CASE REPORT / OLGU SUNUMU

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From Giant Coronary Aneurysm-related Acute Coronary Syndrome to the Diagnosis of Behçet's Disease

Dev Koroner Anevrizmaya Bağlı Akut Koroner Sendromdan Behçet Hastalığı Tanısına

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Abstract

Behçet's disease is a multisystemic vasculitis that rarely involves the coronary arteries. This case report presents a 19-year-old female with no traditional cardiovascular risk factors, who was admitted with acute chest pain. Electrocardiography revealed inferior ST-segment elevation myocardial infarction, and emergent coronary angiography demonstrated giant aneurysms involving all three major coronary arteries, along with total occlusion of the distal right coronary artery. Despite percutaneous interventions and thrombolytic therapy, distal flow was not restored. Extensive diagnostic evaluation was performed to identify underlying systemic causes. Differential diagnoses such as Kawasaki disease, lupus, Takayasu arteritis, polyarteritis nodosa, immunoglobulin G4-related disease, and infectious vasculitides were systematically excluded. Mucocutaneous findings, including multiple aphthous ulcers and hyperpigmented plaques, combined with vascular involvement, led to the diagnosis of Behçet's disease based on the International Study Group criteria. The patient was treated with intravenous cyclophosphamide followed by maintenance therapy with infliximab. Due to the extensive and diffuse nature of the aneurysms, surgical intervention was not pursued. Oral anticoagulation was maintained long term. During a four-year follow-up period, no new cardiac events occurred, and inflammatory markers remained well-controlled. This case underscores the importance of considering systemic vasculitides, particularly Behçet's disease, in young patients presenting with acute coronary syndrome without conventional risk factors. Early diagnosis and appropriate immunosuppressive therapy are critical in preventing life-threatening cardiovascular complications in such rare presentations.

Keywords: Behçet's disease, giant coronary aneurysm, acute coronary syndrome, vasculitis

Öz

Behçet hastalığı, nadiren koroner arterleri tutan multisistemik bir vaskülittir. Bu olgu sunumunda, bilinen kardiyovasküler risk faktörü olmayan 19 yaşında genç bir kadın, ani başlayan göğüs ağrısı ile başvurdu. Elektrokardiyografide inferior derivasyonlarda ST elevasyonu izlendi ve akut miyokard enfarktüsü tanısıyla acil koroner anjiyografi yapıldı. Anjiyografide sağ koroner arterin distalinde total oklüzyon ve üç ana koroner arterin tamamında dev anevrizmalar saptandı. Yapılan perkütan girişimlere ve trombolitik tedaviye rağmen distal akım sağlanamadı. Altta yatan sistemik nedenleri araştırmak amacıyla geniş kapsamlı bir ayırıcı tanı süreci yürütüldü. Kawasaki hastalığı, sistemik lupus eritematozus, Takayasu arteriti, poliarteritis nodoza, immünoglobulin G4 ilişkili hastalıklar ve enfeksiyöz vaskülitler dışlandı. Mukokutanöz bulgular (aftöz

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ülserler, hiperpigmente plaklar) ile birlikte vasküler tutulumun varlığı, Uluslararası Çalışma Grubu kriterlerine göre Behçet hastalığı tanısına yol açtı. Hastaya altı ay boyunca intravenöz siklofosfamid uygulanıp sonrasında infliksimab ile idame tedavisi başlandı. Anevrizmaların yaygın ve difüz olması nedeniyle cerrahi girişim uygun bulunmadı. Oral antikoagülan tedavi uzun süre devam ettirildi. Dört yıllık takip süresince yeni kardiyak olay gelişmedi ve inflamasyon belirteçleri kontrol altında kaldı. Bu olgu, geleneksel risk faktörleri bulunmayan genç hastalarda akut koroner sendromla başvuran olgularda sistemik vaskülitlerin, özellikle Behçet hastalığının ayırıcı tanıda mutlaka göz önünde bulundurulması gerektiğini vurgulamaktadır. Erken tanı ve uygun immünosupresif tedavi, kardiyovasküler komplikasyonları önlemede kritik öneme sahiptir.

