Case reports
Huge femoral artery pseudoaneurysm in a patient with Behçet's disease

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ABSTRACT

Background: Behçet’s disease (BD) is a variable vessel vasculitis and vascular involvement is one of its life threatening manifestations. Arterial involvement frequently occurs with male predominance with pseudoaneurysms being the most common presentation. Immunosuppressive therapy is the mainstay of treatment in vascular involvement.

Case presentation: The case we report here is a 40 year old Iraqi BD patient with manifestations of recurrent oral and genital ulcers, bilateral anterior uveitis, and deep vein thrombosis. The pathergy test was positive. The HLA-B51 was negative, erythrocyte sedimentation rate 102 mm/1st h and C-reactive protein was 48 mg/L. After discontinuation of his medications for about 9 months, the disease presented with leg pain and swelling that was diagnosed as huge left superficial femoral artery pseudoaneurysm by Doppler ultrasonography. CT angiography revealed a 90 × 88 × 70 mm pseudoaneurysm with partial mural thrombosis. He was scheduled for emergency surgery due to severe intractable pain. He received a pulse of methylprednisolone 1 g/day for 3 days and then surgery was done in the form of exclusion, repair and femorofemoral bypass were done. Post-operatively, the patient had an uneventful course; distal pulses became palpable, pain and swelling subsided. Post-operation, prednisolone 1 mg/kg was continued and he received cyclophosphamide 750 mg intravenously. His blood homocysteine level was higher than normal 23.8 μmol/L. He was discharged with a high dose of steroid and monthly cyclophosphamide treatment.

Conclusion: Arterial pseudoaneurysm is life-threatening in BD and should be kept in mind to prevent major complications. Vascular involvement in BD patients is probably associated with hyperhomocysteinemia.

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1. Introduction

Behçet’s disease (BD) is a variable vessel vasculitis that involves multiple systems [1]. The diagnostic features of the disease are recurrent oral and genital aphthosis, skin lesions and eye involvement. It can affect all vessels with various sizes and both arterial and venous lesions could be seen in BD patients [2] and subclinical atherosclerosis has been reported [3,4]. Arterial involvement occurs in about 2.2–18% of patients with mostly male predominance [5,6].

The vascular involvement has various clinical presentations such as aneurysm and pseudoaneurysm formation in the arteries, thrombotic occlusion in arteries, and venous thrombosis [7]. Pulmonary artery aneurysms have also been frequently reported [8]. The most common arterial manifestation in BD patients is pseudoaneurysm with a remarkably higher frequency than aneurismal formation [6,9]. In some pathological reports, infiltration of the media and adventitia with neutrophils and mononuclear cells were seen in arterial involvements [5]. It seems that the first phenomenon in the affected artery is active arteritis mainly around the vasa vasorum leading to transmural necrosis and gradual vessel wall thickness and aneurysmal dilatation and at last perforation of the vessel wall and pseudoaneurysm formation [9]. When there is deficiency in the tunica media and intima, out-pouching of the vessel occurs, called pseudo-aneurysm, but in true aneurysm all the three layers have defects [10]. Pseudoaneurysms are more prone to rupture than aneurysms and can be a cause of mortality in BD patients due to bleeding and ischemia [11,12]. We here report a patient with Behçet’s disease presented with huge left superficial femoral artery pseudoaneurysm.
2. Case presentation

In March 2015, a 40 year old Iraqi male patient with blurring of vision and left lower extremity swelling was admitted in Rheumatology ward of Hafez Hospital, Shiraz University of Medical Sciences for further work-up. He had a positive history of recurrent oral ulcers (Fig. 1) and genital lesions with scar of his last genital aphthous lesion on his testis. His work-up sonography showed left popliteal vein non- obstructive sub-acute deep vein thrombosis (DVT). The pathergy test was positive. Ophthalmologist exam reported bilateral anterior uveitis. The HLA-B51 was negative, erythrocyte sedimentation rate (ESR) 102 mm/1st h and C-reactive protein (CRP) was 48 mg/L. He fulfilled the ISG and ICBD criteria for BD [13] and was discharged with prednisolone 1 mg/kg/day with slow tapering, warfarin and azathioprine 100 mg/day.

He did not show-up for his next follow-up visit due to good response and discontinued his medications after 3 months. His general condition was well for near 1 year till 20 days before his last admission (September 2016) when he developed severe left thigh pain and swelling, so was referred to rheumatology clinic and color Doppler ultrasonography of the left thigh vessels showed a large aneurysmal dilatation in the anteromedial part of the thigh with a diameter of 80 mm and thrombosis within the canal. CT angiography of both lower extremities revealed a 90 × 88 × 70 mm pseudoaneurysm in the inferior distal part of the left superficial femoral artery with partial mural thrombosis; the lumen of the artery in this area was 76 mm with 46 mm patent lumen with evidence of peripheral soft tissue edema (Fig. 2).

