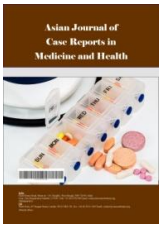

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Iliac Primitive Aneurysm Revealing Behçet Disease

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Authors' contributions

This work was carried out in collaboration among all authors. Authors MS and CD wrote the manuscript. Authors MJ, FF and ZB analyzed the manuscript. Author IF performed the surgical operation. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Thirty-one year old woman with a history of recurrent oral aphtosis presented a leg swelling, pain and functional impairment. A venous thrombosis of the left ilio-femoro-popliteal was diagnosed and the patient received an anticoagulation therapy. One month later, she presented an arterial aneurysm of the right primitive iliac artery confirmed by an angio-CT exam. Physical examination revealed necrotic pseudofolliculitis, a bilateral posterior uveitis with papillitis. The diagnosis of BD was made. A corticosteroid treatment was conducted (solumedrol 1g/day for 3 days then 1 mg/kg/day of prednisone equivalent for 6 weeks then progressively tapered until 10 mg/day) associated with cyclophosphamide (1g/month for 12 months). An angio CT scan 12 months later was performed and objectivated the progression in size of the aneurysm. Therefore, a surgical treatment was decided and consisted on a lay flat of the aneurysm and graft interposition with polytetrafluoroethylene prosthesis. The post-operative outcome was favorable.

Keywords: Behçet disease; arterial aneurysm; surgery.

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1. INTRODUCTION

Behçet disease (BD) is a multisystemic disorder of unknown etiology. It is a primitive vasculitis that affects the eyes, skin, joints, blood vessels, and nervous system. Vasculo- Behçet is characterized by aneurysm formation and occlusion of the large vessels [1]. Arterial aneurysms have been reported with a frequence of 0.15 to 2.5% among patients with Behçet's Disease. The aneurysm could be located in the visceral or peripheral arteries, in the aorta or its major branches. They are leading to mortality if not carefully and rapidly treated [2]. Here, we report the case of iliac primitive aneurysm in a young woman with successful surgical treatment.

2. CASE REPORT

Thirty-one year old woman with a history of recurrent oral aphtosis presented a leg swelling, pain and a functional impairment. A doppler ultrasonography of the lower extremities showed a venous thrombosis of the left ilio-femoro-popliteal axis extended to the arch of the long saphenous vein. She was treated with anticoagulant treatment (low molecular weight heparin then anti vitamin K). A month later, it occurs a swelling of the right inguinal area. A doppler ultrasonography of the lower extremities showed an arterial aneurysm of the right primitive iliac artery measuring 40*30 mm compressing the homolateral vein. The Angio-CT exam confirmed the aneurysm in the iliac artery (Fig. 1). Therefore, a Behçet's disease was suspected and she was hospitalized in internal medicine department. Physical examination revealed a young woman in a good general condition, full conscious and afebrile (37°C). She had also an extensive oral ulcer without genital ulcer, necrotic pseudofolliculitis lesions in thighs. The pathergy test was negative. Ophthalmological examination revealed a bilateral posterior uveitis with papillitis and iridocrystalline synechiae. The HLA-B51 was negative, erythrocyte sedimentation rate (ESR) 120 mm/1st h and C-reactive protein (CRP) at 85 mg/L. Laboratory analysis showed an erythrocyte sedimentation rate (ESR) at 102 and serum fibrin at 7.42 g/l. The diagnosis of BD was made according to the ISG of BD criteria [2] basing on the presence of recurrent oral ulcer, necrotic pseudofolliculitis lesions and uveitis. The patient had two serious manifestations of the disease which are posterior uveitis with papillitis and venous thrombosis and peripheral arterial aneurysm. Therefore a corticosteroid treatment was conducted (solumedrol 1 g/day for 3 days then 1mg/kg/day of prednisone equivalent for 6 weeks then progressively tapered until 10 mg/day) associated with cyclophosphamide (1 g/month for 12 months). An angio CT scan 12 months later was performed and objectivated the progression in size of the aneurysm: 6.5*10.3 cm (Fig. 2a and 2b). Consequently, a surgical treatment was decided. She was transferred to the vascular surgery department and the operation consisted on a lay flat of the aneurysm and graft interposition with polytetrafluoroethylene prosthesis. Post-operatively, the patient had an uneventful course; distal pulses became palpable. Post-operation, maintenance therapy was based on cyclophosphamide (1g every 3 months for one year) and 10 mg of prednisone equivalent with a good clinical and radiological response. The angio-CT six months after surgery showed a normal caliber of iliac artery with no thrombosis.

3. DISCUSSION

Behçet disease is a multisystemic vasculitis in that affect small, medium, and large vessels. It is an auto-inflammatory disease which causes oro-genital ulcerations and other features including cutaneous, ocular, vascular, articular and neurological manifestations [3]. Arterial involvement has been reported in1.5 to 2.2% of patients with Behçet's disease and mostly involves the main arteries in the form of pseudo-aneurysms, aneurysms, occlusion and thrombosis [4-5]. The arterial involvement occurs almost 5 to 9 years after the diagnosis of Behçet disease with male predominance [6]. The present case has the particularity of being discovered in a young woman and has also revealed the BD. As in our patient, venous thrombosis is associated with arterial lesions in most reports [7]. In our patient, the aneurysm interested the primitive iliac artery. This location was rarely reported in the literature and was described mainly in the internal iliac artery. Arterial aneurysm are located mostly in the aorta, pulmonary, femoral, popliteal and carotid arteries [3]. Arterial aneurysm in BD is usually associated to oral and genital ulcers which help clinician to make the diagnosis of BD [8]. In our patient, the vascular involvement was associated to severe ocular posterior uveitis with papillitis. High CRP and ESR levels as in the present case have been reported in patients with arterial aneurysm in BD [9]. Behçet aneurysm is a serious complication with risk of irruption and the tendency to multiply [10].

