

# Pulmonary Thromboembolism and Intracardiac Thrombus as a Presentation of Behcet's Disease

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## Abstract

Behçet's disease is a multi systemic vasculitis whose presentation does not always meet the typical triad of orogenital ulceration, skin and ocular lesions. Right ventricular thrombus, thromboembolism and pulmonary artery aneurysms are rare manifestations and associated with poor prognosis. The authors describe the clinical case of a 38 year old man with multiple admissions for respiratory infection and erythema nodosum. A chest angiotomography revealed pulmonary thromboembolism and right ventricular thrombus, without other changes in laboratory tests. The patient did not take the prescribed oral anticoagulation and abandoned follow up, was admitted two years later with a similar clinic and recurrent oral ulceration. New thromboembolic and angiography showed aneurysmal dilatation of the pulmonary arteries. Behçet's disease was diagnosed. Corticosteroid therapy was started with a good response after one month.

**Keywords:** Behçet; Pulmonary thromboembolism; Intracardiac thrombus; Pulmonary artery aneurysms

## Introduction

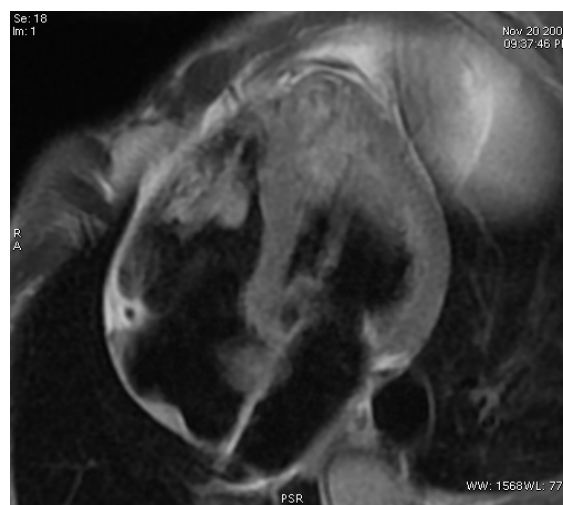
Behçet's disease (BD) is a chronic, multi systemic vasculitis of unknown etiology that primarily affects men, in the third and fourth decades of life. It is more prevalent in the middle and far east and the Mediterranean bay, where a high prevalence of 80-370 cases per 100,000 habitants has been described [1]. It characteristically presents with recurrent orogenital ulceration, skin and ocular lesions, but may also be associated with neurological, gastrointestinal, joint and cardiovascular involvement. The vascular involvement is common (5-30%), particularly venous involvement [2-4]. Aneurysmal formations are more frequent than arterial occlusion, and are one of the most serious complications, due to the risk of rupture. The abdominal aorta is the most common site of aneurysms (60% of cases), followed by the pulmonary artery [2,3]. Pulmonary involvement is rare, occurring in 1 to 7.7% of cases, and includes, among others, pulmonary artery aneurysms, arterial and venous thrombosis, pulmonary infarction and recurrent pneumonia [1,3,5]. Pulmonary artery aneurysms are the most common pulmonary manifestations, and Behçet's disease is the most common cause of those [3,6-8]. Pulmonary arterial thromboembolism is a very rare manifestation, almost always associated with vasculitic phenomena in pulmonary artery and with right intra cavitory thrombus, and with no relation to venous thrombosis, despite its high prevalence [4]. Cardiac manifestations are also rare, about 1-5%, and are associated with poor prognosis [3,4]. Intra cavitory thrombus is read in the literature only 50 cases [3,4,9].

## Clinical Case

We present the case of a 38 year old Caucasian male who was a smoker but otherwise previously healthy. In 2008 he had three hospital admissions for lower respiratory tract infections with chestpain, dyspnoea, fever, cough with occasional haemoptysis and erythemanodosum. At the last admission, chest radiograph showed peripheralhy po transparent lesions, with triangular shape, which raised the suspicion of pulmonary embolism, confirmed by chest CT angiography. This exam also detected a mass with 30 × 22 mm, adherent to the free wall of the right ventricle, compatible with intracavitory thrombus, later confirmed by cardiac magnetic resonance imaging (Figure 1). The doppler ultrasound of the lower limbs showed no changes and serological screening for autoimmune diseases or thrombophilic

changes was negative. Oral anticoagulation was prescribed, but the patient failed to attend follow-up appointments.