He was transferred to the vascular surgery department and was scheduled for emergency surgery due to severe intractable pain. Because of the inflammatory nature of previous disease and the highly acute phase reactants on arrival, he received a pulse of methylprednisolone 1 g/day for 3 days and then surgery was done. Pre-operatively, left distal pulses were absent and the patient had severe edema up to the thigh level with a huge pulsatile tender mass in the medial aspect of the left thigh. Intraoperative finding was a huge pseudoaneurysm 9 cm in diameter originating from the distal part of the left superficial femoral artery with pressure effect on the left femoral vein. So, exclusion and repair of pseudoaneurysm and femorofemoral bypass was done with externally reinforced expanded polytetrafluoroethylene (ePTFE) vascular graft (Fig. 3). Post-operatively, the patient had an uneventful course; distal pulses became palpable, pain and swelling subsided, and he was transferred to the rheumatology ward with pathology report of dissected artery and pseudo-aneurysm with intramural thrombosis. Post-operation, prednisolone 1 mg/kg was continued and he received cyclophosphamide 750 mg intravenously. His blood homocystein level was higher than normal 23.8 μmol/L (normal < 15 μmol/L). He was discharged with a high dose of steroid and monthly cyclophosphamide treatment.

3. Discussion

Arterial involvement has been reported in patients with Behçet’s disease which mostly involves the main arteries in the form of pseudo-aneurysms, aneurysms, occlusion and thrombosis [9,11]. Peripheral as well as main arteries can be involved, as well. There have been reports of involvement of the superficial femoral artery in the form of pseudoaneurysm in BD [11,14].

We here reported a young man with Behçet’s disease presenting with vascular involvement, previously left lower extremity DVT and then arterial pseudo-aneurysm. A similar case report has been presented of a BD patient with previous lower extremities DVT presenting with superficial femoral artery pseudo-aneurysm [15]. Another 44 year old BD case in remission with previous DVT and left brachial artery aneurysm was reported and suddenly presented with chest pain and a diagnosis of a large 32 × 30 mm aneurysm in the proximal Left Anterior Descending (LAD) artery was found [16].

Vascular inflammation and endothelial dysfunction play roles in the pathogenesis of vasculitis [2]. Large vessel involvement is seen in up to 40% of BD patients. Veins are more frequently involved than arteries [11]. However, venous thrombosis coexists with arterial lesions in most reports [1]. Vascular involvement is one of the major causes of morbidity and mortality in BD [7,12]. Behçet’s aneurysm has the tendency to multiply and can involve any arteries [10,17]. The most affected sites reported for arterial aneurysm were the aorta, pulmonary, femoral, popliteal and carotid arteries [2]. Arterial aneurysmes were also associated with high CRP and ESR levels [18,19]. Also in a study by Bartlett et al., it was recommended that BD patients during exacerbation, especially men, should be screened for an abdominal aortic aneurysm. New, oral and genital lesions may be predictors of a new arterial complication [20].

The decision of how to manage a peripheral artery aneurysm in BD is determined by the location of the artery involved and the clinical presentation, whether ruptured or impending to rupture exists and whether the disease is active or in remission phase [21]. Immunosuppressive therapy should be considered in patients with arterial disease before the surgical intervention and should be continued after surgery to avoid post-operative complications [22]. Acute arterial involvement is a medical emergency and it was recommended that the treatment should be started with pulsed intravenous glucocorticoid and immunosuppressive like pulsed IV cyclophosphamide followed by steroid as maintenance therapy. Arterial aneurysms should be corrected by intervention due to increased risk of aneurysmal rupture [23]. Non-pulmonary artery involvement may necessitate endovascular or open surgical interventions [24]. The traditional treatment of aneurysmal lesions in BD patients is surgical repair and graft insertion, with reports of graft occlusion, anastomotic pseudo-aneurysm formation or post-operative infection [9,22]. Therefore, close follow-up with immunosuppressive medications and periodic surveillance are the only way to prevent arterial complications in BD patients.

The patient we reported had one episode of DVT in the left popliteal vein and after one year he developed left superficial femoral artery pseudoaneurysm formation and associated with a high ESR and CRP. On follow-up after receiving immunosuppressive and graft insertion, after 3 months he had no complications.

Hyperhomocysteinemia was present the BD patient Hyperhomocysteinemia is an independent risk factor for atherosclerosis and associations with arterial aneurysms without atherosclerosis has been reported [25]. The pathogenesis of arterial involvement

Fig. 1. Oral ulcer of the lower lip in the Behçet’s disease patient at his presentation with superficial femoral aneurysm.
in BD patients is different from atherosclerosis [9]. Hyperhomocysteinemia has been reported with involvement of both arteries and veins in BD and also with disease activity [26]. There have also been reports of the association between thrombosis and hyperhomocysteinemia in BD patients [2]. Further studies are recommended to detect any association of hyperhomocysteinemia and the presence of both arterial and venous involvement in BD patients.

In conclusion, arterial pseudoaneurysm is a life-threatening manifestation of vasculo-Behçet disease that always should be kept in mind to prevent major complications, especially in men with previous vein thrombosis. Vascular involvement in BD patients is probably associated with hyperhomocysteinemia.

Conflict of interest

None.

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