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Pulmonary Artery Aneurysm (PAA) Revealing Behçet Disease

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Abstract: The arterial illness is rare in Behçet's disease (MB), it constitutes a major evolving turning point and darkens the prognosis. We report a case of Pulmonary Artery Aneurysm (PAA) in the course of Behçet's disease that affects a 51 year old patient who consulted for haemoptysis. The chest X-ray showed a left hilar projection pulmonary opacity. The chest CT angiography showed an aneurysm of the trunk of the left pulmonary artery measuring 65 X 98 mm. Aneurysms of the pulmonary arteries constitute an infrequent illness (1-7%) in the course of Behçet's disease and typically of poor prognosis that we have to consider in the presence of evocative signs of the disease.
Keywords: Aneurysm, Pulmonary Artery, Behçet's disease, CT angiography

INTRODUCTION

Behçet's disease is an unknown vascular wilting affecting vessels of any calibre more often veins than arteries [1]. It is a dreadful affection due to its complications. Indeed, the risk of Behçet's disease is not manifested in an increased mortality except few seldom arterial illnesses but especially a progressive deterioration of the functional prognosis due the cumulative after-effects of neurological and ocular affections. Avowedly, the arterial lesions are unusual, but sporadic publications in this trend continue to come into view. We report a case of aneurysm of the trunk of the left pulmonary artery in a 51 years old patient.

OBSERVATION

Mr. M. D. is 46 years having a personal history of orchitis, treated 20 years ago, with concept of contagion tuberculosis. The history of the disease dates back to 03 years by the appearance of a recurrent but not followed-up bipolar ulceration. Admitted for dyspnea, and hemoptysis of low abundance, evolving for one month. The clinical examination revealed a bipolar ulceration, with no notion of polyarthralgia, nor that of pseudofolliculitis. Ophthalmologic checking-up was normal. The rest of the physical examination was without particularities. In biology, haemoglobin with12 g / dl, lymphopenia with 5OO elements/mm^3, erythrocyte sedimentation rate is 12 mm in the first hour, CRP with1.3 mg / l. Serum electrolytes, liver and lipid tests were normal. Serological tests for hepatitis B and C, TPHA, VDRL and HIV were negative. Just as well, the IDR with tuberculin and distilled water were negative. The chest X-ray displayed opacity of hydric toning of the left hilum at polycyclic contours, occupying the aorto-pulmonary window that can bring up several aetiologies including (Figure 1):

- A cheesy home Tuberculosis (endemic countries and the concept of contagion).
- A hydatid cyst.
- Adenocarcinoma or a single metastasis.
- An aneurysm of the pulmonary artery (within the context).

The thoracic scanner without and after the injection of contrasting product in bolus (Fig.2(a) and (b)) presented an aneurysm of the trunk of the left pulmonary artery measuring 65 X 98 mm without endoluminale thrombosis. The branches of the pulmonary arteries are of normal calibre. The likely lower straight bronchiolar Acinar lesions with discrete pleural thickening. Lung parenchyma of normal appearance,without mediastinal adenopathy, nor pericardial effusion. Echocardiography: indicating dilatation of main pulmonary artery 39 mm without pulmonary hypertension. Not dilated cardiac chambers with good systolic function of the left ventricle. The patient was put under bolus solumedrol during three consecutive and relaying days by oral corticosteroid the bolus ENDOXAN adapted to renal function associated to good clinical and biological improvement with a decline of 06 months.

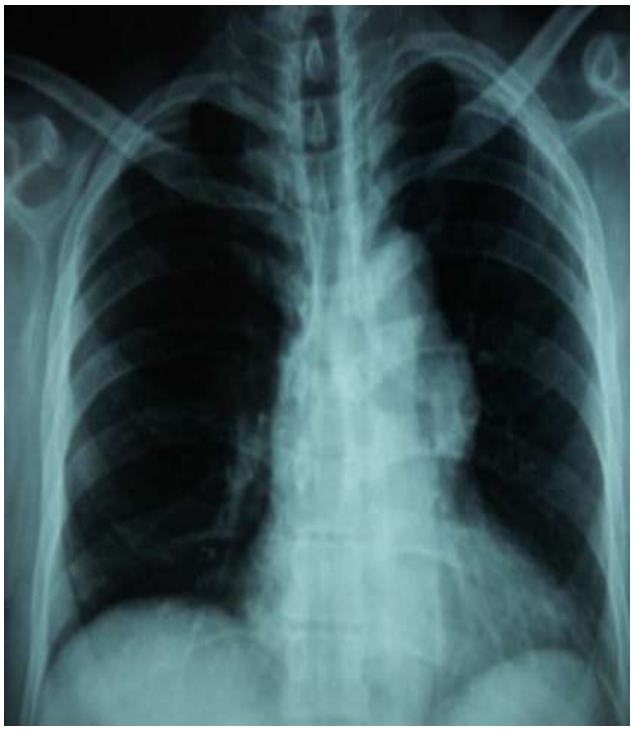
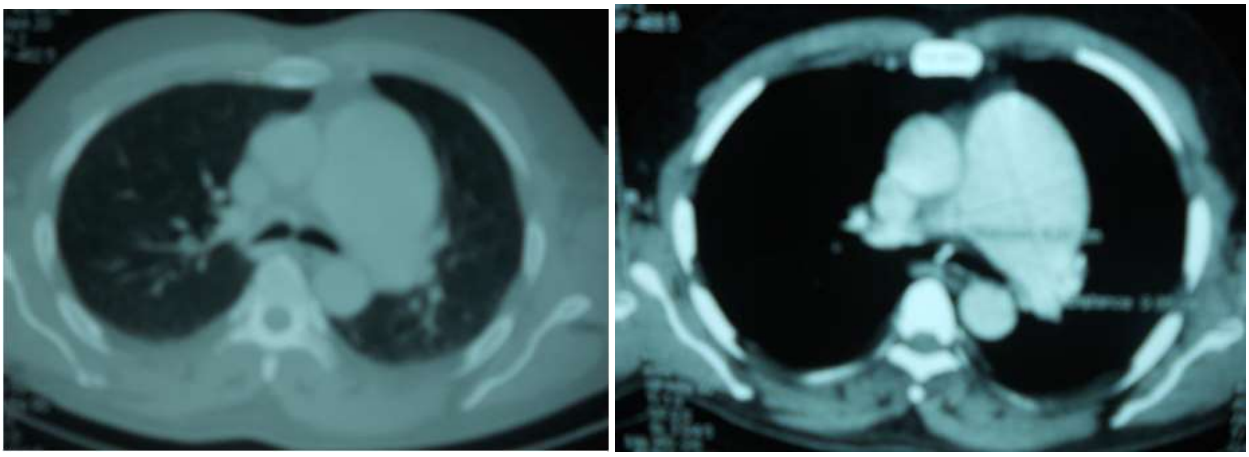


