

Abdominal aneurysms in Behçet's Disease

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Behçet's Disease (BD) is an uncommon systemic disorder affecting the skin, mucous membranes, eye, joints, central nervous system and the blood vessels. We report two cases of abdominal aneurysm in patients with BD and review the literature.

Case 1

A 39-year-old Saudi man, known to have multiple sclerosis and BD, previous DVT of the left lower limb, and no risk factors for cardiovascular disease, complained of abdominal pain radiating to the left side of the back in the previous 6 months. The abdomen was soft and lax, with no tenderness, and pulses were present in all the peripheral arteries. He had old scars on the scrotum and in the mouth, his ESR was high (48 mm/h), and ultrasound (US) of the abdomen showed a gallbladder stone and abdominal aneurysm. Spiral CT angiography confirmed the presence of an infrarenal saccular abdominal aortic aneurysm (AAA) with inflamed walls (Figure 1). The echocardiogram was normal. The patient underwent resection of the AAA and an aorto-bi-iliac bypass using a bifurcated dacron graft. Histology showed acute and chronic inflammation. Follow-up duplex scan of the abdominal aorta was conducted every 6 months. The last one, 2 years after the operation, showed no pseudoaneurysm or occlusion/stenosis.

Case 2

A 44-year-old Saudi man, known to have recurrent oral and scrotal ulcers presented with abdominal pain radiating to the back. The abdominal examination discovered a small pulsatile mass in the left side of the umbilicus. The pulses were palpated on the peripheral arteries. The ESR was high (59 mm/h) and ANA was positive (1:80). Abdominal US revealed an infrarenal AAA measuring 55 x 48 mm. CT angiography and angiography proved the presence of a saccular AAA starting 8 cm below the renal arteries and an AAA involving the left side of the distal abdominal aorta just above the bifurcation (Figure 2). The rheumatologist was consulted and the patient was started on steroids and Immuran. The patient underwent resection of the AAA and a dacron tube graft size 16 mm was inserted. In follow-up, the patient underwent a duplex scan every 6 months and the last one (20 months after the operation) showed no evidence of leakage, pseudoaneurysm or stenosis/occlusion of the graft.

Discussion

Behçet's disease was described by the Turkish physician, Hulusi Behçet, in 1937. BD is a multisystemic and inflammatory disease of uncertain etiology. Because there is no pathognomonic laboratory test or histologic findings specific to the disease, the diagnosis is based on clinical criteria. The international study group criteria require recurrent oral ulceration plus at least two of the following: recurrent genital ulceration, an eye lesion, a skin lesion, a positive pathergy test, vascular

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Figure 1. Spiral CT angiography demonstrated the presence of saccular AAA with inflammatory walls.



Figure 2. Preoperative angiography showed AAA involving the left side of the distal abdominal aorta just above the bifurcation.

involvement in the form of a DVT, arterial occlusion or aneurysm.¹

The incidence of vascular involvement reported in the literature ranges from 7% to 29% with a male/female ratio of 4-5:1. There is a high frequency of venous involvement (25% to 85%), arterial involvement (7% to 10%), and combined arterial and venous involvement (5% to 68%).²⁻⁵ The first two years of BD are the most critical period for vascular complications.^{2,8} DVT is the most frequent venous manifestation and the most commonly involved site is the lower limbs followed by the superior vena cava, the inferior vena cava and the upper limbs.^{2,3,8} Pseudoaneurysms are more common than arterial occlusion/stenosis or true aneurysm in BD and the rupture of a pseudoaneurysm is a major cause of death.⁴ The aorta is the most frequently affected site of pseudoaneurysm followed by the pulmonary, femoral, subclavian and popliteal arteries.^{3,4} All pseudoaneurysms are of the saccular type. Arterial occlusion/stenosis is common in the distal run off arteries of the lower limbs.^{4,5}

The insertion of an arterial or venous catheter may induce either a thrombosis or pseudoaneurysm formation in the puncture site. Duplex scan and CT angiogram or MRA must be done to confirm the diagnosis before surgery and every 6 months after the treatment for discovery of a new pseudoaneurysm or occlusion.⁶

Standard surgical repair and endovascular treatment are the two methods used in treating the arterial lesion in BD, particularly a pseudoaneurysm involving the aorta and the iliac arteries. Pseudoaneurysm treatment should be done as soon as possible because rupture of an inflammatory aneurysm can occur even when it is small because of the arterial wall fragility in BD. The surgical repair is often difficult, the suture tends to cut out, the healing is delayed and the graft is affected by occlusion or pseudoaneurysm. A recurrent pseudoaneurysm in anastomotic sites occurs in 30% to 50% of the cases.^{7,8}

