SAGE Open Medical Case Reports
Volume 12: 1–3

DOI: 10.1177/2050313X241257441
journals.sagepub.com/home/sco
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Behçet's disease detecting by hemoptysis and recurrent epididymo-orchitis: A case report from Syria

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Abstract

Behçet's disease is a chronic systemic inflammatory vasculitis of unknown etiology. It is characterized by recurrent episodes of oral aphthous ulcers, genital ulcers, skin lesions, ocular lesions, and other manifestations. This disease affects many organs and systems, showing a wide range of clinical features. Although pulmonary artery involvement is not common in Behçet's disease, its presence carries a substantial risk of mortality. This report provides a detailed history of a 25-year-old male who was admitted with productive cough, hemoptysis, dyspnea on minimal exertion, fever, and chest pain. He had recurrent orchitis and epididymitis for 7 years, as well as oral and genital ulcers and severe headache. Clinical examination revealed decreased breath sounds at the right middle lung. Thoracic computed tomography angiography confirmed multiple pulmonary artery aneurysms bilaterally. The patient was diagnosed with Behçet's disease, and immunosuppression therapy was initiated. During follow-up, the patient did not report any complications. This case report underscores the significance for clinicians to consider Behçet's disease as a differential diagnosis in patients presenting with hemoptysis and a history of orchitis and epididymitis, given that Behçet's disease rarely causes pulmonary artery aneurysms.

Keywords

Behçet's disease, pulmonary artery aneurysm, hemoptysis, vasculitis, epididymitis, orchitis

Date received: 3 February 2024; accepted: 9 May 2024

Introduction

Behçet's disease (BD) is a rare type of systemic vasculitis that manifests with a diverse range of symptoms. These symptoms include uveitis, oral and genital ulcers, as well as various skin lesions such as erythema nodosum, acneiform lesions, and cutaneous hypersensitivity thrombophlebitis.[1] BD is prevalent among people living along the Silk Road, which encompasses regions such as the Mediterranean, the Middle East, and Asia. Notably, Turkey has the highest prevalence rate among these regions.[2] The condition predominantly affects males, with a male-to-female ratio ranging from 1.5 to 5:1. Typically, BD manifests between the age brackets of 20 and 30.[3] Several published criteria exist for diagnosing BD. However, in our case, we have adopted the new International Criteria for BD, which enables physicians to diagnose the disease early.[4] In this article, we present a special case of BD involving a pulmonary artery aneurysm (PAAs). The patient exhibited symptoms such as productive cough, hemoptysis, fever, and weight loss over a period of 4 months. Additionally, the patient had a history of orchitis and epididymitis spanning 7 years.

Case presentation

A 25-year-old male, an active smoker with a 15-pack-year history, presented with a productive cough, hemoptysis (approximately 150 ml per day), dyspnea on minimal exertion, fever, and chest pain. Two months prior, he had been admitted to an outside hospital with similar symptoms,

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excluding hemoptysis, where he was diagnosed with pneumonia based on bronchoalveolar lavage. He received Ceftriaxone and was discharged after a brief hospital stay. His past medical history included recurrent orchitis and epididymitis over 7 years, treated with Ceftriaxone. Additionally, he reported oral and genital ulcers over the last year, severe headaches, weight loss, and night sweats. Later, the patient presented to the Emergency Department due to hemoptysis and severe chest pain. His vital signs were as follows: body temperature 39°C, respiratory rate 24/min, blood pressure 110/60 mmHg, heart rate 117/min, and oxygen saturation 94% in room air. Clinical examination revealed decreased breath sounds in the right middle lung. Cardiovascular, ophthalmological, and neurological examinations, including head magnetic resonance imaging (MRI), showed normal results. Chest X-ray showed pulmonary nodules in the right middle zone and the left pulmonary hilum (Figure 1). Laboratory results were as follows: hemoglobin 11.5 g/dl, white blood cells 9200/mm^3, platelets 479×10^5/mm^3, hematocrit 34.6%, and C-reactive protein 280 mg/l. Antineutrophil cytoplasmic antibodies (ANCA) testing was negative, with an increase in C3 and IgG. ECG, echocardiography, and urine tests were normal. Computed tomography angiography was consistent with multiple PAAs bilaterally (Figure 2). Pathergy testing and diagnostic investigations for tuberculosis were negative. The patient received various antibiotics and antifungals without improvement. He fulfilled the diagnostic criteria for BD. Daily oral mycophenolic acid, combined with a high dose of corticosteroid therapy (three boluses of methyl-prednisone followed by 0.5 mg/kg/day of prednisolone), was initiated. His condition improved, and he was discharged a week after admission. The patient did not experience any new complications during their last visit to the outpatient clinic.

Discussion

In this case, the patient exhibited recurrent oral ulcers, genital ulcers, and vascular manifestations, fulfilling the required clinical criteria for BD with a total score of 5 points.[4] BD is believed to be a vascular disorder that can manifest as occlusive lesions in veins or arteries and can also present as arterial aneurysms. These lesions are thought to be caused by perivascular or endovascular inflammation.[5] Vascular lesions occur in fewer than 40% of patients with BD. Vascular BD predominantly affects males and remains a significant contributor to mortality. Venous system involvement accounts for approximately 85% of vascular cases, while arterial system involvement is rare, occurring in only 10%–15% of cases. The

