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Hemorrhagic Shock Revealing Aortic Aneurysm Rupture due to a Behçet's Disease

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ABSTRACT

The diagnosis of Behçet's disease is based on clinical criteria, mainly those defined by the International study group for Behçet's disease. These criteria typically combine bucco-genital aphthosis, uveitis and systemic manifestations. Vascular complications are dominated by venous thrombosis. Arterial complications are rare and severe. We report two cases of Behçet's disease diagnosed following haemorrhagic shock due to a ruptured aortic aneurysm.

KEYWORDS

Behçet's disease; Aortic aneurysm; Hemorrhagic shock

INTRODUCTION

The Behçet's disease is a multi-systemic disease [1]. Its diagnosis is based on clinical criteria, defined by the "International study group for Behçet's disease" [1].

These criteria typically combine oral aphtosis, uveitis, and systemic skin manifestations. Behçet's disease is mainly characterised by vascular thrombosis [2].

However, diagnosis of this disease in patients with hemorrhagic shock is exceptional [2].

We report the case of two patients in whom Behçet's disease was discovered by hemorrhagic shock caused by rupture of aortic aneurysm.

CASE 1

A 53-year-old man, consulted in the emergency department for abdominal pain. The patient reported no pathological history. He suffered from abdominal pain that appeared since 3 days. Medical examination showed a pulsatile abdominal mass, with De Bakey sign, an oral aphtosis and noueux erythema. The patient was hospitalized.

Biological examination showed anemia (hemoglobin = 8.6 g/dl) and an inflammatory syndrome (ESR = 110 in the first hour and a CRP = 201 mg/l). Abdominal CT scan was emergently performed. It revealed a huge saccular aneurysm of the supra renal aorta with signs of fissuration (Figure 1A).

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Two hours later, the patient developed severe abdominal pain, awareness disorder, blood hypotension (80/50 mm Hg), tachycardia, pallor and coldness of the extremities. The patient was resuscitated, intubated, ventilated, transfused and transferred to the operating room.

Intraoperative examination revealed a great peritoneal effusion, and a ruptured aneurysm of the supra-renal aorta measuring 10 cm of diameter (Figure 1B).

The patient died during the surgery because of an incontrollable bleeding.

Histologic examination of the aortic wall revealed hypertrophy of the muscular fibers, with infiltration of inflammatory cells and neutrophil polynuclear, and blood vessels confirming the diagnosis of Behçet's disease.

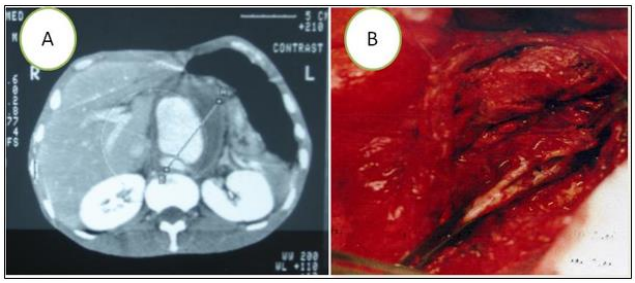

Figure 1: A) A huge sacciform aneurysm of the suprarenal abdominal aorta; B) Intraoperative view: Hemoperitoneum with loss of substance in the aorta, reminiscent of arterial sore throat.

CASE 2

A 27-years old woman, was admitted to the emergency department for acute chest pain quickly complicated by hemodynamic shock. Clinical and biological examination revealed tachycardia, hypotension with signs of shock and anemia (hemoglobin level = 7 g/dl), necessitating catecholamines and blood transfusion. CT scan was emergently performed.

It revealed a fissured false aneurysm of the aortic arch (Figure 2). The patient underwent emergent implantation of covered endoprosthesis. The patient had a clinical improvement within few days. In the post-operative period, the patient reported episodes of recurrent herpetiform mouth ulcers that she ignored.

We completed with a genital examination that objectified a minor ulcer.

The ophthalmological examination was carried out as part of the etiological assessment, it concluded to the diagnosis of of retina vasculitis.

The diagnosis of Behçet's disease was confirmed. So, the patient received corticosteroids and immunosuppressive treatment.

Two months later, the patient was admitted to the emergency department for chest pain and blood hypotension. CT scan revealed a ruptured false aneurysm of the extremity of the endoprosthesis. The patient was transferred in the operating room for emergent surgery, but she died before incision.

