Dramatic vascular course of Behcet's disease

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ABSTRACT

Vascular involvement in Behcet's disease is rare (approximately 14% venous and 1.6% arterial), serious and recurrent. We report a case of Behcet's disease with deep venous thrombosis and right iliac pseudoaneurysm which was repaired with polytetrafluoroethylene (PTFE) graft. The patient received warfarin, aspirin, clopidogrel, immunosuppressive and corticosteroids. Two months later the patient developed manifestations of superior vena cava thrombosis and the graft was blocked. Three months later, ischemia of the right foot deteriorated and left femoral artery to right femoral artery crossover (PTFE) graft was performed.

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Behcet's disease is a multisystemic disorder characterized by recurrent ulcers of the mouth and genitalia and relapsing ocular inflammation.[1] Although vascular involvement in Behcet's disease is rare (approximately 14% venous and 1.6% arterial), they are serious, recurrent and carry the poorest prognosis.[2] We report a very rare case of Behcet's disease with iliac pseudoaneurysm, arterial and venous thrombosis. The aim of this report is to evaluate the clinical appearance and reoperation of the arterial involvement of such a case among the other vascular manifestations.

Case Report. A 50-year-old Sudanese male patient presented with a large (15 X 10 cm) tender pulsatile mass at the right iliac fossa. He also had significant loss of weight (26 kg) over the preceding month and tingling sensation followed by loss of sensation of the right big toe. The patient was also complaining of recurrent ulcers in the scrotum and oral cavity. He is a smoker. Hemoglobin was 7.9 g/dl and erythrocyte sedimentation rate was 148/hour. Computed tomographic (CT) angiography showed false aneurysm of right common iliac artery, dilated ureter and right-sided hydronephrosis. There was erosion of the bodies of lumbar vertebrae 4 and 5. In addition there was right femoral vein thrombosis (**Figure 1a & b**). The patient underwent repair of the aneurysm with polytetrafluoroethylene (PTFE) graft. Right double J stent was inserted. Histopathology of the wall of the aneurysm revealed necrotic wall of blood vessel with thrombus formation with no evidence of specific infection (**Figure 2**). The patient was discharged home with good general condition and normal pedal pulses. He received warfarin, aspirin and clopidogrel. He was later started on immunosuppressive and corticosteroids.

Two months following this he developed manifestations of superior vena cava thrombosis. A CT with contrast confirmed the diagnosis (**Figure 3**). It showed also pulmonary embolism. There were no palpable pulses in the right leg. Computed tomographic angiography showed the graft was blocked (**Figure 4**). Initially the leg was not critically ischemic and it was managed conservatively. Three months later, the patient started to have rest pain and right ankle pressure dropped to 50 mm Hg. Left

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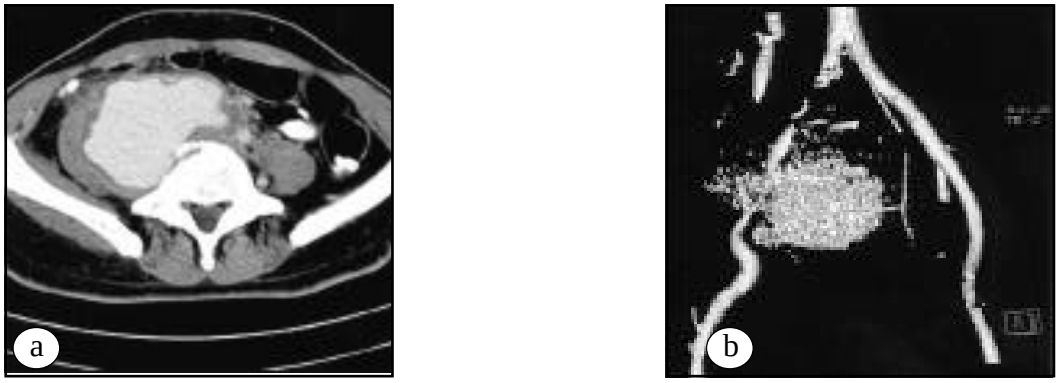

Figure 1 - Computed tomography (CT) angiography **a)** Axial CT scan showing huge right common iliac artery aneurysm with concentric mural thrombus. Pressure erosion of adjacent vertebral body. **b)** Post-contrast CT scan showing false right common iliac artery aneurysm with normal other arteries.

