

Endovascular treatment of aortic aneurysms in patients with Behcet's disease: report of two cases

Tratamento endovascular de aneurismas da aorta em pacientes com doença de Behçet: relato de dois casos

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Abstract

Behcet's disease, a systemic vasculitis of unknown etiology, may be the cause of aortic aneurysmal diseases in some patients. We report our experience with two Behcet's disease patients who presented with aortic aneurysms and were submitted to endovascular therapy, and describe their respective follow-ups. Current pathophysiology, diagnosis, and treatment approaches were reviewed. Our experience suggests that the endovascular approach, combined with adequate immunosuppressive treatment, is an excellent therapeutic option for some patients with Behcet's disease suffering from aneurysms.

Keywords: Behcet's disease, aortic aneurysm, endovascular treatment.

Resumo

A doença de Behçet, uma vasculite sistêmica de causa desconhecida, pode ser causa de doença aneurismática da aorta em alguns portadores dessa patologia. Nós apresentamos nossa experiência com dois casos de aneurismas aórticos em pacientes com doença de Behçet submetidos à terapêutica endovascular, descrevendo seus respectivos seguimentos. A terapêutica atual, a patofisiologia e os critérios diagnósticos vigentes foram revisados. Concluímos que a técnica endovascular é uma excelente opção terapêutica para certos pacientes com doença de Behçet e que esta deve ser acompanhada de tratamento imunossupressivo adequado.

Palavras-chave: Doença de Behçet, aneurisma aórtico, terapia endovascular.

Introduction

Behcet's disease, a systemic vasculitis of unknown etiology, is characterized by recurrent oral and genital ulcers, ocular manifestations and skin lesions.¹ Cardiovascular manifestations may occur in around 7-38% of patients;¹ it is, thus, the most common cause of death among these patients.²

Peripheral vascular lesion is rare, and arterial lesions are less common than venous disease; its prevalence is around 1.5-3% in the whole world.² In venous involvement, thrombosis is the most frequent event, occurring even in adequate anticoagulation.³ Aneurysm is more common than occlusion in Behcet's disease arterial involvement, and

the most common site for aneurysm formation is the abdominal aorta, followed by the femoral artery and the pulmonary arteries.⁴

Among all the vascular lesions that may occur in these patients, arterial aneurysms represent the most difficult and challenging pathology for the vascular surgeon due to its technical difficulties and high recurrence.⁵ Due to its rapid expansion, with a high rate of rupture, Behcet's disease aneurysm should be readily treated. The most suitable treatment for aortic aneurysms in Behcet's disease patients is still a controversial issue. Endovascular treatment is an important alternative for high-risk patients, once morbidity and mortality rates after conventional surgery remain high.⁶

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Two cases of aortic aneurysms in Behcet's disease patients, corrected with endovascular technique, are presented, reporting their respective mid-term follow-ups, and current physiopathology, diagnostic criteria and treatment are reviewed.

Case 1

A 28-year-old Caucasian woman diagnosed with Behcet's disease at 23 years of age, due to recurrent oral and genital ulcers, pseudofolliculitis and erythema nodosum, presented reporting pain in the lower posterior part of the abdomen for 6 months. She had no ophthalmologic abnormalities or positive family history of the disease. At physical exam a pulsatile abdominal mass was found.

Initial ultrasonography revealed a saccular aneurysm of the abdominal aorta above the celiac trunk. Angiotomography confirmed a 5.7 x 4.7-cm saccular aortic aneurysm above the celiac trunk, in addition to occlusion of this ramus (Figures 1 and 2). Superior mesenteric artery (SMA) was patent with stenosis in its origin (Figure 3).

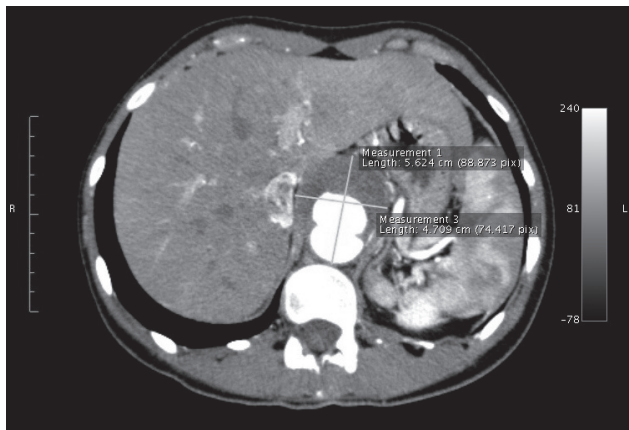


Figure 1 - Angiotomography of the aorta: 5.7-cm diameter saccular aneurysm of abdominal aorta above the celiac trunk

