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Case Report

Unusual Localization of Behcet's Disease Revealed by A False Aneurysm

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Abstract

Behcet's disease is an inflammatory disorder with multisystemic involvement, diagnosed on the basis of clinical criteria established by the International Study Group for Behcet's. Vascular involvement is rare and may be inaugural in many cases. It affect both arteries and veins, manifesting as thrombophlebitis, arterial occlusion, aneurysms and false aneurysms. Computed tomographic angiography is essential for diagnosis and study of the characteristics of aneurysms and false aneurysms. The management of aneurysms in Behçet's disease is delicate, and requires the combination of corticosteroids and immunosuppressive drugs in severe forms supplemented by open or endovascular repair, in order to reduce complications risk.

We present a two exceptional case of Behçet's disease revealed by false aneurysm of right subclavian artery and popliteal artery.

Keywords:

False aneurysm, Behçet's disease, immunosuppressants, Vasculitis, Open surgery.

Introduction

Behçet's disease is a rare multisystemic chronic disease. A vasculitis of vessels of all calibers that affects the arterial and venous areas with a preponderance of venous lesions **[**1**]**. Arterial involvement in Behcet's disease is rare, most often marked by aneurysms and false aneurysms, which is a severe form of the disease given the high risk of rupture **[**2**]**.

We report two cases, singular by their localization, of Behçet's disease revealed by false aneurysm of right subclavian artery and popliteal artery, for the purpose of discussing approach to this unusual complication of Behcet's disease.

Case 1

A 47 years old moroccan man, was admitted for a swelling in the right supraclavicular fossa, that has been gradually increasing in volume in the past 2 months. His medical history was notable for a recurrent orogenital aphthosis.

Physical examination showed an hemodynamically stable patient with an expansive, pulsatile and beating mass measuring 5cm located above the collarbone. The rest of the clinical examination found an oral aphthosis and pseudo folliculitis in his back. The peripheral pulses were palpable on both sides.

Computed tomographic angiography and arteriography revealed a false aneurysm of the right subclavian artery (figure 1,2).

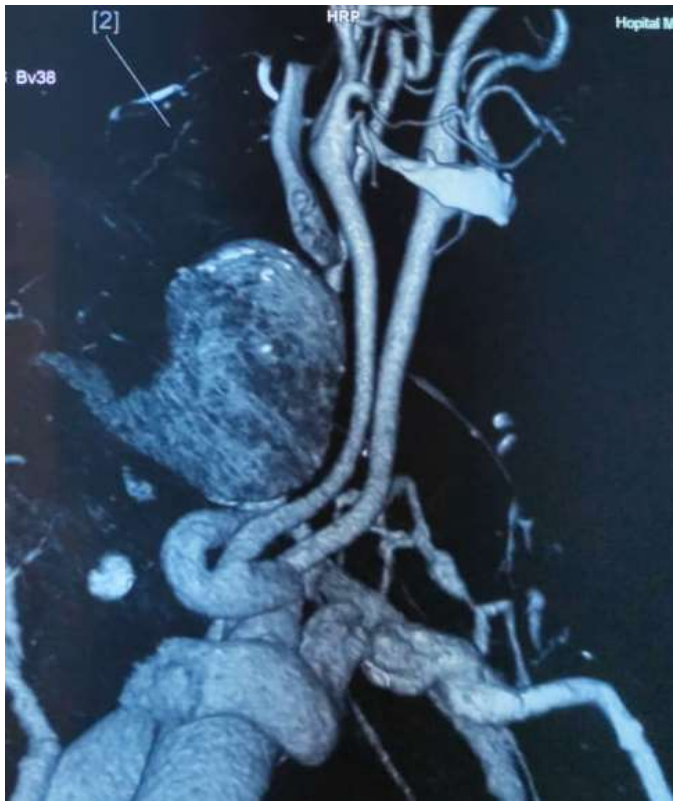

Figure 1 : Computed tomographic angiography showing a false aneurysm of the right subclavian artery.

