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(CASE REPORT)

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Thoracic manifestation of Behçet's disease

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Abstract

Behçet's disease is a chronic, multisystemic, and recurrent vasculitis that can affect several organs in the body, but it rarely involves the thorax. The most common pleuropulmonary manifestations are: pulmonary artery aneurysm, arterial thrombosis, aneurysm of the aortic arch and subclavian arteries, superior vena cava obstruction, innominate and subclavian vein occlusion, pulmonary infarction, recurrent pneumonia, organizing pneumonia, and pleuritis. The multiplicity of pulmonary embolisms can lead to pulmonary arterial hypertension. Chest CT with angiographic sequence is currently the preferred examination for exploring thoracic involvement in Behçet's disease. Several CT abnormalities related to parenchymal lung involvement may be observed. The most commonly encountered features are alveolar opacities, round, triangular, or poorly defined opacities, a decrease in lung volume, triangular or linear opacities, nodular or reticular opacities, collapse, labile opacities, or ground-glass opacities. Behçet's disease progresses in flare-ups of varying severity. Mortality is primarily related to vascular involvement. Pulmonary artery aneurysm is the most detrimental factor for the prognosis of the disease due to the catastrophic hemoptysis it can cause. The treatment of thoracic manifestations of Behçet's disease is not well standardized, but corticosteroids and/or immunosuppressants, tailored to the severity of the disease, remain the cornerstone of treatment. We report a case of a patient with bilateral pulmonary embolism revealing Behçet's disease.

Keywords: Behçet's disease; Vasculitis; Chest CT angiography; Corticosteroid therapy; Immunosuppressant; Evolution; Prognosis

1. Introduction

La maladie de Behçet est une maladie inflammatoire, chronique, multisystémique et récidivante. Il s'agit d'un désordre multisystémique caractérisé par une vascularite. Cette pathologie entraîne plusieurs atteintes : cutanéomuqueuses, oculaires, vasculaires, articulaires, gastro-intestinales, urogénitales, pulmonaires et neurologiques. Son diagnostic repose sur plusieurs critères émis en 1990 par l'International Study Group (ISG) [1]. L'atteinte thoracique est fréquente au cours des vascularites, mais rare au cours de la maladie de Behçet. Parmi les atteintes thoraciques les plus fréquentes on trouve : l'anévrisme de l'artère pulmonaire (AP), l'embolie pulmonaire, l'atteinte des petits vaisseaux du poumon et l'atteinte pleurale. Peu de données concernant le traitement et l'évolution de l'atteinte thoracique de la maladie sont fournies par la littérature médicale. Néanmoins, les données existantes suggèrent un mauvais pronostic de cette localisation de la maladie [2].

2. Case presentation

Mr. M.Z., a 31-year-old man with no toxic habits, never treated for tuberculosis, and without recent contact, not known to have chronic bronchitis or chronic dyspnea, consulted for massive hemoptysis (a single episode) associated with atypical chest pain and a history of oral and genital aphthae. The symptoms developed in the context of febrile

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sensations and general malaise. The general examination found a conscious patient, stable hemodynamically and respiratorily. The pleuropulmonary examination showed no particularities. The cutaneous-mucosal examination revealed an oral aphtha, three genital aphthae *(Figure 1)*, and a pseudofolliculitis on the back *(Figure 2)*. The pathergy test was negative.

The infectious workup was positive with leukocytosis of 13,550, predominantly neutrophilic (10,180), and a CRP of 160. The search for Mycobacterium tuberculosis (BK) and the x-Pert gene in the sputum was negative, as was the aspergillus serology. Serologies for hepatitis B and C, syphilis, and HIV were all negative. The thrombophilia workup was negative. les sérologies hépatite B et C, syphilis et VIH étaient négatives. Le bilan de thrombophilie était négatif.

