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A Case Report Of Behçet's Disease With Coronary Artery Involvement

Cardiac Involvement Due to Behçet's Disease

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Case Report

History

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ABSTRACT

We present a case of coronary artery thrombosis due to Behçet's disease. A 49-year-old female patient, diagnosed with Behçet's disease and followed up by us, applied to the emergency department with sudden onset of pressure-type chest pain, and was evaluated as acute myocardial infarction due to elevated cardiac enzymes and ST segment elevation in the anterior leads on electrocardiography (ECG). Coronary angiography revealed near-total thrombosis in the lumen of the left anterior descending artery (LAD) and in the circumflex (CX) artery,it was intervened with anticoagulant therapy. The patient's cardiac risk was found to be low and differential diagnosis was made in terms of myocardial infarction. Thereupon, the causes of hypercoagulability were investigated in the patient who was consulted to us, and acute phase reactants were found to be high. This was related to the coronary artery involvement of Behçet's disease and in addition to the treatment containing antiaggregant, statin, angiotensin receptor blocker, intravenous (IV) high-dose methylprednisolone and cyclophosphamide-mesna treatment was given for immunosuppressive purposes. It should not be forgotten that there may be vascular involvement due to Behçet's disease and coronary arteries may also be affected.

Keywords: Behçet's disease, coronary angiography, coronary artery disease, cyclophosphamide, myocardial infarction

Koroner Arter Tutulumu Olan Behçet Hastalığı: Olgu Sunumu

Süreç

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ÖZ

Behçet hastalığına bağlı koroner arter trombozu olgusu sunuyoruz. Behçet hastalığı tanısı almış ve tarafımızca takipli 49 yaşında kadın hasta, ani başlayan baskı tipi göğüs ağrısı ile acil servise başvurmuş, kardiyak enzim yüksekliği ve elektrokardiyografide (EKG) anterior derivasyonlarda ST segment elevasyonu gelişmesi üzerine akut miyokard enfarktüsü olarak değerlendirilmiştir. Koroner anjiografide sol ön inen arter (LAD) lümeninde ve sirkumfleks (CX) arterde totale yakın tromboz saptanmış ve antikoagulan tedaviyle müdahale edilmiştir. Hastanın kardiyak riski düşük bulunmuş olup; myokard enfarktüsü açısından ayırıcı tanıları yapılmıştır. Bunun üzerine tarafımıza konsulte edilen hastada hiperkoagulabiliteye neden olacak patoloji saptanmamış ve akut faz reaktanları yüksek bulunmuştur. Bu durum Behçet hastalığının koroner arter tutulumuna bağlanmış ve antiagregan, statin, anjiotensin reseptör blokorü içeren tedaviye ek olarak immunsupresif amaçlı intravenöz (IV) yüksek doz metilprednizolon ve siklofosfamid-mesna tedavisi verilmiştir. Behçet hastalığına bağlı vasküler tutulum olabileceği ve koroner arterlerin de etkilenebileceği unutulmamalıdır.

Anahtar sözcükler: Behçet hastalığı, koroner anjiografi, koroner arter hastalığı, miyokard enfarktüs, siklofosfamid

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Introduction

Although Behçet's disease is defined as a syndrome with recurrent oral aphthae, genital ulcer, ocular inflammation and articular involvement, it can cause inflammation in almost all organs. Although central nervous system and gastrointestinal system involvements create life-threatening clinical pictures; cardiopulmonary involvement and vasculitis are important causes of mortality [1]. Vascular findings are thrombophlebitis, pseudoaneurysm, arterial stenosis and occlusion [2]. While cardiovascular symptoms are observed in 7-46% of the cases; 20% of these may be mortal [1]. Cardiac involvement in Behçet's disease can take various forms such as coronary vasculitis, endocarditis, myocarditis, pericarditis, aortic aneurysm, ventricular thrombosis, congestive heart failure, aortic or mitral failure [1,3,4]. Coronary artery involvement is complicated by aneurysm and/or thrombosis, hemorrhage, acute myocardial infarction, and sudden death [4]. In this case, we will present a case of coronary artery thrombosis due to Behçet's disease.

