

## A case report of coronary artery aneurysm in a patient with Behçet's disease

### Behçet hastalığı bulunan hastada saptanan koroner arter anevrizması: Olgu sunumu

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**Summary**– Behçet's disease (BD) is a multisystem vasculitis that may involve vessels of all sizes. Acute coronary syndrome (ACS) due to secondary involvement of BD is rare and its management less clear. In this case, a 29-year-old man admitted to the emergency room with ongoing chest pain was interned to the coronary care unit with a diagnosis of ACS. The patient had been diagnosed 1 year before with BD and had been without regular follow-up, despite the suggested use of colchicum. An immediate coronary angiography revealed a fresh thrombus image in the proximal segment of the right coronary artery (RCA), an aneurysm of the left anterior descending artery (LAD) at proximal segment, and a hemodynamically significant lesion following the aneurysm. Intervention was ended because of normal flow (TIMI III) of distal RCA. An intravenous infusion of glycoprotein IIb/IIIa receptor inhibitor (tirofiban) was administered, and a control angiography showed dissolution of a thrombus in RCA, but enlarged aneurysm of LAD and a new aneurysm in RCA.

First described 1937, Behçet's disease (BD) is characterized by the triad of oral ulcers, genital ulcers and ocular lesions.<sup>[1]</sup> Involvement of the central nervous system, musculoskeletal system, joints, connective tissue, and vascular tissue are also seen. Only 3-6% of patients with BD have cardiac involvement, which includes conduction system abnormalities, pericarditis, myocarditis, coronary artery disease, myocardial infarction, congestive heart failure, valvular insufficiency, endomyocardial fibrosis, and intracardiac thrombus.<sup>[2]</sup> Acute coronary syndromes (ACS) in patients with BD can be complicated by thrombus formation or coronary aneurysmal compli-

**Özet**– Behçet hastalığı, tüm boyutlardaki damarları etkileyebilen çoklu sistemi tutan bir vaskülitir. Behçet hastalığına sekonder gelişen akut koroner sendrom (AKS) ise nadir olup nasıl tedavi edileceği konusu açıklığa kavuşmamıştır. Bu yazıda bir yıl önce Behçet tanısı konan ve kolşisin önerilmesine rağmen düzenli ilaç kullanmayan ve acile göğüs ağrısı ile başvuran 29 yaşındaki erkek hasta sunuldu. AKS tanısı ile koroner yoğun bakım ünitesine alınan hastaya yapılan koroner anjiyografide sağ koroner arterde (RCA) taze trombus ve sol ön inen arterin (LAD) proksimal segmentinde bulunan anevrizma ve sonrasında ciddi lezyon saptandı. RCA akımının normal olması (TIMI III) üzerine işlem sonlandırıldı. İntravenöz glikoprotein IIb/IIIa inhibitörü (tirofiban) başlanan hastaya yapılan kontrol koroner anjiyografide RCA'da trombus görülmedi ve LAD'de bulunan anevrizmanın büyüdüğü ve RCA'da da bir anevrizmanın olduğu saptandı.

cations. There are 30 cases previously described in the literature, and here we describe the management of a 29-year-old BD patient with thrombus in the right coronary artery (RCA) and rapid progression of aneurysm of left anterior descending artery (LAD).

#### Abbreviations:

ACS	Acute coronary syndrome
BD	Behçet's disease
LAD	Left anterior descending
RCA	Right coronary artery

#### CASE REPORT

A 29-year-old male patient presented with chest pain, ongoing for a period of 3 h before admission. The

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character of the pain was a retro-sternal squeezing sensation, radiating to the neck, jaw and shoulder. No history of coronary artery disease, hypertension, diabetes mellitus, obesity and hyperlipidemia was detected as coronary artery disease risk factors. He had smoked at least one pack of cigarettes/day for 10 years. After careful questioning for opiates and marijuana addiction, no addiction was indicated. The patient had been diagnosed with BD 1 year previously, due to relapsing orogenital ulcers and papulopustular eruption. Colchicine 1.5 mg/oral twice a day had been started. Due to the healing characteristics of the lesion, the patient discontinued the medical treatment.

Physical examination revealed normal vital findings with blood pressure of 123/76 mmHg, heart rate of 80 bpm and body temperature of 37.1°C. On his chest auscultation, heart sounds and both of the lung areas were normal. A chest X-ray also was normal. Serial electrocardiography recordings showed alternating left bundle branch block with normal sinus rhythm. The transthoracic echocardiography showed inferior wall hypokinesis of the left ventricle with mild mitral regurgitation. As soon as ACS diagnose was made, intravenous unfractionated heparin, nitroglycerin, aspirin, clopidogrel and  $\beta$ -blocker therapies were begun.

