Behcet’s disease was first described by Hulusi Behcet in 1937.\textsuperscript{1,2} It is an autoimmune multisystemic disease, characterized by vasculitis, presenting with chronic relapsing course of oral and genital ulcers, ocular lesions and venous or arterial manifestations. Etiology of the disease is still unknown. The most important predictor of morbidity and mortality is the vascular complications. Vascular complications are mainly associated with thrombosis and pseudoaneurysm formation. It is not always easy to decide for the best treatment option, surgical or medical. In this report, a case with recurrent aneurysmal lesions, attacks of deep venous thrombosis and ruptured iliac pseudoaneurysm due to active Behcet’s disease, is presented. In the treatment of the patient, the implantation of an endovascular stent graft as an alternative to direct surgery to reduce the risk of subsequent pseudoaneurysm formation is discussed.

**Key Words:** Behcet syndrome; aneurysm

**ABSTRACT**

Behcet’s disease is an autoimmune multisystemic disease, characterized by vasculitis, presenting with chronic relapsing course of oral and genital ulcers, ocular lesions and venous or arterial manifestations. Etiology of the disease is still unknown. The most important predictor of morbidity and mortality is the vascular complications. Vascular complications are mainly associated with thrombosis and pseudoaneurysm formation. It is not always easy to decide for the best treatment option, surgical or medical. In this report, a case with recurrent aneurysmal lesions, attacks of deep venous thrombosis and ruptured iliac pseudoaneurysm due to active Behcet’s disease, is presented. In the treatment of the patient, the implantation of an endovascular stent graft as an alternative to direct surgery to reduce the risk of subsequent pseudoaneurysm formation is discussed.

**Key Words:** Behcet syndrome; aneurysm

**ÖZET**


**Anahtar Kelimeler:** Behçet sendromu; aneurizma

**Tüm Yazarlar:**

Soner YAVAŞ, MD,\textsuperscript{a} Fahrettin KÜÇÜKAY, MD,\textsuperscript{b} Ahmet Tulga ULUS, MD\textsuperscript{a}

Departments of
\textsuperscript{a}Cardiovascular Surgery\textsuperscript{b}Radiology, Türkiye Yüksek İhtisas Training and Research Hospital, Ankara

Geliş Tarihi/Received: 18.12.2008
Kabul Tarihi/Accepted: 23.01.2009

Yazışma Adresi/Correspondence:
Soner YAVAŞ, MD
Türkiye Yüksek İhtisas Training and Research Hospital, Department of Cardiovascular Surgery, Ankara, TÜRKİYE/TURKEY
dsoneryavas@yahoo.com.tr

**Copyright © 2009 by Türkiye Klinikleri**

**Is Endovascular Intervention for the Treatment of Pseudoaneurysm in Active Behcet’s Disease with Arterial and Venous Involvement, Successful?: Case Report**

Aktif Dönemdeki Arteriyel ve Venöz Tutulumlu Behçet Hastalığında, Yalanç Anevrizma Tedavisinde Endovasküler Girişim Başarılı mıdır?

**OLGU SUNUMU**

**Is Endovascular Intervention for the Treatment of Pseudoaneurysm in Active Behcet’s Disease with Arterial and Venous Involvement, Successful?: Case Report**

Aktif Dönemdeki Arteriyel ve Venöz Tutulumlu Behçet Hastalığında, Yalanç Anevrizma Tedavisinde Endovasküler Girişim Başarılı mıdır?

**Is Endovascular Intervention for the Treatment of Pseudoaneurysm in Active Behcet’s Disease with Arterial and Venous Involvement, Successful?: Case Report**

Aktif Dönemdeki Arteriyel ve Venöz Tutulumlu Behçet Hastalığında, Yalanç Anevrizma Tedavisinde Endovasküler Girişim Başarılı mıdır?

**Is Endovascular Intervention for the Treatment of Pseudoaneurysm in Active Behcet’s Disease with Arterial and Venous Involvement, Successful?: Case Report**

Aktif Dönemdeki Arteriyel ve Venöz Tutulumlu Behçet Hastalığında, Yalanç Anevrizma Tedavisinde Endovasküler Girişim Başarılı mıdır?

**Is Endovascular Intervention for the Treatment of Pseudoaneurysm in Active Behcet’s Disease with Arterial and Venous Involvement, Successful?: Case Report**

Aktif Dönemdeki Arteriyel ve Venöz Tutulumlu Behçet Hastalığında, Yalanç Anevrizma Tedavisinde Endovasküler Girişim Başarılı mıdır?

**Is Endovascular Intervention for the Treatment of Pseudoaneurysm in Active Behcet’s Disease with Arterial and Venous Involvement, Successful?: Case Report**

Aktif Dönemdeki Arteriyel ve Venöz Tutulumlu Behçet Hastalığında, Yalanç Anevrizma Tedavisinde Endovasküler Girişim Başarılı mıdır?
complications. Vascular complications are mainly associated with thrombosis and pseudoaneurysm formation, which makes it very difficult to decide for the best treatment option, surgical or medical.

A case with recurrent aneurysmal lesions and venous thrombosis due to active Behçet’s disease and the implantation of an endovascular stent graft as an alternative to direct surgery to reduce the risk of subsequent pseudoaneurysm formation, is presented.