To avoid these complications anastomosis should be done in macroscopically disease-free segments outside the inflammatory area. This means that intervention should be avoided in the active stage of disease. The use of steroids and immunosuppressives in the pre-operative and in post-operative periods reduces inflammation and prevents recurrent pseudoaneurysm. Some surgeons recommend wrapping the anastomosis in a supplementary prosthetic sheet or using pledgeted sutures, while other surgeons do only an aneurysmorrhaphy or simple ligation of the

aneurysm when possible to prevent further anastomotic false aneurysm.^{7,8} It is preferable to use a synthetic graft for bypass because the vein will be affected in BD. The use of antiaggregant or anticoagulant drugs does not always prevent thrombosis of the graft.^{7,8}

Endovascular grafting for AAA provides an important alternative to invasive surgical procedures for high-risk patients, especially during the active stage of BD, and may even be performed without pre-procedural angiography.

Multiple studies have confirmed that patients who had endovascular stent graft placement had lesser operating room time, a shorter hospital stay and less blood loss than open surgery. The success rate of endovascular grafting is 80% in moderate to high-risk patients and 90% in low risk patients.^{10,11}

A false aneurysm could appear secondarily after the endovascular technique because the self-expanding stent exerts continuous radial force on the arterial wall. Late femoral pseudoaneurysm may occur because of the femoral cutdown or puncture, but it is easier to treat in the femoral location than in the aorto-iliac position.¹²

It would have been possible to treat our two cases by endovascular grafting, but this method was not available in 2001 in our hospital. Close follow up with antiinflammatory medication and surveillance with regular intervals are the only current methods for the prevention and/or treatment of an arterial complication in a patient with BD. When aneurysm has been found in a patient with BD, it is important to screen for a possible multiple aneurysm and for venous lesions since they may be found together.¹³

References

- ¹ International study group for behçet's disease: Criteria for diagnosis of behçet disease. *Lancet*. 1990; 335:1078-1080.
- ² Koc Y, Giillii, Ak Pek G et al: vascular involvement in Behçet Disease. *J. Rheumatology*. 1992; 19:402-10.
- ³ Kabbaj N, Ben Jelloun G vascular involvement in Behçet Disease. *J. Radial*. 1993; 74:649-56.
- ⁴ Gurler A, Boyvat A, Clinical manifestations of Behçet's Disease. An analysis of 2147 patients. *Yousei Med J*. 1997; 38:423-7.
- ⁵ G-Y Ko, MD, J Y Byun, MD: the vascular manifestations of Behçet Disease. Angiographic and CT findings. *The British Journal of Radiology*. 7(2000) 1270-1274.
- ⁶ Kingston M, Ratcliff JR: Aneurysm after arterial puncture in Behçet's disease. *BMJ*. 1979;30:1766-7.
- ⁷ Takagi A., Kagiura N, Surgical Treatment of Non Specific Inflammatory Arterial Aneurysm. *J. Cardiovascular Surg*. 1986; 27:117-24.
- ⁸ Tuzun H, Besirli K, Management of Aneurysm in Behçet's Disease. *S Syndrome Surgery*. 1997; 121:150-6.
- ⁹ Tuzuner A, Umcu H: A case of Behçet's Disease with an Abdominal Aortic Aneurysm and Two Aneurysms in the Common Carotid Artery. *Angiology*. 1996; 47:1173-1180.
- ¹⁰ Ramazan Oner G: Endovascular Treatment of Huge Saccular AAA in a Young Behçet Patient: Midterm Result. *BMC Med Imaging*. 2002;Mar 22:2
- ¹¹ May, J White G: Endoluminal Repair of AAA: Strengths and Weakness of Various Prosthesis observed in a 4-5 year experience. *J. Endovascular Surgery*. 1997; 4:147-151.
- ¹² Vasseur MA Haulon S Endovascular Treatment of AAA in Behçet Disease. *J Vas Surg*. 1998; May 27(7):974-6.
- ¹³ Ceyran H., Akcali Y, Kahraman C. Surgical Treatment of Vasculo-Bahçet Disease: Review of patients with concomitant multiple aneurysms and venous lesions. *Vasa*. 2003; Aug, 32 (3) 149-153.