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Case Report

Beyond the Silk Road: Pulmonary Aneurysms as a Severe Complication of Behçet's Disease

I Hadj Hsain*, Ch Lahlou, Khaoula Boumeriem, J Boutaleb, Ezzaky Sara, S Marrakchi, Fz Laamrani, L Jroundi and O Eloufir

Radiology Department, University Mohammed V Rabat, Morocco

***Corresponding Author:** I Hadj Hsain, Radiology Department, University Mohammed V Rabat, Morocco.

Received: September 23, 2024; **Published:** November 13, 2024

Abstract

Behçet's syndrome (BS) is a multisystem inflammatory disorder of unknown etiology, characterized by recurrent oral ulcers, genital ulcers, and uveitis. This condition, which shows a particular prevalence in countries along the Silk Road, including Turkey, where the incidence is highest, represents a significant challenge in terms of diagnosis and management. The syndrome may also involve various organ systems, leading to a wide range of clinical manifestations. Here, we discuss the case of a patient presenting with multiple pulmonary artery aneurysms (PAAs), a rare but severe complication of BS. The discussion focuses on the diagnostic process, therapeutic options, and the importance of a multidisciplinary approach in managing such complex cases.

Keywords: *Silk Road; Pulmonary Artery Aneurysms (PAAs); Behçet's Disease*

Introduction

Behçet's Syndrome (BS) is most frequently observed in the Mediterranean region, the Middle East, and the Far East. The highest prevalence rate has been reported in Turkey (80-370 per 100,000 inhabitants) [1]. The prevalence varies from 2 to 30 cases per 100,000 inhabitants in other Asian countries, with lower figures in Europe and the United States [2].

Recurrent oral and genital ulcers and uveitis constitute the clinical triad of the disease. According to the diagnostic criteria of the International Study Group for Behçet's Disease [3], the diagnosis is established based on the detection of oral ulcers and at least two of the following criteria: recurrent genital ulcers, eye lesions, notably uveitis and retinal vasculitis, skin lesions (folliculitis, erythema nodosum) and a positive pathergy test (i.e. the formation of pustules 24 to 48 hours after a skin prick).

Pulmonary artery aneurysms (PAAs) are well-known causes of mortality and morbidity in Behçet's Disease (BD) [4,5]. However, pulmonary artery involvement is not limited to PAAs. The other main type of pulmonary artery involvement in patients with Behçet's Disease is pulmonary artery thrombosis (PAT) [4,5]. In this review, we report the case of a patient followed for Behçet's Disease presenting several aneurysms of both pulmonary arteries.

Observation

A 28-year-old patient, with a history of multiple oral ulcer episodes and three genital ulcer episodes over two years, developed hemoptoic sputum for two months followed by moderate hemoptysis and a general deterioration in health one month later. Cardiovascular and pleuropulmonary examinations were normal. Examination revealed an oral ulcer on the inner cheek and a genital ulcer scar on

the scrotum. Chest X-ray showed rounded, well-defined mediastinal and parahilar opacities. A thoracic CT scan revealed two well-defined liquid-density lesions. Angio-CT showed multiple lesions enhancing significantly in the arterial phase with contrast, indicating communication with the trunk and branches of the pulmonary arteries (Figure 1) diagnosed as aneurysms of both pulmonary arteries and their division branches. On the right, there were three aneurysms, measuring 35.8, 15.3, and 6 mm in major axis; on the left, two large aneurysms measured 38.3 and 18 mm, respectively. The diagnosis of Behçet's disease with pulmonary artery aneurysms was confirmed. Echocardiography revealed a moderate pericardial effusion without intracavitary thrombus. Bronchoscopy showed blood spots in places and abundant bleeding of bright red blood from the right paracardiac orifice and the left basal pyramid. Abdominal ultrasound, thrombophilia screening, and ophthalmological examination were normal. The treatment included six boluses of cyclophosphamide (600 mg/m^2 every 15 days) and daily colchicine, combined with hemostatic treatment.

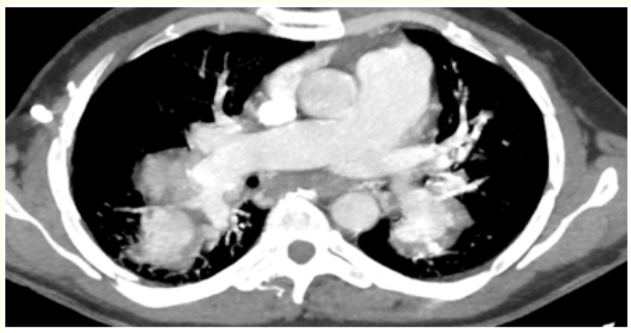

Figure 1: *Axial section of Angio-CT showing multiples lesions enhancing significantly in the arterial phase with contrast, indicating communication with the trunk and branches of the pulmonary arteries.*

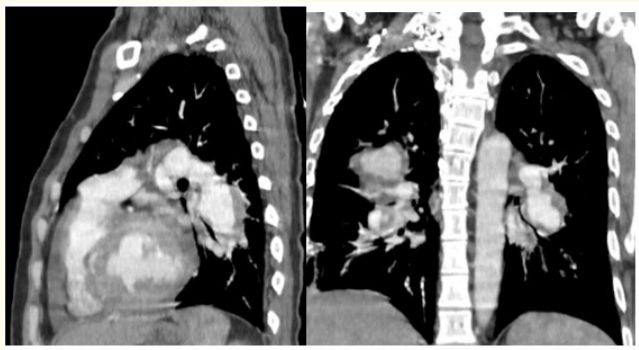

Figure 2: *Coronal and sagittal section of Angio-CT showing multiples lesions enhancing significantly in the arterial phase with contrast, indicating communication with the trunk and branches of the pulmonary arteries.*

Discussion

Behçet's disease is a multisystemic vasculitis. It is a rare chronic multisystem inflammatory condition of unknown cause. A genetic predisposition (human leukocyte antigen [HLA], HLA-B51) and certain microorganisms are thought to play a role in its onset. Affected individuals are typically young, aged 20 to 30 years, and the proportion is predominantly male, as is the case in our observations. The mortality rate associated with this disease can reach 5%. This is mainly due to nerve damage and aneurysm rupture [6].

