ARTERIAL APHTA: WHICH VALUE IN BEHCET DISEASE?

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ABSTRACT

The Behcet disease is a vasculitis with polyorganic attacks whose diagnosis is established by the international study group for behcet disease (ISGBD). The asynchronous occurrence of symptoms makes sometimes the diagnosis more difficult, leading to diagnosis and therapeutic delay. The arterial ulcer or aphta has currently, no diagnostic value in the criteria of ISGBD. Its morphological similarity and most likely pathophysiological also, with bipolar ulcers deserves attention.

This article is an observation relating the history of a patient who does not respond to the criteria for ISGBD. The tradive occurrence of minor criterion was a cause of diagnostic and therapeutic delay. The early presence of arterial aphta did not change the therapeutic attitude even if the Behcet disease is known by its vascular tropism. The occurrence and the value of such manifestation remains not discussed in the literature.

KEY WORDS: Behcet disease, arterial aphta, arterial aneurysm

1. INTRODUCTION

Behcet's disease is a systemic vasculitis characterized by the occurrence of aphtas in the mouth and genital area. Diagnosis is based on clinical criteria established by the international study group for Behcet disease (ISGBD)[2] (Tab 1).

Behcet's disease presents a diagnostic challenge; in fact, the diagnosis is not always easy since the criteria established by ISGBD can be spread over time or incomplete. In case of incomplete criteria, the lack of treatment expose to the occurrence of complications.

We report the case of a patient who had an arterial lesion characterized by the presence of aphtoide ulceration at the wall of an aneurysm of the abdominal aorta. This lesion reveals the existence of Behcet's disease. The incomplete criteria delayed the specific treatment enhancing risks.

2. OBSERVATION

A 40-year old patient of Mediterranean origin, without any cardiovascular risk factor, who had a sural phlebitis, 8 years later, that favorably responds to medical treatments.

This patient had also a history of recurrent oral aphtas, and underestimated the need for a medical check-up. The thrombophilia tests did not reveal any anomaly. Abdominal ultrasound performed to check out an abdominal pain, allowed to find out an aneurysm of the abdominal aorta confirmed by angiography.

The abdominal CT angiography had shown two aneurysms (FIG 1), the first is from the subrenal abdominal aorta, which is with a 5 cm diameter. The second is from the right common iliac artery.

In addition, we noticed the presence of two arterial ulcers giving an aspect of "tubercles" on the lateral sides of the aortic aneurysm.

The patient was referred to us to deal with his two aneurysms.

The etiological investigation was very poor, except the history of thrombophlebitis and recurrent oral aphthous ulceration.

The active surgical treatment decision was based on the voluminous and painful aspect of the aneurysm.

The presence of arterial ulcer was considered as a pre-rupture factor.

The surgical management consisted in an open approach to perform an aorto biiliac bypass grafting.

The opening of the aneurysmal dome showed two arterial aphthous lesions (FIG 2) corresponding to the ulcerations seen on CT scan images. The postoperative course was uneventful.

The young age of the patient, the Mediterranean origin and the presence of oral aphta and arterial ulcer were suspicious for Behcet disease without further confirmation.

The patient was treated by antiplatelet therapy with favorable evaluation to a month consultation.

After 6 months, the patient was examined, presenting a recent genital aphta and polyarthralgia.

Thus, the diagnosis of Behcet disease was clearer taking into consideration the new completed diagnosis triad. The patient was treated with corticosteroids and immunosuppressants.

3. DISCUSSION

Behcet's disease is a vasculitis of unknown etiology, affecting especially young adult males. It is more common in the Mediterranean, the Middle East and Asian countries [6,4]. Diagnosis is based on clinical criteria established by ISGBD (FIG).

The prevalence of vascular involvement in Behcet's disease (angio-behcet) is estimated between 6

and 38% of cases [4], it includes even venous or arterial lesions.

Venous disease is most common in all series except in autopsy series of Matsumoto and al [3], which indicate that artery disease is the most common probably due to its asymptomatic nature. All arteries of the body can be involved. The large trunks are more exposed [1].

The arterial lesion is most often aneurysmal [8].

The arterial aneurysm is secondary to the local inflammatory process initiated by the deposition of circulating immune complexes.

This results in a disruption of the internal and external elastic lamina, an intimal thickening and degeneration of the tunica media.

The association with vasculitis of the vasa vasorum generates complete rupture of the tunica media and the constitution of the aneurysm (7).