CASE REPORT

In January 2008, a 39 years old male was admitted to the hospital with the suspicion of perforated abdominal aorta aneurysm. Previously, he was operated for carotid artery pseudoaneurysm in 2005. He had some of major and minor symptoms of Behçet’s disease and had a history of recurrent oral ulcerations (2-3 times a week), inflammatory skin lesions and deep venous thrombosis (DVT) attacks, and described genital ulceration for only one time. His paternity test was negative and free of ophthalmic involvement. He also had a history of chronic obstructive pulmonary disease. He was under medical treatment for being in active phase and was receiving warfarin, colchicine, atorvastatin, metoprolol, acetyl salicylic acid, clopidogrel, azathioprine therapy.

Duplex ultrasound revealed total occlusion of right common, internal and external carotid arteries. Transthoracic echocardiography was normal. Abdominal ultrasonography reported a 60 x 40 mm thrombus formation around aortic bifurcation and an aortic aneurysm with a luminal diameter of 30 x 46 mm, located below renal arteries. Subsequent thoracic computed tomography (CT) demonstrated ascending, arcus and descending aorta to be in normal diameters. Abdominal aorta angio-CT confirmed the presence of a ruptured iliac pseudoaneurysm (65 mm in diameter), but no abdominal aortic aneurysm.

The patient was transferred to the angiography unit and the exact location of the pseudoaneurysm was confirmed (Figure 1). A covered stent (10 x 60 mm) (Boston Scientific Ireland Limited, Galway, Ireland) was placed in right iliac artery pseudoaneurysm. Control angiography demonstrated technically successful insertion and no residual extravasation (Figure 2).

Control aortic angio-CT was performed on the 15th day and demonstrated that the stent was patent without any stenosis, extravasation or pseudoaneurysm.

On the postoperative 45th day, arterial duplex ultrasonographic examination of the lower extremities revealed triphasic normal flow patterns.

Eleven month follow-up of the patient is uneventful.
DISCUSSION

Vascular Behçet’s disease is rare and presents approximately 14% venous and 1.6% arterial lesions, occurring in various locations with multiple clinical expressions. If multiorgan involvement is exist, cardiac event incidence will increase to a level between %7 and %29. The most common lesions are superficial thrombophlebitis and deep venous thrombosis of the extremities followed by the involvement of vena cavae and suprahepatic veins. When Behçet’s disease is associated with large arteries, mainly the aorta and iliac arteries, lesions are usually in the form of aneurysms or occlusions. Aneurysms have worse prognosis than occlusive lesions. They have a sudden onset and are prone to progress rapidly to a pseudoaneurysm in many cases and often resulting with rupture.

In patients with Behçet’s disease, vascular assessment should be done routinely for early diagnosis and treatment. When an aneurysm has been found in a patient with Behçet’s disease, the patient should be scanned for possible multiple aneurysms. All types of arterial punctures for angiography or blood gases should be minimized because of the risk of new aneurysm formation.

Best medical treatment for Behçet’s disease is still unknown. The treatment modalities today include azathioprine, aspirin and anticoagulation for deep vein thrombosis or prednisolone and cyclophosphamide for arterial disease.

Surgical treatment of arterial lesions associated with Behçet’s disease is difficult because of the aortic wall fragility and often complicated by graft occlusion or early occurrence of anastomotic pseudoaneurysm which is characteristic of the disease. Surgical treatment should be avoided, if possible, in the acute phase of the disease and systemic therapy should be considered for patients.

The establishment of remission before surgery will decrease the incidence of postoperative complications which is sometimes is not possible because of the emergent nature of these problems. An aggressive surgical approach can be life-saving in such patients. Early and late results can be improved by individualizing the risks for the patients and selecting a disease-free area for reconstruction.

Surgery, when feasible, is indicated for aneurysms due to the fact of high risk of rupture, but must be limited only for the patients presenting with a growing aneurysm, acute rupture or severe ischaemia. Postoperative corticosteroids and/or immunosuppressive therapy and/or anticoagulation should be considered to prevent arterial relapse or graft thrombosis.

Iscan and coworkers, in their series of 20 patients operated in our hospital, concluded that, after the initial operation 10 year survival rate was 30%, 10 year complication-free survival rate was 13%, and 5 year repeat operation-free survival rate was 26%.

Endovascular repair seems to be a promising alternative treatment of aneurysmal manifestations in Behçet’s disease. The endovascular approach can be used during the inflammatory phase. Implantation of an endovascular stent graft is also an alternative modality in the treatment of pseudoaneurysms with previously well described advantages such as avoidance of general anesthesia and surgical dissection can resulting with shorter hospitalizations, thus diminishing morbidity and will have predictable outcome in high-risk groups.

In this case we preferred implantation of an endovascular stent graft via femoral artery because if a femoral artery pseudoaneurysm occurs at the graft insertion site, repair of it will be easier than an abdominal or iliac artery anastomotic pseudoaneurysm.

Because of our knowledge about the disease is limited mainly with case reports in Behçet’s disease with vascular involvement, a note of caution should be kept in mind, since the inflammatory response to stent-grafts is yet to be clearly defined.
REFERENCES