Case Report

A 49-year-old female patient with no known chronic disease was diagnosed with Behçet's disease about 1 year ago with recurrent oral and genital ulcers, bilateral posterior uveitis, and HLAB51 positivity. There were bilateral erythema nodosum-like rashes on the anterior surface of the tibia, and when the biopsy was performed, the pathology report showed that "epidermis is normal; subcutaneous septal panniculitis, perivascular neutrophilic infiltration were observed and findings consistent with early erythema nodosum were found. When evaluated together with the clinic, it is significant for Behçet's Disease." The patient, who was followed up with azathioprine treatment, presented to the emergency department with a sudden onset of severe, pressing chest pain in the last 1 week. It was evaluated as acute myocardial infarction due to the development of cardiac enzyme elevation and ST segment elevation in anterior leads in ECG. Coronary angiography revealed total thrombotic occlusion in the LAD artery and in the CX artery, angiographic intervention and anticoagulant therapy were applied (Figure-1). No vascular aneurysm was observed in angiography. When the cardiovascular risk assessment of the patient is performed, the low-density lipoprotein (LDL) is 62 mg/dl, HbgA1c: 5.6. ; found low. The causes of hypercoagulability were investigated in the patient who was consulted by us. Laboratory findings of the patient were Erythrocyte Sedimentation Rate: 50 mm/h, C-reactive protein: 40 mg/L, Anti-Nuclear Antibody: negative, Anti-Myeloperoxidase and Anti-Proteinase 3 were negative, Anti-Cardiolipin Antibody immunoglobulin M and G, Anti-beta 2 Glycoprotein and lupus anticoagulant were negative. When the patient was interviewed by hematology about the causes of hypercoagulation, hematological causes were excluded as a result of the examinations and genetic analysis. The patient, who did not have clinical, laboratory and imaging findings suggestive of vasculitis and systemic lupus erythematosus, was transferred to our service with the thought of coronary artery involvement due to Behçet's disease. In addition to the treatment including antiagregant, statin, angiotensin receptor blocker, pulse methylprednisolone iv and mesna treatment was applied together with iv cyclophosphamide for 3 days for immunosuppressive purposes. Methylprednisolone was gradually tapered off. Clinical and laboratory response was obtained with this treatment in the patient. After 6 cycles of cyclophosphamide-mesna treatment, maintenance treatment with azathioprine and low-dose methylprednisolone was planned.

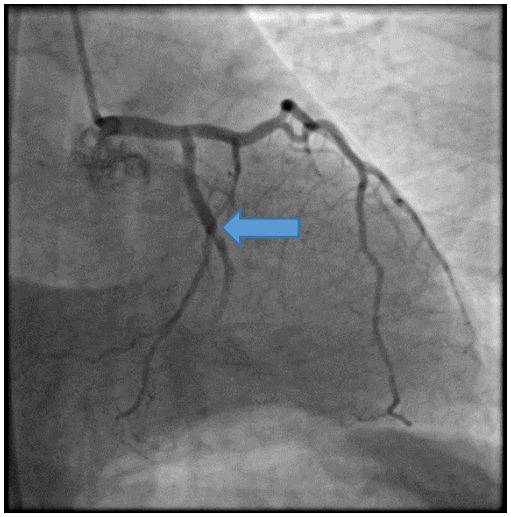

Figure-1: There were total thrombotic occlusion in the circumflex artery in patient's coronary angiography

Discussion

Behçet's disease can affect vessels of any size and cause aneurysm or thrombosis. Vascular involvement is reported between 15% and 40%; venous involvement is more common, arterial involvement is rarer. It most commonly progresses with superficial thrombophlebitis and deep vein thrombosis in the lower extremities [5]. Arterial involvement due to Behçet's disease often progresses in the form of aneurysms and pseudoaneurysms [6]. Cardiac involvement due to Behçet's disease is rare; often the main vascular structures of the heart, coronary arteries, as well as the endocardium and myocardium are affected [7]. While examining our case with a history of oral aphthae, genital ulcer and uveitis; she was diagnosed with Behçet's disease with pathergy positivity and HLAB51 positivity, and had acute coronary syndrome even though she did not have any chronic disease or coronary artery disease in herself or in her family. This situation has been associated with Behçet's disease because of near-total occlusion due to thrombosis in the LAD and CX arteries in coronary angiography and high acute phase reactants.

Studies have shown that the risk of cardiovascular disease increases in Behçet's disease; Geri et al. They described cardiac pathologies in 52 (6%) of 807 Behçet's patients. Of these 52 patients, 86.5% were male; the mean age of all patients was around 30 years [8]. Our case differed in that it was a 49-year-old female.

Cardiac involvement in Behçet's disease; includes endocarditis, myocarditis, pericarditis, intracardiac thrombus, endomyocardial fibrosis, coronary arteritis, coronary and ventricular aneurysm, myocardial infarction and valve diseases [1,2]. Also; Interatrial septum aneurysm, mitral valve prolapse, mitral regurgitation, sinus valsalva dilatations are rare lesions [1]. In the study of Geri et al., pericarditis was the most common heart involvement with 38.5%, and the frequency of myocardial infarction was reported as 17% [8]. In the case series of Marzban et al., it was reported that coronary vessels are frequently involved as coronary artery aneurysm, stenosis or coronary arteritis [6]. In our case, no aneurysm was observed in the coronary arteries, however; thrombosis-related myocardial infarction developed.

Considering the treatment options in vascular involvement of Behçet's disease, interferon-α is effective, but the frequency of its side effects limits the treatment. The effectiveness of infliximab, one of the tumor necrosis-α (TNF-α) inhibitors, in cardiovascular involvement is controversial [9]. Cyclophosphamide and corticosteroids are included in the treatment according to EULAR recommendations in pulmonary and peripheral artery involvement due to Behçet's disease [10]. We used iv high-dose methylprednisolone and iv cyclophosphamide treatment in our case.

Conclusion

It should be kept in mind that the most important cause of mortality in Behçet's disease is vascular thrombosis or aneurysms. Antiagregant and anticoagulant treatments are insufficient to prevent recurrence in thrombotic events in Behçet's disease; It should be remembered that these conditions occur due to the inflammatory process, and it should be kept in mind that they should be mainly treated with immunosuppressive agents (steroid, azathioprine, cyclophosphamide and others).

Ethical Rules: Written consent was obtained from the case; the Declaration of Helsinki has been complied with.

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