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Case Report

Aneurysm of the Ascending Aorta, Rare and Serious Complication of Behçet's Disease

Khalloq R*, Bouamoud A, Zahidi Alaoui H, Malki M, Pr Asfalou I and Pr Benyass A
Mohamed V Military Instruction Hospital, Rabat, CHU Avicenne Rabat, Morocco

***Corresponding authors:** Khalloq R, Mohamed V Military Instruction Hospital, Rabat, CHU Avicenne Rabat, Morocco

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Abstract

Arterial damage during Behçet's disease occurs in 2 to 12% of patients and results in obliterating and/or aneurysmal lesions predominating on the large trunks. Cardiac complications are rarer (1 to 6%) affecting all three tunics.
Ascending aortic aneurysm is a very rare complication of Behcet's disease; the few cases described in the literature were almost always associated with aortic insufficiency.
We report the observation of a 75-year-old patient followed for Behcet's disease, in whom the diagnosis of an asymptomatic ascending aortic aneurysm was retained on transthoracic echocardiography and CT scan, with absence of a leak. aortic something which makes our observation particular.
Through this clinical case we discuss the epidemiological, histological therapeutic and evolutionary particularity of this rare entity.

Keywords: Behcet; Aneurysm; Ascending thoracic aorta; Surgery

Introduction

Behçet's disease (BD) is a systemic vasculitis, described in 1937 by Behçet, with mainly venous tropism. Arterial involvement is only present in 2 to 8% of cases. It has three aspects; these may be stenoses, thrombosis or more often arterial aneurysms.
Since the publication of the first case of aortic aneurysm during Behcet's disease in 1961, the number of published cases has continued to increase [1].

Ascending aortic aneurysm is a very rare complication of Behçet's disease; the few cases described in the literature were almost always associated with aortic insufficiency. We report the case of an aneurysm of the ascending aorta in the context of Behcet's disease, with absence of an aortic leak, which is the particularity of our case.

Clinical Case

Patient aged 75, with cardiovascular risk factors of high blood pressure (hypertension) on dual therapy (ACE inhibitor and amlodipine) well controlled, known carrier of Behcet's disease on colchicine, asymptomatic patient, presents to the department of cardiology for a control transthoracic echocardiography (ETT) as an assessment of the impact of his hypertension. The ETT reveals an aneurysm of the ascending thoracic aorta from the sinotubular junction measuring 59 mm with a sinus of valsalva of normal size and with no aortic leak.

We completed with a cervicothoracic CT angiogram which showed an aneurysm of the ascending thoracic aorta measuring 59 mm in section, extended from the sino-tubular junction to the level of the horizontal thoracic aorta, with circumferential thickening of an inflammatory nature.
The surgical indication was retained, however the patient refused any surgery.

Discussion

The vascular involvement of Behcet's disease is common with male predominance and can present in very varied forms, 37% venous involvement, 12% arterial involvement and 6% cardiac manifestations [1].
Arterial damage is currently better recognized and observed in 3% to 5% of cases depending on the series. This frequency is probably underestimated if we take into account autopsy data where arterial damage is observed in one in three patients.
The time between diagnosis and the appearance of aneurysmal vascular damage is variable; in the literature this period varies from 18 to 53 months [2].

They are described as a panvasculitis affecting small and large arteries with a possibility of aneurysmal and occlusive expression.
Aneurysms are the most common form of arterial expression. They generally complicate a true arterial ulcer and they constitute the first cause of mortality in Behçet's disease. They are

