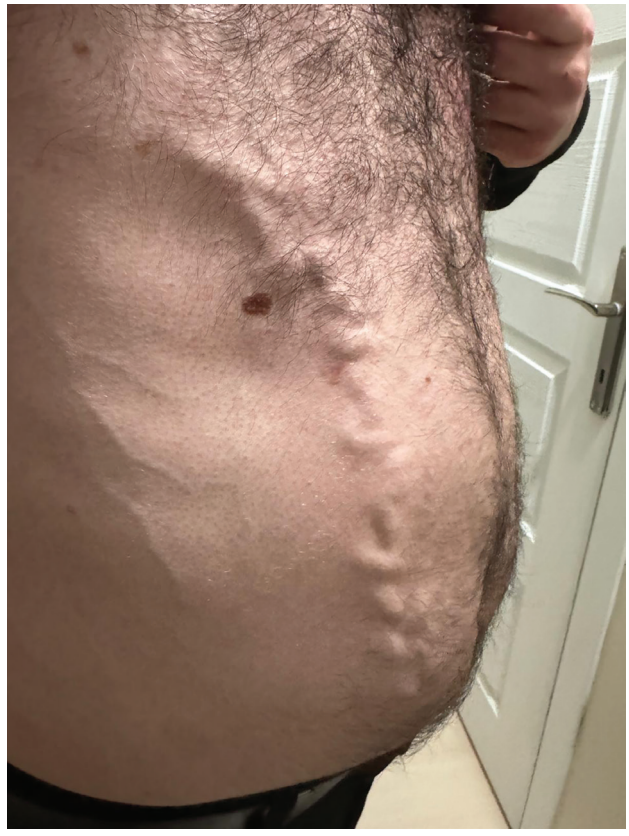

Figure 2. Enlarged and firm left calf with non-pitting edema, suggestive of long-standing venous insufficiency

The main pulmonary artery measured 22.3 mm (normal ≤25 mm) in the parasternal short-axis view (Figure 5).

These findings were consistent with segmental PE originating from extensive iliac vein thrombosis, most likely attributable to the underlying vasculitic pathology of BD. The clinical and imaging findings also raised the possibility of CTEPH. However, after three months of optimized anticoagulation therapy, follow-up echocardiography showed normalization of pulmonary pressures, and repeat CT angiography revealed no residual perfusion defects, effectively excluding CTEPH.

May-Thurner syndrome was initially considered due to the anatomic distribution of the thrombus but was excluded by imaging. A complete thrombophilia panel was negative, further supporting BD as the primary etiology. Informed consent was obtained from the patient.

DISCUSSION

This case illustrates a rare but clinically significant presentation of PE in a patient with BD and extensive iliac vein thrombosis. Vascular involvement is among the most severe manifestations of BD, with venous thrombosis occurring far more frequently than arterial lesions. BD-associated thrombi are characteristically adherent to the vessel wall due to intense neutrophil-rich inflammation and endothelial dysfunction, which theoretically reduce the likelihood of embolization (1,2). However, as demonstrated in this case, embolic events can still occur, particularly in patients with extensive thrombosis.

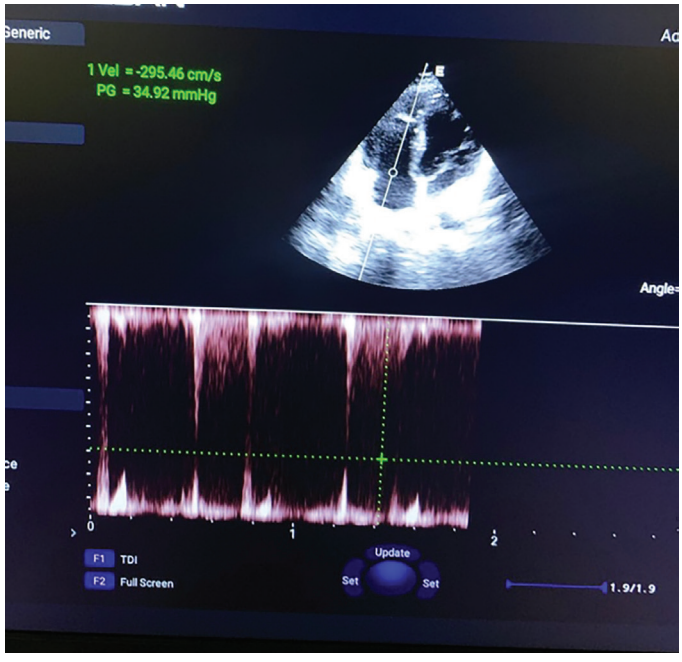

Figure 3. Transthoracic echocardiography (apical four-chamber view) demonstrating mild right ventricular dilation and an estimated systolic pulmonary artery pressure of ~40 mmHg

The prominent abdominal wall collaterals observed in this patient (Figure 1) indicated chronic iliocaval obstruction with possible recent progression. Such findings should prompt evaluation for iliocaval syndromes—especially May-Thurner syndrome—although this was excluded in our patient. Marked asymmetry in the appearance of the lower limbs and prominent varicosities (Figures 2 and 3) further supported chronic venous stasis.

