



Ruptured Suprarenal Abdominal Aortic Pseudoaneurysm with Superior Mesenteric and Celiac Arteries Occlusion, Revealing Behçet's Disease: A Case Report

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Behçet's disease (BD) is a multisystemic, chronic autoimmune inflammatory vasculitic disease with an unknown etiology. Although the literature reports that vascular involvement occurs in 7% to 38% of all BD cases, the arteries are rarely involved; however, arterial involvement is usually associated with significant mortality and morbidity. We report the case of a young female patient who presented to the emergency department with severe abdominal pain and a history of weight loss. The patient was evaluated using computed tomography angiography, which revealed a ruptured suprarenal aortic pseudoaneurysm with occlusion of both the superior mesenteric and celiac arteries. Urgent surgery was performed with aortic repair with an interposition graft and superior mesenteric artery embolectomy. The patient's clinical history and radiological imaging findings were strongly suggestive of the diagnosis of BD with vascular involvement.

Key Words: Behcet disease, False aneurysm, Superior mesenteric artery, Suprarenal aorta, Vasculitis

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INTRODUCTION

Behçet's disease (BD), originally described by Hulusi Behçet in 1937, is a chronic inflammatory relapsing vasculitis of an unknown etiology that involves multiple organ systems [1]. In 1990, the International Study Group for BD recommended a set of diagnostic criteria requiring the presence of oral ulceration plus any two of the following: genital ulceration, typical defined eye lesions, typical defined skin lesions, or a positive pathergy test [2]. BD peaks during the third and fourth decade of life and affects both sexes, with a female predominance [3]. The highest prevalence of the disease was reported in Turkey and Iran. Nevertheless, it has been reported worldwide with different

prevalence rates [4,5].

Vascular involvement in BD is referred to as vasculo-BD and has been reported in 7% to 38% of patients with BD [2]. Both veins and arteries of any size can be affected, with venous thrombosis and occlusion being the most common pathology. Although arterial involvement is uncommon, it is associated with severe complications, with ruptured arterial aneurysm being the leading cause of death in patients with BD [6,7].

Cardiac involvement is a rare but life-threatening complication of BD. It has been reported in 1% to 5% of cases, with pericarditis being the most common pathology. Other manifestations, such as coronary artery disease, valvular heart disease, and intracardiac thrombus, are considered to

be very rare [8].

The surgical repair of aneurysms in BD is considered a challenge to vascular surgeons because of technical difficulties and the potential formation of anastomotic false aneurysms [9]. Surgery is best avoided during the active stage of the disease; however, in our case, surgery was performed as an emergency procedure and BD was not diagnosed at presentation of the patient [5,6].

CASE

A 27-year-old female patient presented to the emergency department with severe epigastric and central abdominal pain radiating to the back, with exacerbation over the last 3 days. There was no history of trauma. Her medical history included recurrent ill-defined oral and genital ulcers, chronic postprandial abdominal pain, and weight loss of 20 kg in the last 12 months. She also reported a positive family history of BD.

On physical examination, there was diffuse abdominal tenderness, no palpable masses, and sluggish peristalsis. Her blood pressure was 110/65 mmHg, heart rate was 110 beats/min, and body temperature was 37.2°C. The laboratory results showed a hemoglobin level of 8.9 g/dL, white blood cell count of 6,000/ μ L, and a high erythrocyte sedimentation rate of 50 mm/h.

Abdominal ultrasound revealed an epigastric mass, with no established diagnosis. Chest and abdominal computed tomography angiography (CTA) showed a contained rupture of a suprarenal aortic aneurysm (4.7×5.1 cm) with the surrounding hematoma tracking upward along the diaphragmatic crus, which is highly suggestive of a ruptured pseudoaneurysm. The superior mesenteric artery (SMA) and celiac artery (CA) were both occluded, which explains the mesenteric ischemic symptoms (Fig. 1, 2). The thoracic aorta and the rest of the abdominal aorta were normal, with no other remarkable findings seen on CTA.

