

Coronary artery ectasia in a patient with Behçet's disease

Ersan Tatli, MD, Huseyin Surucu, MD, Meryem Aktoz, MD, Mutlu Buyuklu, MD.

ABSTRACT

Behçet's disease is a multisystemic disease of unknown etiology. Disease manifestations consist of recurrent oral and genital ulceration, skin lesions, and relapsing ocular inflammation. Arterial involvement is an uncommon complication of Behçet's disease, and it most frequently affects the abdominal aorta followed by femoral artery, and the pulmonary artery. Coronary lesions in Behçet's disease have been little reported in the literature. A 36-year-old female with 6-year history of Behçet's disease was hospitalized with ectasia of the left main coronary artery. This unusual vascular complication Behçet's disease is presented.

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From the Department of Cardiology (Tatli, Aktoz, Buyuklu), Trakya University School of Medicine, Edirne, and the Department of Cardiology (Surucu), Erdem Hospital, Istanbul, Turkey.

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Address correspondence and reprint request to: Dr. Ersan Tatli, Associated Professor, Department of Cardiology, Trakya University School of Medicine, Edirne, Turkey. Tel. +90 (284) 2357641 Ext. 2150. Fax. +90 (284) 2357652. E-mail: ersantatli@yahoo.com/ersantatli@hotmail.com

Behçet's disease is recognized as a chronic multisystem disorder affecting systemic organs and characterized by oral and genital ulceration, dermatitis, and recurrent uveitis. The vascular involvement of Behçet's disease is seen in 1-7%. Vascular Behçet's disease mainly affects the venous system while arterial lesions are known to be less common.¹ The most frequently observed arterial lesions of vascular Behçet's disease are aneurysms, or obstruction of the abdominal aorta, or both, and the pulmonary artery, or the femoral artery, or both.² Coronary lesions are rare complications of vascular Behçet's disease. Only a few coronary aneurysms have been reported in the literature.³⁻⁵ We report ectasia of the coronary arteries on the left main coronary artery in a 36-year-old patient with Behçet's disease.

Case Report. A 36-year-old female, diagnosed with Behçet disease since she was 30 years old, with recurrent oral aphthae, genital ulcer, and iridocyclitis, developed angina pectoris. She had class II (Canadian Cardiovascular Society) angina. The Behçet's disease had been controlled with colchicine (1.5 mg/day) and corticosteroid (2 mg/day). On physical examination, she was afebrile, pulse rate was 76 per minute and blood pressure was 120/80 mm Hg. Cardiovascular system examination was normal. Her chest x-ray, liver, and kidney function tests were normal. C reactive protein concentration was raised; antinuclear antibody and anti-DNA tests were negative. Her human leucocyte antigen-B5 and pathergy tests were positive. Electrocardiogram demonstrated ST-T changes in the anterior derivations. Normal anterior wall motion was shown on transthoracic echocardiogram. Coronary angiography showed ectasia of the coronary artery (20 mm x 8 mm), proximally including the left anterior descending artery and circumflex coronary artery, beginning from the ending of the left main coronary artery (Figure 1). She was not considered a candidate for surgery, and was discharged on corticosteroids, colchicine and antiaggregant treatment.

Discussion. Arterial lesions of Behçet's disease may present as an occlusive disease, an ectasia, or a combination of the occlusive lesion and aneurysm.² The prevalence of coronary involvement in Behçet's disease is 0.5%.⁵ Coronary aneurysms are more frequent than stenotic lesions. Aneurysms are believed to occur due to weakened adventitia secondary to lymphocyte infiltration to the vasa vasorum.⁶ Aneurysms have poorer prognosis than occlusive disease in Behçet's disease because of the risk of rupture. The aneurysms most often involved are the aorta, and pulmonary arteries, followed by femoral, subclavian, and common carotid arteries. Involvement of the coronary arteries is very rare. Behçet's disease continues with remissions and attacks. There is still no definite medical treatment. Frequent monitoring of the inflammatory markers, including C-reactive protein and white blood cells, are essential for the management of Behçet's disease. The most common approach is the use of immunosuppressive agents.⁷ These include

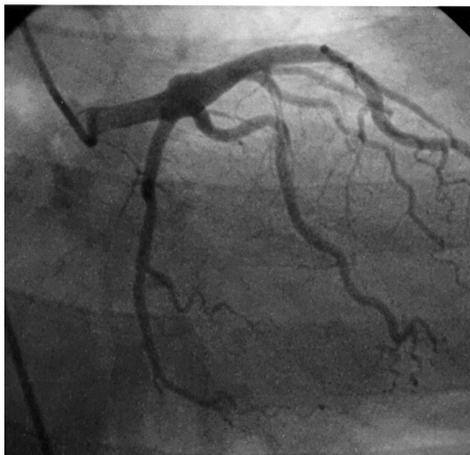


Figure 1 - Coronary angiography showing ectasia of the coronary artery of size 20 mm × 8 mm that includes proximally the left anterior descending artery and circumflex coronary artery, beginning from the end of the left main coronary artery.

corticosteroids, cytotoxic agents, and cyclosporine. The dose of corticosteroid must be adjusted appropriately.

In conclusion, young patients presenting with any type of arterial ectasia should also be investigated for Behçet's disease, among other etiologies.

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Case Reports

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