

Right Ventricular Thrombus Formation and Pulmonary Embolism as a Complication of Behçet Disease: Case Report

Behçet Hastalığının Bir Komplikasyonu Olarak Sağ Ventriküler Trombüs Oluşumu ve Pulmoner Emboli Olgusu

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ABSTRACT Behçet disease is a chronic, inflammatory, multisystemic vasculitis mainly characterized by orogenital ulcers. It is mainly observed in Turkey, other Mediterranean areas, and Japan. Viral, bacterial, genetic, immunologic, and environmental factors have been implicated in the pathogenesis of the disease. Although disease is multisystemic, cardiac involvement is rare and often associated with poor prognosis. Hypercoagulopathy is one of the important manifestation of the Behçet disease. Thromboembolic events such as myocardial infarction, stroke and pulmonary embolism risk is higher in patients with Behçet compared to normal population. Right ventricular thrombus is a quite rare. In this paper we present a 26-year-old male patient who presented with pulmonary embolism and was subsequently diagnosed with a right ventricular thrombus and completely recovered after treatment. We also discussed diagnostic and therapeutic options.

Key Words: Behçet syndrome; thrombosis; pulmonary embolism

ÖZET Behçet hastalığı başlıca orogenital ülserlerle seyreden kronik inflamatuvar multisistemik bir vaskülitir. Ülkemizle beraber Akdeniz ülkelerinde ve Japonya'da sık görülmektedir. Etyopatogenezinde genetik, çevresel, viral, bakteriyel ve immünolojik vb. çeşitli faktörlerin rol oynadığı düşünülmektedir. Hastalık multisistemik tutulum göstermektedir. Kardiyovasküler tutulum ise nadir olmakla birlikte kötü prognozla ilişkilidir. Hiperkoagüabilite Behçet hastalığının bir diğer önemli manifestasyonlarından birisidir. Behçet hastalarında akut miyokard infarktüs, serebrovasküler ve pulmoner emboli gibi tromboembolik olaylar normal popülasyona göre daha fazla görülmektedir. Sağ ventriküler trombüse ise oldukça nadir rastlanmaktadır. Bu sunumda pulmoner emboli ile başvuran, takibinde sağ ventriküler trombüs tespit edilip, tedavi ile tamamen düzelen 26 yaşında erkek hasta sunulmuş olup tanı ve tedavi yöntemleri tartışılmıştır.

Anahtar Kelimeler: Behçet sendromu; tromboz; pulmoner emboli

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Behçet disease (BD) is a multisystem chronic inflammatory disorder with unclear etiology. The disease manifests mainly with oral aphthous ulcers, genital ulcerations, and uveitis.¹ Apart from this classical triad, there may be neurological, gastrointestinal, and cardiovascular involvement.^{2,3} Cardiac involvement is quite rare and associated with poor prognosis. Cardiac involvement may present with myocarditis, pancarditis, acute myocardial infarction, conduction system abnormalities, and various vascular disorders such as aneurysmal dilatation.⁴ In this paper we present a 26-year-old male admitted to hospital and treated for right ventricular

thrombus and acute pulmonary embolism. We also discussed the relationship between BD and thrombosis.

CASE REPORT

A 26-year-old man treated for BD for 3 years admitted to emergency department with sudden-onset dyspnea and right sided chest pain during sleep. He stated that his current symptoms began during sleep but admitted that he had experienced occasional exertional dyspnea in the past, for which he had never seen a doctor. He denied any previous chest pain or hemoptysis. He had no other systemic disease, either. He had been on colchicine therapy for 2 years. Owing to increased severity of oral and genital ulcerations, azathioprine 50 mg/day had been added 4 months before the index admission. On admission his blood pressure was 100/60 mmHg and pulse rate was 96 bpm. Oxygen saturation measured with pulse oximetry was 97%. Physical examination only revealed 2 oral aphthous lesions. Cardiac examination was within normal limits. His electrocardiogram was normal except for right bundle branch block. A telecardiogram was also normal. Echocardiography showed a right ventricular mass of 28x30 mm located at the origin of papillary muscles, which was consistent with a thrombus lesion (Figure 1). Right ventricle was mildly dilated but had preserved systolic functions. A mild tricuspid insufficiency was accompanied by a pulmonary artery systolic pressure of 25 mmHg. The mass had an amorphous appearance and was immobile (Figure 1). The patient was admitted to hospital ward and begun on low molecular weight heparin. A thoracic computed tomography with contrast enhancement revealed patchy areas of consolidation in anteromedial segment and middle lobe of the right lung. Thus, a ventilation perfusion (V/P) scintigraphy was performed. The test revealed a high probability scan for pulmonary embolism with multiple defects for both right and left lungs which are most prominent in right upper and lower lobes (Figure 2). The patient was begun on heparin infusion and acetyl salicylic acid for 10 days. Because of the patient was not in hypotension and shock status, thrombolytic therapy was

