

## Endovascular repair of a thoracoabdominal pseudoaneurysm in a patient with Behçet's disease

### *Correção endovascular de pseudoaneurisma toracoabdominal em paciente com Doença de Behçet*

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#### Abstract

Behçet's disease is an inflammatory disease characterized by recurrent oral and genital ulcers, uveitis and skin lesions. Arterial involvement is rare, but when present aneurysmal degeneration is more common than occlusive disease. This report describes the clinical case of a female patient who had been receiving treatment for Behçet's disease for twenty years before presenting with abdominal pain that progressed for 2 months before suddenly worsening significantly. A CT scan revealed a thoracoabdominal pseudoaneurysm. She was successfully treated with endovascular repair.

**Keywords:** Behçet's disease; aortic aneurysm; dissecting aneurysm; endovascular procedures.

#### Resumo

A Doença de Behçet é uma doença inflamatória caracterizada por úlceras genitais e orais recorrentes, uveítes e lesões cutâneas. O envolvimento arterial é raro, sendo mais comuns as degenerações aneurismáticas do que as oclusivas. Neste caso clínico, paciente do sexo feminino, em tratamento de doença de Behçet havia 20 anos, iniciou com dor abdominal progressiva por dois meses, com piora súbita importante, foi submetida à tomografia computadorizada, que mostrou pseudoaneurisma toracoabdominal. A paciente foi submetida a tratamento endovascular com sucesso.

**Palavras-chave:** Doença de Behçet; aneurisma aórtico; aneurisma dissecante; procedimentos endovasculares.

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## INTRODUCTION

Behçet's Disease (BD), or syndrome, is a systemic inflammatory disease characterized by recurrent urogenital or oral ulcers, uveitis and cutaneous lesions.<sup>1</sup> In around 8% of cases there is arterial involvement, in which aneurysmal degeneration is more common than occlusive disease.<sup>2,3</sup> Aneurysms have been reported in several arterial locations, such as the aortic arch and pulmonary artery,<sup>4</sup> celiac trunk,<sup>5,6</sup> superior mesenteric artery,<sup>7</sup> carotid arteries,<sup>8</sup> brachiocephalic trunk,<sup>9</sup> abdominal aorta,<sup>10-12</sup> iliac artery,<sup>13</sup> deep femoral artery<sup>14</sup> and popliteal artery.<sup>15</sup> There are few descriptions of aneurysms involving the thoracoabdominal segment of the aorta,<sup>11,13,16</sup> or of aortic pseudoaneurysms.<sup>2,16</sup> We therefore report on such a case, which was successfully treated using endovascular techniques.

## CASE DESCRIPTION

The patient was a 36-year-old black female who had been receiving treatment for BD from a rheumatologist for 20 years and was on a continuous prescription for 20 mg/day of prednisone. She also had systemic arterial hypertension and reported that she did not smoke and did not have diabetes mellitus, infections or traumas. The signs and symptoms characteristic of BD exhibited by the patient were a history of recurrent painful oral ulcers and genital ulcers and pustules, but she did not have uveitis or other cutaneous lesions. She presented at the Hospital with abdominal pain in an area between the mesogastrium and epigastrium, with onset around 2 months previously and progressive worsening over time. Four days previously she had received medical attention in her home town and a Computed Tomography (TC) scan without contrast had revealed a thoracoabdominal aneurysm. She was then referred to our service.

On physical examination at admission the patient was hemodynamically stable, normal mucosal color, with blood pressure of 180/100 mmHg in both upper limbs and all peripheral pulses were symmetrical. Cardiac auscultation revealed normal sounds and pulmonary auscultation was free from adventitial sounds. She was not suffering from any type of neurological deficit. Routine laboratory work-up test results were normal. An angiotomography scan with contrast showed three saccular dilations with diameters of 0.9 cm in the proximal thoracic aorta (2.0 cm), the thoracic descending aorta (1.0 cm) and the abdominal aorta (1.0 cm). The scan also showed a pseudoaneurysm that started at the thoracoabdominal transition and continued to 1.0 cm above the celiac trunk, 7.0 × 7.8 cm in diameter, with mural thrombus and compression of the arterial lumen (0.9 cm in diameter) (Figures 1, 2 and 3).



Figure 1. 3D angiotomography reconstruction showing thoracoabdominal pseudoaneurysm.



Figure 2. Computerized angiotomography slice showing thoracoabdominal aortic pseudoaneurysm.



Figure 3. Angiotomography slice showing images of small saccular aneurysms in initial portion of thoracic aorta.

In view of the considerable abdominal pain, with lumbar involvement, and the spontaneous and symptomatic aortic pseudoaneurysm, urgent treatment was indicated. Both open and endovascular surgical treatment options were possible and an aortic endoprosthesis was available on site.

After completion of a free and informed consent form, the patient underwent endovascular repair of the aortic pseudoaneurysm. Via surgical access obtained in the right common femoral artery and the left brachial artery, an aortography was conducted using a pigtail catheter with a centimeter scale (via the femoral access). Another pigtail catheter was advanced into the thoracic descending aorta via the brachial access. The endoprosthesis that was available in the department was implanted (a tubular proximal abdominal extension AFX 95X25MM – ENDOLOGIX®). A control arteriography showed that endoprosthesis placement had been successful and was free from leaks. The patient was sent to the ICU, where she remained in intensive care for 2 days. She was extubated early on during this period, did not need vasoactive drugs and did not

exhibit neurological deficit. She was discharged from hospital 6 days after the surgical procedure and an angiogram scan showed that endovascular repair of the pseudoaneurysm had been successful (Figures 4 and 5). Over a 24-month follow-up period the patient attended routine clinical consultations and was assessed using CT of the thorax, remaining free from intercurrent conditions.