Anahtar Kelimeler: Behçet hastalığı, dev koroner anevrizma, akut koroner sendrom, vaskülit

INTRODUCTION

Behçet's disease is a chronic, multisystemic condition characterized by recurrent oral and genital ulcers, ocular involvement, and systemic vasculitis (1). While vascular involvement is more commonly seen in the venous system, arterial involvement may also occur and significantly increases the mortality rate. Coronary artery involvement is extremely rare (<0.5%) but can lead to serious clinical outcomes such as myocardial infarction (MI), arrhythmia, or sudden cardiac death (2). In this report, we present the case of a young female patient with giant coronary aneurysms who presented with acute coronary syndrome (ACS), was subsequently diagnosed with Behçet's disease following extensive evaluation, and was successfully managed through long-term follow-up.

Case Presentation

A 19-year-old woman with no known cardiovascular risk factors presented to the emergency department with sudden-onset chest pain. On arrival, she was hemodynamically stable. Electrocardiography (ECG) revealed ST-segment elevation in the inferior leads, consistent with acute inferior MI. Laboratory testing showed elevated troponin at 3500 ng/L (reference: 0-14 ng/L), while hemogram, renal and liver function tests were within normal limits.

Immediate coronary angiography revealed total occlusion of the distal right coronary artery (RCA) and giant, beaded aneurysms in all three major coronary arteries (Figure 1), with the proximal RCA measuring 12 mm in diameter. Balloon dilations (3,4,4.5 mm), failed to restore distal flow, and during the procedure, the patient experienced ventricular fibrillation. Due to persistent ischemic changes on ECG and malignant arrhythmias, intravenous thrombolytic therapy (tPA) was administered.

Follow-up coronary computed tomography (CT) angiography confirmed persistent distal RCA occlusion and marked aneurysmal dilatation in the RCA, left anterior descending (LAD), and circumflex (CX) arteries; each measuring 11-12 mm. Due to high thrombus burden in aneurysmal segments, dual antiplatelet therapy (aspirin + clopidogrel) and oral anticoagulation with warfarin were initiated, with international normalized ratio (INR) monitoring for optimal anticoagulation. Non-vitamin K oral anticoagulants (NOACs) were avoided because the thrombus burden was high and precise INR control was required.

Differential Diagnosis and Diagnostic Process

Extensive evaluation excluded Kawasaki disease, systemic lupus erythematosus, polyarteritis nodosa, Takayasu arteritis, and immunoglobulin G4-related disease. Dermatologic and

