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Case Report

Dyspnea and hemoptysis revealing Behçet's syndrome: Pulmonary artery aneurysms in a case report ☆

Sara Ez-zaky, MD, Ihssane Laasri, MD, Laila Jroundi, PhD, Fatima Zahra Laamrani, PhD*

Emergency Radiology Department, Ibn Sina University Hospital Center, Mohammed V University, Rabat, Morocco

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ABSTRACT

Behçet's disease is a systemic vasculitis that primarily affects young adults. Arterial involvement in Behçet's disease is rare and associated with a poor prognosis. This manifestation is rarely the initial presentation, often leading to a delayed diagnosis. We present the case of a 27-year-old man who arrived at the emergency department with dyspnea and hemoptysis. An initial chest X-ray revealed bilateral opacities, and subsequent thoracic CT angiography confirmed the presence of pulmonary artery aneurysms. Pulmonary artery involvement, though rare, is a serious condition that manifests as hemoptysis (airway bleeding) caused by pulmonary aneurysms. These aneurysms, often challenging to diagnose, can result in severe hemorrhages and carry a low 2-year survival rate. CT pulmonary angiography is the most effective imaging method for identifying thoracic manifestations of Behçet's disease. Treatment includes early immunosuppression, surgery, or embolization in severe cases, such as those involving massive hemoptysis.

Introduction

Behçet's disease is a chronic multisystemic vasculitis affecting vessels of various sizes in different organs, with thoracic manifestations involving multiple chest structures. Pulmonary artery involvement is a typical and significant contributor to mortality in these patients [1]. Pulmonary artery aneurysms, which can cause life-threatening hemoptysis due to bronchial rupture, require early diagnosis and treatment. Given the risks associated with invasive imaging, CT is recommended for diagnosing vascular involvement in Behçet's disease [2].

This study reviews the imaging findings of pulmonary artery aneurysms in Behçet's disease.

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

* Corresponding author.
E-mail address: sara92ezzaky@gmail.com (S. Ez-zaky).

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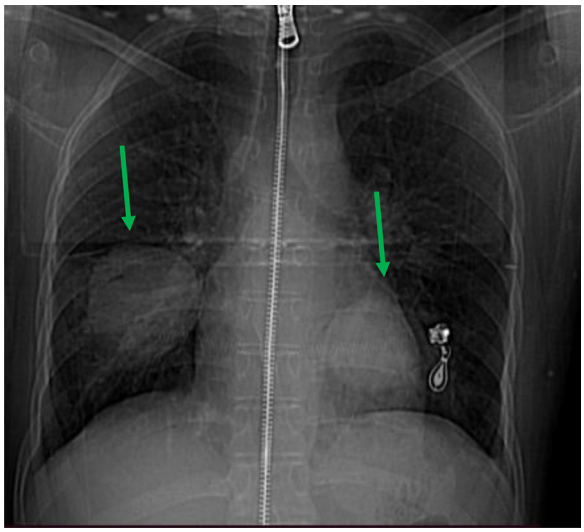

Fig. 1 – Frontal chest X-ray showing 2 well-defined water-density opacities located in the bilateral lower lobes (green arrow).

Case report

We present the case of a 27-year-old male with no notable medical history, who presented to the emergency department with progressively worsening dyspnea and mild hemoptysis.

A chest X-ray (Fig. 1) was performed, revealing 2 bilateral rounded opacities in the lower lobes, with a water-density tone and well-defined borders. A thoracic CT angiography (Figs. 2-3) was then requested, showing 2 bilateral aneurysmal formations in the branches of the pulmonary arteries, rounded, with well-defined contours and partially thrombosed, located in the medial lobe (measuring 62 × 22 mm) and the left posterior-basal segment (measuring 44 × 42 mm).

The diagnosis of pulmonary artery aneurysms was confirmed, and Behçet's syndrome was suspected. Upon further questioning, it was noted that the patient had a history of recurrent oral and genital aphthosis. He was then transferred to the internal medicine department for further management.

A comprehensive clinical examination revealed scars from genital aphthosis, and the pathergy test was positive. The patient was started on cyclophosphamide and prednisone, with ongoing monitoring of the pulmonary artery aneurysms.

