

Case report

An unusual case with vasculo-Behçet's disease: peripheral arterial aneurysm in an elderly woman

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ABSTRACT

Behçet's disease (BD) is a chronic, multi-systemic disorder that can affect all sizes of arteries and veins. Vascular involvement of BD is generally observed in young men and is an important cause of morbidity and mortality. Arterial lesions can appear as aneurysms, stenosis and occlusions in BD. We, here, present the case of a woman who developed a peripheral arterial aneurysm in her sixth decade, 20 years after disease onset. BD patients with aneurysms should be consulted at any age for pre- and post-operative assessment for immunosuppressive treatment to reduce recurrences, complications and disease activation.

Introduction

Behçet's disease is a chronic, systemic, inflammatory disorder characterised by mucocutaneous, ocular, neurological, musculoskeletal and vascular disease (1). Vascular involvement is more frequent in young age and male gender (2, 3). Most vascular lesions are venous (80%) and in a two-decade outcome survey of 387 BD patients by Kural-Seyahi *et al.*, 7.6% of males and only 0.8% of females had arterial lesions, and only four males had peripheral arterial aneurysms (4). A median of seven years between the diagnosis of the disease and arterial onset is reported, compatible with two other studies (2, 5). Herein, we report an unusual case of Behçet's disease, a 57-year old woman with a peripheral arterial aneurysm.

Case report

A 57-year-old woman was diagnosed with BD after presenting with oral and genital ulcers, deep vein thrombosis of right inferior extremity and erythema nodosum like lesions in 1992. She had a stable disease course later on. When referred by the Cardiovascular Surgery

Clinic in 2012, she was complaining of pain and swelling on her right thigh region for two months. Her MR angiography and Doppler ultrasonography revealed a 49*44 mm sized pseudoaneurysm originating from right superficial femoral artery. Aorto-femoro-popliteal arteriography of the patient was also performed (Fig. 1) to evaluate the feasibility of the aneurysm to be repaired with an endovascular stent graft, however the localisation of the aneurysm was not suitable for the procedure. She was symptom-free for other manifestations of Behçet's disease and biochemistry tests were negative for acute phase reactants. The patient did not have any risk factors associated with atherosclerosis such as hyperlipidemia, smoking or family history. The patient was taking colchicine 1.0 mg/day and never used any immunosuppressive therapy. Before surgery, methylprednisolone 1 mg/kg was started and

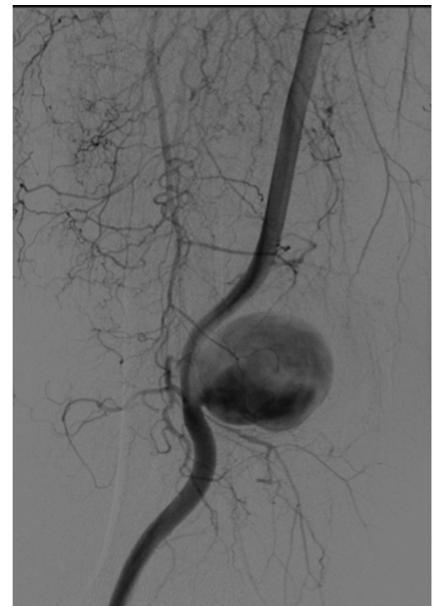


Fig. 1. Superficial femoral artery pseudoaneurysm demonstrated with a aorto-femoro-popliteal arteriography.

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the patient received four cycles of cyclophosphamide. Due to serious nausea and vomiting, cyclophosphamide was switched to azathioprine 150 mg/day after the fourth cycle. During the medical treatment, the size of aneurysm, thrombosis in its lumen and velocity of blood flow were followed with Doppler ultrasonography at every visit. At the ninth month of immunosuppressive therapy, Doppler ultrasound showed an enlargement of the aneurysm and the patient was referred to Cardiovascular Surgery. Aneurysmectomy and end-to-end anastomosis by using a PTFE (poly-tetrafluoroethylene) graft was done. Pathology of the operational material was suitable with a chronic vasculo-Beçet disease. There were irregularities of intimal surface, thrombus formation in the lumen of the aneurysm, thinning of media layer and fragmentation and splitting of elastic fibers and smooth muscles in almost whole of the vessel wall. Fibrous thickening of the adventitia leading to the occlusion of vaso vasorum and perivascular lymphocytes and plasma cell infiltrations were also present (Fig. 2-3). An atheromatous plaque or associated localised degenerative changes were not observed in the vessel wall.

The patient performed well after the operation in seven months of follow-up, and continued immunosuppressive therapy with azathioprine 100 mg/day.

Discussion

Vascular involvement is a major cause of morbidity and mortality in Behçet's disease. Patients with vasculo-Beçet's are likely to develop recurrent vascular lesions, therefore they should be identified and followed regularly with appropriate methods (4). We, here, report the development of a superficial femoral arterial aneurysm in a female, 57-year old patient. Although atherosclerosis due to patient's gender and age is a possibility for differential diagnosis, the lesion was reported as a pseudoaneurysm with imaging which is highly unusual for an aneurysmatic pattern of atherosclerosis. The patient did not have any classical risk factors for atherosclerosis. There was also no history of trauma or vascular intervention which can effect