This systemic vasculitis affects virtually all types and sizes of vessels [7]. The pulmonary manifestations of patients with Behçet's disease are mainly related to vasculitides involving the arteries, veins, and pulmonary septal capillaries [8]. Pulmonary vascular involvement can lead to the formation of an aneurysm (PAA), thrombotic occlusion, pulmonary infarction, and pulmonary hemorrhage. However, thoracic involvement remains quite rare with a prevalence of (1 to 8%). These include thrombosis, particularly of the superior vena cava, pulmonary artery aneurysm, and, rarely, pulmonary infarction or alveolar hemorrhage [9].

Pulmonary artery aneurysms in Behçet's disease are rare, occurring in 15% of cases [10], and can be an obvious manifestation of Behçet's disease, as was the case in our observation. They affect the large pulmonary or lobar trunks, more rarely they are partial and are often bilateral [11]. The pulmonary artery represents the second location after the abdominal aorta for vascular involvement of Behçet's disease, these aneurysms most often occur after 3 to 8 years of the evolution of this disease and are exceptionally a revealing mode of the condition, as in our observation. Overall, they occur in less than 10% of cases and represent the most threatening cause of death in MB [12,13].

Clinically, the warning signs of a PAA are dyspnea, chest pain, or cyanosis. In rare cases, vascular complications such as hemoptysis, sometimes with devastating consequences, can reveal the disease before aphthosis, making the initial vascular symptoms critical [14,15].

The first-choice radiological examination is the standard pulmonary radiography showing unilateral or bilateral hilar masses at the mediastinal level. Pulmonary parenchymal involvement is common and manifests as nodules, large circular opacities, and even transient alveolar infiltrates associated with hemorrhagic foci or infarcts [13]. Spiral CT angiography is the gold standard examination to study pulmonary vascularization thanks to its good resolution and spatial reconstruction, avoiding artifacts from the movement of vascular structures. The acquisition begins without contrast and first shows the dilation of the trunk of the pulmonary artery and one of its branches. Contrast-enhanced spiral images allow for optimal representation of the pulmonary vessels, determine the topography of these aneurysms (central or peripheral), as well as the number, size, density, and fusiform or saccular appearance of the aneurysms. This also allows for the detection of possible thromboses of the dilated pulmonary arteries as intraluminal hypodensities that can be more precisely delineated after injection. Finally, inflammation around the aneurysm is also sought, which is characterized by thickening of the aneurysm wall with blurring of the surrounding adipose tissue, as well as contrast extravasation in case of evident rupture, it also allows for the assessment of parenchymal alterations that may manifest as diffuse reticular nodular opacities associated with capillaritis lesions or, in some cases, may present in association with extensive or triangular infiltrates due to hemorrhage or infarction [16].

The association between PAA and another vascular involvement is quite frequent, a study conducted by Celik., *et al*. on 207 patients, followed for Behçet's disease, in whom pulmonary artery aneurysm was diagnosed showed that 57% of the cases had concurrent extra-pulmonary venous thrombosis, which was the most frequent vascular involvement. In addition, intracardiac thromboses were found in 20 patients, i.e. 10%, and they were more widespread among patients from Morocco. Cerebral vasculitis was reported in 2 patients (1%) from India and Saudi Arabia, in addition, arterial aneurysms at other sites were described in 10 patients (5%). The popliteal artery thrombus was an accompanying lesion in 2 patients (1%) from the United States [17]. The spontaneous development of these aneurysms leads to an increase in their size and rupture into the bronchi with massive hemoptysis leading to sudden death as in the case of our patient, in some cases, a spontaneous recurrence can occur due to intra-aneurysmal thrombosis, The prognosis depends essentially on the evolution of the aneurysms [17].

There is no standard treatment for these aneurysms. Surgical treatment consists of removing the latter. The regression of aneurysms has been achieved under corticosteroids, cyclophosphamide, or thalidomide. Aneurysm embolization is currently the most promising conservative treatment [11].

Conclusion

Arterial involvement, and especially pulmonary artery aneurysms (PAA), remains a formidable complication of Behçet's disease, with a high risk of mortality and an unpredictable prognosis linked to the occurrence of sudden massive hemoptysis. Therefore, the appearance of hemoptysis in a young person, presenting as an initial symptom without an obvious etiology, should alert the physician to search for other pathognomonic signs of this vasculitis, as the appearance of an aneurysm can reveal this disease.

Acknowledgements

The authors are grateful to the patient for granting permission to publish the case.

Conflict of Interest

No conflicts of interest.

Funding Support

The authors did not receive support from any organization for the submitted work.

Ethical Approval

The study was conducted in accordance with the Declaration of Hadj Hsain and its later amendments.

Patient Consent

I declare the following description "Written informed consent was obtained from the patient for the publication of patient data and associated images".

Data Availability

The datasets used and/or analyzed during the present study are available from the corresponding author on reasonable request.

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Volume 7 Issue 12 December 2024