Noncompaction of the ventricular myocardium in a patient with Behçet's disease showing multiple thrombus formations in the right atrium, inferior vena cava and right iliac vein: A case report

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A case of noncompaction of the ventricular myocardium in a 24-year-old man with Behçet's disease is reported. Transthoracic echocardiographic examination revealed two cardiac masses in the right atrium, pericardial fluid behind the right atrium and multiple muscular trabeculation in the apical part of the left ventricle. Subsequent transesophageal echocardiography clearly showed two thrombus formations in the right atrium and prominent ventricular

trabeculations with deep intertrabecular recesses perfused from the ventricular cavity, which are characteristic findings of ventricular noncompaction. In addition, abdominopelvic computed tomography showed thrombus formation in the inferior vena cava and right iliac vein. These multiple thrombus formations were treated successfully with steroid, immunosuppressive and anticoagulant therapies. This is the first reported case of left ventricular noncompaction in association with Behçet's disease showing pericarditis and multiple thrombus formations in the right atrium, inferior vena cava and right iliac vein.

Key Words: *Behçet's disease; Intracardiac thrombus; Ventricular noncompaction*

Noncompaction of the ventricular myocardium (NVM) is a rare and unclassified congenital cardiomyopathy (1). The disease is thought to result from an arrest of the physiological intrauterine compaction process of the ventricular myocardium during embryonic heart development. NVM may occur as an isolated cardiac lesion, and it has also been reported in association with many other cardiac and extracardiac congenital anomalies. However, there have been no reported cases of NVM in association with Behçet's disease.

Behçet's disease is a vasculitis of unknown origin. It is characterized by recurrent oral and genital aphthous ulcers, arthritis, uveitis and thrombophlebitis. Venous thrombosis is a common complication of this disease (2). Similarly, thromboembolic events have also been reported as one of the clinical manifestations of NVM. Although cardiac involvement is extremely rare in Behçet's disease, pericarditis, intracardiac thrombosis, valvular disease, arrhythmias and conduction disturbance may occur (3-7). Prognosis is poor in the presence of cardiac involvement. The pathogenetic mechanisms underlying the thrombotic disposition in Behçet's disease are not well known. We describe a case of NVM in a 24-year-old man with Behçet's disease, characterized by pericarditis and multiple thrombus formations in the right atrium, inferior vena cava and right iliac vein.

CASE PRESENTATION

A 24-year-old man was admitted to the Eskisehir Osmangazi University Hospital, Eskisehir, Turkey, with complaints of fever and chest pain for two months. The patient did not have dyspnea, palpitations, syncopal episodes or systemic embolic events, and there was no family history of cardiomyopathy. He had no history or clinical signs of chronic pulmonary disease.

Physical examination showed tachycardia, normal blood pressure and a temperature of 39°C. Aphthous ulcers were observed in the oral cavity, and there were active ulcers and ulcer scars on the scrotum; the patient had a history of recurring lesions. A pericardial friction was heard at the left sternal border. Electrocardiography demonstrated sinus rhythm with T wave inversion in leads II, III, aVF and V4 to V6. Chest radiography was normal.

Laboratory findings were as follows: hemoglobin 76 g/L, erythrocyte sedimentation rate greater than 140 mm/h, C-reactive protein 136 mg/L, D-dimer 3.25 mg/dL, ferritin 1505 mg/dL. Liver and kidney function tests were normal. Antinuclear antibody, Anti-DNA, Sm, RNP, Ro, La, Jo-1, HLA B5, pathergy and tuberculin skin tests were negative. Thrombophilia screening revealed normal antithrombin III, protein C and protein S levels, and he was not a carrier of factor V Leiden or prothrombin G20210A mutations.

Transthoracic echocardiography revealed two cardiac masses in, and pericardial fluid behind, the right atrium. A dilated left ventricle with mildly impaired systolic function was observed. The left ventricular end-diastolic and end-systolic diameters were 60 mm and 46 mm, respectively. The ejection fraction (EF) was 42%. Mild mitral and tricuspid regurgitation were present. The left ventricular apical region and mid-portion of the posterolateral walls had an inhomogeneous appearance with trabeculation and numerous recesses with penetration into the myocardium (Figure 1). A subsequent transesophageal echocardiograph confirmed multiple prominent trabeculations and deep intertrabecular spaces perfused from the ventricular cavity within the left ventricular wall, mostly involving the apical and posterolateral regions, and two thrombus formations in the right atrium (Figure 2). An abdominopelvic computed tomography

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Figure 1) Transthoracic echocardiography. Mid-portion of the left ventricular (LV) walls are characterized by the presence of multiple, prominent trabeculations and intertrabecular recesses in the short axis view before (A) and after six months of the treatment (B); echocardiography showed no changes in the structure of the LV cavity

Figure 2) A Transesophageal echocardiography (TEE) showing two thrombus formations in the right atrium (RA) (thick arrow) and the pericardial fluid behind the RA (thin arrow). **B** After six months of the treatment, TEE demonstrates no thrombus in the right atrial cavity (thick arrow) and no pericardial effusion behind the RA (thin arrow). RV Right ventricle

Figure 3) A Abdominopelvic computed tomography showing thrombus formation in the inferior vena cava (arrows). **B** After six months treatment, no thrombus formation is seen in the inferior vena cava

scan showed a thrombus in the inferior vena cava (Figure 3) and right iliac vein (Figure 4).