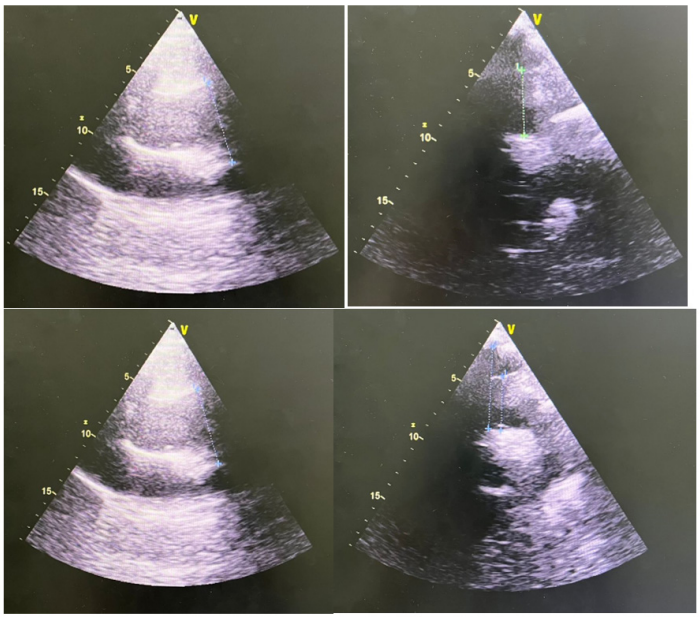

Figure 1: transthoracic echocardiography images showing an aneurysm of the ascending aorta after the sinotubular junction sparing the first portions of the ascending aorta.

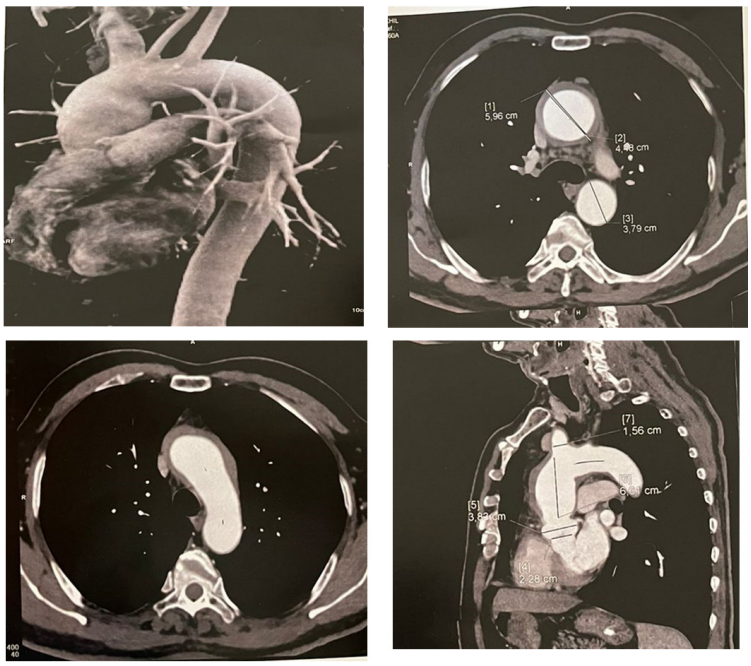

Figure 2: cervicothoracic angioscan images showing aneurysmal dilatation of the ascending thoracic aorta, measured at 59 mm in section, with homogeneous circumferential parietal thickening measured at 5.7 mm and a circulating lumen of 45 mm, originating from the sino-tubular junction to the start of the brachiocephalic trunk arteriosus which measures 15.6 mm in section.

readily multifocal with a great predilection for the abdominal aorta, the femoral and pulmonary arteries, as well as simultaneous aortic and arterial locations. pulmonary or even carotid are not exceptional [2].

Aortic involvement is mainly represented by abdominal aortic aneurysm, however the thoracic location was rarely described in the literature. The cases described in the literature mainly concern the descending thoracic aorta [3]. However, involvement of the ascending thoracic aorta is very rarely described and the few cases described were associated with severe aortic leak [4-7].

Histologically, it is a vasa-vasorum vasculitis devoid of giant cells leading to fragmentation and rupture of the media with deposition of immune complexes. These lesions result either in the formation of a true aneurysm, or in the perforation responsible for a false aneurysm. In affected arteries, infiltrative lesions of the media and adventitia occur initially and are subsequently followed by destructive and fibrous lesions of the media. In the active phase, granulomatous lesions similar to those observed during Takayasu disease are frequently observed. Saccular aneurysms are probably the result of severe destruction of the media by intense inflammatory reactions [2]. The direct role of vascular trauma is well known, illustrated by the occurrence of aneurysms at the arterial puncture point or at the suture point of vascular bypasses, performing a true pathergy test at the level of the artery. The role of overconsumption of tobacco is highlighted in some studies. This iatrogenic risk of arterial punctures must be taken into account in the choice of vascular explorations favoring digitalized venous angiograms, CT angiography and MRI angiography. The PET scanner can also find an interesting indication in these shapes [2].

The main therapeutic objective is to exclude intra-aneurysmal flow, in order to stop the progression of aneurysmal dilatation, and thus prevent rupture [2].