Sometimes these inflammatory phenomena lead to perforation of the aneurysm. The areas of arterial aphtas, which are true deep ulcers of the aneurysmal wall, are, in our minds, the most likely areas for perforation.

Their presence indicates an increased risk of perforation and should prompt an urgent surgical care.

Apart from the numerous writings in the literature discussing arterial aphtas in Behcet's disease, no data exists, to our knowledge, assessing its impact or its true value in Behcet's disease.

Could the particular macroscopic appearance of arterial aphta, very close to the oral and genital aphtas, and the probable inflammatory origin, be allowing to evoke tripolar aphta?

Adjuvant medical treatment based on immunosuppressants and corticosteroids is required to prevent recurrence (8).

4. CONCLUSION

Arterial aphta is a vascular manifestation sometimes encountered in Behcet's disease. Its analogy with oral and genital aphtas raises the question of its true diagnostic value.

Studies are needed to assess its prevalence and to check its relationship with Behcet's disease.

The authors declare no conflict of interest.

Figures and tables

FIGURE 1: CT angiography showing an aneurysm of the abdominal subrenal aorta with two tubercles on the superolateral left side. Also a second aneurysm of the common right iliac artery

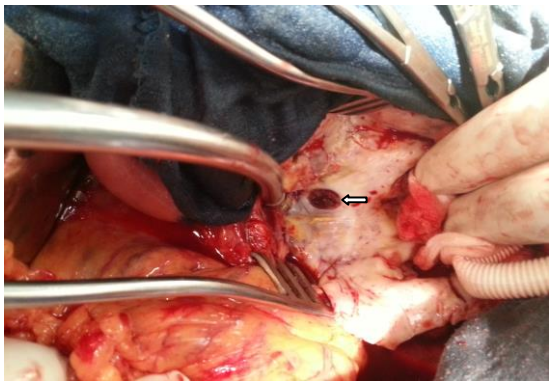

FIGURE 2: arterial aphta corresponding to the tubercle on CT angiography.

Table 1: Criteria of the International Study Group for Behçet's Disease (1990)

Recurrent oral ulceration	Minor aphthous, major aphthous, or herpetiform ulceration observed by physician or patient, which recurred at least 3 times in one 12-month period
Plus 2 of:	
Recurrent genital ulceration	Aphthous ulceration or scarring, observed by physician or patient
Eye lesions	Anterior uveitis, posterior uveitis, or cells in vitreous on slit-lamp examination; or retinal vasculitis observed by an ophthalmologist
Skin lesions	Erythema nodosum observed by physician or patient, pseudofolliculitis, or papulopustular lesions; or acneiform nodules observed by physician in postadolescent patients not on corticosteroid treatment
Positive pathergy test	Read by physician at 24-48 hr

Reference :

1. Hyung-Kee Kim, Hyang Hee Choi, Seung Huh. Anévrysme rompu de d'artère iliaque associé à un faux-anévrysme aortique chez un patient présentant une maladie de Behçet. Annales de chirurgie vasculaire. 2010;Vol 24(2):27e7-27e11.

2. International Study Group for Behcet's Disease. Criteria for diagnosis of Behcet's disease. Lancet. 1990 May 5;335(8697):1078-80.

3. Matsumoto T, Uekusa T, Fukuda Y. Vasculo-Behçet's disease: a pathologic study of eight cases. Hum Pathol. 1991 Jan;22(1):45-51.

4.Ouazzani B, et al. Devenir de la maladie de Behçet en milieu ophtalmologique marocain. J Fr Ophtalmol 1995;18:373-5.

5. S. B'chir Hamzaoui,et al. La maladie de Behçet en Tunisie. Étude clinique de 519 cas. Rev med int 2006 ; Vol 27 - N° 10 P. 742-750 .

6. Suzuki Kurokawa M, Suzuki N. Behcet's disease. Clin Exp Med 2004;3:10-20.

7. Wechsler B, Du LT, Kieffer E. Manifestations cardio-vasculaires de la maladie de Behçet. Ann Med Interne 1999;150:542-54.

8. Younes Bensaid, Brahim Lekehal, Abbès El Mesnaoui, Zakariyae Bouziane, Nabil Sedki . Complications artérielles de la maladie de Behcet : à propos de 47 cas .Arterial complications of Behçet's disease. e- mémoires de l'Académie Nationale de Chirurgie, 2008, 7 (2) : 54-59 .

