

## Behcet's Disease Complicated with Pulmonary Artery Aneurysm, Intra-Cardiac Thrombus and Pulmonary Embolism

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### Abstract

### Case Report

Behcet's disease is a systemic process characterized by oral ulcers, genital ulcers, uveitis, and skin lesions. Furthermore, Behcet's disease can manifest as vasculitis lesions, such as arterial aneurysm, venous thrombosis or thrombophlebitis. We report the case of a pulmonary artery aneurysm thrombosis associated with intracardiac thrombus in a 21-year-old male with a previous diagnosis of Behcet's disease presented with dyspnea and hemoptysis.

**Key words:** Behcet's Disease, pulmonary artery aneurysm, Pulmonary embolism, hemoptysis.

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## INTRODUCTION

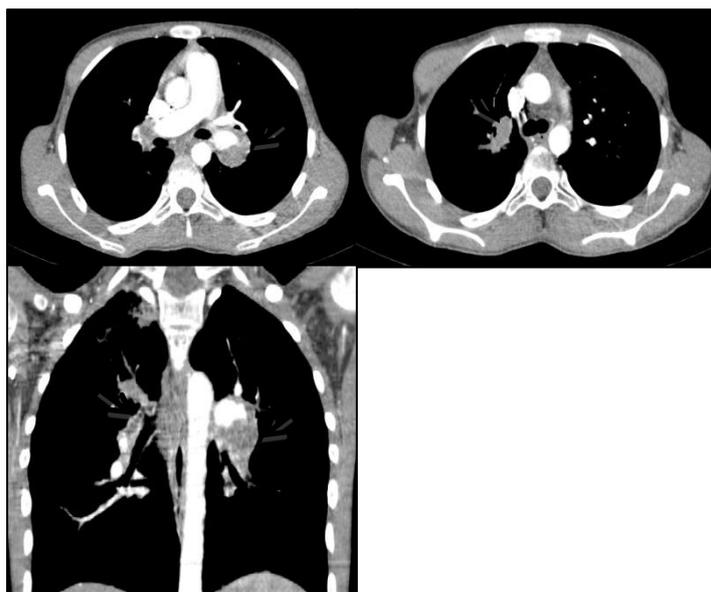
Vascular involvement in Behçet's disease is very common and is observed in 7 to 38% of cases. Small or large vessel vasculitis in Behçet's disease leads to vessel occlusion or aneurysm formation, but arterial involvement occurs infrequently and presents as aortitis or a peripheral arterial aneurysm with arterial thrombosis [1]. Venous thrombosis is also present in one-fourth of Behçet's disease patients [2]. The pulmonary artery is the second most common site of arterial involvement in Behçet's disease in 1 to 7% of cases, and can cause massive hemoptysis by rupture in bronchi [3, 4].

## OBSERVATION

A 21 year-old man was admitted to the hospital with dyspnea at rest, hemoptysis, palpitations and chest pain. The patient had been diagnosed with Behçet's disease about 4 years previously. On physical examination vital signs were stable. Blood tests revealed; hemoglobin 10.2 g/dL, hematocrit 30.6%,

platelet count 445,000/ $\mu$ L. His chest radiogram showed left hilar enlargement. Initial diagnosis of pulmonary artery aneurysm and pulmonary emboli were especially associated with the hilar enlargement and previous diagnosis of Behçet's disease. Dynamic contrasted computed tomography of both pulmonary arteries revealed aneurysmal dilatation and chronic thrombus involving all interior lumen of the left pulmonary artery with a dimension of 2,5  $\times$  2 cm (figure 1). Pulmonary embolism was detected on the right pulmonary artery with peripheral but localized patchy infiltration. A thrombus with a dimension of 2.5  $\times$  1,5 cm was determined in the right ventricle (figure 2).

Immunosuppressive therapy was administered to the patient and he was quickly relieved of her symptoms after a combination therapy with methylprednisolone and low-molecular-weight heparin. In the follow-up light hemoptysis and death of the patient.



**Fig-1:** Axial and coronal contrast-enhanced computed tomography scan shows left pulmonary artery aneurysm (yellow arrow) accompanied by mural thrombus, and pulmonary embolism was detected on the right pulmonary arterie (red arrow)



**Fig-2:** Axial contrast-enhanced computed tomography shows thrombus in the apex of right ventricle (white wellow)

## DISCUSSION

Behcet's disease is a chronic systemic inflammatory disease [1, 2]. Men are substantially more prone to be affected by arterial disease (sex ratio=10/1) [3]. Intrathoracic appearances of Behcet's sickness speaks to 1-8 % and comprise principally of thromboembolism of the superior vena cava as well as other mediastinal veins, aneurysms of the aorta and pulmonary arteries, pulmonary infarct and hemorrhage, pleural effusion, and rarely, myocardial and pericardial involvement [4 ; 6]. Pulmonary artery aneurysm has a poor prognosis and is one of the main sources of death in Behcet's malady patients. Pulmonary vasculitis and thromboses of pulmonary vessels may cause areas of localized necrosis, central or diffuse hemorrhages, and central regions of atelectasis. Hemoptysis, when massive and untreated, has a death rate of >50% [5]. Pulmonary artery aneurysms in Behcet's disease are extremely uncommon in 1 to 7% of patients [3; 7]. They are of interest to large arterial pulmonary or lobar trunks, or more rarely segmental arterial trunks. They are often bilateral [8]. The pathogenesis of pulmonary

artery aneurysm is multifactorial, leading to an immune-mediated vasculitis [9]. This inflammatory process leads to neovascularization and degeneration of the arterial wall and to the formation of the aneurysm [10].

Chest radiogram shows unilateral or bilateral hilar enlargement, peripheral patchy infiltration [2, 4]. New imaging innovations, especially, dynamic thorax computed tomography, can be useful in the exhibition of thrombus of the systemic veins, heart and pulmonary arteries [11, 12]. In our patient dynamic thorax computed tomograms revealed a right ventricular thrombus. Pulmonary angiography confirms aneurysms and extensions and frequently indicates the thrombosis [13].

The indication of anticoagulation therapy is not clearly shown in Behcet's disease and venous thrombus. Treatment involves combination of anti-inflammatory, and immunosuppressive agents, Prednisone, cyclophosphamide. Additionally,

endovascular embolization may be a therapy of choice for treatment-resistant hemoptysis [14].

## CONCLUSION

Thrombosis pulmonary artery aneurysm associated with intracardiac thrombus is a rare complication of Behçet's disease. In this case we demonstrating the difficulty of management of vasculo-Behçet patients with concurrent bleeding and thrombotic complications. Hemoptysis, when massive and untreated, has a high mortality.

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