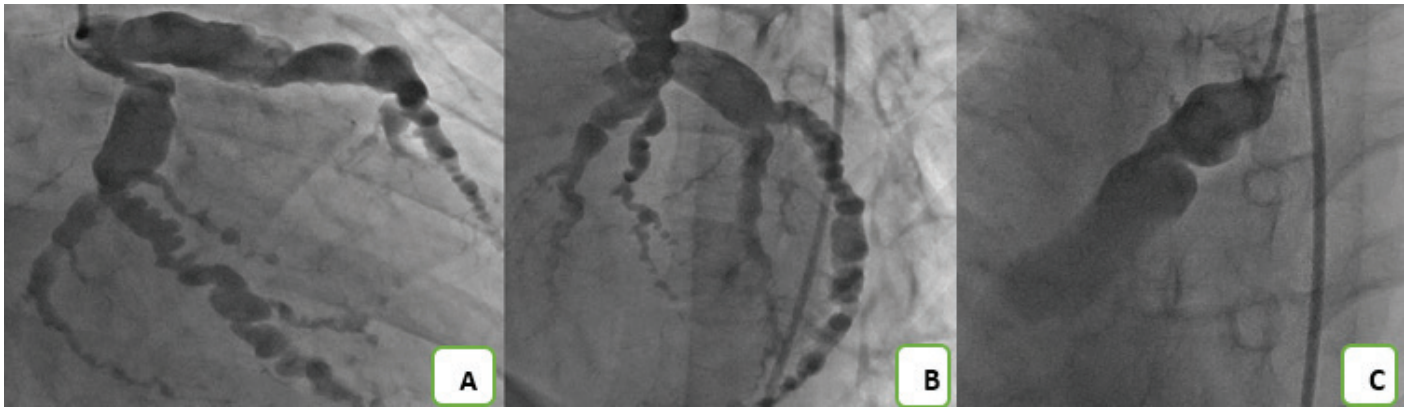

Figure 1. A) Image of giant saccular aneurysm in the right caudal region of LAD and CX coronary angiography. B) Image of aneurysm with thrombus in right cranial coronary angiography. C) RCA where the thrombus interrupts the flow
LAD: Left anterior descending artery, CX: Circumflex artery, RCA: Right coronary artery

rheumatologic assessments revealed two major aphthous ulcers on the upper gingiva, one minor ulcer on the lower lip mucosa, and post-inflammatory hyperpigmented plaques on the intergluteal region and inner thighs. Based on vascular involvement and mucocutaneous findings, Behçet's disease was diagnosed according to the International Study Group criteria (3).

Treatment and Follow-up

The patient received monthly intravenous cyclophosphamide (1,000 mg) for six months, followed by maintenance therapy with infliximab administered every three weeks. Surgical or graft-based interventions were deemed inappropriate due to the diffuse aneurysms and associated thrombotic risk.

For the first month, the patient was treated with a combination of aspirin, clopidogrel, and a vitamin K antagonist (VKA). After this initial period, dual antiplatelet therapy was discontinued, and she was followed by VKA monotherapy for one year.

Over a four-year follow-up, regular infliximab therapy was maintained. Follow-up CT angiography showed chronic RCA occlusion without progression in aneurysmal size or thrombus burden in the LAD and CX arteries (Figure 2). No new coronary events occurred, and C-reactive protein (CRP) levels remained within normal limits (Figure 3).

Detailed information was given to the patient regarding the possible contribution of the case report to the literature. The patient gave written and verbal consent for the publication of the case report.

DISCUSSION

Coronary involvement in Behçet's disease is extremely rare, particularly in young women without conventional cardiovascular risk factors. Giant coronary aneurysms should prompt considering systemic vasculitides (4,5). Interventional

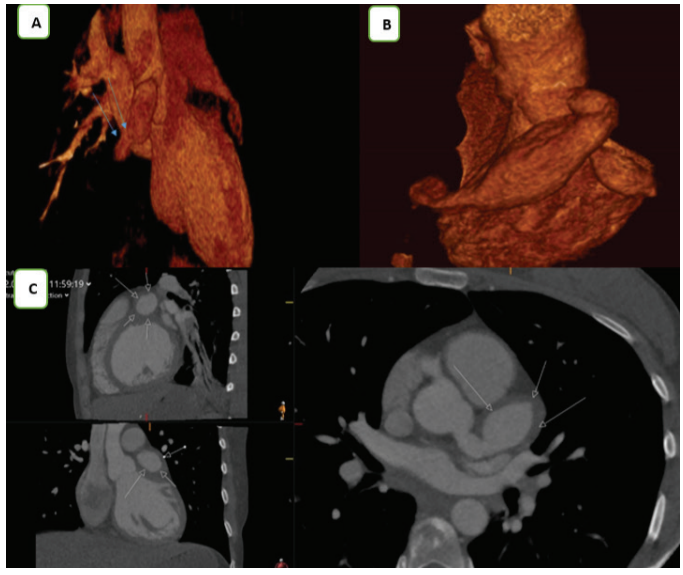

Figure 2: A) The 3D coronary CT angiography shows the aneurysmal and totally occluded image of the RCA, B) 3D Coronary CT angiography: aneurysmal left coronary system (LMCA-LAD and CX) C) The aneurysmal left system appearance with the LAD wall covered by thrombus, indicated by arrows
CT: Computed tomography, RCA: Right coronary artery, LMCA: Left main coronary artery, LAD: Left anterior descending artery, CX: Circumflex artery