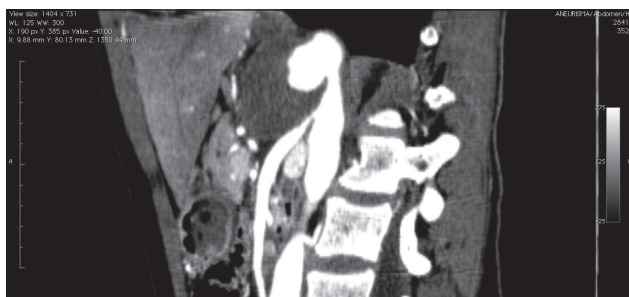


Figure 2 - Angiotomographic reconstruction: stenosis in the origin of superior mesenteric artery

Distal abdominal aorta and iliac arteries had their flow and diameter preserved.

After 1 month of optimized corticotherapeutic treatment (prednisone 30 mg/day) and normalized systemic inflammatory signs, surgery was indicated. Endovascular treatment of aortic aneurysm was performed with a Zenith® (Cook, USA), 22 x 115 mm endoprosthesis implant, associated with SMA angioplasty with a Genius® (Eurocor, Germany), 6 x 39 mm, stent graft. A proximal extension was necessary, due to the presence of type 1 endoleak (Zenith®, 24 x 115 mm). Intraoperative arteriography at the second procedure evidenced no leaks and unobstructed artery.

The patient had a favorable evolution during postoperative. A control angiotomography was performed, which did not evidence any abnormalities. She was discharged on the 8th postoperative day, maintaining immunosuppressive treatment.

On the sixth postoperative month, an angiotomography was performed to confirm aneurysm exclusion (Figure 4) and SMA stent patency (Figure 5). After a 1-year follow up, the patient remains asymptomatic and has no complaints.

Case 2

A 33-year-old man, with a history of visual loss resulting from successive posterior and anterior uveitides,



Figure 3 - Tridimensional reconstruction: thoracabdombinal aorta, relation of aneurysm with superior mesenteric artery

presented with progressive deterioration of the renal function. A systemic vasculitis was suspected and the patient underwent arteriography, which demonstrated a descending thoracic aortic aneurysm with 5-cm-diameter (Figures 6, 7 and 8). The Department of Rheumatology of the FMUSP defined the diagnosis as Behcet's disease.

While hospitalized, the patient complained about pain in the chest and non-specific thoracic pain. Physical exam

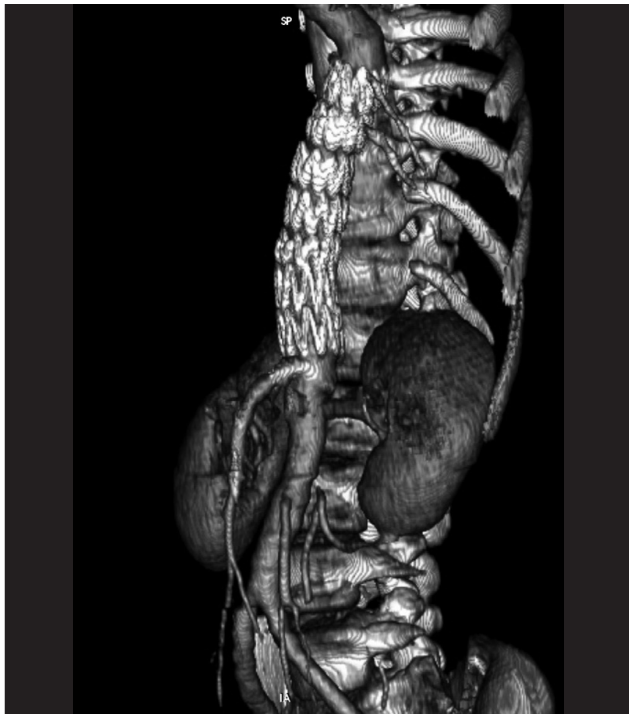


Figure 4 - Tridimensional angiotomography: well-placed endoprostheses, absence of leaks