The coronary angiography urgently done to clarify the case showed a fresh thrombus in the proximal segment of the RCA (Figure 1a) and a 12 mm aneurysm of LAD at the proximal segment, with a hemodynamically significant lesion following the aneurysm (Figure 1b). Intervention was ended because of good blood flow of distal RCA and an intravenous infusion of glycoprotein IIb/IIIa receptor inhibitor (tirofiban) was administered. 3 weeks later, a control coronary angiography showed a new but mild aneurysmal dilatation of RCA (Figure 1c, d) and the enormous transformation of the aneurysm of LAD (Figure 1e, f).

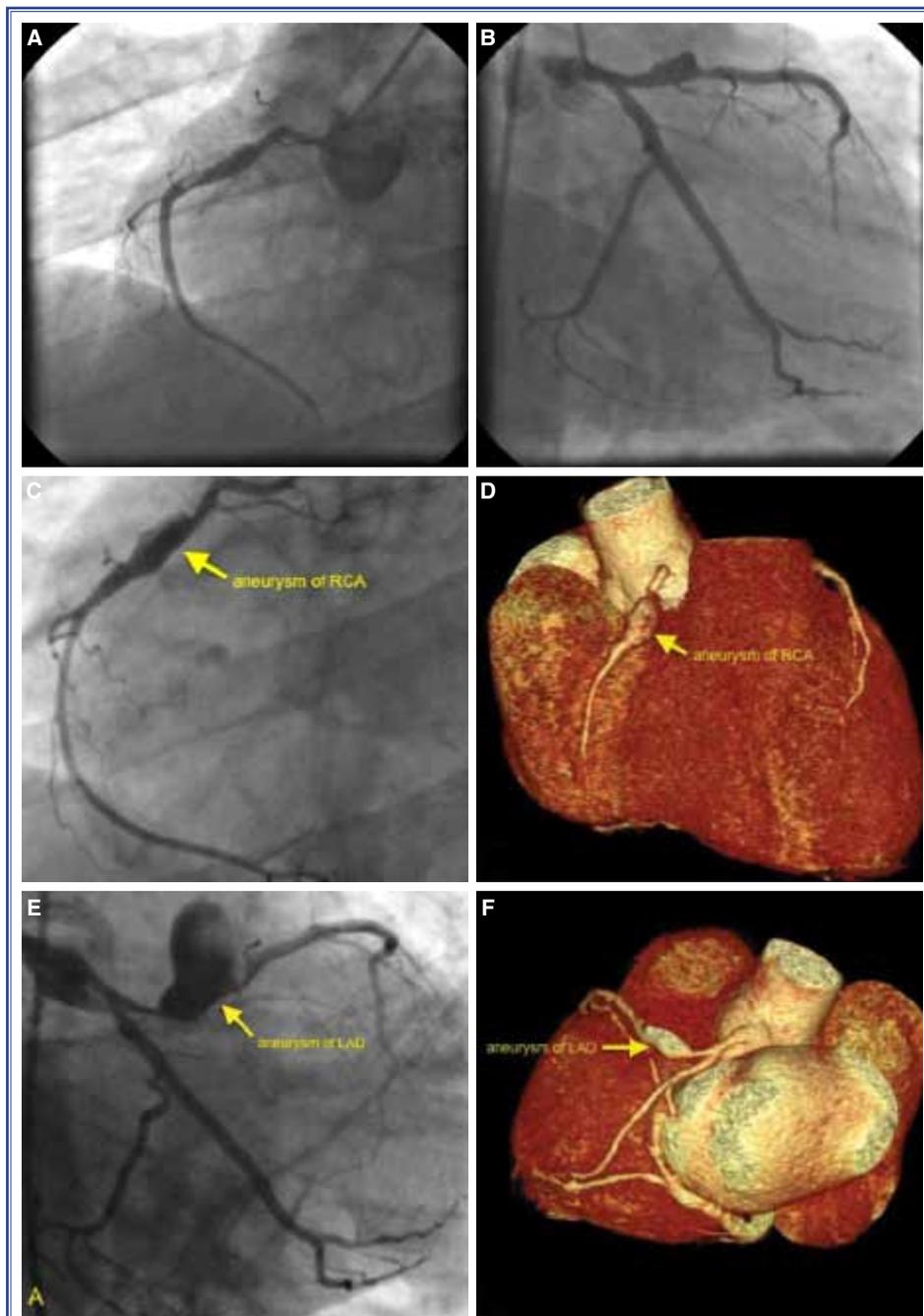
## DISCUSSION

BD is a multisystem vasculitis that can involve vessels of all sizes, and is characterized by recurrent oral and genital ulcers, with variable manifestations affecting the skin, eyes, central nervous system and musculoskeletal system.<sup>[2,3]</sup> In general, BD is seen in the Mediterranean region, the Middle East, and is also known as “Silk Road Disease.”<sup>[3]</sup> Vascular involvement of Behçet (vasculo Behçet) varies from all kinds