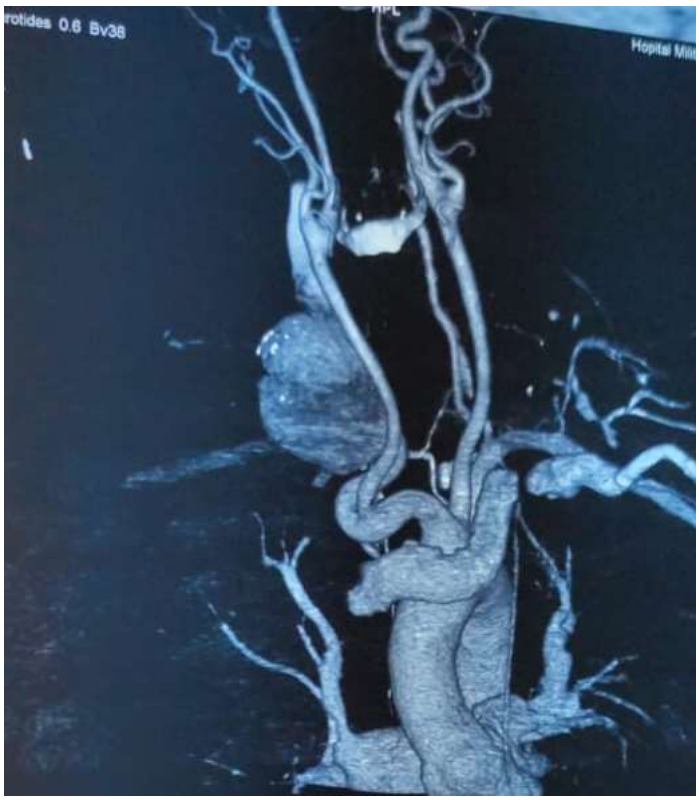

Figure 2 : Computed tomographic angiography showing a false aneurysm of the right subclavian artery.

The laboratory findings objective an inflammatory syndrome with erythrocyte sedimentation rate of 81 mm/h at the first hour, and a markedly increased C-reactive protein level (170 mg/L).

The diagnosis of Behcet disease with vascular involvement was retained.

Before the operation, methylprednisolone was injected intravenously. The patient underwent surgery. We performed open surgery with aneurysm flattening (figure 3), the damaged arterial segment was resected with restoration of vascular continuity. The post operative was simple.

The medical treatment was started, methylprednisolone (1000 mg) was injected intravenously 3 days in a row, and then prednisone orally. Immunosuppressive (azathioprine 100 mg/day), colchicine and antiaggregant therapy (clopidogrel 75 mg/day) were started also after the procedure. The patient was discharged without any complication after a 05 days. The 6-month follow-up visit was unremarkable.

Figure 3: Operative view of the right subclavian artery false aneurysm.

Case 2

A 54-year-old Moroccan man. Presented to the emergency department with a swelling in the right popliteal fossa. His medical history revealed oral and genital ulceration 1 year before admission, indicative of Behçet disease. Ultrasound showed a right popliteal cyst, and computed tomographic angiography showed a ruptured false aneurysm of the popliteal artery (figure 4). The patient was immediately operated, with the same procedure as in the previous case (figure 5). He received intravenous corticosteroid therapy. He was discharged 07 days after admission with a course of oral corticotherapy (prednisone 1mg/Kg) and azathioprine therapy. No postoperative complications had developed after 1 year.