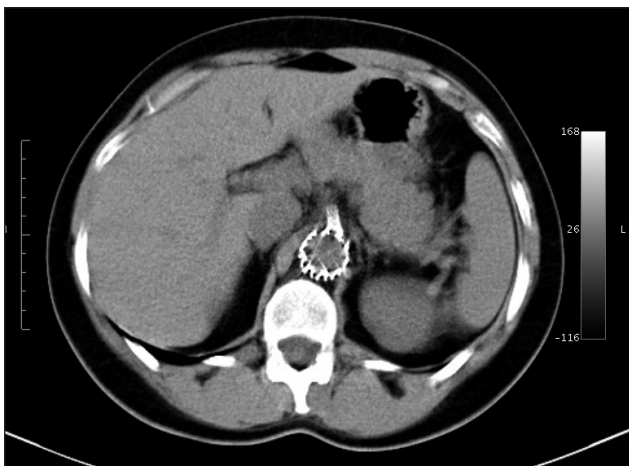


Figure 5 - Axial cut: patent stent graft of the superior mesenteric artery



Figure 6 - Aortic arch: aneurysm below subclavian artery



Figure 7 - Aortography: 5-cm aneurysm in descending thoracic aorta



Figure 8 - Aortography: distal neck of the aneurysm above celiac trunk

indicated bilaterally decreased femoral and distal pulses and normal pulses in the upper limbs. There was no significant difference in blood pressure between the arms. Emergency intervention was chosen due to acute symptoms.

An endovascular treatment of the thoracic aortic aneurysm was performed with the implant of a Talent® (Medtronic,



Figure 9 - Angiotomography after 18 months: absence of leaks

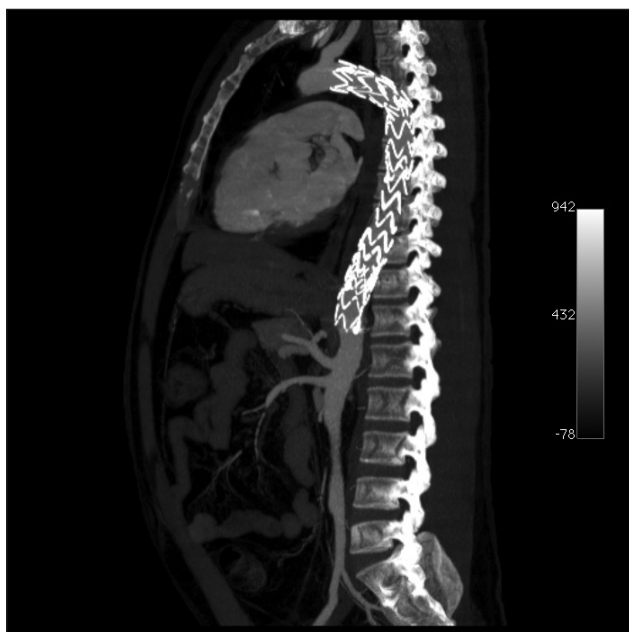


Figure 10 - Angiotomographic reconstruction: patent subclavian artery and celiac trunk

USA), 22 x 130 mm endoprosthesis, and a Talent®, 24 x 130 mm distal extension, maintaining the left subclavian artery's patency. At control arteriography, a type 1 distal leak was found, which was corrected with the implant of a third Talent®, 24 x 130 mm prosthesis, 3 cm above the celiac trunk.

Postoperative had no intercurrents. Follow-up angiotomography was performed on the seventh postoperative day, revealing neither leaks nor patency of the prosthesis (Figures 8, 9, 10 e 11).

The patient was discharged on the eighth postoperative day with prednisone 30 mg/day and monthly pulse therapy with cyclophosphamide for disease remission. Angiotomography performed at 18 months' follow-up showed absence of leaks, endoprosthesis patency and absence of recurrent aneurysm signs.

Discussion

Behcet's disease was first described in 1937 by Turkish dermatologist Hulusi Behcet as a systemic inflammatory disease which classically causes oral and genital ulcers, in addition to ocular inflammation.⁷ It is more common in men. In Japan it is the second most common cause of acquired blindness.⁸ And it may also affect the vascular system. Behcet's disease cause is still not clear. There are no



Figure 11 - Tridimensional reconstruction: overlapping well-placed endoprostheses

pathognomonic laboratorial tests or specific histological finding for Behcet's disease. Therefore, the diagnosis is based on clinical findings, according to criteria established by the International Study Group for Behcet's Disease (Table 1).