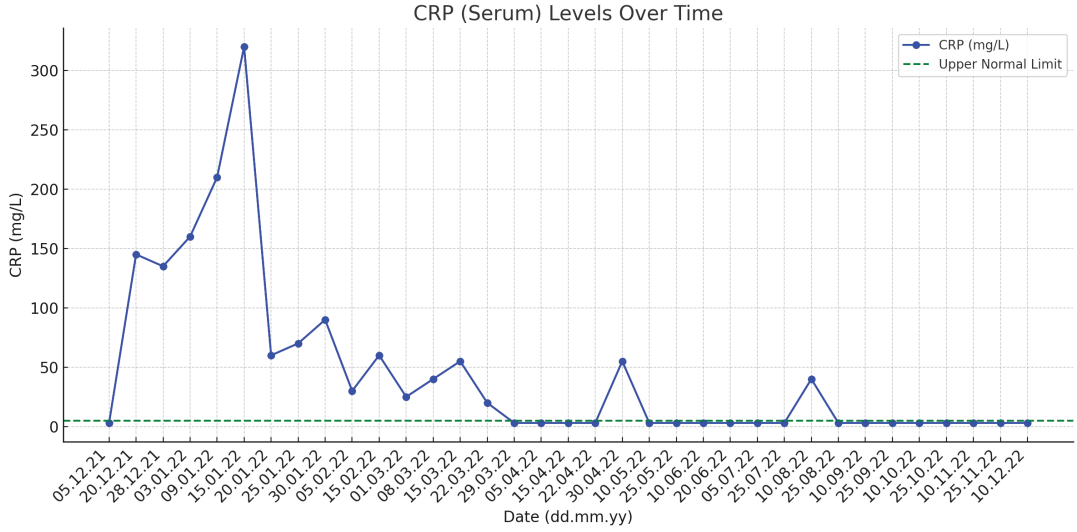

Figure 3. CRP monitoring after anti-inflammatory treatment
CRP: C-reactive protein

approaches such as percutaneous coronary intervention (PCI) or coronary artery bypass grafting carry high restenosis or graft failure rates without immunosuppressive therapy (6,7). In our case, medical management with immunosuppressants and anticoagulation achieved favorable outcomes, highlighting the importance of early recognition and treatment.

The decision to use tPA was based on persistent ST-segment elevation despite failed PCI and the occurrence of ventricular fibrillation, indicating ongoing ischemia. Warfarin was chosen over NOACs to allow precise INR monitoring in the context of high thrombus burden. CRP normalization served as a marker for effective inflammation control.

CONCLUSION

Systemic diseases must be considered in young patients presenting with ACS without traditional risk factors. Giant coronary aneurysms warrant evaluation for Behçet's disease. Early immunosuppressive therapy and tailored anticoagulation can prevent serious cardiac complications. Due to the rarity of such cases, treatment approaches are not fully generalizable.

*Ethic

Informed Consent: Detailed information was given to the patient regarding the possible contribution of the case report to the literature. The patient gave written and verbal consent for the publication of the case report.

Footnotes

Authorship Contributions

Surgical and Medical Practices: C.T., Ş.K., İ.D., Y.Y., Concept: C.T., Ş.K., İ.D., Y.Y., Design: C.T., Ş.K., İ.D., Y.Y., Data Collection or Processing: C.T., Ş.K., İ.D., Y.Y., Analysis or Interpretation: C.T., Ş.K., İ.D., Y.Y., Literature Search: C.T., Ş.K., İ.D., Y.Y., Writing: C.T., Ş.K., İ.D., Y.Y.

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