

Pulmonary Artery Aneurysms Complicating Behçet's Disease about a Case

A. El Moutaallik Billah^{1*}, H. El Madkouri¹, Y. Bouktib¹, A. El Hajjami¹, B. Boutakiout¹, M. Ouali Idrissi¹, N. Cherf Idrissi Ganouni¹

¹Radiology Department, AR-RAZI CHU MOHAMMED VI MARRAKECH Hospital, Faculty of Medicine and Pharmacy Marrakech Cadi Ayad University

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*Corresponding author: A. El Moutaallik Billah

Radiology Department, AR-RAZI CHU MOHAMMED VI MARRAKECH Hospital, Faculty of Medicine and Pharmacy Marrakech Cadi Ayad University

Abstract

Case Report

Behçet's disease is a chronic, multisystem and recurrent vasculitis that can affect several organs of the body, but only rarely affects the thorax. Its main arterial involvement is represented by the aneurysm of the pulmonary artery (PA) which represents the most pejorative element for the prognosis of the disease because of cataclysmic hemoptysis that it can generate. Thoracic CT angiography is the test of choice for positive diagnosis. We report the case of a patient with bilateral pulmonary artery aneurysms during Behçet's disease, revealed by chest pain and confirmed by CT angiography.

Keywords: Behçet's disease, Organs, pulmonary artery (PA), Thoracic CT angiography.

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INTRODUCTION

Behçet's disease is an inflammatory, chronic, multisystemic and relapsing disease [1]. It preferentially affects young people, and affects both men and women. Behçet's disease is ubiquitous but more common in patients from the Mediterranean basin, the Middle East and Asia [2].

Angio-behçet mainly affects the venous system and arterial manifestations are rare but serious, and can reveal the disease and most often result in aneurysms of the pulmonary arteries [3].

Thoracic CT angiography remains the modality of choice for positive diagnosis and therapeutic management is essentially based on immunosuppressive treatment. Endovascular and surgical intervention is rarely necessary [4].

PATIENT AND OBSERVATION

We report the case of a 17-year-old male patient, followed for Behçet's disease since 2015. Hospitalized for treatment of a pulmonary embolism. Currently presents with sudden onset of right chest pain without other associated signs. FC: 124 and FR at 30.

Clinical examination finds a conscious patient tachycardic at 124 beats/min and polypneic at 30 cycles/min. A chest CT angiogram showed three images of saccular aneurysmal dilatations at the expense of the arterial branches of the dorsal segment of the LSD and at the level of the two intermediate trunks with compression of the right intermediate bronchus and significant reduction in the lumen of its two dividing branches responsible for a subtotal atelectasis of the ipsilateral lower lobe. The chest CT also showed areas of nodular ground glass and scattered areas in both pulmonary fields, with a central and peripheral distribution, some of which are condensing in the left basal area in relation to areas of alveolar hemorrhage.

The diagnosis of angio-behçet was made in this patient.

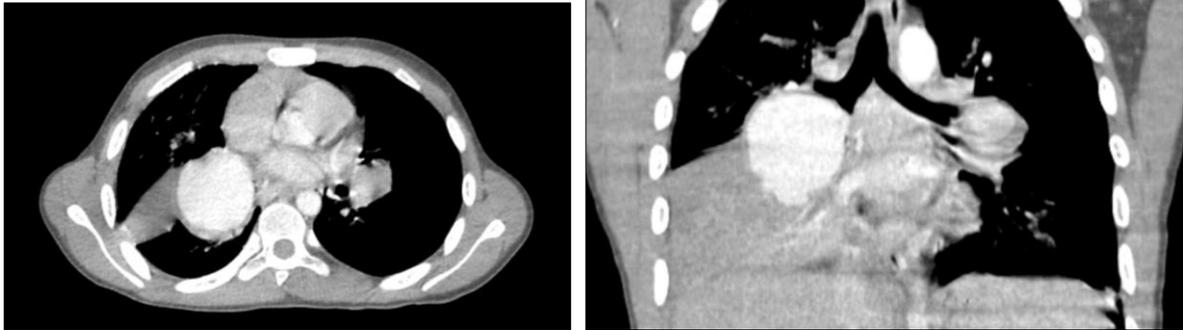


Figure 1: Objective thoracic angioscanning of saccular aneurysms depending on the arterial branches of the dorsal segment of the LSD and at the level of the two intermediate trunks