In 2010 he was admitted at the emergency department, with complaints suggestive of respiratory infection, erythemanodosum and exuberantand recurrent oral ulceration, during the last year. He was not taking any therapy. He was admitted as an inpatient at the for further reevaluation. Diagnostic investigations revealed: sedimentation rate of 93 mm at the 1<sup>st</sup> hour; proteinuria (<1 g/24 h) without changes in



**Figure 1:** Intracavitory thrombus in cardiac magnetic resonance.

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**Received** November 05, 2013; **Accepted** December 09, 2013; **Published** December 11, 2013

**Citation:** Neves J, Andrade L, Saraiva A (2013) Pulmonary Thromboembolism and Intracardiac Thrombus as a Presentation of Behçet's Disease. J Pulm Respir Med 3: 162. doi:[10.4172/2161-105X.1000162](https://doi.org/10.4172/2161-105X.1000162)

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**Figure 2:** Aneurysmatic dilatations of pulmonary artery in a thorax computed tomography.

renal function; antiphospholipid antibodies, antinuclear antibodies and Anti-neutrophil cytoplasmic antibodies were negative; thrombophilia investigation was negative; hyper homocysteinemia of 18 mmol/l (for a normal 5-15 mmol/l); presence of the HLA - B51 allele was identified. Echocardiography revealed a decrease of thrombus dimension (22 × 10 mm) and chest computed tomography demonstrated thromboembolic pulmonary arteries with dilatations in the apical segment of the right lower lobe. Doppler (Figure 2) ultrasound of the lower limbs showed recanalization phenomena of previous thrombosis. The pathergy test was positive and there were superficial phlebitis of the upper arm secondary to vene puncture. There were thus obtained the diagnostic criteria according to the BD "International Criteria for Behçet's Disease" (ICBD) [10,11].

The patient started corticosteroid therapy with prednisolone at a dose of 1 mg/kg body weight, with significant clinical improvement after one month of treatment, but was again lost to follow-up due to socio-economic reasons.

## Discussion

Although venous thrombosis is a frequent manifestation of BD (10-30%), pulmonary thromboembolism is a rare feature [2]. The low incidence of pulmonary thromboembolism in this pathology, despite the prothrombotic state associated with it, is due to the fact that the thrombus ordinarily adherent to the sides of the vasculature [4]. In this case, the presence of thrombus and the intracavitary pulmonary thrombosis *in situ*, associated with pulmonary artery aneurysms are possible sources of thromboembolism [2,3]. Apost-mortem study of 23,796 patients concluded that of the 354 patients who had pulmonary embolism and right intracardiac thrombus, the thrombus was the only source of embolization in 62.1% [12]. In 2000, Mogulkoç et al. published a review of 25 patients with BD with intracardiac thrombus. Among these, pulmonary thromboembolism was observed in 13 patients (52%) [9]. The exact pathophysiological mechanism of the prothrombotic tendency of BD remains unknown, but it is assumed that some thrombophilic factors may contribute, such as hyperhomocysteinemia, present in this patient (homocysteine 18 mmol/L). In a study in patients with BD higher values of homocysteine were observed in the group

with thrombotic events, suggesting that hyperhomocysteinemia may be an independent risk factor for thrombosis [13,14].

Intracardiac thrombosis is an extremely rare event and may precede in years other disease manifestations, including oral ulcers, and should therefore always raise the suspicion of BD in the absence of other characteristics [2,9,15]. The mechanism of formation is still not well understood and may be due to a *in situ* process, secondary to venous thrombosis, to which it is often associated, or even to endomyocardial fibrosis [2]. It is more common in males and the right ventricle is the most common location (78% of cases) [3,15]. The thrombus is usually adherent to the myocardium and its presence is strongly associated with pulmonary artery aneurysms. Several cases in the literature describe the existence of intracardiac thrombosis and pulmonary artery aneurysms in BD, or not associated with pulmonary thromboembolism [15-20]. In 33% of patients, pulmonary artery aneurysms are associated with a right ventricular thrombus [8]. Pulmonary artery aneurysms are typically located in the right lower lobe artery, with arterial thrombosis and associated to inflammatory phenomena [6-8]. Hemoptysis is the most common manifestation, present in 79% of cases. Cough, fever, dyspnea, pleuritic chest pain are also common features and can be interpreted as an infection [8]. In this case, is questionable if the symptoms are due to thromboembolic phenomena or to an early manifestation of the existence of pulmonary artery aneurysms, not observed in the first angiography. Fever (52%), hemoptysis (48%), dyspnea (44%) and cough (20%) can also be the symptoms of intracardiac thrombus [9]. Recommended treatment in severe forms of vasculitis, as presented, is the combination of corticosteroids and immunosuppressive drugs, being cyclophosphamide the first-line therapy for patients with BD and vascular involvement [1,2,9]. The presence of thromboembolic and cardiac thrombus initially imposed hypocoagulation; however, in face of the subsequent diagnosis of aneurysms of the pulmonary artery (which confers a great morbidity and mortality due to the risk of rupture and fistulization when associated with an increased of pulmonary artery pressure), anticoagulants were not restarted because of the increased bleeding risk [8].

We emphasize a case of BD with a rare form of presentation, within frequent but potentially serious cardiopulmonary manifestations, with an atypical progression of symptoms.

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