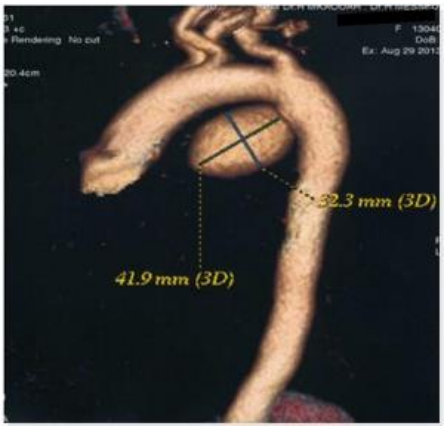

Figure 2: CT scan showing a pseudoaneurysm of the aortic arch.

DISCUSSION

This work report two cases of Behçet's disease revealed by hemorrhagic shock.

Behçet's disease is a systemic vasculitis characterized by oral and genital aphtosis, uveitis, articular, neurologic, and vascular signs [1,3].

It concerns young men between 20 years and 40 years, without cardiovascular risk factors [4]. It is one of the main cause of inflammatory aortites [3].

Vascular involvement in Behcet disease is characterized mainly by deep vein thrombosis [5].

Arterial involvement concerns 1% to 2% of the patients [6]. Huong et al., reported a duration of 7.3 years between symptoms and arterial involvement [3]. The vascular lesions may be aneurysms and occlusions. Arterial aneurysms are most frequent than occlusive lesions. Aorta, renal arteries, popliteal arteries, and pulmonary arteries are the most involved [4].

Diagnosis of Behçet's disease by hemorrhagic shock is exceptional [2].

The main causes of death in Behçet disease are rupture of arterial aneurysms causing hemorrhagic shock; such was the case of one of our patients [3].

Clinical diagnosis of the aortitis in Behçet disease is difficult because of the lack of specific signs in clinical examination [3]. In fact, the symptoms revealing inflammatory aortitis depond on the affected vascular territory, the type of aortic lesion (aneurysm or occlusive disease) and its localization. Sometimes the aortitis is revealed by non-specific systemic signs or it is a fortuitous discovery [3]. In our patients, it was discovered in case of hemorrhagic shock associated to a chest pain or an abdominal pain.

Standard biology can show an inflammatory syndrome with erythrocyte sedimentation rate (ESR) and C reactive protein (CRP) elevation as it was noted in our patients.

Imaging remains the ideal exam for diagnosis. The aneurysm may sometimes be visible on a chest x-ray, an unprepared abdomen or echo-doppler, but the chest or abdominal angioscanner is the reference exam for aortitis diagnosis. It shows inhomogenic contrast of the aortic wall with perivascular inflammation (95%) [7].

If the angioscanner is contraindicated, an angio MRI may be performed.

The management of aortic aneurysm in patients with Behçet's disease is an emergency. It must be carried out in a multidisciplinary center including several medical and surgical specialities [8]. It is based on two therapeutic components: Medical treatment based on corticosteroid therapy and immunosuppressants, and open or endovascular surgical treatment of the aortic lesion.

The treatment of these complications, in particular aneurysm rupture, requires adequate resuscitation, and an emergent surgical or endovascular management.

Endovascular treatment is most often considered first-line to avoid manipulating highly inflammatory tissue [9,10]. In fact, inflammation of the vascular wall may cause postoperative complications, especially suture dehiscence and anastomotic false aneurysms [8,11]. Some teams prefer endovascular treatment in the acute phase, because of the severe local inflammation and the high risk of surgical complications [8]. However, a few cases of aneurysms in the stent-insertion zone have been reported, caused by the residual local inflammation.

Postoperative complications may involve prosthesis thrombosis, prosthetic-enteric fistulas, and anastomotic false aneurysms [8,11]. This last complication was observed in our second patient who had a ruptured anastomotic false aneurysm.

CONCLUSION

Our two cases suggest that face to an unexplained hemorrhagic shock in a young patient free from atherosclerosis, or with inflammatory arterial disease, complicated inflammatory aortitis should be evocated,

particularly Behçet's disease. The severity of this aortic damage justifies both the systematic screening in patients with Behçet's disease and, when possible, their surgical or endovascular correction associated with corticosteroid and immunosuppressive therapy.

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