Discussion

Behçet's disease is a chronic autoimmune inflammatory condition affecting multiple systems, Behçet's disease is a chronic autoimmune inflammatory condition affecting multiple systems, first described by Hulusi Behçet in 1924. It typically affects the eyes, mouth, and genitals [3,4]. This condition is most commonly observed in countries within the Mediterranean region, the Middle East, and the Far East. It typically affects young adults, with males often experiencing more severe symptoms [5].Vascular complications, also known as "vascular-Behçet's disease," impact 25% of patients and are the leading cause of death among individuals with Behçet's disease [6]. Pulmonary artery involvement is a rare occurrence in Behçet's disease, affecting fewer than 5% of patients [7]. It usually occurs 3-4 years after disease onset [8].

The primary symptom is hemoptysis, which can vary in severity and is typically caused by either the rupture of an aneurysm into a bronchus or thrombosis in the pulmonary vessels. These aneurysms can lead to severe bleeding and have a poor prognosis, with a 2-year survival rate under 50% [6]. In Behçet's disease, hemoptysis might be incorrectly diagnosed as pulmonary thromboembolism (PTE). Misattributing this condition to PTE and administering inappropriate anticoagulation could be life-threatening [9].The pathophysiological process behind pulmonary artery aneurysms (PAAs) involves inflammation of the vasa vasorum in the tunica media. This inflammation damages the elastic fibers in the media, leading to dilation of the arterial lumen [1].

Early detection of aneurysms in Behçet's disease is essential, but invasive imaging techniques like angiography and aortography are rarely used due to their complications. Procedures involving venous puncture or contrast injections can exacerbate thrombosis, and aneurysms may form at arterial puncture sites [2].CT pulmonary angiography (CTPA) is the most effective imaging method for detecting thoracic manifestations of Behçet's disease, surpassing older techniques like conventional angiography, CT angiography and MR angiography [1]. CTPA provides accurate visualization of arterial aneurysms through multiplanar vascular reconstruction. It can identify all major features of Behçet's disease, particularly aneurysms, by showing their relationship with surrounding structures and offering valuable information for surgical planning.Although MRI is less sensitive than CTPA for detecting small pulmonary vascular aneurysms, it has advantages for patients with allergies or renal failure. MRI can enhance blood flow visualization using specific sequences without the need for contrast material injection [10].

Pulmonary artery aneurysms can occur as single or multiple entities, may affect one or both sides of the body, and can be either fusiform or saccular in shape [1]. PAAs are usually found in the right lower lobe pulmonary artery, followed by the right and left main pulmonary arteries. Focal enlargement of the pulmonary artery can be an early radiologic sign of a pulmonary aneurysm [6]. These lesions may be completely or partially blocked by a thrombus. Thrombosed aneurysms can lead to ischemia or infarction in the pulmonary tissue [9].

Pulmonary aneurysms on CTPA are classified into 6 patterns:

1. Enhanced aneurysmal wall.
2. Stable pulmonary artery aneurysms or bronchial artery aneurysms with localized thrombosis.
3. Unstable PAAs with leakage.
4. Stable or unstable pulmonary artery pseudoaneurysms (PAPs) with blurred wall definition, prone to rupture.

