Behçet’s disease with aneurysm of internal iliac artery and percutaneous treatment†

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INTRODUCTION

Behçet’s disease (BD) is seen most often in young adults of Middle East or Asian descent. It is characterized by recurrent orogenital ulcers, ocular manifestations and skin lesions. The typical vascular feature is medium- and large-vessel involvement, which causes both arterial occlusive disease and aneurysm formation [1].

Case

A 40-year-old male patient with an 8-year history of BD was admitted to our emergency service complaining of sudden onset pain and numbness in his left leg. From the history of the patient, we learned that he stopped immunosuppressive therapy 2 years ago, despite medical advice. The patient underwent surgery for right femoral artery aneurysm that had occurred after angiography performed 8 months previously.

Physical examination showed a huge pulsatile mass located on left lower abdominal quadrant with tenderness and systolic bruit (grade 2), oral ulcers, cutaneous pustules on the back, hypoesthesia and weak pedal pulses on the left lower extremity. His arterial blood pressure was 120/80 mmHg, and the heart rate was 83 beats/min.

The laboratory findings included a normal leucocyte count, erythrocyte sedimentation rate of 40 mm/h and a markedly increased C-reactive protein level.

Doppler ultrasonography (USG) detected left internal iliac artery pseudoaneurysm. Contrast-enhanced computed tomography (CT) showed a mass with the dimensions of 49 × 90-mm adjacent to the left iliacus muscle, and which was thick, regular walled pseudoaneurysm, originated from proximal internal iliac artery portion. Left hydronephrosis and left ureter dilatation were also detected due to pseudoaneurysm compression.

To reduce the size of the aneurysm, prevent its rupture and eliminate the hydronephrosis, we planned the implantation of a covered stent in his left internal iliac artery. Written informed consent was received from the patient.

Before the operation, methylprednisolone (80 mg) was injected intravenously 30 min prior to the procedure. Digital angiography was performed via the left femoral artery under local anaesthesia. The vessel width was investigated at the periphery of left internal iliac artery and a 10 cm diameter aneurysmatic filling was detected (Fig. 1). Following intravenous administration of a heparin bolus (5000 U), a 12-F vascular access sheath was manoeuvred into the first segment of the left iliac artery under fluoroscopic guidance to accommodate the delivery system. In order to avoid endoleak, anterior and posterior branches of the aneurysm were occluded before deployment of the stent-graft by using two coils. A 10 × 90 mm covered and tubular stent-graft (Jostentgraft; Jomed) was then deployed ~10 mm proximally and distally beyond the neck of the pseudoaneurysm.

Immediate follow-up angiography revealing the luminal aperture was obtained, and it was observed that the pseudoaneurysm remained occluded without no sign of leakage or endoleak (Fig. 2). The diameter of the internal iliac artery was measured ~1 cm. Postprocedurally, the pulsation and bruit of the mass disappeared immediately. Immunosuppressive (azathioprine 100 mg/day) and antiaggregant therapy (clopidogrel 75 mg/day) were started after the implant procedure. The patient was discharged without any complication after a week.

DISCUSSION

Vascular manifestations in BD are a major predictor of mortality and morbidity. The major findings in vascular BD are arterial and venous occlusions, followed by arterial aneurysms [2]. The pathological findings mainly due to vasculitis described in the
an aneurysm wall of BD patients are adventitial thickening and fibrosis, perivascular lymphocytic infiltration, elastin and muscle fibre decrement in media, smooth muscle fibre and fibroblast increment in intima. These vessel wall changes lead to formation of true aneurysms by wall distension or pseudoaneurysm by wall perforation [2]. The aneurysms in BD frequently involve medium- and large-sized vessels such as aorta, femoral, pulmonary, iliac and popliteal arteries. Rupture is the most common presenting symptom besides being the most common cause of vascular-related deaths [3].

Pseudoaneurysm formation at the site of arterial puncture for angiography or endovascular treatment has been reported in BD patients [4]. Consequently, non-invasive interventions such as USG, CT and magnetic resonance angiography imaging are recommended rather than conventional angiography for patients to whom endovascular treatment is not being considered [4].

On physical examination, pulsatile mass on the trace of peripheral artery or an intraabdominal mass can be palpated. A systolic murmur on the mass can also be heard. The clinical findings in aneurysms are related to localization, size and growth rate. In general, patients refer after a vascular surgical intervention or with a pulsatile mass, which makes it easy to diagnose. Delayed cases can have local erythema and tenderness as well as nerve and vein compression symptoms due to growth [5].

Previously, open surgical repair was the definitive treatment for vascular lesions, such as aneurysms, in patients with BD. However, the success rate of surgical management has not been high because a false aneurysm often occurs after surgical repair. Recently, endovascular stent graft seems to be more successful than surgical intervention because of lesser complication risk and is suggested in treatment of peripheral aneurysms in BD [6]. As it is less invasive, the morbidity is also lower than the open surgical technique [6]. These are particularly important advantages in surgical candidates with high risk.

Vascular aneurysms should be kept in mind in patients with BD admitted to emergency services. Leg pain, which is frequently seen and diagnosed as myalgia, can be due to a giant aneurysm in these patients. Checking for peripheral pulses should be an important part of physical examination as it can be weaker in affected side forming an important clue for diagnosis.

CONCLUSIONS

Vascular problems in BD, like aneurysm formation, may occur in various states including leg pain. The findings in our case suggest that endovascular therapy combined with immunosuppressive treatment for aortic pseudoaneurysms in patients with BD appears to be a promising alternative and effective management option. A longer follow-up is required to allow confidence of lasting success.

Conflict of interest: none declared.

REFERENCES