PE has historically been considered uncommon in BD, with reported rates ranging from 2.5% to 5% in patients with vascular involvement (3). In this case, the segmental PE likely resulted from embolization of extensive thrombotic material within the

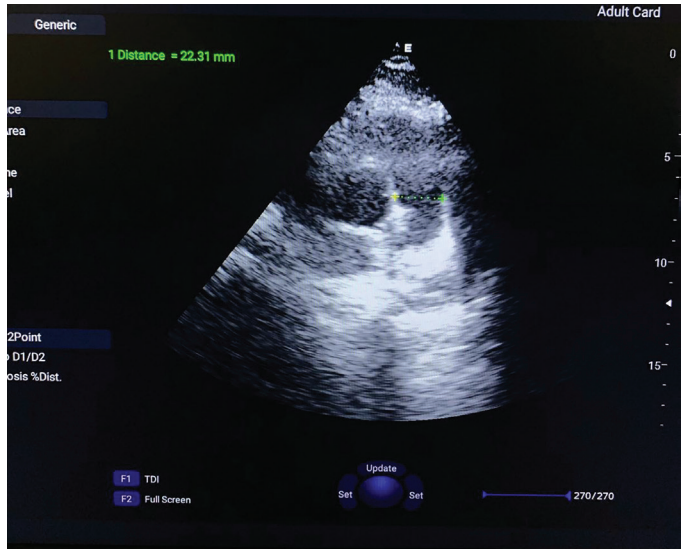

Figure 4. Parasternal short-axis view on transthoracic echocardiography showing a mildly dilated main pulmonary artery (22.3 mm)

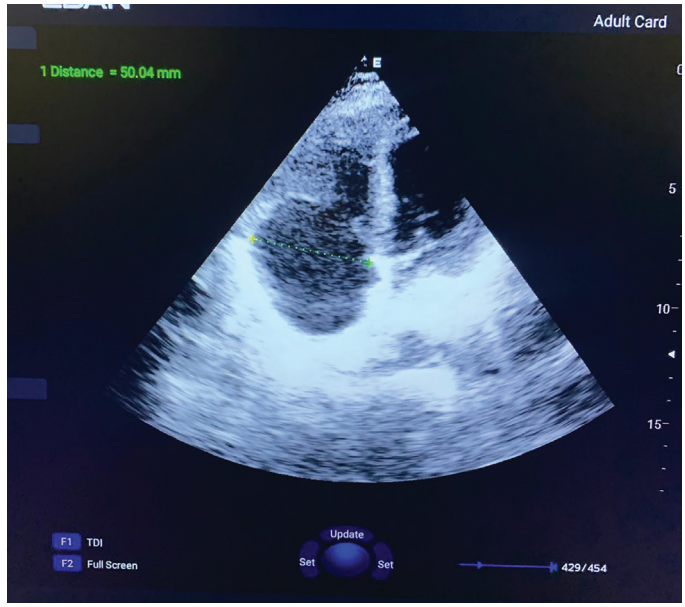

Figure 5. Apical four-chamber view on transthoracic echocardiography showing a dilated right atrium measuring 50.0 mm

iliac veins, consistent with an inflammatory vasculitic form of venous thromboembolism.

TTE was instrumental in identifying mild right ventricular strain and elevated pulmonary pressures, essential components of the non-invasive risk stratification of PE. Current PE guidelines emphasize assessment of right ventricular function and biomarkers to predict adverse outcomes in hemodynamically stable patients (4).

In BD, anticoagulation must be administered cautiously, particularly when pulmonary artery aneurysms are suspected, owing to the risk of fatal rupture (5). No aneurysms were detected in this case. Because BD-related thrombosis is primarily driven by inflammation rather than by hypercoagulability, long-term immunosuppressive therapy plays a central role in preventing recurrence. First-line agents include corticosteroids and azathioprine, while biologics such as tumor necrosis factor-alpha inhibitors or interferon-alpha may be required for refractory cases (6-8). Following rheumatology consultation, this patient was restarted on immunosuppressive therapy in addition to long-term anticoagulation.

This case highlights the diagnostic value of combining physical examination findings (e.g., abdominal wall collaterals), functional assessment (TTE), and cross-sectional imaging (CT angiography) to evaluate both systemic and pulmonary vascular involvement. This also raises an important question: should patients with BD who have known large-vessel thrombosis undergo routine echocardiographic screening for subclinical pulmonary hypertension or silent embolism? Although no formal guidelines currently support this approach, our findings suggest it may be beneficial in selected high-risk patients.

CONCLUSION

In young patients with BD who develop unexplained dyspnea, clinicians should maintain a high index of suspicion for venous thromboembolic events, including PE. TTE is a valuable, non-invasive tool for detecting right ventricular strain and assessing pulmonary pressures, even in hemodynamically stable individuals. The presence of abdominal wall collaterals should prompt further evaluation for chronic venous outflow obstruction. Comprehensive, multimodal imaging enables timely diagnosis and may significantly improve clinical outcomes in patients with BD-related vascular complications.

*Ethic

Informed Consent: Informed consent was obtained from the patient.

Footnotes

Authorship Contributions

Surgical and Medical Practices: S.A., A.S., Concept: S.A., A.S., Design: S.A., A.S., Data Collection or Processing: S.A., A.S., Analysis or Interpretation: S.A., A.S., Literature Search: S.A., A.S., Writing: S.A., A.S.

Conflict of Interest: No conflict of interest was declared by the authors.

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