COMMON CAROTID ARTERY ANEURSYM IN BEHÇET'S DISEASE
Behçet hastalığında arteria karotid kommunis anevrizması

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Summary: *Arterial aneurysms are among the most serious complications of Behçet's disease. Aneurysms due to Vasculo-Behçet's disease very seldomly involve carotid arteries. This complication has been reported in six cases in the literature so far . In these cases , the arteries have been prone to reccurrent pseudo-aneurysm development after vascular reconstructive procedures. Therefore alternative surgical techniques of repair have been suggested. We have successfully treated carotid artery aneurysm with resection and reconstruction in a young patient with Behçet's disease who developed hemiplegia and bilateral visual loss.*

Key Words: *Behçet's disease, Aneurysm*

Özet: *Arteriyel anevrizmalar Behçet hastalığının en ciddi komplikasyonlarındandır.Vaskülo-Behçet hastalığına bağlı anevrizmalar çok nadir olarak karotid arterleri tutar . Bu komplikasyon literatürde altı vakada rapor edilmiştir. Bu olgularda vasküler rekonstrüktif girişimlerden sonra yineleyen psödoanevrizma gelişimine eğilim vardır. Bundan dolayı onarımda alternatif cerrahi teknikler ileri sürülmüştür. Biz, hemipleji ve bilateral görme kaybı gelişen Behçet hastalıklı genç bir hastada oluşmuş karotid arter anevrizmasını anevrizmektomi ve rekonstrüktif vasküler girişimle başarıyla tedavi ettik.*

Anahtar Kelimeler: *Behçet hastalığı, Anevrizma*

Behçet's disease was first described in 1937 by Hulusi Behçet as a syndrome consisting of recurrent oral, genital ulcerations and iritis(1,2). Today it is considered as a chronic and systemic disease involving vascular, pulmonary , intestinal , urogenital , cardiac and neurologic systems (3).

A viral etiology is thought to play a role in the pathogenesis . However , this has not yet been proven. The diagnosis is based on the clinical manifestations and the detection of HLA-B5 antigen(1). Thrombophlebitis is a common initial presenting symptom (4). The arterial system is only occasionally affected resulting in thrombosis and aneurysmal changes of the aorta and peripheral arteries. The current case report belongs to a patient with Behçet's disease who presented with asymptomatic carotid artery aneurysm.

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Case Report: A 31- year - old male patient referred to Erciyes University Medical Faculty , Department of Otorhinolaryngology with the complaints of painful and rapidly enlarging mass in the right side of the neck and difficulty in swallowing . The patient had received the diagnosis of Behçet's disease nine years ago , and had been hospitalized for 2 months in the Neurology Clinic of the same hospital because of left sided hemiplegia two years ago .

On physical examination he had a pulsatile mass of 4x4x5 cm size in the right anterior cervical region with an audible bruit on auscultation. An aneurysm of the carotid artery was diagnosed by duplex ultrasonography (USG) , angiography and cervical computerized tomography (CT) (Figure 1,2). A vertical cervical incision was performed to explore the right common carotid artery and the aneurysm identified (Figure 3). Resection and interposition of an 8 mm polytetraflouroetylene (PTFE) graft was used (Figure 4). The histopathologic study of excised arterial specimen showed polymorphonuclear leu-

kocytes, erythrocytes, fibrin and nuclear remnants. Endothelial hyperplasia and inflammatory cellular infiltration rich in neutrophiles in the vascular wall were noted. Since no apparent postoperative complications were observed, the patient was discharged from the hospital on the fifth postoperative day .

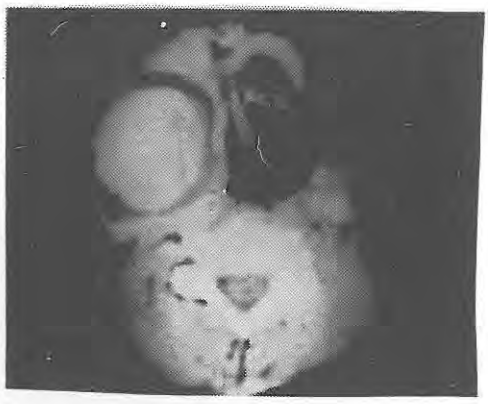

Figure 1. Cervical CT demonstrating right common carotid artery aneurysm

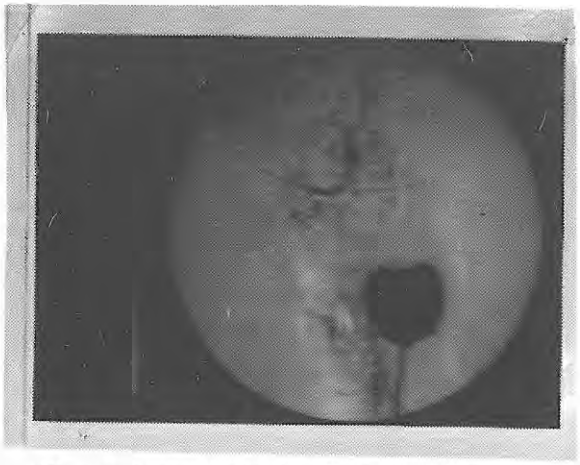

Figure 2. Angiography depicting right common carotid artery aneurysm and distal -proximal parts of the artery neurysm