and all size of vessels. Venous involvement, arterial involvement and aneurysm formation of the vascular tree of all sizes may be seen. Although the main underlying pathophysiological mechanism of the disease is obscure, inflammation of unknown origin of the vascular endothelium is the cause of vascular complications. Additional thrombus formation associated with vasculitis is an important cause of mortality and morbidity<sup>[4]</sup> and is seen at a rate of 20-40%.<sup>[5]</sup>

First described by Schiff et al.<sup>[6]</sup> as a myocardial infarction, coronary involvement in BD is very rarely found, its prevalence being only 0.5%.<sup>[7-9]</sup> To date, about 30 case reports of ACSs associated with BD have been described in the literature.<sup>[2,3]</sup> Generally patients are young, usually male, without vascular risk factors, except smoking in some.<sup>[10]</sup> Like etiopathogenesis, the definite treatment and management of BD-associated myocardial infarction is not clear yet. Some authors reported the use high dose of corticosteroids and immunosuppressives,<sup>[11]</sup> thrombolytic agents,<sup>[12,13]</sup> and primary percutaneous coronary intervention.<sup>[14]</sup> Surgical treatment should be reserved for critical vascular lesions including those of large size, rapid growth, and acute or impending ruptured aneurysm.<sup>[15]</sup> As Ergelen et al.<sup>[13]</sup> described thrombolytic therapy added the tirofiban therapy in a patient with BD whose left main coronary artery obstructed with thrombus in our instruction, the same regimen administered to our case without thrombolytic because of the presence of thrombolysis in myocardial infarction-III distal blood flow. No bleeding complication was observed during tirofiban infusion. As suggested by rheumatology, Colchium was started with the patient. During a 3-week follow-up in the hospital, no anginal episode was recorded. The ACS of this patient was attributed to the RCA lesion, not the LAD aneurysm. A control coronary angiography revealed dissolution of a thrombus in RCA, and a new aneurysmal dilatation of the RCA at the proximal side. Transformation of the LAD aneurysm from 12 mm to 39 mm in diameter was observed, and needed to be corrected with surgery.

Harrison et al.<sup>[16]</sup> described a 40-year-old BD male patient who underwent RCA stenting trans right femoral approach due to acute myocardial infarction. The patient presented with a right femoral artery pseudo aneurysm 4 months later, and left ventricular inferobasal pseudo aneurysm and right coronary aneurysm



**Figure 1.** (A) Coronary angiography showed a fresh thrombus in the proximal segment of right coronary artery. (B) Coronary angiography showed an aneurysm of 12 mm diameter of left anterior descending artery at the proximal segment with a hemodynamically significant lesion following the aneurysm. (C) Control coronary angiography showed dissolution of the thrombus buta new aneurysmal dilatation of right coronary artery, (D) Coronary multidetector computed tomography view. (E) Control coronary angiography showed gigantic transformation of the aneurysm of left anterior descending, (F) Coronary multidetector computed tomography view.

with stent occlusion 5 months later. Due to this documentary evidence in the literature, conservative treatment was preferred in this patient until a control coro-

nary angiography. After this, surgical intervention for the LAD aneurysm became as an obligation, with rheumatology advising a 1-month follow-up with ste-

roid and cyclosporine therapy before the operation.

BD may present with vasculitis that is mortal and bring morbidity. ACS due to secondary involvement of BD is rare, and its management less clear. Percutaneous interventional therapy, medical treatment with high dose steroids combined with added other immunosuppressants, medical treatment with thrombolites, and surgery are the possible options in Behçet's coronary involvement, none of which has proven better than any other, and choice of treatment depends on the patient's situation. Glycoprotein IIb/IIIa receptor inhibitors were found to be efficacious in relieving anginal symptoms and thrombus dissolution in a 29-year-old male BD presented with ACS. However, a new aneurysmal formation and enlargement of the aneurysm was detected. The management was changed to immunosuppressive therapy, followed by correction surgery in this case.

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**Key words:** Acute myocardial infarction; aneurysm; Behçet syndrome/complications; coronary thrombosis.

**Anahtar sözcükler:** Akut miyokart enfaktüsü; anevrizma; Behçet sendromu/komplikasyonlar; koroner trombüs.