Case Report
A Behcet’s Disease Patient with Right Ventricular Thrombus, Pulmonary Artery Aneurysms, and Deep Vein Thrombosis Complicating Recurrent Pulmonary Thromboembolism

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1. Introduction

Behcet’s disease (BD) is a multisystem disorder presenting with recurrent buccal aphthosis, genital ulcer, and uveitis with hypopyon [1]. Pulmonary involvement in Behcet’s disease is rare, occurring in 1 to 7.7% of the patients [2, 3]. Pulmonary artery aneurysms, arterial and venous thrombosis, pulmonary infarction, recurrent pneumonia, bronchiolitis obliterans organized pneumonia, and pleurisy are the main features of pulmonary involvement in Behcet’s disease [4]. Cardiac involvement causes coronary artery disease, recurrent pericarditis, myocarditis, and endocarditis abnormalities. Intracardiac thrombus formation is very uncommon [5]. We present a Behcet’s disease patient with intracardiac thrombus, pulmonary artery aneurysms, and deep vein thrombosis complicating recurrent pulmonary embolism.

2. Case Report

A twenty-year-old woman was admitted to the hospital with complaints of cough, fever, palpitations, and chest pain. It was learned that, four years ago the patient had been diagnosed with Behcet’s disease and received irregular colchicine treatment but not regularly. The patient was hospitalized. On the transthoracic echocardiogram, a thrombus with a dimension of 4.2×1.6 cm was recognized in the right ventricle. On abdomen CT, aneurysmal iliac veins and deep vein thrombus on Doppler ultrasonograms were diagnosed. At the controls after three months of immunosuppressive and anticoagulant therapies, some clinical and radiological improvements were recognized. The patient suspended the treatment for a month and the thrombus recurred. We present our case in order to show the effectiveness of immunosuppressive and anticoagulant therapies and rarely seen pulmonary thromboembolism in recurrent Behcet’s disease.
3. Discussion

Intrathoracic manifestations of Behcet's disease consist mainly of thromboembolism of the superior vena cava and/or other mediastinal veins, aneurysms of the aorta and pulmonary arteries, pulmonary infarct and hemorrhage, pleural effusion, and rarely, myocardial and pericardial involvement, cor pulmonale, and mediastinal or hilar lymphadenopathy [6]. Pulmonary infarction is a stage in the natural course of the disease. Pulmonary vasculitis and thromboses of pulmonary vessels may cause infarctions, focal or diffuse hemorrhages, and focal areas of atelectasis [6–8]. Although vascular involvement is seen only in 25% of the patients, it is the most common cause of mortality in Behcet's disease [6, 9, 10]. New imaging technologies, especially, dynamic thorax CT, can be helpful in the demonstration of thrombus of the systemic veins, heart and pulmonary arteries [8]. Dynamic thorax CT revealed a right ventricular thrombus in our patient. The thrombus was confirmed by echocardiography. Thromboembolism stemming from a cardiac cavity has been previously deemed to be relatively uncommon [9]. A review by Moğulköç et al. regarding intracardiac thrombi in 25 patients with Behcet's disease was previously published [5]. The authors noted that they had seen pulmonary embolism in 13 patients (52%). In seven of these 13 patients, thrombophlebitis was observed in the major vessels which might have been the source of the embolism. Although deep venous thrombosis of the lower extremities frequently accompanies pulmonary artery aneurysms, pulmonary thromboembolism is very rare in Behcet's disease because the thrombi in inflamed veins are strongly adherent [11]. In a review study done by Houman et al. on 113 Behcet's disease patients, vein involvement had been detected in 49 patients (43.3%), and deep vein thrombus had been observed in 44 of them (38.9%). Deep vein thrombosis was more frequently observed among males (40 males and 4 females) [12]. Another review consisting of reports of Turkish authors revealed one intracardiac thrombus out of 56 (1.78%) Behcet's disease patients [13]. Recently, two Behcet's disease patients with intracardiac thrombi and pulmonary artery aneurysms have been reported [14, 15]. Luo et al. analyzed the clinical characteristics of Behcet's disease with intracardiac thrombus [16]. The data of 8 patients diagnosed with Behcet's disease with intracardiac thrombi in Peking Union Medical College Hospital from January 1990 to January 2011 were studied retrospectively. Intracardiac thrombus associated

*Figure 1: Chest radiogram demonstrating bilateral hilar enlargement and patchy infiltration.*
with Behcet’s disease most commonly occurs in young men and usually involves the right side of the heart [16].

The pathologic mechanism of microvascular thrombus formation in vasculitis is believed to be caused by endothelial cell ischemia or disruption that leads to enhanced platelet aggregation [4, 5]. Decreased release of vascular tissue plasminogen activator has been reported in systemic and cutaneous vasculitis [9]. Impaired fibrinolysis as a result of endothelial cell injury from deposited immune complexes is another possible mechanism. Prolonged euglobulin lysis times and abnormal fibrin concentrations were found in several types of vasculitis, including Behcet’s disease [6, 8]. In the present case, intracardiac thrombus, deep vein thrombosis, and pulmonary embolism were detected. Considering the possible mechanisms leading to thrombus, and recurrent emboli due to intracardiac thrombus and deep vein thrombosis, immunosuppressive and antithrombotic medications were started. Warfarin was not the preferred
treatment option due to the risk of bleeding. Although the first line treatment is medical, thrombus can become massive and may demand surgical treatment.

We presented infarct centers and new emboli centers on peripheral divisions showing chronic thrombus observed related to the repeated emboli on pulmonary arteries. During the follow-up, there was in change on thrombus divisions. Newly formed emboli were not observed and infarct centers regressed. This situation showed the effectiveness of the treatment. Our patient's pulmonary artery pressure was not high. Since pulmonary artery aneurysm decreases the load on the right side of the heart, rise in the pulmonary artery pressure might be observed in cases with severe pulmonary embolism. There was cardiac and deep vein thrombus inside the right ventricle of our patient. As these two diagnoses might cause recurrent embolisms per se, anticoagulant treatment needs to be used concomitantly with the immunosuppressive treatment. Some publications have asserted that these patients were subjected to bleeding episodes, and anticoagulant treatment is contraindicated for them. On the contrary, we saw that this treatment prevented occurrence of recurrent embolisms.

We have initiated methylprednisolone and cyclophosphamide combination therapy as suggested for the management of other severe forms of systemic vasculitis. We added an anticoagulant treatment into this combination. We have observed clinical and radiological improvement with this treatment.

We kindly deemed suitable to present this case report in order to show the necessity of anticoagulant treatment to be added to the immunosuppressive therapy in such complicated cases.

**Conflict of Interests**

There is no actual or potential conflict of interests.

**Acknowledgment**

The authors declare that they have no affiliation with or financial involvement in any organization or entity with a direct financial interest in the subject matter or materials discussed in the paper.

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