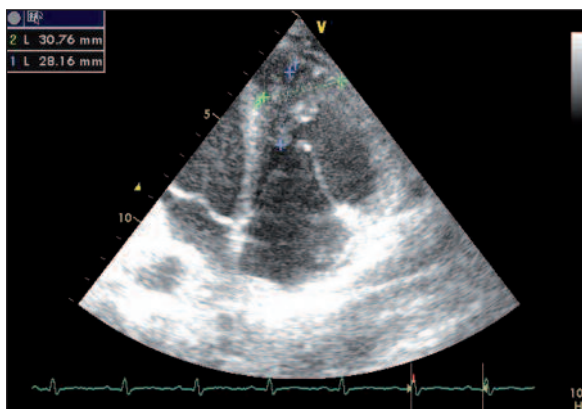


FIGURE 1: A right ventricular thrombus at the origin of papillary muscles.

not considered. On follow-up, he was recommended surgery to remove right ventricular thrombus but he refused it. Cyclosporin and coumadin were commenced on follow-up. Bilateral lower extremity venous Doppler revealed no thrombus. Protein C and S levels were normal, and factor 5 Leiden mutation was negative. His antinuclear cell antibodies, antiphospholipid antibodies, and von Willebrand factor antigen were negative. Homocysteine level was normal. Erythrocyte sedimentation rate was 30 mm/h, CRP level was 13 mg/L. His INR level was kept between 2 and 3. A control echocardiography at 3 months revealed complete resolution of thrombus (Figure 3). Right ventricle diameter was normal. Pulmonary artery systolic pressure was between normal ranges.

DISCUSSION

Cardiac involvement is very rare in BD, with a prevalence of 1% to 5%. In an autopsy study in Japan the prevalence of cardiac involvement was 16.5%.⁵ In those with vascular involvement, arterial and venous thrombus formation is one of the most common manifestations. The arterial thromboses being later in the course than the venous ones. Although vascular injury is quite common, intracardiac thrombosis is substantially rare. It is more common in men and those with young BD patients with eye involvement.⁶ It may sometimes be confused with other intracardiac masses like myxoma. In such cases, transesophageal echocar-

