



## Behçet Hastasında Non-Anevrizmatik Rekürren Pulmoner Tromboemboli Recurrent Pulmonary Thromboembolism Without Aneurysm In Behcet's Disease

Turan Aktas<sup>1</sup>, Fatma Aktas<sup>2</sup>, M.D Handan İnönü Köseoğlu<sup>3</sup>, M.D Zafer Ozmen<sup>4</sup>, M.D Yalçın Baş<sup>5</sup>

1 Gaziosmanpasa University, Faculty of Medicine, Department of Pulmonary Diseases, Tokat

2 Gaziosmanpasa University, Faculty of Medicine, Department of Radiodiagnostics, Tokat

3 Gaziosmanpasa University, Faculty of Medicine, Department of Pulmonary Diseases, Tokat

4 Gaziosmanpasa University, Faculty of Medicine, Department of Radiodiagnostics, Tokat

5 Gaziosmanpasa University, Faculty of Medicine, Department of Dermatology, Tokat

### ÖZET

Behçet Hastalığı (BD) etyolojisi net olarak bilinmeyen , kronik inflamasyonla seyreden ve multisistemik tutulum gösterebilen bir vaskülitir. Çoğunlukla Akdeniz, Ortadoğu ve Asya ülkelerinde görülmektedir. İlk defa 1937 yılında Türk dermatolog olan Hulusi Behçet tarafından tanımlanmıştır. Behçet hastalarında tüm sistemlerde tutulum görülmesinin yanında en çok cilt , oral mukoza, göz, renal, pulmoner, eklem ve santral sinir sistemi tutulumu saptanmaktadır. Bunlar içerisinde pulmoner tutulum %1-18 oranında tespit edilmektedir. Pulmoner tutulum genellikle pulmoner arterlerde anevrizma ve buna eşlik eden trombüs şeklinde görülmekte olup bunun yanında diğer vasküler sahada da trombus izlenebilmektedir. Ancak anevrizma olmadan pulmoner arter içerisinde trombüsle seyreden BD çok nadir olup literatürde şimdiye kadar bildirilmemiştir.

**Anahtar Kelimeler:** Behçet Hastalığı, Tromboz, Pulmoner Arter Anevrizması

### ABSTRACT

Behçet's disease(BD) is a multi-systemic vasculitis with unknown etiology and chronic inflammation. It is seen in Mediterranean, Middle East and Asian countries in general. It was firstly described by Hulusi Behçet, a Turkish dermatologist, in 1937. Although the multisystem involvement can be seen in BD, the involvements of skin and oral mucosa are seen mostly. The ratio of pulmonary involvement is %1-18 in BD. The most common pathological finding in pulmonary involvement is pulmonary artery aneurysm with or without thrombosis formation, but the thrombus in pulmonary artery without aneurysm is a very rare clinical manifestation in BD . Therefore, it is aimed to report a case of BD with thrombosis in pulmonary artery without aneurysm.

**Key Words:** Behçet's Disease, Thrombosis, Pulmonary Arter Aneurysm.

**Corresponding Author:** Turan Aktas

**Adress:** Gaziosmanpasa University, Faculty of Medicine Department of Pulmonary Diseases, Tokat, Turkey

**E-mail:** turanaktas79@yahoo.com

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## INTRODUCTION

Behçet's Disease is firstly defined by Hulusi Behçet, a Turkish dermatologist, in 1937. Its etiology is still unknown. It is a multi-systemic vasculitis and it especially involves vascular systems in skin, oral mucosa, joints, kidneys, brain, eyes and lungs. The ratio of pulmonary involvement is %1-18 in BD (1). The most common clinical manifestation of the pulmonary involvement in BD is pulmonary artery aneurysm with or without protective thrombosis formation. It is claimed that the thrombosis formation in vascular system is secondary to the chronic inflammation of vascular bed. The tendency of thrombosis increases due to the fibrinolytic system activation and endothelial damage caused by the factor V laden and prothrombin gene mutation with anti-cardiolipin and anti-endothelial antibodies formation (2). The pulmonary artery aneurysm occurs as a result of the chronic inflammation in pulmonary vascular bed and especially in pulmonary arterial system. Also, following that the protective thrombosis formation can be seen. However, thrombosis formation in pulmonary arterial system without aneurysm is seen very rarely. For this reason, the main aim of this report is to present a 47 years old male patient with recurrent pulmonary thromboembolism without pulmonary artery aneurysm being followed and taking treatment for about 8 years because of BD with involvements of skin, oral mucosa and eyes (üveitis).