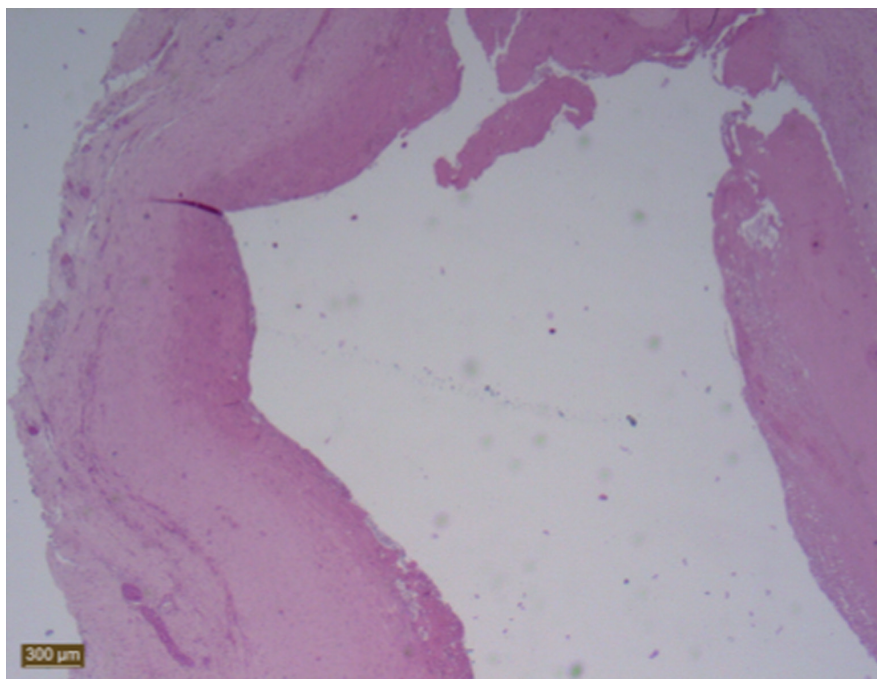


Fig. 2. Fibrosis in aneurysmal vessel wall (H&Ex40).

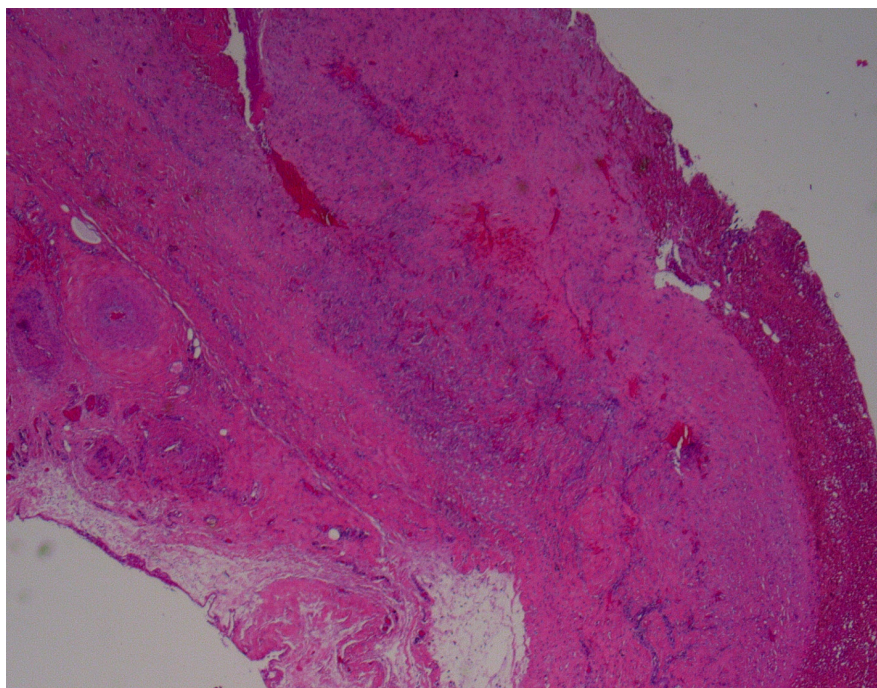


Fig. 3. Fibrosis in aneurysmal vessel wall and thrombus formation in the lumen (H&Ex100).

the formation of this pseudoaneurysm. Furthermore, pathological assessment of the operation material revealed features compatible with a chronic vasculo-Beçet's disease.

The pathogenesis of aneurysms in Behçet's disease is different from the aneurysms due to atherosclerosis. The cellular infiltrates at media, adventi-

tia and around the vaso vasorum usually compose of neutrophils, lymphocytes, plasma cells, histiocytes and eosinophils at active arteritis phases of Behçet's disease. The lumen of artery is commonly thrombosed. The destruction or interruption of medial elastic fibers, proliferation of fibroblasts and severe inflammation also

weaken the vessel wall. The obliterative endarteritis and thickening of vaso vasorum accelerate the aneurysmal dilatation process with transmural necrosis and these episodes result in pseudoaneurysm formation (6, 7). In a retrospective analysis of 2319 patients, Sarica-Kucukoglu *et al.* reported the prevalence of vascular involvement 14.3% and more usual in males (8). The mean age at the onset of vascular involvement was 30.6 ± 7.9 and 3.6% of 332 vasculo-Behçet patients had arterial lesions. These results are supported by other studies (2, 3). Arterial involvement of BD is reported as 15-20% of vascular lesions and can be observed as occlusions or aneurysms at every size of arteries (9, 10). They are mostly seen at aorta, followed by pulmonary, femoral, subclavian, popliteal and carotid arteries (11). Aneurysms are one of the most serious complications of Behçet's disease and aggressive immunosuppressive treatment is the standart approach. Due to high risk of hemorrhagic complications, anticoagulation therapy is controversial (12, 13). Onset of arterial lesions in BD patients

are usually reported around fifth years of follow-up, which then declined (2, 6). Our case is diagnosed as BD at the age of 37, but the peripheral arterial aneurysm was observed twenty years after her BD diagnosis.

In conclusion, physicians following BD patients should be alert to new, unusual vascular presentations at every stage, especially if surgery is planned. This will enable patients to be treated with immunosuppressives pre and post-operatively, which might reduce recurrences and complications.

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