

# SURGICAL MANAGEMENT OF ARTERIAL ANEURYSMS IN BEHCET'S DISEASE

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*Objective: We present our experience of Behcet's disease patients with arterial aneurysms.*

*Methods: All patients had diagnosis of Behcet's disease beforehand. 3 of them were referred for arterial aneurysm and 1 for coronary artery disease. Patients were analysed retrospectively for their demographic characteristics, operative and postoperative courses.*

*Results: Average of patients' ages were 35,2±6,4. They were followed-up for 4,5-10,4 (mean 7,8) years. Patients were operated on elective basis except one with the suspicion of aneurysm rupture.*

*Conclusions: Aneurysmal involvement of arteries may dictate an emergency operation. No-touch technique should be preferred in order to prevent progression of a possible aneurysm.*

*Key words: Behcet's disease, Behcet's aneurysm, surgical treatment*

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**B**ehcet's Disease, which was initially described in 1937, is a systemic disorder characterised by urogenital ulcerations, chronic eye inflammation and skin lesions<sup>1</sup>. The etiology is unknown. Although viruses, streptococcal infections, autoimmune mechanisms and endothelial cell dysfunction have been postulated in the pathogenesis, no definite cause has been identified. The diagnosis depends on identification of several of its typical features. Six different criteria sets have been described for the diagnosis of the Behcet's disease<sup>2</sup>. Although Behcet's disease has a worldwide distribution, individuals of Eastern Mediterranean and Far East descent have the highest incidence. Behcet's Disease affects most often in the third or fourth decades of life, more frequently in males and males express more rapid clinical progress than females<sup>2,3</sup>. In Behcet's Disease; locomotor, central nervous and gastrointestinal systems are also involved. Vascular system is occasionally involved and called "Vasculo-Behcet" and recognized since 1946 which is mostly the thrombosis of the veins.

Arterial involvement is an uncommon complication of Behcet's Disease characterised by saccular aneurysms or occlusions of multiple large vessels in young adults<sup>4,5</sup>. Arterial involvement frequently affects the abdominal aorta followed by the femoral, pulmonary, brachiocephalic and the visceral arteries<sup>2</sup>. Many arterial complications have been recognised for Behcet's Disease, but rupture of abdominal aortic aneurysm is one of the most severe.

We herein present the surgical treatment of the abdominal aortic and the femoral artery aneurysms in 4 patients with known Behcet's disease.

## METHODS

From 1985 till the end of 2003, we operated 4 patients with Behcet's disease who had aneurysm and pseudoaneurysm. Patients' demographic characteristics, the operative measures and the follow-ups are analysed.

## RESULTS

All patients were male. The average of their ages were  $35,2 \pm 6,4$ . Patients were followed up for 4,5 to 10,4 years (mean 7,8 years). Medical condition of the patients and the operations are listed in Table I.

**Table I:** Vascular involvement and treatment

	Age	Diagnosis	Treatment
Case 1	40	CAD	Medical
		AAA	Graft interposition
Case 2	36	AAA	Graft interposition
Case 3	39	Ruptured AAA	
		Vertebral coloumn destruction	Graft interposition
Case 4	26	CAD	Medical
		CABG (LIMA-LAD, Ao-D1)	
		Right femoral artery pseudoaneurysm	Primary treatment
		Ascending Aortic aneurysm	Primary treatment
		Left femoral artery aneurysm	Primary treatment

CAD: Coronary artery disease

AAA: Abdominal aortic aneurysm

LIMA: Left internal mammary artery

LAD: left anterior descending

branch of Left coronary artery Ao:aorta

D1: First diagonal branch of LAD

Patients with abdominal aortic and femoral artery aneurysms had an uneventful postoperative course. Patient with coronary artery disease, after an uneventful initial course and discharge, he was diagnosed to have a pseudoaneurysm of the ascending aorta and the saphenous vein graft was found to be occluded<sup>6</sup>.

