Case Report,

**A Rare Case of Angio-Bechct Revealed by Hemoptysis of Great Abundance**

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Abstract:
Behçet's disease (BD) is a chronic, relapsing vasculitis of unknown etiology that is characterized by mucocutaneous, ocular, articular, vascular, gastrointestinal, and central nervous system. It mainly affects young subjects, generally 20 to 30 years old with a predominance of men. Thoracic involvement of Behçet's disease is unusual but serious. It is related to the well known vascular tropism of the disease. It may involve the superior vena cava, pulmonary arteries, aorta and subclavian vessels. These vascular attacks can be arterial or venous thrombosis, aneurysms or pseudo-aneurysms. In Behçet's disease, pulmonary artery aneurysms are considered exceptional. These aneurysms manifest themselves by recurrent hemoptysis as is the case with our patient who consulted in pneumology department 20 august 1953 at the university hospital Ibn Rochd Casablanca, for a moderate to great abundance hemoptysis revealing an angiobechet. This is a negative element in patients with BD. It is a diagnostic and therapeutic emergency. The treatment of BD is not well codified but it depends on the severity of the systemic manifestations. Regarding vascular involvement, surgical treatment of an aneurysm with all its risks in patients with Behçet's disease frequently results in aneurysm recurrence. The arterial aneurysms have a poor prognosis, especially pulmonary aneurysms.

Keywords: hemoptysis, thoracic CT angiogram, angio-bechct, Pulmonary artery aneurysms, rupture, treatment.

Introduction:
Bechet's disease (BD) is a systemic vasculitis of unknown etiology which mainly affects young subjects, generally 20 to 30 years old with a predominance of men. BD is ubiquitous but more common in patients from the Mediterranean basin, the Middle East and Asia. It is not a persistent chronic inflammatory disease, but rather a disease characterized by recurrent acute inflammatory flares. The anatomical bedrock of this disease is vasculitis affecting both veins and arteries in multiple organs [1]. Less frequent arterial damage can be life-threatening [2]. Pulmonary artery aneurysms dominate thoracic vascular damage can be fatal by rupture. This article describes a rare case of a young patient who consulted for hemoptysis revealing an angio-bechct.

Patient and observation:
The patient is Mr. A.A, 19 years old, with no toxic habits, no particular pathological history, a notion of painless right monocular redness without recurrent loss of visual acuity. He was admitted to our structure, for a hemoptysis of moderate to great abundance for two months, associated with a purulent bronchial syndrome, without dyspnea or extra-thoracic signs, all progressing in a context of apyrexia and mild deterioration of general condition.

Somatic examination found a conscious patient that was hemodynamically and respiratory stable. Without detectable abnormality except for a few discreet diffuse high-sounding rales. The frontal chest radiograph (Figure 1) revealed two dense homogeneous opacities with regular contours with bilateral hilar larynx (aspect of bilateral hilar lymphoma).

Performing a chest CT scan with a contrast product (Figure 2) showed that these were most likely lesions of vascular origin. As a result, the extensive clinical questioning and clinical examination found the notion of an...
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aphthosis flare-up a hyperpigmented scar of genital ulcers on the inner side of the right thigh. A pathergy test was positive after 48 hours. Slit lamp examination did not find any abnormalities including no uveitis. The diagnosis of Behçet's disease is therefore based on these elements.

A thoracic CT angiogram (Figure 3) revealed the appearance of partially thrombosed bilateral lobar pulmonary aneurysms, likely associated with other distal aneurysms. It’s about a pulmonary aneurysms revealing Behcet’s disease.

On the biological assessment, the patient deglobulized from 12.7 g / dl to 9.4 g / dl with an inflammatory syndrome. After consertation with the internists, this case is in a life-threatening emergency. A pre-therapeutic assessment carried out beforehand. The patient thus received 1g of methylprednisolone as a bolus

Figure 1: The frontal chest radiograph shows 2 dense homogeneous opacities with regular contours with bilateral hilar projection (aspect of bilateral hilar lymphoma).