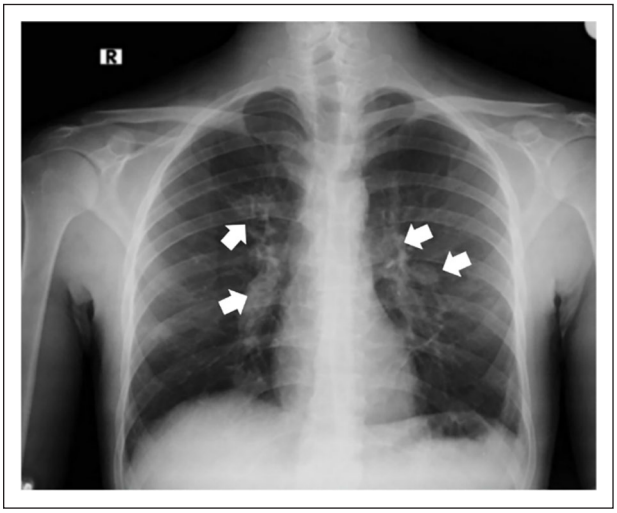

Figure 1. Chest X-ray findings revealed pulmonary nodules in the left pulmonary hilum and the right middle zone.

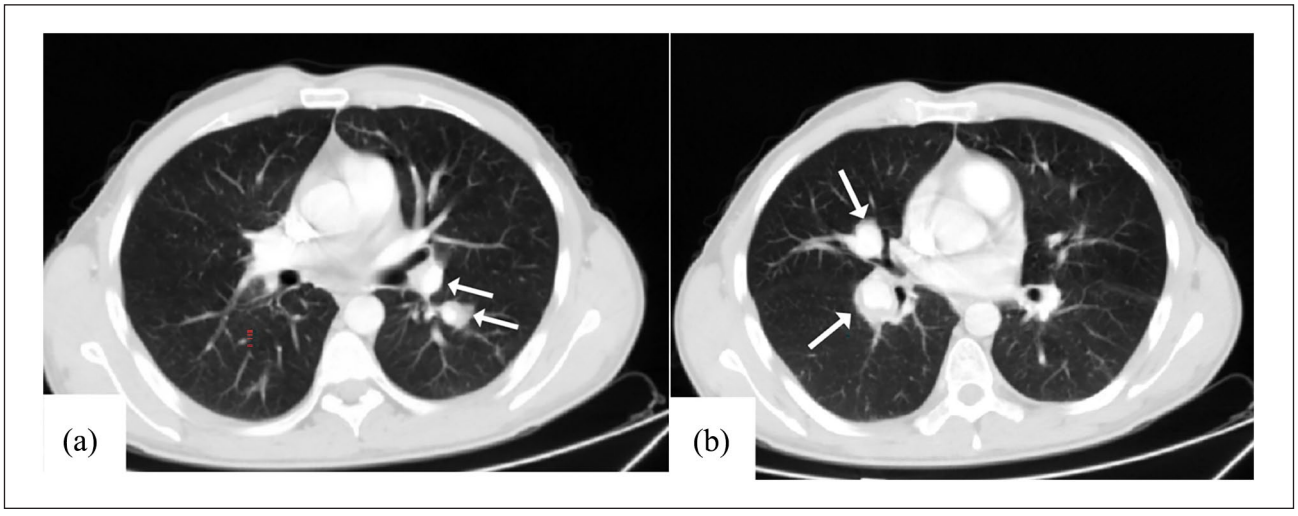

Figure 2. Computed tomography angiography was consistent with multiple pulmonary artery aneurysms (PAAs) bilaterally. (a) Left PAAs in the left pulmonary hilum. (b) Right PAAs in the right middle lobe.

abdominal aorta is the most commonly affected artery, whereas the pulmonary artery and thoracic aorta are less frequently involved.[6,7] A study conducted in China on 923 BD patients revealed a 17.98% rate of vascular involvement, with males being affected at a rate 1.868 times higher than females.[8] PAAs, with a prevalence rate of approximately 1%, are considered a rare complication in Behçet's syndrome.[2] Hemoptysis, the most prevalent symptom of pulmonary BD, is associated with a poorer prognosis. Additionally, individuals may experience other symptoms such as dyspnea, cough, chest pain, and pleural pain.[9,10] What sets our case apart is the manifestation of BD as recurrent epididymo-orchitis. Epididymitis accompanying BD is rare, occurring in only 4%–11% of patients.[11] Vascular BD is effectively managed using conventional immunosuppressants, including azathioprine, cyclosporine A, and cyclophosphamide. These medications, when complemented by high-dose glucocorticoids and monoclonal tumor necrosis factor alpha inhibitors, have demonstrated their usefulness and effectiveness in controlling the condition. However, it is important to note that there is currently no consensus regarding the efficacy of anticoagulants in the treatment of vascular BD.[12] Furthermore, clinicians should be aware that rheumatic diseases, such as scleroderma, may contribute to the development of PAAs. Therefore, these conditions should be considered as part of the differential diagnosis when evaluating patients with vascular BD.[13] Despite these advancements, it is essential to acknowledge the limitations of this report, including the inability to access a comprehensive patient history.

Conclusions

BD, a chronic, multisystem, relapsing inflammatory condition, can manifest with non-specific findings across various organ systems. Clinicians should maintain a high index of suspicion for BD, particularly in patients diagnosed with PAAs, especially when accompanied by orchitis and epididymitis.

Acknowledgements

The authors would like to thank the patient for his great cooperation with us.

Author contributions

A.A.B., M.B., S.K., R.Y., E.A., N.A.S., Z.F., and A.E. took part in writing the manuscript. All authors read and approved the final manuscript.

Availability of data and materials

The laboratory tests and imaging results are available from the corresponding author on reasonable request.

Declaration of conflicting interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

Ethics approval

Our institution does not require ethical approval for reporting individual cases or case series.

Informed consent

Written informed consent was obtained from the patient for his anonymized information to be published in this article.

Consent for publication

We got written informed consent from the patient to publish this article.

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