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A Case of Behcet's Disease with Bilateral Pulmonary Artery Aneurysm

Bilateral Pulmoner Arter Anevrizmasi ile Seyreden Behçet Olgusu

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Abstract

Behçet's disease is an autoinflammatory disease characterized by recurrent painful oral ulcers with small, medium and large diameter vessel involvement. Although the pathogenesis is not clearly understood, the hypersensitivity of T-cells to antigens is known to play an important role. The hyperactivation of T-cells leads to an increase in such proinflammatory cytokines as IFN-gamma and TNF-alpha, which are responsible for the symptoms of Behçet's disease. The primary clinical concerns are mucosa, skin and ocular lesions, while minor concerns are joint, neurological, gastrointestinal, vascular and pulmonary anomalies. Vascular involvement and pulmonary involvement, while rare, are the prominent clinical presentations. We present this case to literature as a rare example of bilateral pulmonary artery aneurysms.

Keywords: *Behcet's disease, vasculitis, clinical manifestations, pulmonary artery aneurysm.*

Öz

Behçet Hastalığı, tekrarlayan ağrılı oral ülserler ile karakterize ve küçük, orta, büyük çaplı damarları tutan otoinflamatuar bir hastalıktır. Patogenezi net olarak bilinmemekle birlikte, T hücrelerin antijenlere karşı aşırı duyarlılığı patogenezde önemli rol oynamaktadır. T hücrelerinin aşırı aktivasyonu, IFN-gama ve TNF-alfa gibi proinflamatuvar sitokinlerin miktarında artışa yol açar. Bu sitokinler de Behçet Hastalığındaki semptomlardan sorumludur. Başlıca klinik tutulumlar mukozal lezyonları, cilt lezyonları ve oküler lezyonlardır. Minör tutulumlar ise eklem tutulumu, nörolojik tutulum, gastrointestinal tutulum, vasküler tutulum ve pulmoner tutulumdur. Vasküler tutulum ve pulmoner tutulum en nadir ancak en mortal seyreden klinik prezentasyonlardandır. Bilateral pulmoner arter anevrizması literatürde nadir görüldüğünden olgumuz sunulmuştur.

Anahtar Kelimeler: *Behçet hastalığı, vaskülitler, klinik belirtiler, pulmoner arter anevrizması.*

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Behçet's disease is a multisystemic disease with an unknown etiology and a unique geographical distribution (1), being referred to as "Silk Road Disease" due to its frequent observation along the route of the Silk Road between China and the Mediterranean (2). Behçet's disease is relatively rare, with a frequency of 80/100,000 in Iran, 20–420/100,000 in Turkey and 0.64/100,000 in the United Kingdom (3). A meta-analysis of 45 population-based studies revealed prevalence rates of 10.3/100,000 cases for all countries, 119.8/100,000 for Turkey and 3.3/100,000 for Europe (4). Being a multisystemic disease, clinical manifestations involve almost the entire body, although the most common presentations are skin lesions in the form of oral and genital aphthous lesions and ocular involvement in the form of uveitis (5). Pulmonary involvement in Behçet's disease may take the form of pulmonary artery aneurysm, arterial and venous thrombosis, pulmonary infarction, recurrent pneumonia, bronchiolitis obliterans organizing pneumonia and pleurisy (6). Pulmonary artery aneurysms are rare in literature, with an estimated incidence of 1 in 14,000 in autopsies (7).

Clinical knowledge is limited, and the available data are generally derived from autopsy reports. The case presented here featured a bilateral pulmonary aneurysm with oral aphthae, genital ulcers and hemoptysis, and is presented to literature due to its rarity.

CASE

A 25-year-old male patient was admitted to another hospital with a complaint of hemoptysis for 13 months, and underwent a thorax Computed Tomography (CT) Angiography revealing aneurysmatic enlargements in the pulmonary artery branches in the lower lobes of both sides, the largest of which was on the left and measured approximately 3 cm in diameter (Figure 1). The patient was admitted after presenting first to the outpatient clinic.