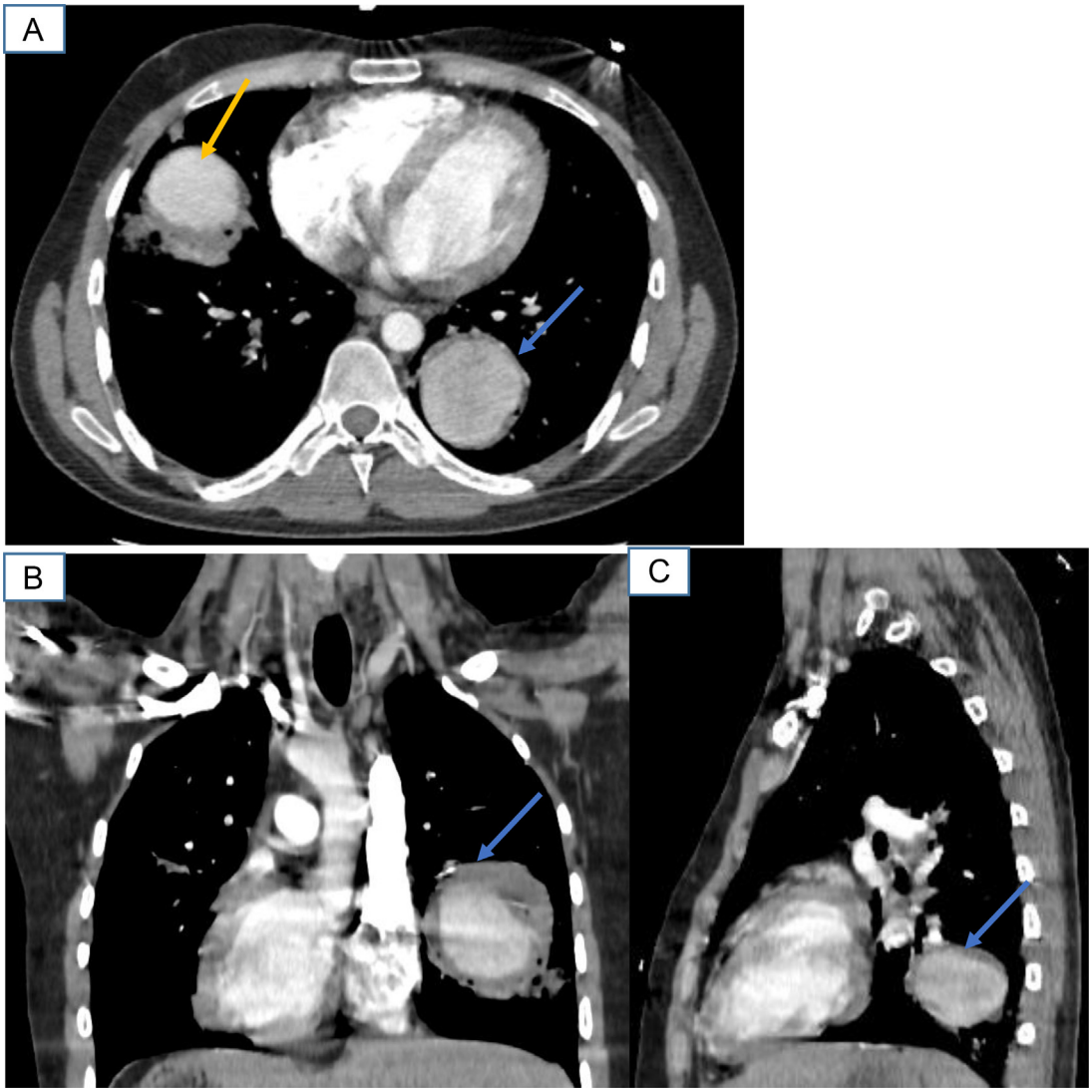

Fig. 2 – Axial (A), coronal (B), and sagittal (C) slices from a thoracic CT angiogram showing 2 aneurysmal formations in the middle lobe (yellow arrow) and the left lower lobe (blue arrow).

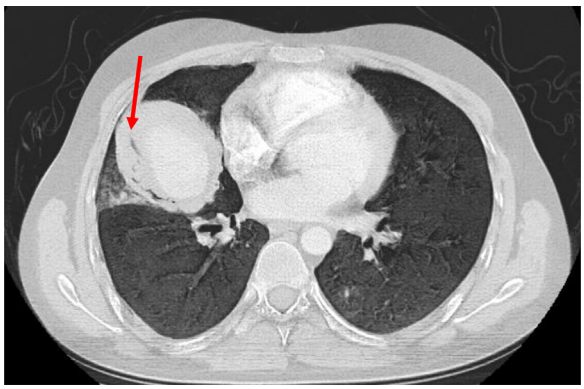

Fig. 3 – Axial slice from a thoracic CT angiogram in lung window showing the bronchial communication and the pulmonary artery aneurysm (red arrow).

5. Right ventricular strain.
6. Right ventricular strain with or without intra-cardiac thrombosis [10].

It is essential to differentiate between in situ thrombosis in true PAAs and marginal extraluminal thrombosis in PAPs, the latter of which suggests a chronic leak and higher rupture risk. CT imaging should also reveal any connections between PAAs and nearby bronchi [10]. Therapeutic options for managing PAAs in Behçet's disease include medical treatment, surgical repair, and transcatheter embolization [6]. Early immunosuppressive therapy is most effective for progressive aneurysm regression, ideally before irreversible damage occurs. Treatment response and duration vary. Medical treatment is preferred for multiple or recurrent PAAs, except in cases of massive hemoptysis, where surgical resection may be necessary [6].

Conclusion

In Behçet disease, vascular system involvement is the main cause of mortality.

Pulmonary artery aneurysm is typical for Behçet disease and is a significant sign of a poor prognosis. CT is a valuable imaging technique in the diagnostic work-up of Behçet disease [11].

Patient consent

Written informed consent was obtained from the patient for the publication of this case report.

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