The spectrum of the vascular disease is broad and singular in Behcet's disease. The frequency of vascular involvement is estimated in 7-38% of cases.¹ Vascular lesions may be arterial or venous. Usually the most affected artery is the aorta, followed by femoral and pulmonary arteries; 65% of these involvements are aneurysms and 35%, occlusions.^{9,10}

Aneurysms in Behcet's disease are in many aspects different from degenerative aneurysms. Incidence is higher among younger persons; the suprarenal type is the most common one and the aneurysm's format is, as described in case 1, usually saccular. They are often multiple, presenting urgent symptoms.¹¹⁻¹³ The commonest complication of aneurysm is rupture, which is the most frequent cause of death related to vascular complication.

Pathogenesis of arterial lesions is considered to be related to arterial vessels and wall vasculitis. Then, the formation of a pseudoaneurysm caused by the obliteration of the vessels of the vasa vasorum due to the inflammatory process follows, which results in the rupture of the nutrition flow and in necrosis of the aortic wall.¹⁴

Immunohistochemical studies have confirmed an accumulation of complement and immunoglobulin in the medial and adventitial layer. There is a thickening of the intima, where the internal and the external elastic lamina, medial and adventitial are ruptured. The destruction of the medial layer seems to be responsible for the development of aneurysm dilation. Lesions of the aortic wall are frequently located and observed in the saccular type.¹³ Reports indicate that these aneurysms are usually ruptured, with no correlation with the size of the dilation.¹⁵

External manifestations, such as active uveitis, aphthas and genital ulcers, and also internal manifestations, such as cardiovascular complications, are strongly related to vascular disease exacerbation. If really necessary, surgeries should be performed when the disease is totally controlled with corticoids and immunosuppressants, because any intervention may lead to posterior complications, such as new occlusions, aneurysms or pseudoaneurysms.¹⁶ In emergency cases, as a rupture, the remission state of the patient should be reevaluated. It has been shown that, before intervention, the attack dose of corticoids/immunosuppressants in bolus (e.g., prednisolone) may decrease perioperative and postoperative vascular complications.¹⁷ However, the role of immunosuppressant and antiinflammatory medications in the

Table 1 - International criteria for Behcet's disease

Criterion	Characteristics
Major criterion (mandatory)	
Recurrent oral ulceration	Big, small or herpetiform,* 3 times in 12 months
Minor criteria	
Recurrent genital ulceration	Aphthous scarification
Eye lesion	Anterior/posterior uveitides <i>in vitreo</i> (Slit lamp), retinal vasculitis*
Cutaneous lesions	Erythema nodosum*† Pseudofolliculitis, papulopustular lesions, acneiform noduli* (after adolescence, without corticosteroid use)
Positive pathergy test	24-48 hours*, oblique insertion of 20-caliber needle

* Observed by the physician.

† Reliable report by the patient.

prevention of recurrent arterial lesion after intervention in Behcet's disease also needs posterior evaluations. In both cases, pharmacological control of the disease's activities involves the use of corticosteroids and immunosuppressants.

Open surgical repair was previously the definitive treatment for vascular lesions in Behcet's disease patients, but surgery's poor result in vascular/Behcet's disease is usually attributed to postoperative vascular complications, such as pseudoaneurysm and graft occlusion.^{18,19} Aiming at preventing surgical complications, endovascular methods, e.g. endoprostheses, have recently been recommended due to being less invasive.²⁰

Recent studies reveal that patients receiving endovascular treatment present lower operative time, hospital stay and blood loss than those who undergo open surgical repair, with a lower mortality rate (0.6-3.5%). Success rate in prostheses release is high (90% in low-risk patients and 80% in moderate-to-high-risk patients), although some limitations still exist, e.g. the size of the delivery systems, leaks and the position of main trunks.²¹ There is a clear possibility of relapse in Behcet's disease after endovascular treatment, in which interrupted immunosuppressive therapy is a risk factor.²² However, Park et al.⁹ indicate that relapsing may be controlled through the repeated use of endoprostheses.

We conclude that endovascular treatment is certainly an excellent choice for selected cases of Behcet's disease. However, the state of the disease should be carefully controlled with corticosteroid and/or immunosuppressant medication. Long term follow-up is fundamental for evaluating interurrences and complications of endovascular therapy, besides recurrence surveillance inherent to Behcet's disease.

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