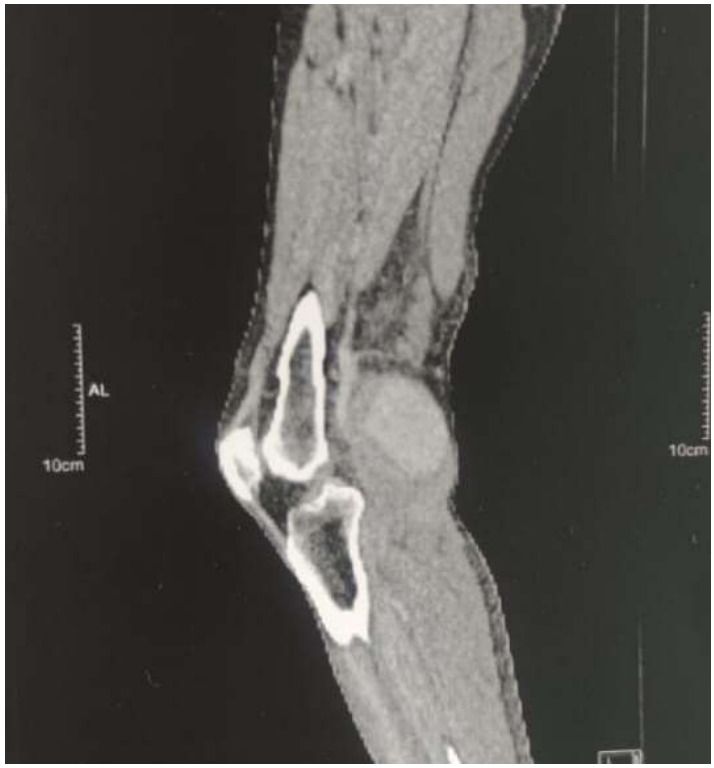

Figure 4: Computed tomographic scan a ruptured false aneurysm of the popliteal artery.

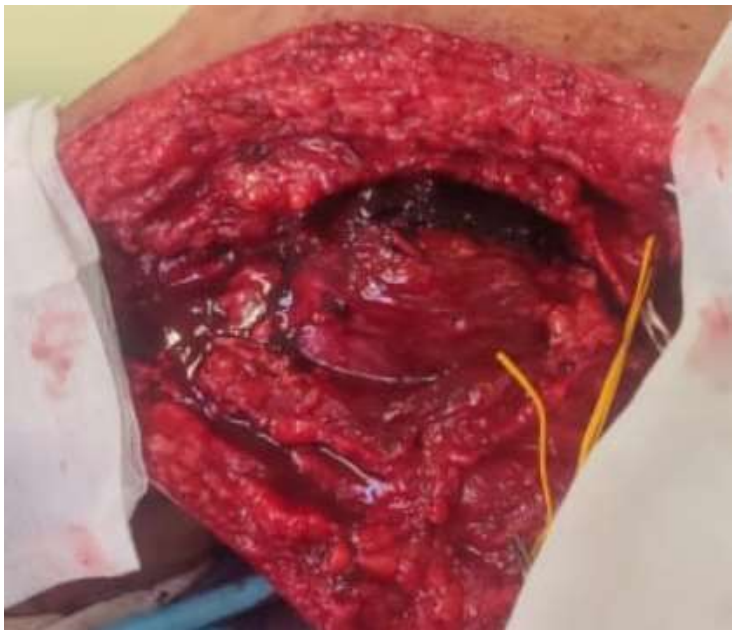

Figure 5: during the open surgery of the ruptured false aneurysm of the popliteal artery.

Discussion

Behcet disease (BD) has a peculiar geographic distribution. It is more common in Mediterranean regions **[**3**]**, where an association with HLA B51 has been proved. Vascular involvement of behcet's disease is characterized by arterial and venous occlusions, and arterial aneurysms. The frequency of aneurysmal lesions is very rare: 0.94% of patients with popliteal artery **[**6**]**.

The pathologic process appears to involve a vasculitis of the vasa vasorum with adventitial thickening 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 **[**4**]**. 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 **[**5**]**.

Various treatment modalities in artery aneurysm include immunosuppressive therapy, surgery, and endovascular surgery. Cyclophosphamide azathioprine and glucocorticoids constitute the basic treatment of aneurysms on Behcet **[**7**]**. TNF alpha cures have been reported in refractory cases or with severe aneurysmal lesions **[**8**]**

The treatment of aneurysms can be by open surgery with aneurysm flattening. The restoration of vascular continuity can be performed by sleeve anastomosis, it consists of wrapping around the anastomosis with a short prosthetic graft segment To prevent secondary pseudoaneurysm **[**7**]**. Endovascular treatment with stent could be the first choice treatment because arterial inflammation increases the risk of perioperative hemorrhage **[**9**]**.

Ideally, administration of immunosuppressant therapy before the intervention is useful to reduce this inflammation and to prevent further anastomotic false aneurysms **[**9**]**.

Conclusion

The findings in our cases suggest that Surgery combined with immunosuppressive treatment in patients with Behcet disease appears to be a promising and effective management option. A longer follow-up is required to allow confidence of lasting success.

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