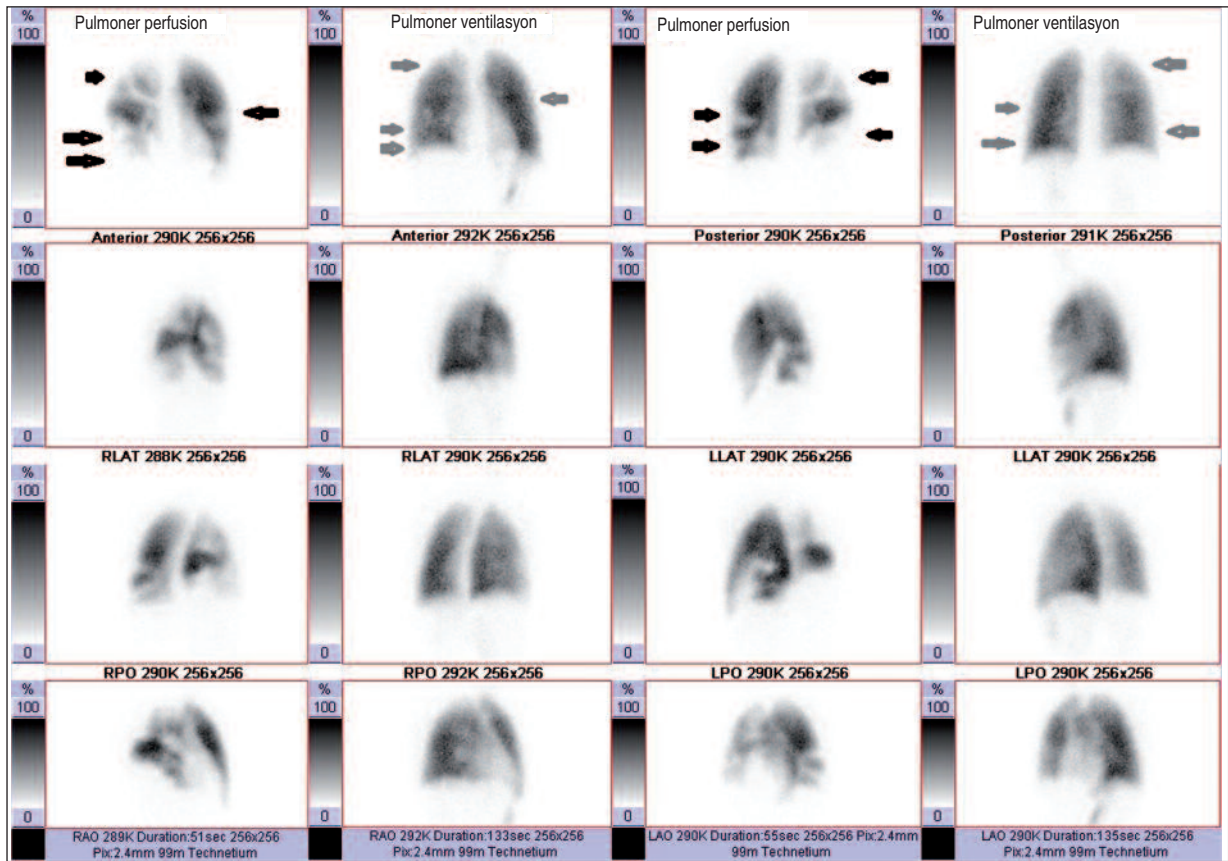


FIGURE 2: Findings consistent with pulmonary embolism in a ventilation perfusion scintigraphy. (black arrows indicate ventilation, gray arrows indicate ventilation images).

diography (TEE) or magnetic resonance imaging (MRI) can be utilized.⁷ Thrombi have a heterogeneous appearance and are more common in ventricles than atria.⁵ Our case was also a young male having a right ventricular thrombus that had caused pulmonary embolism. Transthoracic echocardiography images were compatible with a thrombus, which had an amorphous appearance with a broad base and was immobile. As a consequence of these features consistent with a thrombus, no TEE or MRI was needed.

The cause of thrombotic tendency in BD is not entirely clear although many factors have been implicated. Vascular inflammation, endothelial cell ischemia and dehiscence increase platelet aggregation, impair fibrinolysis, and lead to thrombus formation.^{5,8} Other factors that have been implicated include presence of antiphospholipid antigens, prothrombotic factors like protein C-

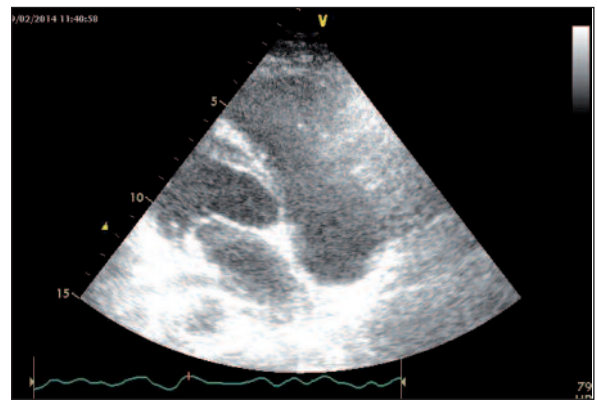


FIGURE 3: Complete resolution of thrombus, as seen on an echocardiography at the third month of therapy.

S and antithrombin, von willebrand factor antigen, and hyperhomocysteinemia.^{9,10} These parameters were all between normal ranges in our patient.

Optimal treatment of thrombi in BD is controversial. Size, mobility, and localization of thrombi are important factors for deciding optimal therapy. Medical therapy should be given a priority; however, thrombi can sometimes be removed with aspiration.¹¹ Medical therapy includes antithrombotic therapy, anticoagulation, and immunosuppressives.⁶ Our case was considered to have an active disease despite colchicine and azathioprine treatment. He had elevated CRP and sedimentation rate and his oral aphthous lesions persisted. Our case was treated with cyclosporin and low molecular weight heparin followed by coumadinization, and the thrombus disappeared in as short as 3 months. Moreover, we did not consider thrombolytic therapy. Our case was hemodynamically stable. According to European Society of Cardiology pulmonary embolism guideline, thrombolytic therapy should only be considered in patients presented with hypotension and shock.¹² In patients with shock and hy-

potension; thrombolytic therapy is an effective treatment option. Another treatment strategies of APE are surgical embolectomy and percutaneous catheter-directed treatment. Percutaneous catheter-directed treatment options include; thrombus fragmentation with pigtail or balloon catheter, rheolytic thrombectomy with hydrodynamic catheter devices, suction thrombectomy with aspiration catheters and rotational thrombectomy.¹² We don't have any expertise about percutaneous catheter-directed therapy in pulmonary embolism, so we did not consider this option.

Right ventricular thrombus is extremely rare in BD. In patients with BD it should be remembered that cardiovascular thrombi may develop, especially in active cases. The connection of right ventricular thrombi with BD should not be forgotten and the existence of this disease should be sought for in such cases.

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