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Case Report

Pulmonary Embolism in a Patient with Behçet's Disease: A Case Report

M. Abdelbaki*, N. Khellafi, D. Benmessaoud

Laghouat Mixed Hospital, Cardiology Department; Faculty of Medicine, Amar Tellidji University, Algeria.

*Corresponding Author: Mourad Abdelbaki; *m.abdelbaki@lagh-univ.dz*

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Abstract/Introduction

Behçet syndrome is a systemic vasculitis of unknown etiology, frequently characterized by recurrent oral and genital ulcerations and uveitis. Patients with Behçet syndrome can have both arterial and venous involvement.

We report the observation of a case of a 29-year-old man who was admitted with clinical presentation of pulmonary embolism, for whom investigations for the etiology concluded in the diagnosis of Behçet's disease.

Short Summary of the Case

We present the case of a 29-year-old man who came to the emergency department with acute chest pain, dyspnea, and hemoptysis for 2 weeks. The ECG did not show any abnormalities. The biology found a normal troponin level, a disturbed inflammatory profile with an elevated D dimer level.

An urgent echocardiography performed showed a floating mass in the right atrium (cyst? thrombus? tumor?)

Faced with this picture of acute dyspnea with a high dimer of D, we decided to perform a pulmonary CT angiogram which showed multiple perfusion defects without the presence of pulmonary aneurysms.

Urgent treatment with low molecular weight heparins followed by antivit K drugs was started.

A rheumatology opinion was requested and the thrombophilia evaluation was negative, and the diagnosis of Behçet's disease was confirmed in our patient.

Discussion

Behçet syndrome (BS) is a chronic systemic form of recurrent vasculitis.

Vasculitis involves multiple organs, including the skin, joints, eyes, mucous membranes, veins, arteries, nervous and gastrointestinal systems, etc.

The main histological feature is a unique vasculitis that is not restricted to a specific vessel size [1].

BS is usually diagnosed between the third and fourth decades of life, being slightly more common in men than in women, and has more severe features in young adult.

The classic clinical presentation of BS is painful oral aphthous ulcers, genital ulcers, and ocular involvement, symptoms which together are called the three-symptom complex.

The International Study Group FOR Behcet's Disease (ISG) developed the simplified ISG criteria analysing the specificity and sensitivity of the five diagnostic criteria; Recurrent oral ulcers were classified as the main criterion, recurrent genital ulcers, typical ocular lesions, typical skin lesions, and positive skin pathergy test as the minor criteria.

Behcet's disease can be diagnosed in the presence of a major and two minor criteria according to these criteria [2,3].

The International Criteria for Behcet Disease (ICBD) are the latest diagnosis/classification criteria, created by the participation of 27 countries from different parts of the world [4].

Vascular Behçet syndrome (VBS) is unique because it affects both the venous and arterial systems. DVT is the most common venous thrombosis in patients with BS. Although the incidence of DVT in Behçet syndrome is high, pulmonary artery thrombosis (PAT) is considered a rare complication. Several cases of patients with Behçet syndrome have been reported who experienced pulmonary thrombosis during the pathogenesis of BS.

The pulmonary arteries are the second most common site of arterial involvement in Behcet's disease [5]. Pulmonary vascular involvement such as PAA, in situ PAT, pulmonary hemorrhage, pulmonary infarction, PTE, arteriovenous shunt in the lung and aneurysmal fistula are the most common pulmonary disorders in Behcet's disease [5-8]. Thirty-three cases of PTE have been reported in Behcet's disease (Table 1) [6,8].

Table 1. Reports of pulmonary thromboembolism in patients with Behcet's disease [8]

Study	Year	PTE Cases
Behcet's disease complicated by thrombosis: A report of 93 Chinese cases	2014	15.1% (14/93 patients developed thrombosis) of the 766 patients with BD

Pulmonary embolism and Behcet's disease	2006	Among 153 patients with Behcet's disease according to the criteria of the International Study Group for Behcet's disease, seven (5 men and 2 women mean age at 26.6 +/- 6 years) were diagnosed as having pulmonary embolism. This was the first in 3 cases; for 2 women, pulmonary embolism complicated pregnancy
Thrombolytic therapy in pulmonary embolism of Behcet's disease	1996	A patient with Behcet's disease who presented vein thrombosis and pulmonary embolism

The etiologies of BD complicated with pulmonary embolism include deep vein thrombosis, in situ thrombosis, or intracardiac thrombi [9-11]. In our patient echocardiography showed a floating thrombus in the right atrium and the pulmonary CT angiogram showed multiple perfusion defects without the presence of pulmonary aneurysms.

Conclusion

Vascular involvement in Behçet syndrome is a major contributor to the morbidity and mortality of these patients. Early detection of vascular involvement has a major impact on the prognosis of patients with Behçet syndrome. The mainstay treatment for pulmonary artery thrombosis is immunosuppression and anticoagulation.

Keep in mind that Behcet's disease is one of the reasons to consider in young patients with PE who are not considered to have any risk factors for PE [3].

Conflict of Interest

There was no conflict of interest.

Data Availability

Data would be available upon reasonable request

List of Abbreviations

BS: Behçet syndrome
DVT: Distal venous thrombosis
ECG: Electrocardiogram
ISG: International study group
ICBD: International criteria for Behçet disease
PAT: Pulmonary artery thrombosis
VBS: Vascular Behçet syndrome

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