The chest X-ray revealed a heterogeneous alveolar opacity in the middle third of the right lung field with flattening of the right diaphragm dome *(Figure 3)*. The chest CT angiography showed triangular condensation foci with peripheral bases, likely corresponding to pulmonary infarction foci *(Figure 4)*, associated with bilateral pulmonary embolism *(Figure 5)*. A bronchoscopy revealed a diffuse first-degree inflammatory state with the presence of whitish secretions, fine spurs, and free orifices without any buds or granulomas. A cytodiagnostic study showed reactive inflammatory cytology with no malignant cells. Aspirations for the search of Mycobacterium tuberculosis (BK) and x-Pert gene, as well as Aspergillus, were negative. A cytobacteriological study of the aspirated fluid was sterile.

As part of the workup for underlying conditions, a cardiac evaluation was normal, the ophthalmological examination showed no abnormalities, and a 24-hour proteinuria test was normal. The international classification criteria for Behçet's disease -2013 was 6 (diagnosis is confirmed if >4).

The patient was started on antibiotic therapy with amoxicillin-clavulanic acid for 7 days, followed by a 1g bolus of Solu-Medrol for 3 days, then prednisone at 1mg/kg/day for 3 weeks with gradual tapering. Adjuvant treatment included cyclophosphamide (Endoxan IV bolus, 1g monthly), therapeutic dose anticoagulation, and colchicine 1g/day for his cutaneous-mucosal involvement. The patient showed good clinical and radiological progress.

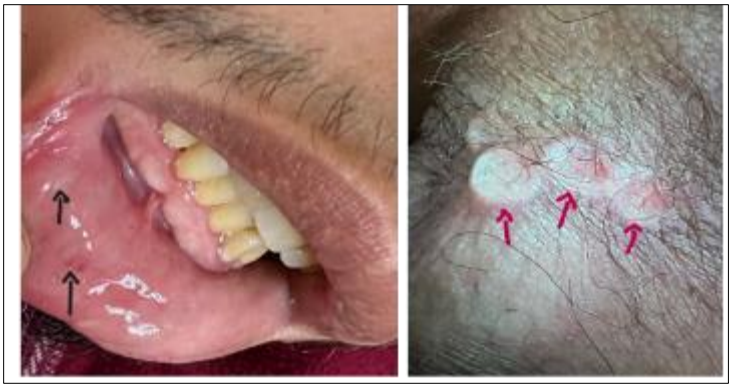

Figure 1 Oral and genital aphthae

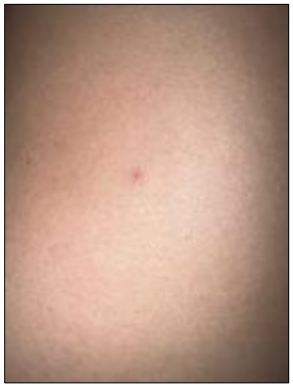

Figure 2 Pseudofolliculitis on the back

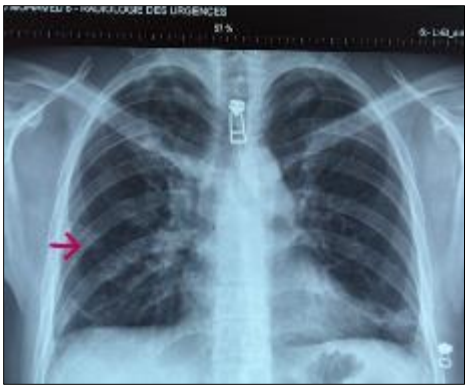

Figure 3 Heterogeneous alveolar opacity in the middle third of the right lung field with flattening of the right diaphragm dome