## CASE

47 years old male patient applied to our clinic with complaints of dispnea, chest pain and

hemoptysis. He has been using colchium and has been followed for around 10 years because of BD with skin and eye involvement. However, he quit the treatment of Colchium in 2 months. His complaints began 2 days before he came to our clinic. Examination of the patient was done with thorax CT angiography due to his serious complaints and existing diagnosis. Thrombosis formations and pulmonary infarctions were detected in the branches of pulmonary artery and parenchyma of the lung as a result of the examination of thorax CT angiography (Figure 1). Also, dopler USG was performed to scan the lower extremity deep venous system to detect source of the thrombosis in pulmonary arterial system. There were thrombosis formations in bilateral lower extremity deep venous system according to the dopler USG imaging. Therefore, it was decided to begin low weight heparin (LWH) and immunosuppressive treatments based on the results of the examinations. The thrombosis formations in both lower extremity deep venous system and pulmonary arterial system disappeared as a result of the follow-up care lasting for 1 year. But, he applied to the emergency service with the same complaints in the second year of his follow-up. The partial thrombosis formations and left sided pleural effusion with the width of 11mm were scanned again in thorax CT angiography (Figure 2). The right sided chronic thrombosis formations were identified after the scanning of lower extremity deep venous system with dopler USG. He was hospitalized and the LWH with immunosuppressive treatment was started. The thrombosis formations were not seen in thorax CT angiography and dopler USG scanning performed in the 6th month of his



**Figure1:** In thorax CT angiography, thrombosis formations and pulmonary infarctions were detected in the branches of pulmonary artery and parenchyma of the lung.



**Figure 2:** In thorax CT angiography, the partial thrombosis formations and left sided pleural effusion with the width of 11mm were detected again at his second application to the hospital in follow-up care.

second follow-up care . His follow- up under the LWH and immunosuppressive treatments has still been going on.

## DISCUSSION

Behcet's Disease is a vasculitis and mostly common in countries located on Ancient Silk Road. Its prevalence is around 1/1000-10000 in the world. It is usually seen in young females at second and third decades. ( Male/Female:2/10) ( 3,4).

The diagnostic criterias of BD were defined again by International Study Group of Behcet's Disease in 1990. According to these criterias, there must be at least two of the clinical evidences which are genital ulcer, uveitis, skin lesions and positive pattery test in addition to the oral apthae (5). It is a multi-systemic vasculitis and it can be life-threatening depending on the location of involvement of vascular systems in body. Involvement of the skin is the most frequently identified involvement type and also vascular involvements in central nervous system, eyes, joints, kidneys and pulmonary system can be seen in clinical manifestation, too. Oral apthae, genital ulcer phollicullitis and uveitis were identified at the beginning of the diagnosis in this case.

The pulmonary involvement in BD is seen rarely (%1-18)(1,6). Generally, the clinical manifestation of pulmonary involvement is identified as pulmonary artery aneurysm or pulmonary vein aneurysm in BD. The thrombosis formation accompany the aneurysm and this situation is accepted as a defense mechanism developed by

the body against the rupture of aneurysm. The frequency of thrombosis is %10-30 in BD (7). It is claimed that the reason for increase in tendency to thrombosis is due to the chronic inflammation and vasculitic reaction in BD. However, a strong relationship between thrombophilic factors and BD has not been confirmed in literature, yet (8). Moreover, the defects in fibrinolytic system have been examined, but they have not had any relation with thrombosis formation in BD (9,10). In a study, Gül et al. suggested that G1691A mutation in factor V can be related to the thrombosis formation of deep veins in BD(11). But, based on the recent evidence, the thrombosis formation in BD has not been confirmed yet (8). In a study conducted in France revealed that mortality is related to the thrombosis formations in BD. This study also claims that mortality is mostly related to the thrombosis formations in arterial system without any relation to venous system (7). Pulmonary involvement and predisposition of thrombosis formation can be life-threatening in BD.

In conclusion, BD is a multi-systemic, life-threatening vasculitis and it is seen in young females in general. The tendency of thrombosis formation increases in circulating system in BD. The pulmonary involvement can be serious and life-threatening in BD, especially in pulmonary artery aneurysm and thrombosis formations in pulmonary vascular bed. Nonspecific clinical complaints like dispnea, hemoptysis and chest pain can be seen in these patients in early periods. The early diagnosis is very important to avoid serious and life-threatening results.

There is no conflict of interest.

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