Surgical Repair of Carotid Artery Pseudoaneurysm and Stenosis in Behçet's Disease

Behçet Hastalığına Sekonder Karotid Arter Psödoanevrizması ve Stenozunun Cerrahi Onarımı

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ABSTRACT Carotid artery pseudoaneurysms are rare vascular lesions that may appear due to trauma, tumor invasion, radiation, infection, and arteritis. They should be treated as soon as possible in order to prevent rupture and neurological sequelae. Behçet's disease is one type of arteritis that typical involves medium and large vessels, causing arterial occlusive disease and aneurysm formation. We hereby report a patient with carotid artery pseudoaneurysm and stenosis due to Behçet's disease.

Key Words: Behcet syndrome; carotid stenosis; aneurysm, false

ÖZET Karotid arter psödoanevrizmaları; travma, tümör invazyonu, radyasyon, enfeksiyon ve çeşitli arteritlere sekonder gelişebilen nadir vasküler lezyonlardır. Rüptür veya nörolojik komplikasyonların ortaya çıkmasını önlemek için tanı konur konmaz tedavi edilmeleri gerekir. Behçet Hastalığı, tipik bulgusu tıkayıcı arter hastalığına ya da anevrizma gelişimine yol açan orta-büyük damar tutulumu olan bir patolojidir. Biz bu yazıda, Behçet Hastalığına sekonder karotid arter psödoanevrizması ve stenozu olan bir olgumuzu sunmaktayız.

Anahtar Kelimeler: Behçet sendromu; karotis stenozu; anevrizma, yalancı

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Carotid artery pseudoaneurysms (CPAs) are rare vascular lesions that may appear due to trauma, tumor invasion, radiation, infection and arteritis.[1] They usually appear as pulsatile neck masses, and may be fatal if hemorrhage or neurological deficits occurs. Behçet's disease (BD) is a chronic, relapsing inflammatory disorder that involves multiple organs including mucocutaneous, genital, ocular, skin, articular, gastrointestinal, neurological, and vascular systems. The incidence of vascular involvement ranges between 12.8 and 35.1%, and various vascular pathologies have been reported in patients with BD.[2,3] Vascular involvement tends to appear in males.[2,3] BD arteritis typically involves medium and large vessels, and results in arterial occlusive disease and aneurysm formation.[4] Those complications may be evident in 1-7% of BD patients, and 7.1% of the arterial complications involve carotid artery.[5-7] Arterial involvement is usually treated surgically. There is no clear consensus on the optimal timing of intervention; however, it was demonstrated that postponing surgical intervention until

remission may decrease the incidence of later vascular complications.[5,7] However this delay may lead to fatal complications. Herein, we report a rare case of left CPA and stenosis in a patient with BD who underwent urgent surgical intervention. This patient was successfully managed with surgery and postoperative immunosuppressive treatment.

CASE REPORT

A 29-year-old male admitted to our hospital with painless mass on the left side of his neck. His complaints had begun ten days before his admission. He had no history of neck trauma. He had been hospitalized for deep venous thrombosis and pulmonary thromboemboli (PTE) in another hospital one year ago, and he had been on warfarin for 6 months. During that hospitalization, he had been investigated for immunologic diseases, but no specific disease had been diagnosed. He was using only 100 mg acetylsalicylic acid on admission. He had no dyspnea, or swelling in the lower extremities.

His physical examination revealed an approximately 2x2cm sized, painless, pulsatile mass on the left side of the neck. There was no murmur on the carotid artery. His vital signs and other physical examination findings were normal. Color Doppler ultrasound of left carotid artery revealed a 2x2 cm CPA originating from the common carotid artery (CCA), and significant carotid stenosis. Computerized tomography (CT) showed 2x2 cm sized CCA pseudoaneurysm, and significant stenosis distal to the CPA (Figure 1). Post-contrast thoracic CT revealed a filling defect in the left inferior pulmonary artery due to chronic pulmonary emboli. Color Doppler ultrasound showed decreased diameter and irregular intraluminal thromboses compatible with the chronic stage in the left femoral vein. Partial recanalization was also well documented on color Doppler ultrasound. After hospitalization, the patient was investigated for immunological diseases. We found out that he had recurrent genital and oral ulcers. His ophtalmological examination was normal, but he had positive pathergy test. His complete blood count and biochemical parameters were normal. Laboratory tests showed normal erythrocyte sedimentation rate and C-reactive protein

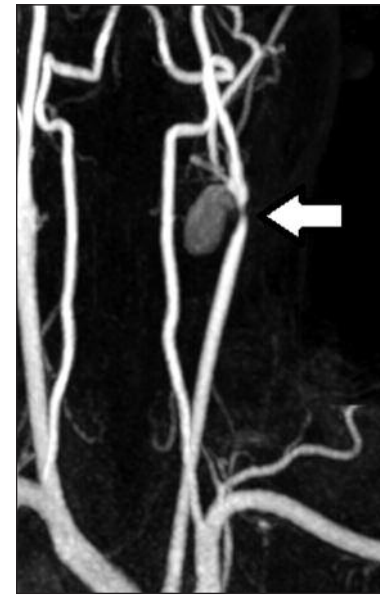

FIGURE 1: Computerized tomography showing carotid artery pseudoaneurysm and significantly stenotic segment of the artery.

level. All his immunologic markers were normal except for a positive lupus anti coagulant test. He was diagnosed with BD, and urgent surgical treatment was decided for CPA.