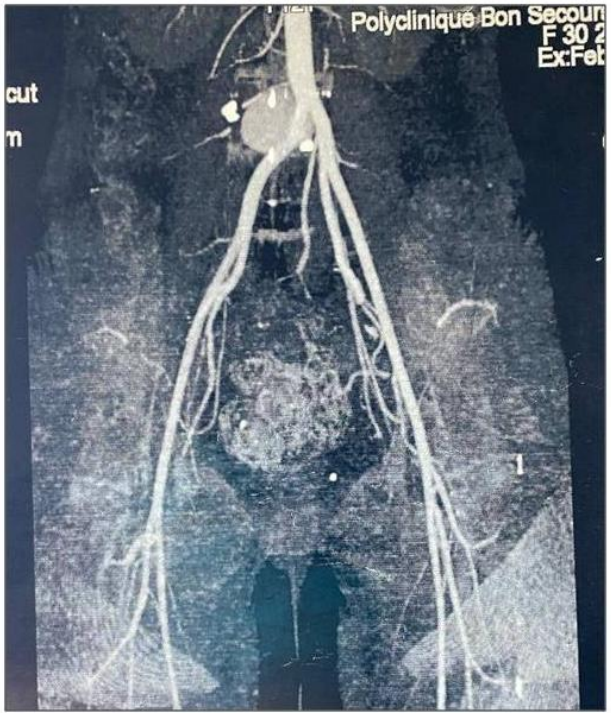
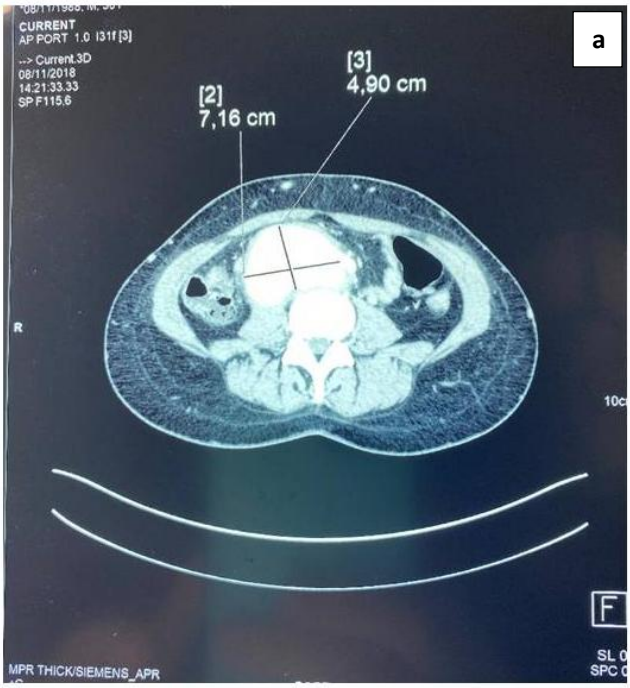


Fig. 1. Aneurysm in the iliac artery in reconstruction tomography



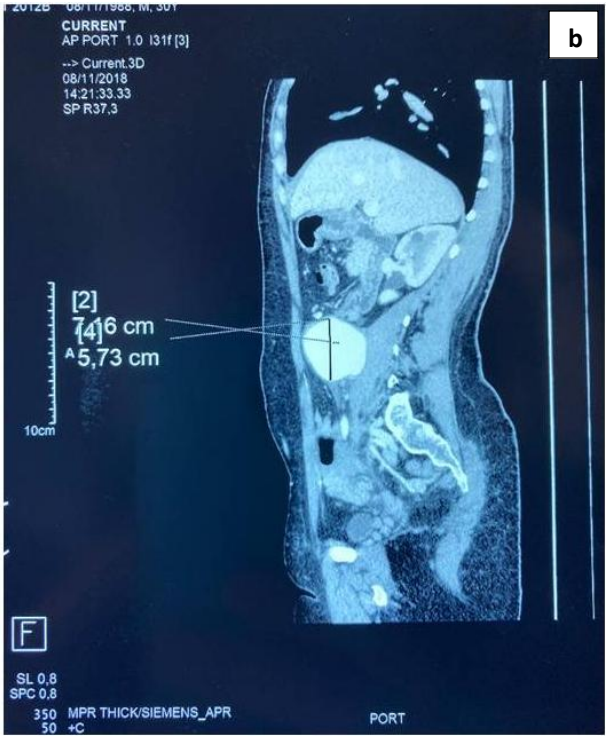


Fig. 2. Accentuation of iliac aneurysm in axial (a) and sagital (b) section

The management of arterial aneurysm in BD depends on the site of the lesion and the remission of the disease. Medical treatment based on corticosteroids and even immunosuppressant agent is indicated to control vessel inflammation and also to avoid post operative complications as graft occlusion and recurrence of aneurysms leading to death [11]. Surgical treatment options in patients with arterial aneurysm include endovascular stent implantation and surgical bypass. The main advantages of endovascular intervention are lower mortality rates (0.6%-3.5%) and higher success [12]. However, these surgical techniques are poorly documented in BD. To succeed the surgical procedure it is better to postpone the intervention until the remission of the BD as in the present case. Extra-anatomic bypass procedures to avoid anastomotic complications are preferred by surgeons [13]. Nevertheless, re-thrombosis and repeating aneurisms at the anastomosis regions are frequently reported [14].

4. CONCLUSION

Arterial aneurysm is a life-threatening manifestation of BD that should be kept in mind mainly in young patients. Its early diagnosis allow a better medical control until the remission of the disease and then to decrease postoperative complications.

CONSENT

As per international standard or university standard, patient's written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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