

## Surgical repair of carotid artery pseudoaneurysm and stenosis in Behçet's disease

Behçet hastalığında karotis arter psödoanevrizması ve darlığının cerrahi onarımı

Emrah Uğuz<sup>1</sup>, Muhammet Fethi Sağlam<sup>1</sup>, Emre Boysan<sup>2</sup>, Ünsal Erçelik<sup>1</sup>, Murat Canyığıt<sup>3</sup>, Mete Hidroğlu<sup>1</sup>, Erol Şener<sup>1</sup>

<sup>1</sup>Department of Cardiovascular Surgery, Atatürk Training and Research Hospital, Ankara, Turkey

<sup>2</sup>Department of Cardiovascular Surgery, Etimed Hospital, Ankara, Turkey

<sup>3</sup>Department of Radiology, Atatürk Training and Research Hospital, Ankara, Turkey

### ABSTRACT

Carotid artery pseudoaneurysms are rare vascular lesions which may occur due to trauma, tumor invasion, radiation, infection, and arteritis. They should be treated as soon as possible 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. Herein, we report a case of a carotid artery pseudoaneurysm and stenosis secondary to Behçet's disease.

**Keywords:** Aneurysm; Behçet's syndrome; carotid stenosis; false.

### ÖZ

Karotis arter psödoanevrizmaları; travma, tümör invazyonu, radyasyon, enfeksiyon ve arteritlere bağlı gelişebilen nadir vasküler lezyonlardır. Rüptür veya nörolojik sekelleri önlemek için mümkün olan en kısa sürede tedavi edilmeleri gerekir. Behçet hastalığı, tipik olarak orta ve büyük damarları tutan ve arteriyel tıkaçıcı arter hastalığına ve anevrizma gelişimine yol açan bir arterit türüdür. Bu yazıda, Behçet hastalığına sekonder karotis arter psödoanevrizması ve darlığı olan bir olgu sunuldu.

**Anahtar sözcükler:** Anevrizma; Behçet sendromu; karotis darlığı; yalancı.

Carotid artery pseudoaneurysms (CPAs) are rare vascular lesions that may occur due to trauma, tumor invasion, radiation, infection, and arteritis.<sup>[1]</sup> 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.<sup>[2,3]</sup> Vascular involvement tends to appear in males.<sup>[2,3]</sup> Behçet's disease arteritis typically involves medium and large

vessels, and results in arterial occlusive disease and aneurysm formation.<sup>[4]</sup> Those complications may be evident in 1 to 7% of BD patients, and 7.1% of the arterial complications involve carotid artery.<sup>[5-7]</sup> Arterial involvement is usually treated surgically. There is no clear consensus on the optimal timing of intervention; however, it has been demonstrated that postponing surgical intervention until remission may decrease the incidence of later vascular complications.<sup>[5,7]</sup> 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. The patient was successfully managed with surgery and postoperative immunosuppressive treatment.

*Received:* December 09, 2015 *Accepted:* February 18, 2016

**Correspondence:** Emrah Uğuz, MD. Atatürk Eğitim ve Araştırma Hastanesi Kalp ve Damar Cerrahisi Kliniği, 06800 Bilkent, Ankara, Turkey.  
e-mail: emrahuguz@gmail.com

### Citation:

Uğuz E, Sağlam MF, Boysan E, Erçelik Ü, Canyığıt M, Hidroğlu M, et al. Surgical repair of carotid artery pseudoaneurysm and stenosis in Behçet's disease. Damar Cer Derg 2018;27(1):51-54.

## CASE REPORT

A 29-year-old male patient was admitted to our hospital with a painless mass on the left side of his neck. His complaints began 10 days before his admission. He had no history of neck trauma. He was hospitalized for deep venous thrombosis and pulmonary thromboembolism (PTE) in another hospital one year ago, and he was on warfarin for six months. During hospitalization, he was further investigated for immunological diseases, although no specific disease was diagnosed. On admission, he was using only 100 mg acetylsalicylic acid. He had no dyspnea or swelling in the lower extremities.