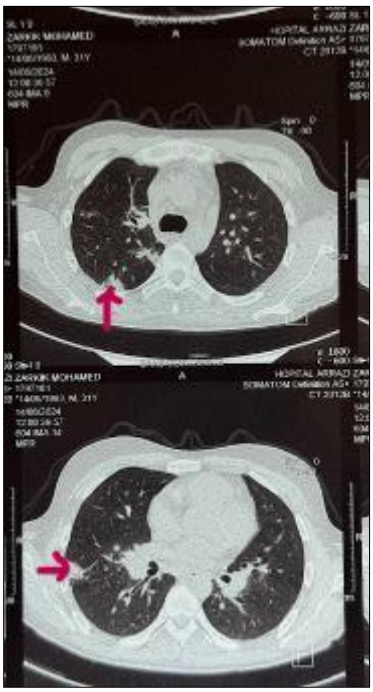

Figure 4 Chest CT angiography with parenchymal window showing triangular condensation foci with peripheral bases, likely related to pulmonary infarction foci

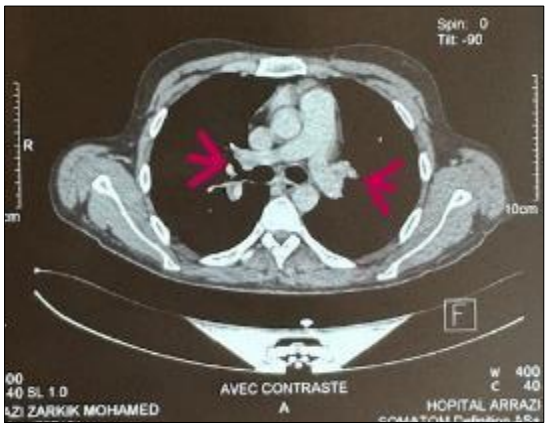

Figure 5 Chest CT angiography with mediastinal window showing bilateral pulmonary embolism

3. Discussion

Behçet's disease (BD) is a systemic vasculitis of unknown origin, typically associated with a high incidence of HLA-B51. It is, however, known that this condition predominates along the Silk Road, the ancient route extending between the Mediterranean, the Middle East, and the Far East [3].

The average age of onset of the disease is most often between 20 and 30 years old [4-5]. The average age of patients with Behçet's disease (BD) and pulmonary manifestations ranges from 17 to 50 years, with an average of 31.8 years [6]. Men are affected two to five times more than women. However, this ratio is evolving, and increasingly [7], women are affected as frequently as men [5].

Behçet's disease is characterized by a variety of clinical manifestations, including oral ulcers, skin lesions, and ocular involvement. Among these manifestations, thoracic complications, although less common, can have significant clinical implications. Pulmonary manifestations of Behçet's disease primarily include pulmonary embolisms and parenchymal involvement. Patients may develop pulmonary nodules, infiltrates, or pulmonary hemorrhages, often related to vascular inflammation [8].

The presence of these abnormalities underscores the need for a thorough clinical and radiological evaluation in patients presenting with respiratory symptoms. The pathophysiology of thoracic manifestations in Behçet's disease appears to be linked to the inflammation of blood vessels, known as vasculitis. Endothelial dysfunction and the activation of immune cells play a central role in the development of thoracic lesions [9]. Immunological mechanisms, including the production of pro-inflammatory cytokines, contribute to thrombus formation and pulmonary involvement.

The diagnosis of thoracic manifestations is often complex, requiring advanced imaging techniques. Studies like that of Duzgun et al. (2018) highlight the importance of chest computed tomography (CT) to detect pulmonary abnormalities associated with Behçet's disease [10]. Symptoms can be nonspecific, making early diagnosis challenging.

The treatment of thoracic manifestations of Behçet's disease relies on the use of immunosuppressive medications and anticoagulants. Corticosteroids are often effective in controlling inflammation, although an individualized approach is necessary depending on the severity of the manifestations [11]. Additionally, biological agents such as TNF-alpha inhibitors have shown promising results in managing more severe forms of the disease.

4. Conclusion

Thoracic manifestations of Behçet's disease, although less common, can lead to significant complications. The disease progresses in flare-ups. The treatment of thoracic involvement in Behçet's disease is not yet well standardized but relies on corticosteroids and immunosuppressants. Early identification and appropriate management are essential to improve the prognosis of patients.

Compliance with ethical standards

Disclosure of conflict of interest

The authors declare no conflicts of interest.

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