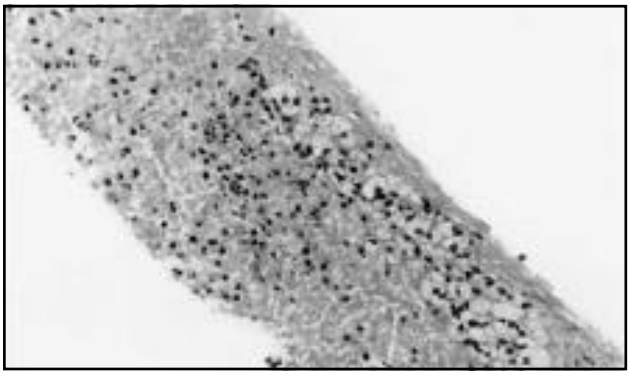

Figure 2 - Histopathology of the wall of the aneurysm revealed necrotic wall of blood vessel with thrombus formation with no evidence of specific infection.

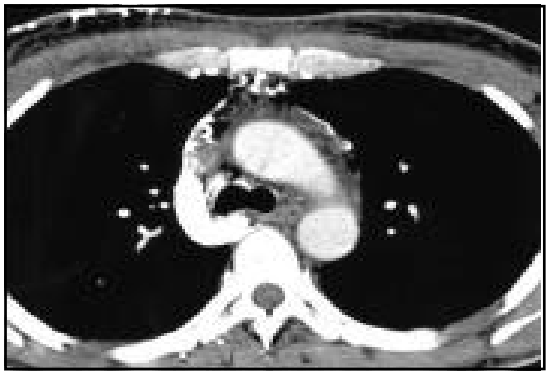

Figure 3 - Post contrast computed tomography scan of the chest showing thrombosis of superior vena cava, dilated azygos vein, numerous mediastinal and chest wall collaterals.

Figure 4 - Coronal computed tomographic angiography showing blocked right common iliac interposition graft.

femoral artery to right femoral artery crossover (PTFE) graft was performed. The circulation in the right foot was back to almost normal and the patient was discharged with antiplatelet, anticoagulant and immunosuppressives.

Discussion. Behcet's disease is a multisystemic vasculitis of unknown origin affecting mainly middle aged males in the Mediterranean, Middle East and Japan. Aside from the classical triad of oral, genital and ocular symptoms, it may affect the neurological, gastrointestinal and vascular systems.[2] The most common vascular involvement is superficial and deep thrombophlebitis (12-25%) followed by superior caval thrombosis (8%).[3] The etiology of vessel occlusion is still controversial. It has been suggested that low thrombomodulin levels in Behcet's patients increase the risk for thrombotic complication,[4] others proposed endothelial dysfunction.[5] The early venous manifestation of our patient was femoral thrombosis. Despite receiving a therapeutic dose of anticoagulant and antiplatelet, the patient developed superior vena cava thrombosis complicated by pulmonary embolism and graft thrombosis.

Arterial involvement in Behcet's is rare (2.2-6.8%)[2] but has serious and life-threatening problems.[6] Aneurysm is more common than occlusion. The pathology of the arterial aneurysm is inflammatory obliterative endarteritis of the vasa vasorum causing destruction of the media and fibrosis and thus weakening the arterial wall. This will predispose to aneurysm formation and eventual rupture.[7] The most common site is the aorta followed by pulmonary, femoral and popliteal arteries.[8] Our patient, and other cases have been reported with false[8] and true iliac aneurysm.[9]

The clinical presentation of the aneurysm depends on the stage of the disease. Usually, thoracic and abdominal aneurysms are discovered in the chronic stage with vague symptoms such as back pain and abdominal discomfort. However iliac artery aneurysm, like our patient, is usually diagnosed in the active stage of the disease.[10] Anemia, hypotension and ureteral involvement are common. For assessment of the arterial lesions, false aneurysm was described at the site of femoral catheterization. Magnetic resonance angiography was used, instead of catheter angiography in Behcet's disease, with high success.[11] We used CT angiography and were able to demonstrate the lesions. Various authors have reported rapid progression of the disease process, high rates of recurrence of true and false aneurysm at the anastomotic sites, disruption of the grafts, vessel and graft occlusion with an example of a patient that required 7 operations within 8 months.[2,10,11] The graft in our patient occluded after 2 months. In the view of the high incidence of graft occlusion, many authors do not recommend any redo surgery as graft occlusion is well tolerated in these patients.[3]

In summary, Behcet's disease should be investigated in young or middle aged patients presenting with arterial aneurysm. Early and midterm complications are high. Surgery should be restricted to symptomatic or false aneurysm, critical ischemia or acute bleeding.

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