Intracardiac Thrombosis and Coronary-to-Pulmonary Artery Fistula with Pulmonary Embolism and Budd-Chiari Syndrome in Behçet's Disease: A Case Report

Behçet Hastalığı (BH) multisistemik, kronik ve inflamatuar vaskülit tablosudur. [1] Although large venous and arterial lesions occur with it, intracardiac thrombus formation is uncommon.

We report a manifestation of BD with intracardiac thrombosis (ICT) and coronary-to-pulmonary fistula, a pulmonary embolism (PE), and Budd-Chiari syndrome (BCS) in which the patient recovered.
Completely after being treated with colchicine, methylprednisolone, cyclophosphamide, and intravenous heparin.

**CASE REPORT**

A 29-year-old male patient with BD was admitted to our clinic with dyspnea, palpitations, and chest pain. The diagnosis had been established three years earlier on the basis of mucocutaneous lesions and gastrointestinal disorders. The patient had then been treated with colchicine and corticosteroids. His past medical history also included deep venous thrombosis (DVT) in his calf. On his physical examination, a left parasternal murmur resembling the sound of a machine was detected along with abdominal ascites. His electrocardiography (ECG) and chest X-ray results were normal. Post-contrast thoracic computed tomography (CT) revealed right pulmonary effusion and decreased calibration together with a filling defect of the left inferior pulmonary artery due to a chronic PE (Figure 1a). On the abdominal ultrasononography, hepatic venous thrombosis was detected. The portal venous system was patent, but no enhancement was detected in the hepatic veins or proximal inferior vena cava (IVC) on the abdominal CT. The findings were concordant with BCS (Figure 1a). Two-dimensional transthoracic echocardiography revealed a mass on the right atrium protruding into the right ventricle (Figure 2a). The suprasternal view showed a turbulent and abnormal flow surrounding the arcus aorta and pulmonary artery (Figure 2b). Doppler interrogation demonstrated a continuous flow signal at that location. In addition, coronary angiography was performed to evaluate coronary involvement because of intractable angina pectoris, and a coronary artery-to-pulmonary artery fistula was detected arising from the left coronary artery (LCA) (Figure 3). On cardiac catheterization and oximetry study, there was a 5% step-up between the right ventricle and distal pulmonary artery, and the left-to-right shunt was 1.3. Since there was no evidence of a significant shunt, the fistula was not thought to be the cause of the angina, and the patient did not undergo corrective intervention. During the sixth month of treatment with colchicine, methylprednisolone, cyclophosphamide, and heparin, thoracic CT revealed complete lysis of the pulmonary thrombosis and ICT.

**DISCUSSION**

Cardiovascular involvement has been reported in 7-46% of BD patients. There are three categories of vascular lesions in BD: arterial occlusion, arterial aneurysms,
and venous occlusions.[3] Pulmonary involvement is secondary to vasculitic lesions and includes stenosis, occlusions, and aneurysms.[4] Data regarding treatment and outcomes of patients having BD-related pulmonary emboli and small-sized vasculitis is limited. Computed tomography angiography is noninvasive and provides excellent delineation of the vessel lumen and wall as well as the perivascular tissues for the diagnosis of vascular complications.

Coronary-to-pulmonary artery fistulas are usually congenital in origin. Edwards[5] classified them into two types: the primary fistula, in which the fistula is the main pathological lesion, and the secondary fistula, which occurs as a consequence of other malformations. In our case, we hypothesized that the fistula formation may be a collateral circulation due to a chronic PE. The abnormal vessel, as detected in our patient, may arise from the coronary artery at any level,[6] and the hemodynamic effect of the left-to-right shunt is related to the size of the shunt and pulmonary vascular resistance. The presence of a fistula may lead to a decreased amount of blood perfusing the myocardium, and this may result in angina, myocardial infarction (MI), or papillary muscle dysfunction.[7] Selective coronary angiography is the most accurate diagnostic tool and usually demonstrates evidence of the origin, course, size, and termination of the fistula. Closure treatment is usually indicated in patients with severe symptoms that cannot be controlled by medical treatment. However, the method of treating asymptomatic patients with coronary artery fistulas is more controversial.[7]

Our BD patient had ICT and a PE with coronary-to-pulmonary artery fistula in conjunction with BCS. There was no significant shunt in our case, so no intervention was considered.
Intracardiac thrombosis is an uncommon complication of BD. Only 66 cases of BD patients with ICT have been reported in the literature. The right heart is the most common localization for the thrombosis, but the reason for the frequency of right-sided ICT associated with BD is still unknown. Soulami et al. postulated that endomyocardial fibrosis has a role in the development of ICT in some patients. Endothelial dysfunction, the release of von Willebrand factor (vWF), platelet activation, enhanced thrombin and fibrin generation, antithrombin deficiency, and impaired fibrinolysis close the pathological chain of enhanced thrombocoagulation associated with vasculitis (perivasculitis) in BD.

However, it is difficult to know whether the thrombi are secondary to the underlying endocarditis or endomyocardial fibrosis. There is no consensus regarding the treatment of cardiac thrombosis in association with BD. The aim of treatment is to control the underlying disease and to resolve the cardiac thrombus. Anticoagulant and antithrombotic agents are the first line of therapy. Routine anticoagulation with heparin or oral anticoagulants is not advised, but anticoagulant treatment together with immunosuppressive therapy is an advised treatment option in Behçet’s cases with ICT. As with pulmonary artery aneurysm (PAA), a combination of cyclophosphamide and methylprednisolone is most frequently prescribed for patients with severe vasculitis, but in patients with BD, the PAA may become smaller or disappear with medical treatment. Occlusion of the hepatic portion of the IVC, major hepatic vein, or both can lead to BCS, and the prevalence of BCS in BD is 3.2%. Behçet’s disease is an uncommon cause of BCS, and only 56 cases of BD complicated by BCS have been published. However, the BCS can be treated medically, interventionally, or surgically.

In conclusion, when investigating the possible etiology of ICT, pulmonary PE, and BCS in a patient, BD should be kept in mind. In addition, immunosuppressive treatment along with anticoagulants can be used in BD patients with vascular disease.

Declaration of conflicting interests
The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding
The authors received no financial support for the research and/or authorship of this article.

REFERENCES