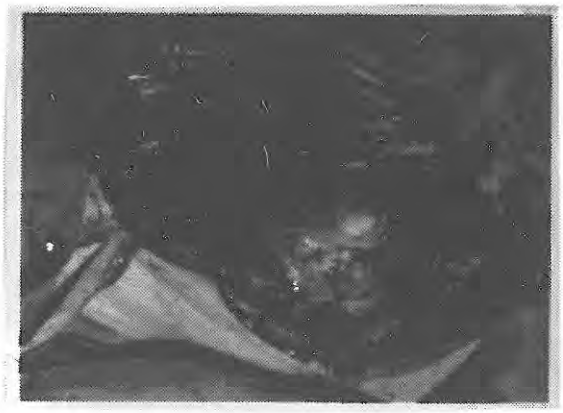

Figure 3. The view of the aneurysm after the exploration of peripheral tissue

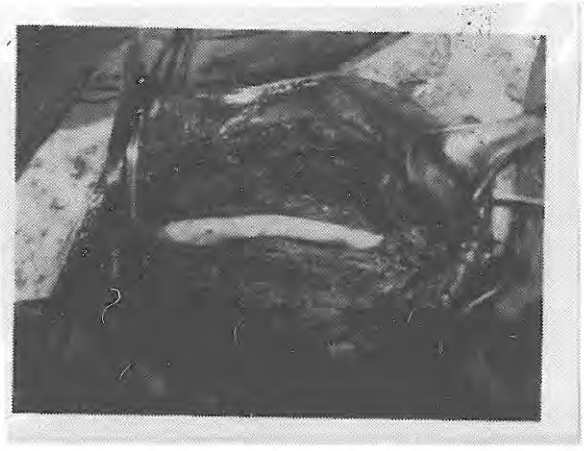

Figure 4. The interposition of a 8 mm PTFE graft after the excision of the common carotid artery

DISCUSSION

Behçet's disease is a clinical syndrome described by the Turkish dermatologist Hulusi Behçet in 1937. It has a worldwide distribution although the major series are still coming from the Mediterranean , middle East and Asian countries. Behçet's disease is most frequently seen in males between 20 and 40 years of age.

The most common pathologic lesion in Behçet's disease is a vasculitis with perivascular infiltration consisting of lymphomononuclear cells. Major symptoms of Behçet's disease are recurrent aphthous ulcers of the oral mucosa, ocular lesions, genital ulcers and skin lesions (erythema nodosum, folliculitis). Minor criteria include subcutaneous or deep venous thrombophlebitis, arterial occlusion and/or aneurysm, arthralgia , artritis, gastrointestinal, central nervous system and cardiovascular involvement.

Behçet's disease associated with vascular lesions is called vasculo-Behçet's disease.Vasculo-Behçet's disease occurred in 24.3% of a large series of patients with Behçet's disease from Turkey and 18.7 % of those from Japan. Vascular lesions can be classified into four types as arterial and / or venous occlusions, arterial aneurysm and varicose formations which are rarely seen.

Vascular involvement which is a minor criterion of Behçet's disease is particularly seen in venous system (1, 5). These lesions look like superficial or deep venous thrombophlebitis. In venous occlusions the prognosis is favorable due to the development of collateral veins and recanalization. Most authors agree that arterial and neurologic lesions occur at the late stages of the disease and particularly in young men(4). These lesions include arterial occlusions and / or aneurysms and A-V fistulas which are discovered as a result of arteriographic studies and are thought to occur in 1.5 to 2 % of the cases (4).

Arterial aneurysms commonly have atherosclerotic, postoperative and traumatic origins. Local infections, syphilitic aneurysms, Behçet's disease and congenital pathologies are less common causes of arterial aneurysms .

Aneurysms due to Behçet's disease are true or false, fusiform or saccular.The most freqently involved arteries are femoral , popliteal , tibial , brachial , iliac and subclavian arteries whereas the involvement of carotid arteries, aortic arch, descending and abdominal aorta and pulmonary arteries are rarely reported (6) . The prognosis of aorta, pulmonary artery and carotid artery aneurysms is poor (4,7). Death occurs as a result of the rupture of the aneurysm (2). Operative mortality is also high in pulmonary and aortic aneurysms. Early diagnosis and early surgical intervention frequently decreases the likelihood of the development of these complications.

Development of an aneurysm in Behçet's disease is thought to be due to medial degeneration , vasculitis of the vasovasorum, rupture of internal and external elastic lamina with thickening of the intima and with perivascular lymphocyte infiltration (5). Immunoglobulins (IgA, IgG, IgM) and C 3 , C 4 deposits on arterial wall indicate that these pathologic changes are correlated with an immunologic phenomenon (6)

The arterial aneurysm in this case was a true , saccular aneurysm. Clinical experience suggests that arterial lesions are usually seen within 8-20 years after initial diagnosis as our case .

The relatively high risk of aneurysm rupture and embolization from mural thrombus may indicate reconstructive surgery.

The treatment of the aneurysm due to Behçet's disease is controversial (8). A common idea of the authors for the surgical correction is bypass grafting on the intact artery and extra-anatomic bypassing in cases of recurrence (3, 9, 10).

Some of the surgical techniques employed include : aneurysmography (5) , arterial support by prosthetic wrapping , ligation (7) and embolization in some aneurysms before surgery.

Reported recurrences are seen in 25 % of cases after these surgical interventions (8). For this reason we have preferred to use PTFE grafts and / or patch grafting for the interpositon of autogenous vein grafting for surgical correction. In our case an 8 mm PTFE tube graft was used.

In the postoperative period ,the risk of anastomotic aneurysm development is high. Therefore , a long period of follow- up is necessary.

Behçet's disease is an uncommon condition that is

managed primarily by medical therapy. However, surgical intervention is necessary when life-threatening vascular complications occur.

Because of a potential lethal nature of the vascular problems associated with Behçet's disease , physicians should have a high index of suspicion for these problems in patients with longstanding history of this disease.

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