## DISCUSSION

Behcet's disease is diagnosed on the basis of clinical symptoms,<sup>1</sup> using criteria set out by the International Study Group for Behcet's Disease (Table 1). This patient's diagnosis and treatment were managed by a rheumatologist and, of the diagnostic criteria, she only had recurrent oral and genital ulcerations (one required criterion + one minor criterion). The images of multiple saccular aneurysms on the angiogram scan conducted at our service provided further evidence of this diagnosis.

Vascular involvement is estimated to occur in 7 to 29% of BD cases and can be in arterial or venous territories.<sup>17,18</sup> The most often involved artery is the aorta, followed by the femoral and pulmonary arteries,<sup>4</sup>



Figure 4. 3D angiogram reconstruction showing endoprosthesis repair of aortic pseudoaneurysm.

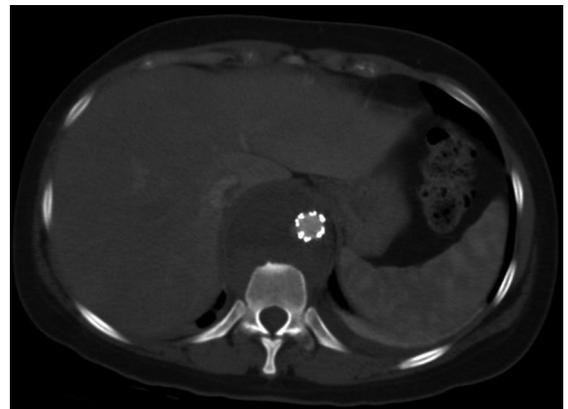


Figure 5. Angiogram slice showing endoprosthesis repair of aortic pseudoaneurysm.

Table 1. International criteria for classification of Behçet's disease.

Criterion	Characteristics
<b>Major criterion (required)</b>	
Recurrent oral ulceration	Major, minor or herpetiform*, 3 times in one 12-month period
<b>Minor criteria</b>	
Recurrent genital ulceration:	Aphthous scarring
Eye lesion	Anterior uveitis, posterior uveitis, or cells in vitreous (slit lamp), retinal vasculitis
Skin lesions:	Erythema nodosum*, pseudofolliculitis, papulopustular lesions, acneform nodules* (in post-adolescent patients not on corticosteroid treatment)
Positive pathergy test	24-48 hours*, oblique insertion of 20 caliber needle

\*Observed by physician. †Reliable patient report.

with aneurysmal degeneration accounting for 65% and occlusive disease for 35%.<sup>19</sup> The abdominal aorta is the segment of the aorta most often involved, and saccular aneurysms are most frequent among BD patients.<sup>20</sup> The pathophysiology of vascular involvement is related to vasculitis that causes occlusion of the vasa vasorum and necrosis of the vessel wall, provoking weakness that can lead to dilation or occlusion.<sup>21</sup> Immunohistochemical studies confirm the presence of complement and immunoglobulins in the tunica media and tunica intima of arteries.<sup>10</sup> Occlusion of the vasa vasorum and/or intramural hematoma are considered the most important factors in this type of aortic condition.<sup>22</sup> The pseudoaneurysm in the case described here was possibly formed by the rupture of a small, localized, saccular aneurysm, with contained blood leakage. Reports in the literature of spontaneous pseudoaneurysms of the aorta are rare,<sup>2</sup> and anastomotic pseudoaneurysms after open reconstruction of the aorta are more common.<sup>23</sup> Clinical treatment of BD is based on corticosteroids and/or immunosuppressants, which can protect patients from arterial inflammatory aggression. Notwithstanding, this patient developed vascular complications despite prolonged treatment with corticosteroids. As endovascular surgery has developed, it has become the preferred choice for surgical treatment of arterial aneurysms, including those related to BD.<sup>2,10,24</sup> However, there is not yet consensus in the literature on this indication. The presence of significant symptoms that did not undergo remission with clinical treatment and the risk of rupture were determinant factors in prescribing emergency treatment. In this case, the long proximal tube extension of an abdominal endoprosthesis was used because the thoracic endoprostheses available at the time of treatment did not offer suitable dimensions. In principal, the primary shortcoming would be the shorter length of the delivery systems of these proximal abdominal extensions, but this was not a problem in this case because the patient was brevilineal. The delivery system was long enough and the endoprosthesis fitted the thoracoabdominal aorta well, since its length and diameter were suitable.

As a routine, antiplatelet drugs should be prescribed after endovascular treatment of aortic aneurysms.<sup>25</sup> On the other hand, clinical control of BD is dependent on the degree of activity of the underlying disease and on correct corticosteroid and immunosuppressant dosages. Some authors suggest routine maintenance of immunosuppression to avoid recurrence of pseudoaneurysms at the extremities of endoprostheses.<sup>2,23</sup>

## CONCLUSIONS

Arterial aneurysms and pseudoaneurysms are not common conditions in BD, but, on the basis of reports in the literature, endovascular treatment appears to be an ever more widely employed option. In the case described here, endovascular treatment proved to be an appropriate and effective option.

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