The patient was diagnosed as having NVM with Behçet's disease showing multiple thrombus formations. The diagnosis

of Behçet's disease was based on the findings of aphthous ulcers within the oral cavity, active ulcers and ulcer scars on the scrotum, and vascular involvement with vascular thrombosis, in agreement with the diagnostic criteria for Behçet's disease

Figure 4) Abdominopelvic computed tomography showing that the thrombus formation in the right iliac vein (arrow) before the treatment (A) is not seen after treatment (B)

described by Dilsen et al (8). The diagnosis of NVM was based on the characteristic echocardiographic findings of the disease. Intravenous heparin, intravenous prednisolone 1000 mg/day for three days and 1 g of cyclophosphamide were prescribed. The fever returned to normal on the second day of treatment. Treatment was continued with oral anticoagulant and prednisolone (1 mg/kg/day). Pericardial friction as well as oral and genital ulcers improved after seven days. After six months, transthoracic and transesophageal echocardiographic examinations showed that the intracardiac thrombi had disappeared and EF increased to 48%.

DISCUSSION

The report of the World Health Organization Task Force on the definition and classification of cardiomyopathies (1) has categorized NVM as an unclassified cardiomyopathy. The prevalence of this disorder was previously reported (9,10) to be 0.014% of patients referred to the echocardiography laboratory. Transthoracic echocardiography, transesophageal echocardiography, magnetic resonance imaging and left ventriculography can be used for the diagnosis of NVM. The disease may be associated with neuromuscular disorders such as facial dysmorphism, cleft palate, obstructive lesions of the right or left ventricle, complex cyanotic heart disease and bicuspid aortic valve. In Behçet's disease, cardiac involvement is reported to be extremely rare. This is the first reported case of ventricular noncompaction in association with Behçet's disease showing multiple thrombus formations in the right atrium, inferior vena cava and iliac vein.

Heart failure, ventricular arrhythmias and cardioembolic events are the most common reported clinical manifestations of NVM. Heart failure is caused by systolic and diastolic dysfunction. Systolic dysfunction is most probably the result of relative ischemia of the myocardium due to the mismatch of myocardial oxygen supply and demand (11). NVM is a congenital myocardial disorder but the onset of symptoms and clinical presentation may be delayed until the sixth decade. Many asymptomatic patients are identified during routine echocardiographic evaluations. In our patient, ventricular noncompaction was diagnosed during routine echocardiographic examination performed for the evaluation of pericardial friction. Although coronary artery disease is usually considered a cause of left ventricular dysfunction in Behçet's

disease, inflammatory mechanisms may play a role in the development of left ventricular dysfunction (12). Because left ventricular EF increased after immunosuppressive therapy, our patient's left ventricular dysfunction at initial presentation was most likely related to myocardial involvement during an activated phase of Behçet's disease, combined with the presence of noncompacted myocardium.

Thrombotic tendency is a well-known feature of Behçet's disease. Vascular involvement is seen in one-third of patients with Behçet's disease. The most frequent type of vascular involvement in Behçet's disease is venous thrombosis. Intracardiac thrombosis is rarely reported in the literature. In these cases, the thrombus formations are reported to be within the ventricle or atrium, mostly in male patients (13). Thromboses can be treated successfully with steroid, immunosuppressive and anticoagulant therapies (14). Endothelial injury and dysfunction have been shown to be the main factors causing thrombosis in Behçet's disease (15). Hereditary risk factors such as factor V Leiden and prothrombin G20210A mutations likely have a role in thrombus formation in Behçet's disease. However, some studies (16,17) emphasized the increase in thrombin-activatable fibrinolysis inhibitor and factor VIII levels in these patients. Impaired fibrinolytic activity or relative hypofibrinolytic state have also been reported in the prothrombotic state of Behçet's disease (18). However, all of the thrombophilic factors examined in our patient were found to be in the normal range.

Thromboembolic complication is one of the clinical characteristics of NVM, and is observed during long-term follow-up in these patients. Thromboembolic events most frequently occur in the arterial system, including cerebrovascular accidents (the most common event), transient ischemic attacks and mesenteric infarction. However, pulmonary embolism has also been reported in NVM and has been the cause of death in some patients (19). The most frequent causes of death in NVM are reported to be ventricular tachycardia and thromboembolic events (20). It is clear that there is a tendency to thrombosis in both NVM and Behçet's disease. The present case report emphasizes that patients with NVM in association with Behçet's disease may have a higher risk for thrombosis and for the probability of thromboembolism, and anticoagulant therapy has a special importance in the prevention of embolic complications in these patients.

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