## DISCUSSION

Vascular involvement is the leading cause of death in Behcet's disease with an approximate prevalence of 25%<sup>7</sup> and is seen more frequently in males (36% vs 14%)<sup>8</sup>. The vasculitis may involve large, medium and small vessels of the arterial and venous circulation. There is a tendency to thrombus formation with thrombi in the lumen of the vessels showing features of inflammation and focal areas of lymphocytes. Three forms of vascular disease are found in BD: Venous occlusions, arterial aneurysms and/or arterial occlusions. Venous occlusions occur more frequently than the arterial lesions. (88% vs 12%)<sup>7</sup>. Superficial thrombophlebitis is the most common disorder. Symptoms of vascular disease vary depending on the sites of involvement<sup>2,3,9,10</sup>. Venous disorders with higher morbidity like deep venous thrombosis, (DVT) and arterial aneurysms may appear, even in the lungs. Aneurysm and/or occlusion of the large arteries of the upper (axillary artery) or lower extremities (femoral artery) are the most common types of arterial lesions.

Femoral pain, intermitant claudication and avascular necrosis of the head of femur may occur due to either the aneurysm or occlusion of the femoral artery. Involvement of the common carotid artery may result in development of hemiplegia. Ruptures of large arteries may lead to death.

Vasculo-Behcet disease is a type of BD with clinical features based on vasculitis of the arter-

ies or deep veins, and is frequently life threatening<sup>11</sup>. Early diagnosis and treatment are essential for the management of vasculo- Behcet's disease. Recent advances in the diagnosis of the vasculo- Behcet's disease have been brought about by the use of radionuclide venography for DVT and CT (computerised tomography) for arterial lesions. In diagnosis of the aneurysms, we performed CT in 2 and MRI (magnetic resonance imaging) in 1 patient. In the treatment of vasculo- Behcet's disease, anticoagulants and the fibrinolytic agents are beneficial for the control of DVT.

Operative therapy is often recommended for the management of arterial aneurysms, because rupture of an arterial aneurysm is the leading cause of death in patients with BD. We have presented a successful repair of ruptured abdominal aneurysm before<sup>12</sup>.

Angiography and CT are the methods commonly used to evaluate cardiovascular involvement, but they carry risk for complications. Venous puncture, intravenous infusion, rapid injection of a large bolus of contrast media and insertion of a venous catheter may initiate venous thrombosis in BD<sup>13,14</sup>. Venipuncture for any reason, even in the absence of a contrast injection, may increase the risk for venous thrombosis. An increased incidence of aneurysm formation at the puncture site has been reported after venography and arterial puncture<sup>9,15</sup>. Therefore, it is desirable to avoid intravenous injections, arteriography and venography in these patients. Doppler ultrasonography, MRI and MR angiography are safe and noninvasive methods that can be used to confirm and monitor cardiovascular involvement in BD. Digital subtraction angiography has also been used in the diagnosis but it may be inadequate if the aneurysms or vessels are completely thrombosed. The prognosis of aneurysms is worse than that of

occlusive lesions<sup>3</sup>.

Current treatment is tailored according to the site and severity of BD. Since BD usually runs an undulating course of exacerbations and remissions, it is generally difficult to evaluate the efficacy of the therapy. Treatment of BD is symptomatic and empirical and varies according to the clinical manifestations. The mainstay of treatment in BD is immunosuppressive therapy as in other severe forms of vasculitis. Other treatment modalities should be used in combination with this therapy and as palliative measures for specific complications. Although definitive data do not exist, there is a tendency among physicians to prescribe colchicine to all patients<sup>14</sup>. Thalidomide, simple analgesics, antiinflammatory agents, interferon- $\gamma$ , prophylactic benzathine penicilline, azathioprine and cyclosporine are treatment options for other manifestations<sup>3,15,16</sup>. Many papers stress the importance of antiinflammatory therapy as an adjuvant to surgery<sup>17,18</sup>. A controlled, long term study regarding to the treatment of vascular BD has not yet been reported.

Surgical therapy of BD always challenges cardiovascular surgeons with a high frequency of complications. Aneurysms limited to the extremities could be ligated without disabling ischemia. Tube graft insertion is suggested for the abdominal aortic aneurysms<sup>19</sup>. Surgery, when feasible, is indicated for aneurysms because they entail a high risk of rupture. The main problem facing the vascular surgeon is the 25% incidence of recurrent anastomotic aneurysms after both inlay graft repair and patching. Appropriate operative procedures, including an adequate choice of anastomotic sites and reinforcement of the suture, may reduce the incidence of complications in patients with arterial aneurysms. Postoperative corticosteroids and/or immunosuppressives are

necessary to prevent arterial relapse. After bypass for lower limb arterial lesions, anticoagulation is warranted to prevent graft thrombosis<sup>20</sup>.