His physical examination revealed an approximately 2×2 cm 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 the left carotid artery revealed a 2×2 cm CPA originating from the common carotid artery (CCA), and significant carotid stenosis. Computed tomography (CT) showed a 2×2 cm CCA pseudoaneurysm, and significant stenosis distal to the CPA (Figure 1). Post-contrast thoracic CT revealed a filling defect in the left inferior pulmonary



**Figure 1.** Computed tomography showing a carotid artery pseudoaneurysm and significantly stenotic segment of the artery.

artery due to chronic pulmonary embolism. Color Doppler ultrasound showed a decreased diameter with 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 re-investigated for immunological diseases. We found that he had recurrent genital and oral ulcers. Although his ophthalmological examination findings were normal, he had a positive pathergy test. His complete blood count and biochemical parameters were normal. Laboratory tests showed normal erythrocyte sedimentation rate and C-reactive protein level. All immunological markers were normal, except for a positive lupus anticoagulant test. He was diagnosed with BD, and urgent surgical treatment was decided for CPA.

A written informed consent was obtained from the patient. Surgery was performed under general anesthesia. A skin incision following the anterior border of the left sternocleidomastoid muscle was made. The left CCA was exposed, and isolated with tapes. Then, pseudoaneurysm was exposed and the adjacent reactive lymph node was excised. The external carotid artery (ECA) and internal carotid artery (ICA) were also exposed and isolated with tapes. After systemic heparinization, ICA, ECA, and CCA were clamped and the pseudoaneurysmal sac was opened. The perforated segment of the CCA was identified. Arteriotomy was, then, extended toward the ICA and an inflammatory plaque which 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 the contralateral carotid artery nor a compromised flow in the circle of Willis.

Histopathological examination of the pseudoaneurysm wall and lymph node showed polymorphonucleated 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 three days after surgery. His medical treatment included warfarin, acetylsalicylic acid, sulfasalazine, and prednisolone. Control color Doppler ultrasound showed a patent left carotid artery at six months.

## DISCUSSION

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

There may be two underlying mechanisms for the 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. These two mechanisms are also blamed for occlusive disease in BD.<sup>[11]</sup>

The main goal of treatment is to prevent aneurysmal rupture, and stroke due to embolism originating from the aneurysmal sac. Therefore, an aggressive treatment strategy including surgery and endovascular intervention is needed for management of such aneurysms. Surgery includes restoring the blood flow with a bypass, or repairing with a patch. Endovascular intervention includes stent intervention. Kim et al.<sup>[11]</sup> reported the results of endovascular technique in treatment of arterial aneurysms and indicated that endovascular approach might be a treatment option for arterial involvement associated with BD.

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.<sup>[12]</sup> There is also a 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, although it carries the risk for embolism, access site pseudoaneurysm, or vascular rupture during procedure.<sup>[13]</sup> In addition, graft thrombosis is the most common complication of both methods.<sup>[13]</sup> The treatment strategy for an arterial aneurysm associated with BD is based on 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.<sup>[5-7]</sup>

We preferred surgical treatment in our patient, since he had not only pseudoaneurysm, but also significant stenosis. We, therefore, chose a polyester carotid patch instead of an autogenous graft for our patient. Koksoy et al.<sup>[6]</sup> and Hosaka et al.<sup>[14]</sup> reported that graft choice made no difference for graft thrombosis or pseudoaneurysm formation in BD. However, some authors have proposed that prosthetic grafts should be preferred instead of vein grafts to prevent vascular complications, since the major manifestation of BD is thrombophlebitis.<sup>[5,15]</sup> 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 to prevent occlusion of the vessels. Immunosuppressive therapy has been shown to delay this outcome.<sup>[16]</sup> We discharged our patient with immunosuppressive therapy in the light of these data. Immunosuppressive medications seemed to control the inflammation associated with the disease without vascular complications or progression of the concomitant peripheral aneurysms. Although our 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 report this case to call attention that occlusive disease may accompany pseudoaneurysms in BD.