A detailed anamnesis revealed a history of amphetamine and marijuana use 2 years previously. The patient had contracted COVID-19 approximately 13–14 months earlier and suffered from a persistent cough afterward that did not change during the day or with posture. There was no sputum. The patient also suffered from long-term occasional hemoptysis in minimal amounts that were bright red in color. Dyspnea that increased with effort had been present for the last 5–6 months. The patient had no chest pain or chronic disease in his medical history but had previously undergone an eardrum operation and an adenoidectomy. There was no history of tuberculosis or contact with tuberculosis patients. Behçet's disease was considered as a pre-diagnosis in the patient, who was identified with bilateral pulmonary artery aneurysms on radiologic imaging. An oral examination revealed frequent healing and recurring oral aphthae. A lower extremity venous Doppler ultrasound provided no evidence of thrombophlebitis or deep vein thrombosis.

A dermatology consultation was requested revealing healed wound scarring in the scrotal region, and a pathergy test was positive, and a diagnosis of Behçet's disease was made based on the sum of the findings. The patient was started on Cyclophosphamide 500 mg, with a total of four cures planned every 15 days. The patient was also started on Mesna for the treatment of hemorrhagic cystitis. The first dose of Cyclophosphamide led to a remarkable decrease in the patient's symptoms. The patient was transferred to the Rheumatology Clinic, preventing any further post-treatment control examinations as the patient was no longer part of our follow-up.

DISCUSSION

Behçet's disease is characterized by periods of exacerbation and remission that diminish in time. Vascular involvement is most commonly superficial thrombosis and deep vein occlusions, and more rarely, arterial aneurysms (8). Pulmonary artery involvement, although rare, is the main cause of morbidity and mortality in Behçet's disease and takes two forms: pulmonary artery aneurysms and pulmonary artery thrombosis.

The case presented here had developed a bilateral pulmonary artery aneurysm.

Patients with pulmonary artery involvement account for a small percentage of those with Behçet's disease. Aneurysms are usually accompanied by venous thrombosis; however, the use of anticoagulants may cause aneurysm ruptures and massive hemoptysis. Behçet's disease treatments prioritize the prevention of relapses and the rapid suppression of inflammation in vital organs through the use of immunosuppressants. Cases with pulmonary artery involvement are generally treated with a combination of steroids and cyclophosphamide, while vascular interventional methods can be applied in an emergency, such as in cases with massive hemoptysis due to aneurysm rupture, although these techniques may be ineffective if the aneurysm is too large or may lead to such complications as cavity/abscess formation (9).

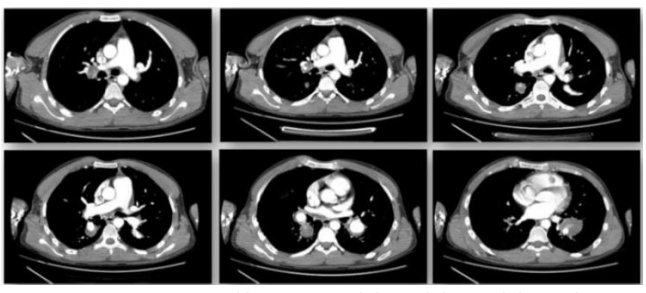

Figure 1: Aneurysmatic dilatations in bilateral lower lobe pulmonary artery branches, the largest of which was approximately 3 cm in diameter on the left

CONCLUSION

The case of Behçet's disease presented here, diagnosed with bilateral pulmonary artery aneurysm, oral aphthae and genital ulcerated lesions, responded well to immunosuppressive treatment.

CONFLICTS OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

Concept - Ş.I., T.K., H.S.Ş.; Planning and Design - Ş.I., T.K., H.S.Ş., D.Ö.K.; Supervision - Ş.I., T.K., H.S.Ş.; Funding - Ş.I., T.K., H.S.Ş.; Materials - Ş.I., T.K., H.S.Ş.; Data Collection and/or Processing - Ş.I., T.K., H.S.Ş.; Analysis and/or Interpretation - Ş.I., T.K., H.S.Ş.; Literature Review - Ş.I., T.K., H.S.Ş.; Writing - Ş.I., T.K., H.S.Ş.; Critical Review - Ş.I., T.K., H.S.Ş.

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