## REFERENCES

1. Behçet H. Über rezidivierende Aphthose, durch ein Virus verursachte Geschwüre am Mund, am Auge und an den Genitalien. *Dermatol Wochenschr* 1937; 105: 1152-7.
2. Yurdakul S, Yazıcı H. Management of aneurysm in Behçet's Syndrome: an analysis of 24 patients. *Surgery* 1997; 121: 1501-6
3. Matsumoto T, Uekusa T, Fukuda Y. Vasculo-Behçet's Disease: a pathologic study of eight cases. *Hum Pathol* 1991; 22: 45-51
4. Wechsler B, Lee Thi Huong Du LT, Gennes C et al. Arterial manifestations of Behçet's Disease. 12 cases. *Rev Med Interne* 1989; 10: 303-311
5. Wechsler B, Du LT, Kieffe E. Cardiovascular manifestations of Behçet's disease. *Ann Med Interne* 1999; 150: 542-554
6. İpek G, Ömeroğlu SN, Mansuroğlu D, Kıralı K, Uzun K, Şişmanoğlu M. Coronary artery bypass grafting in a 26-year-old man with total occlusion of the left main coronary artery related to Behçet's disease. *J Thorac Cardiovasc Surg* 2001; 122: 1247-9
7. Sasaki SH, Yasuda K, Takigami K, Shiiya N, Matsui Y, Sakuma M. Surgical experiences with peripheral arterial aneurysms due to vasculo-Behçet's Disease. *J Cardiovasc Surg (Torino)* 1998; 39: 147-150
8. Lakhanpal S, Tani K, Lie JT, Katoh K, Ishigatsubo Y, Ohokubo T. Pathologic features of Behçet's Syndrome: a review of Japanese autopsy registry data. *Hum Pathol* 1985; 16: 790-5
9. Koç Y, Güllü İ, Akpek G et al. Vascular involvement in Behçet's disease. *J Rheumatol* 1992; 19: 402-10
10. Güler M, Kıralı K, Erentuğ V, Balkanay M, Gürbüz A, Yakut C. Behçet Hastalığında Kolumna vertebraliste destrüksiyona yol açan abdominal aort anevrizması ve cerrahi tedavisi. *Damar Cer Derg* 1998; 3: 155-57
11. Sasaki Sh, Yasuda K, Takigami K, Shiiya N, Matsui Y, Sakuma M. Surgical experiences with peripheral arterial aneurysms due to vasculo-Behçet's disease. *J Cardiovasc Surg (Torino)* 1998; 39: 147-50
12. Erentuğ V, Bozbuğa N, Ömeroğlu SN, Ardal H, Eren E, Eevli MG et al. Rupture of abdominal aortic aneurysms in Behçet's disease. *Ann Vasc Surg* 2003; 17: 682-685
13. Berkmen T. MR angiography of aneurysms in Behçet's disease: a report of four cases. *J comput Assist Tomogr* 1998; 22: 202-6
14. Tunacı A, Berkmen YM, Gökmen E. Thoracic involvement in Behçet's disease: pathologic, clinical and imaging features. *Am J Roentgenol* 1995; 164: 51-6
15. Yazıcı H, Barnes CG. Practical treatment recommendations for pharmacotherapy of Behçet's Syndrome. *Drugs* 1991; 42: 796-804
16. Çalgüneri M, Ertenli İ, Kiraz S, Erman M, Çelik I. Effect of prophylactic benzathine penicilline on mucocutaneous symptoms of Behçet's disease. *Dermatology* 1996; 192: 125-8
17. Barlas S. Behçet's disease. An insight from a vascular surgeon's point of view. *Acta Chir Belg* 1999; 6: 274-81
18. Moro H, Hayashi J et al. Surgical management of cardiovascular lesions caused by systemic inflammatory diseases. *Thorac Cardiovasc Surg* 1999; 47(2): 106-110
19. Tüzün H, Besirli K, Sayın A et al. Management of aneurysms in Behçet's syndrome: an analysis of 24 patients. *Surgery*

1997; 121: 150-6

20. Le Thi Huong D, Wechsler B, Papo T et al.  
Arterial lesions in Behcet's disease. A study in  
25 patients. J Rheumatol 1995; 22: 2103-13