Drug therapy with corticosteroids and immunosuppressants adjuvant to surgery has shown its effectiveness and superiority compared to surgical treatment alone in terms of reduction in the percentage of recurrence in the short and long term. Cyclosporine, azathioprine, tumor necrosis factor (TNF) inhibitors and interferon have revolutionized the treatment of Behcet's disease. The ideal chronology of treatment is to "cool" the active phase by initiating boli of corticosteroids and immunosuppressants as soon as possible with the aim of reaching a quiescent phase as quickly as possible before surgery [2, 5]. Apart from critical patients, it is suggested that the operation be performed until the inflammatory markers are diminished.

Surgery of the ascending aorta poses a problematic tripe. It includes, on the one hand, the treatment of the aorta itself, on the other hand that of the aortic valve and the management of the coronary ostia. The surgical arsenal is broken down into multiple interventions ranging from supracoronary replacement of the ascending aorta to the intervention, described by Bentall in 1968, removing the entire ascending aorta, the aortic valve and requiring the reimplantation of the coronary ostia. Finally, during the 1980s and 1990s, Sir Magdi Yacoub [7] then Tiron David [8] introduced the notion of preserving the aortic valve during these radical surgeries of the aortic root, thus avoiding prosthetic valve replacement.

Aortic surgery in the context of Behcet's disease requires special technical precautions, in particular suturing techniques due to the high risk of reported postoperative complications such as suture release with the appearance of a false aneurysm or even detachment of the valve. [9].

In a recent Korean study, aneurysmal recurrence was closely correlated with the presence of a positive pathergy test.[10]

The prognosis of arterial damage is mainly aortic and extremely severe, arterial damage is the main cause of death during Bechet's disease, however the prognosis has improved with immunosuppressive treatment, allowing the reduction of post complications. surgery and recurrences [1].

Conclusion

Arterial damage during Behcet's disease is the main cause of death. The thoracic aorta is rarely affected, especially the ascending aorta which is almost exclusively associated with damage to the aortic valve. The development of new suturing techniques and the coverage of the surgical procedure with immunosuppressive treatment have improved the results of Bechet's aortic surgery.

References

1. Ajili F, et al. A saccular aneurysm of the abdominal aorta revealing Behçet disease: when to operate?]. Pan Afr Med J, 2014; 19: 252.
2. Mleyhi S, et al. A rare localization of angio-Behçet revealed by a false aneurism of a gluteal artery treated surgically]: J Med Vasc, 2019; 44(3): 228-232. doi: 10.1016/j.jdmv.2019.02.006.
3. Wang M, Bartolozzi LM, Riambau V. Total endovascular treatment for thoraco-abdominal aortic aneurysm in a patient with Behçet's disease: Case report and literature review. Vascular, 2021; 29(5): p. 661-666.
4. Chetoui A, et al, Aneurysm of the ascending aorta associated with massive aortic regurgitation: rare and serious complication of Behçet disease]. Pan Afr Med J, 2015; 21: 85.
5. Tang Y, Xu J, Xu Z, Supra-annular aortic replacement in Behcet's disease: a new surgical modification to prevent valve detachment: Int J Cardiol, 2011; 149(3): 385-386. doi: 10.1016/j.ijcard.2011.03.007.
6. Okita Y, et al. Multiple pseudoaneurysms of the aortic arch, right subclavian artery, and abdominal aorta in a patient with Behçet's disease. J Vasc Surg, 1998; 28(4): p. 723-726.
7. Yacoub MH, et al. Late results of a valve-preserving operation in patients with aneurysms of the ascending aorta and root. The Journal of Thoracic and Cardiovascular Surgery, 1998; 115(5): p. 1080-1090.
8. David TE, Feindel CM. An aortic valve-sparing operation for patients with aortic incompetence and aneurysm of the ascending aorta. J Thorac Cardiovasc Surg, 1992; 103(4): p. 617-621.
9. Ando M, et al. Surgical treatment of Behçet's disease involving aortic regurgitation. Ann Thorac Surg, 1999; 68(6): p. 2136-2140.
10. Tazi-Mezalek Z, Ammouri W, Maamar M. Les atteintes vasculaires au cours de la maladie de Behçet. la revue de médecine interne, 2009; 1496(1004): p. S225-S492.