Figure 2: The chest CT scan with a contrast product shows an aspect of thrombotic aneurysms of two pulmonary arteries, associated to a pulmonary node in upper right lobe with a vascular nature (aneurysms? Arteriovenous malformation?)
per day for 3 consecutive days, then 1 bolus of cyclophosphamide, and no use of anticoagulants. Then switched to oral treatment (30 mg of oral prednisolone per day associated to hygienic diet, also the azathioprine and the colchicines).

The evolution was good, no incidents, disappearance of the bronchial syndrome after antibiotic therapy, cessation of hemoptysis.

**Discussion:**

The diagnosis of Behcet’s disease is mainly based on a set of arguments, while the diagnosis of Angio-Behcet is mainly based on tomodensitometric examinations [3]. It is a rare disease with a very polymorphic expression, requiring multidisciplinary management and prolonged follow-up.

There are several classification criteria for MB. The most widely used international classification criteria for MB are those of the International criteria for the classification of Behcet’s disease revised in 2013 [4]. Behcet’s disease is retained if ≥ 4 points:

<table>
<thead>
<tr>
<th>Symptoms</th>
<th>Points</th>
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<tbody>
<tr>
<td>Mouth ulcers</td>
<td>2</td>
</tr>
<tr>
<td>Genital ulcers</td>
<td>2</td>
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<tr>
<td>Ocular involvement</td>
<td>2</td>
</tr>
<tr>
<td>Skin involvement</td>
<td>1</td>
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<tr>
<td>Vascular involvement</td>
<td>1</td>
</tr>
<tr>
<td>Neurological impairment</td>
<td>1</td>
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<tr>
<td>Pathergy test positive</td>
<td>1</td>
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</tbody>
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Based on the table above, our patient got more than four points due to the following symptoms: namely oral aphthosis, genital aphthosis and a positive pathergic test with vascular disease.

Angio-Behçet is common and affects up to 40% of patients depending on the series [5]. This is a negative element in patients with BD [3]. These vascular attacks can be arterial or venous thrombosis, aneurysms or pseudo-aneurysms, all of which may occur together. In Behçet's disease, pulmonary artery aneurysms are considered exceptional [6]. Clinically, these aneurysms manifest themselves by recurrent hemoptysis as is the case with our patient. Arterial involvement is rare, yet, it can complicate BD in 1 to 7% of cases, or reveal the disease, as our case confirms, and cause life-threatening complications [7,8]. The most commonly affected artery is the aorta, followed by the pulmonary artery [9].

These aneurysms are usually multiple, bilateral and of proximal site involving the trunks and the lobar or segmental bronchi of the pulmonary arteries, and which may in certain circumstances be complicated by sudden hemoptysis involving the trachea and may result in death.

Figure 3: A thoracic CT angiogram revealed the appearance of partially thrombosed bilateral lobar pulmonary aneurysms, likely associated with other distal aneurysms.
the patient's life-threatening condition. The treatment of BD is not well codified but it depends
on the severity of the systemic manifestations. Regarding vascular involvement, surgical
treatment of an aneurysm with all its risks in patients with Behçet's disease frequently results in
aneurysm recurrence [9]. It usually consists of reconstruction using vascular grafts. But it is often
difficult. Neo-aneurysms have been observed at vascular puncture sites [3].
Thrombotic involvement in angio-Behçet is inflammatory in nature. It justifies anti-
flammatory treatment with systemic corticosteroids and possibly an immunosuppressant
(azathioprine, cyclophosphamide) or an immunomodulator (anti-
TNF), as well as the use of an expert center. The prescription of an anticoagulant is controversial, it
is formally contraindicated, but must be discussed in adults in the acute phase, in the absence of
hemorrhagic risk, particularly linked to associated arterial aneurysms [4], which is consistent with
the therapeutic attitude in our case.

Conclusion:-
Angio-Behçet is a vascular disease which most commonly occurs in young men, usually in the
eyears of development of BD. It is a diagnostic and therapeutic emergency. The arterial
aneurysms have a poor prognosis, especially pulmonary aneurysms.

Conflicts of interest
The authors declare no conflicts of interest

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