Figure 1: Chest X-ray: opacity of waterborne tone left hilum polycyclic contours.



(a) (b)

Figure 2(a) and (b): aneurysm of the trunk of the left pulmonary artery, measuring 65 X 98 mm.

DISCUSSION

The MB is a disease of young adult with a frequency peak between 20 and 30 years. It is rare after age of 60 years and only a few cases have been reported in children [3]. Age intercedes in the clinical expression and in disease severity. The arterial lesions are unusual from 3 to 5% of cases with male predominance (8H/1F) [2]. Classically, it is later than the venous lesion to which it is often associated [3]. Arterial lesions are located preferentially on large trunks rather than distal arteries with reduced caliber like in our case. It may be about aneurysm thrombosis or real aneurysm "arterial ulcer," often multiple, sitting on the pulmonary vessels, aorta, renal vessels, popliteal and radial with a risk of major rupture. The AAP are rare: 1.1 to 19% of cases of Behçet's disease [4]. The time period of their appearance is 3 to 8 years after the onset of the disease.

In terms of pathophysiology [6], it is about a destruction of the ar terial wall due to deposition of immune complexes with the intima thickening and mucoid degeneration of the media. These lesions are combined with an impairment of the endothelium of distal arteries and capillaries with thrombosis and inflammatory infiltrate non-specific perivascular lymphocyte. AAP were revealed most often by hemoptysis that can be moderate or lightning, as in our patient case . In the course of Behçet's disease, these haemoptysis associated with pulmonary arterial aneurysm have a poor prognosis either immediate or

secondary. The differential diagnosis [8] is mainly made with médiastinohilaires lymphadenopathy, post-traumatic pseudoaneurysm of the pulmonary arteries, pulmonary hypertension, bronchopulmonary cancer, and lung hydatid cyst. The clinical history of the patient affected by Behcet's disease should suspect a PSA. The chest radiograph is of low sensitivity and specificity. It can show a hilar enlargement or unilateral or bilateral para-cardiac opacity especially in large aneurysms [7] as in our observation. But it may remain normal even in the presence of certain segmental pulmonary aneurysms. The fluoroscopy can show the character of pulsation or expansion of the ground or mass. These opacities are often rounded with neat contours, sometimes poly- lobed and, with vascular connections and associated with hypovascularisation of the downstream. Following an episode of haemoptysis, the contour line of these opacities may become blurred, resulting in bleeding of the surrounding parenchyma. The Mediastinal enlargement in a patient with the MB suspected upper cava thrombosis, mediastinal lymphadenopathy or fibrosis rather than PAA. The chest CT [7, 9] confirms the diagnosis by showing saccular or fusiform formations increasing uniformly and simultaneously with the pulmonary arteries. It studies the type, the seat, the shape, the number and the size of the aneurysm, affirms the presence or the absence of aneurismal thrombosis, identifies lung thromboembolism, analyzes the changes of the lung parenchyma. The Magnetic Resonance Imaging can highlight proximal PAA, and estimate the flow and the limits of the aneurysm without vascular opacification. It is the examination choice in thrombosed aneurysms that angiography can not detect. It can be a means of monitoring the PAA. Pulmonary angiography has a diagnostic and therapeutic role which through pulmonary artery embolization, specifies the number, the size, the shape of aneurysms and their relationship with pulmonary arterial branches.

Medical treatment combines corticosteroids, immunosuppressant, and anticoagulants: due to the associated thromboembolic disease. Surgery is pointed out in case of PAA alone or urgent in case of massive haemoptysis [5]. Some teams use the N-butyl - cyanoacrylate at high concentration (75%) for a super-selective embolization if there is a contraindication to surgery or multiple PAA with good results [10]. Mortality was 30% in the two years after the start of bloodletting. This risk appears to be linked to the volume of the aneurysm. A regression and even disappearance of PAA after medical treatment combining corticosteroids and immunosuppressers have been reported, as in the case of our patient.

CONCLUSION

The AAP constitute a serious complication of Behcet's disease that may stake the vital prognosis. Their positive diagnosis is often difficult which justifies a systematic enquiry for medical care, being interventional or surgical. This research will be performed optimally and non-aggressive by chest angioscanner.

Conflict of Interests

Authors have no conflict of interests.

Authors' Contributions

All authors planned and conducted the study procedure and wrote. All authors read and approved the final draft of the manuscript.

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