Surgery was performed under general anesthesia. A skin incision following the anterior border of the left sternocleidomastoid muscle was made. Left CCA was exposed, and isolated with tapes. After that, pseudoaneurysm was exposed and the nearby reactive lymph node was excised. External carotid artery (ECA) and internal carotid artery (ICA) were exposed, and isolated with tapes. After systemic heparinisation, ICA, ECA and CCA were clamped and the pseudoaneurysm sac was opened. The perforated segment of CCA was identified. After that, arteriotomy was extended towards the ICA and an inflammatory plaque that caused stenosis was exposed. Endarterectomy was performed, and the debris was washed out. Carotid patch repair was performed using a polyester carotid patch, sutured continuously with 6/0 polypropylene. We did not use any shunt in our case since he neither had a stenotic lesion in contralateral carotid artery nor a compromised flow in the circle of Willis.

Histopathological examination of the pseudoaneurysm wall and lymph node showed polymor-

phnucleated and mononucleated inflammatory cell infiltrations in the outer layer of artery, and desquamation of the endothelium. The patient had an uneventful postoperative course. He was discharged 3 days after surgery. His medical treatment included warfarin, aspirin, sulfasalazine and prednisolone. Control color Doppler ultrasonography was performed six months later, and showed patent left carotid artery.

DISCUSSION

Extracranial CPA is a rare condition that may result from trauma, infection, radiation and arteritis.[1] BD is an autoimmune, inflammatory, multisystemic disorder. It is characterized by recurrent oral and genital ulcers, and uveitis. Various immunological markers may be positive in BD.[8] Presence of the lupus anticoagulant, as in our case, was shown to be related with occlusive disease in BD.[8] Vessel involvement is not rare in BD, but approximately 95% of vascular involvement occurs in the venous system, and appear as thrombophlebitis and venous thrombosis.[9] Arterial involvement is rare, and although any artery has the potential to be affected, aorta, pulmonary artery and femoral arteries are the most commonly affected arteries.[9] Occlusive arterial disease is more common in femoral arteries, and aneurysm formation is more common in abdominal aorta, iliac arteries and pulmonary arteries.[10] Carotid artery aneurysm and stenosis are very rare in BD.[5-7]

There may be two mechanisms for pathogenesis of pseudoaneurysms in BD. The first one involves thinning of the tunica media, which precipitates rupture of the elastic lamina. The second one is vasculitis with lymphocytic infiltrations in the vasa vasorum. Those two mechanisms are also blaimed for occlusive disease in BD.[11]

The aim of treatment is to prevent aneurysmal rupture, and stroke due to emboli coming from the aneurysmal sac. Therefore, an aggressive treatment strategy including surgery and endovascular intervention is needed for management of such aneurysms. Surgery includes restoring blood flow with a bypass, or repairing with a patch. Endovascular intervention includes stent intervention. Kim et al. reported results of endovascular technique in treatment of arterial aneurysms. The endovascular approach may be a treatment option for arterial involvement associated with BD.[11]

Surgery has various technical difficulties. The inflammatory fibrous tissue makes isolation of the carotid artery difficult. The adhesions of the pseudoaneurysmal sac to the surrounding tissues may increase the risk of injury to the cranial nerves, including hypoglossal and vagus nerves during dissection.[12] There is also risk of injury to the intact artery, and clamping the carotid artery may cause diminished cerebral blood flow. An anastomotic pseudoaneurysm is a life-threatening surgical complication. Weakening of the arterial wall caused by fulminant inflammation may lead to anastomotic fragility. Endovascular intervention does not have such anatomic difficulties, but it carries the risk for emboli, access site pseudoaneurysm, or vascular rupture during procedure.[13] In addition, graft thrombosis is the most common complication of both methods.[13] The treatment strategy for an arterial aneurysm associated with BD is determined by the clinical presentation and the anatomical location. Rupture or impending rupture, and active or remission stage of the disease may affect outcome. The surgical outcomes of arterial involvements are highly influenced by vascular complications based on the severe inflammatory nature of this disease. Some authors recommended avoiding surgery in the active phase, and suggested aggressive immunosuppression as a key to prevent later pseudoaneurysm formation.[5-7]

We preferred surgical treatment in our patient because since he had not only pseudoaneurysm, but also significant stenosis. We chose a polyester carotid patch instead of autogenous graft for our patient. Akihiro and Köksoy reported that graft choice made no difference for graft thrombosis or pseudoaneurysm formation in BD.[6,14] However, some authors stated that prosthetic grafts should be preferred instead of vein grafts to prevent vascular complications since the major manifestation of BD is thrombophlebitis.[5,15]

Thus, there is no consensus regarding the most suitable graft material for arterial involvement associated with BD.

Patients with BD and arterial involvement should be administered a more aggressive treatment regimen in order to prevent occlusion of the vessels. Immunosuppressive therapy is shown to delay this outcome.[16] We discharged our patient with immunosuppressive therapy in the light of those data. Immunosuppressive medications seemed to control the inflammation associated with the disease, without vascular complications or progression of the concomitant peripheral aneurysms. Although the present case showed an unfavorable location of the pseudoaneurysm, he was fortunately in the remission stage.

In conclusion, BD should always be considered in young patients with aneurysms at unusual locations. Pulsatile neck masses without any history of chronic disorder or trauma should remind the possibility of CPA. We reported this case in order to call attention that occlusive disease might accompany pseudoaneurysms in BD. Immediate surgical or endovascular intervention is recommended to prevent a potentially fatal outcome.

Conflict of Interest

Authors declared no conflict of interest or financial support.

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