Immediate surgical or endovascular intervention is recommended to prevent a potentially fatal outcome.

#### Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

#### Funding

The authors received no financial support for the research and/or authorship of this article.

### REFERENCES

- Nadig S, Barnwell S, Wax MK. Pseudoaneurysm of the external carotid artery--review of literature. *Head Neck* 2009;31:136-9.
- Kural-Seyahi E, Fresko I, Seyahi N, Ozyazgan Y, Mat C, Hamuryudan V, et al. The long-term mortality and morbidity of Behçet syndrome: a 2-decade outcome survey of 387 patients followed at a dedicated center. *Medicine (Baltimore)* 2003;82:60-76.
- Fei Y, Li X, Lin S, Song X, Wu Q, Zhu Y, et al. Major vascular involvement in Behçet's disease: a retrospective study of 796 patients. *Clin Rheumatol* 2013;32:845-52.
- Posacioglu H, Apaydin AZ, Parildar M, Buket S. Large pseudoaneurysm of the carotid artery in Behçet's disease. *Tex Heart Inst J* 2005;32:95-8.
- Kalko Y, Basaran M, Aydin U, Kafa U, Basaranoglu G, Yasar T. The surgical treatment of arterial aneurysms in Behçet disease: a report of 16 patients. *J Vasc Surg* 2005;42:673-7.
- Koksoy C, Gyedu A, Alacayir I, Bengisun U, Uncu H, Anadol E. Surgical treatment of peripheral aneurysms in patients with Behçet's disease. *Eur J Vasc Endovasc Surg* 2011;42:525-30.
- Yang SS, Park KM, Park YJ, Kim YW, Do YS, Park HS, et al. Peripheral arterial involvement in Behçet's disease: an analysis of the results from a Korean referral center. *Rheumatol Int* 2013;33:2101-8.
- Pereira RM, Gonçalves CR, Bueno C, Meirelles Ede S, Cossermelli W, de Oliveira RM. Anticardiolipin antibodies in Behçet's syndrome: a predictor of a more severe disease. *Clin Rheumatol* 1989;8:289-91.
- Kuzu MA, Ozaslan C, Köksoy C, Gürler A, Tüzüner A. Vascular involvement in Behçet's disease: 8-year audit. *World J Surg* 1994;18:948-53.
- Ozyazicioğlu A, Koçak H, Vural U. Carotid artery pseudoaneurysm in Behçet's disease. *Eur J Cardiothorac Surg* 2001;19:938-9.
- Kim HO, Ji YB, Lee SH, Jung C, Tae K. Cases of common carotid artery pseudoaneurysm treated by stent graft. *Case Rep Otolaryngol* 2012;2012:674827.
- Kim DI, Huh SH, Do YS, Shin SW, Joh JH. Surgical experience of carotid pseudoaneurysm. *Yonsei Med J* 2003;44:905-7.
- Bonnotte B, Krause D, Fanton AL, Theron J, Chauffert B, Lorcerie B. False aneurysm of the internal carotid artery in Behçet's disease: successful combined endovascular treatment with stent and coils. *Rheumatology (Oxford)* 1999;38:576-7.
- Hosaka A, Miyata T, Shigematsu H, Shigematsu K, Okamoto H, Ishii S, et al. Long-term outcome after surgical treatment of arterial lesions in Behçet disease. *J Vasc Surg* 2005;42:116-21.
- Berard X, Corpataux JM, Taoufiq H, Sassoust G, Brizzi V, Midy D. Don't trust a vein graft to treat carotid aneurysm in patients with Behçet disease. *J Vasc Surg* 2010;52:471-4.
- Mochizuki M. Immunotherapy for Behçet's disease. *Int Rev Immunol* 1997;14:49-66.