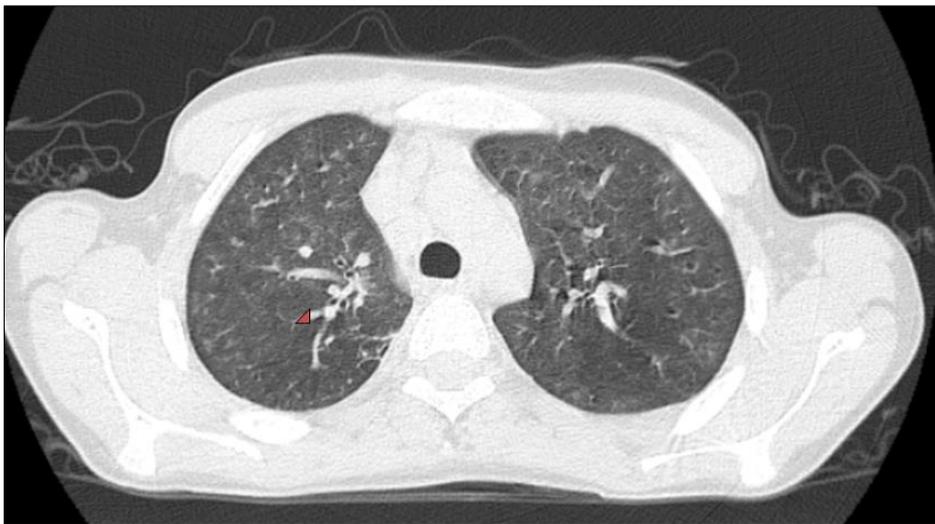


Figure 2: Thoracic CT scan showing on the parenchymal window nodular ground glass foci and scattered areas in the two pulmonary fields

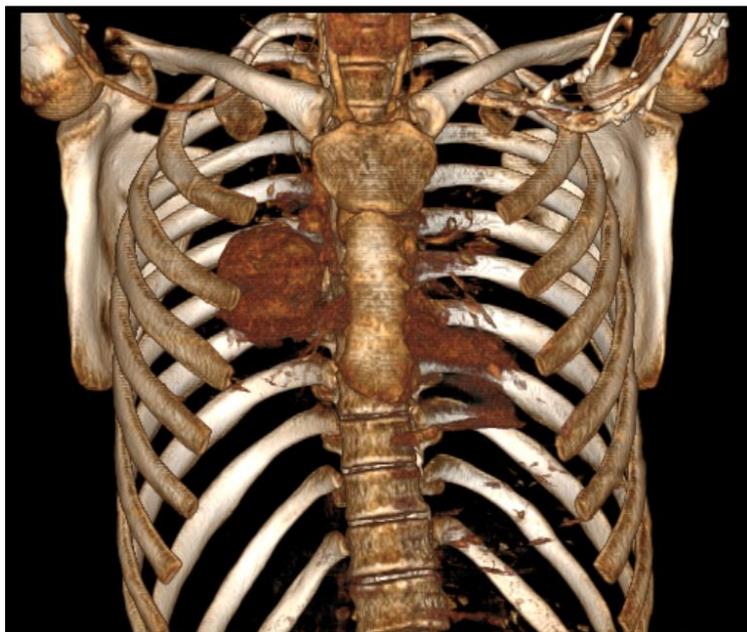


Figure 3: Reconstruction sections showing multiple pulmonary artery aneurysms and their dividing branches

DISCUSSION

Behçet's disease is a multisystem vasculitis. It affects young people, generally 20 to 30 years old with a male predominance. This is the case of our observation. Mortality linked to this disease can reach 5%. It is mainly due to neurological damage and aneurysm rupture [5].

Thoracic involvement is rare (1–8%). It mainly includes thromboses of the superior vena cava, pulmonary arterial aneurysms and rarely pulmonary infarctions and alveolar hemorrhage [6].

Pulmonary artery aneurysms in Behçet's disease are very rare. They constitute a late complication of the disease as is the case in our observation but they can be the revealing manifestation of Behçet's disease. In general, they affect the large pulmonary or lobar arterial trunks, or more rarely segmental. They are often bilateral [5, 6].

Chest x-ray shows rounded hilar or juxta-hilar opacities with clear boundaries. Sometimes, alveolar opacities blur due to hemorrhagic alveolar filling secondary to hemoptysis. CT angiography confirms the vascular nature of the aneurysmal opacities.

The treatment of these aneurysms is not codified. Using cyclophosphamide and high-dose steroids for prolonged periods may also have better results [7]. Aneurysm embolization is a conservative therapeutic modality that currently remains the most promising.

The spontaneous evolution of these aneurysms is towards an increase in their size and the unpredictable occurrence of sudden hemoptysis following an aneurysm rupture, hence the importance of rapid diagnosis and treatment [8].

CONCLUSION

Aneurysms of pulmonary aneurysms are characteristic of Behçet's disease, which must be considered, especially in the presence of associated deep vein and right intracardiac thrombosis. Rapid initiation of immunosuppressive treatment is essential in order to mitigate the risk of rupture which can be fatal to the patient.

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