Although this patient was not known to have BD at presentation, a BD diagnosis was highly suggested by the past symptoms of recurrent oral and genital ulcers, positive family history of BD, and clues found at presentation including the absence of signs of sepsis (which would have indicated a mycotic aneurysm) and findings on CTA that excluded degenerative aneurysm. Investigations to confirm the diagnosis of BD were postponed because of the critical status of the patient and the need for urgent surgery.

Urgent surgery was performed through left thoraco-abdominal exposure with left medial visceral rotation. Proximal aortic control was achieved at the level of the diaphragm and distally below the level of the renal arteries. The pseudoaneurysm was resected, revealing a destructed aortic wall at the anterior and lateral aspects. The resected diseased aortic segment and adjacent healthy tissue measured 5 cm and ended distally just above the origin of the SMA. The origin of the CA was involved in the destructed segment of the aortic wall, and thus could not be localized proximally or distally. A thrombus in the SMA was identified and embolectomized from within the aorta, with good

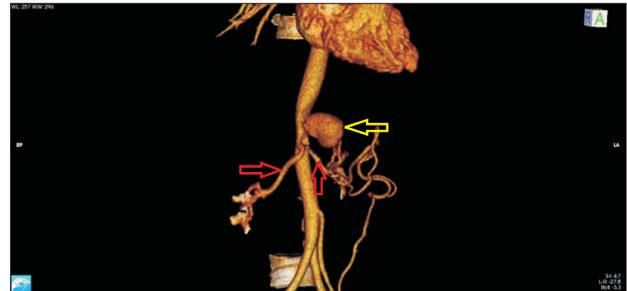


Fig. 2. Preoperative three-dimensional reconstruction of the computed tomographic angiogram showing a false aneurysm (yellow arrow). Both the celiac artery and superior mesenteric artery are occluded (not shown), but both renal arteries are patent (red arrows).

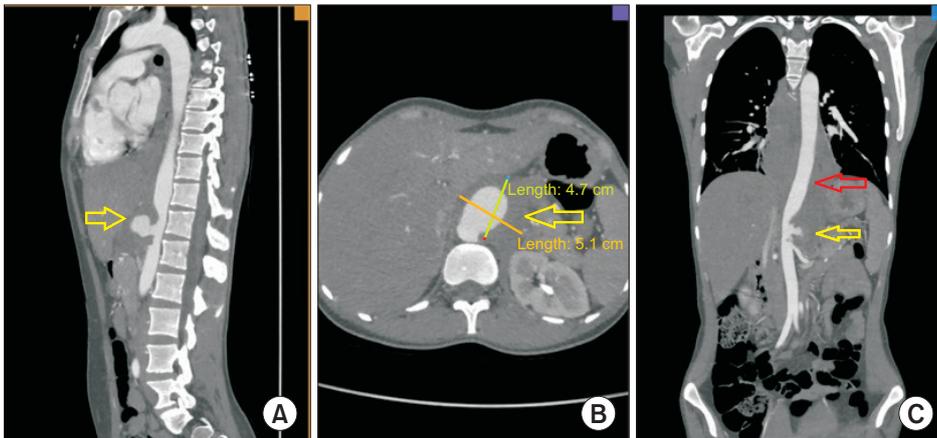


Fig. 1. Preoperative multiplane computed tomographic angiograms, (A) sagittal, (B) axial, and (C) coronal, showing a ruptured aortic false aneurysm (yellow arrows) with hematoma tracking upward to the level of the diaphragm (red arrow).

back flow observed. After cleansing the pseudoaneurysm thrombus, both renal arteries and the SMA were irrigated with cold saline, and the aorta was repaired using a 5-cm-length, 20-mm-diameter Dacron interposition tube graft (MAQUET Cardiovascular; LLC, Wayne, NJ, USA), with end-to-end anastomosis at both ends of the graft. Aortic tissue biopsy specimens were sent for histopathology and culture. The abdominal viscera was explored, and no signs of bowel or organ ischemia were observed. The recorded visceral ischemia time during surgery was 20 minutes.

After the operation, the patient stayed in the intensive care unit for 3 days and started oral intake after being transferred to the ward. She was discharged with an uneventful recovery on the 7th day.

As part of perioperative investigations, two-dimensional echocardiography was performed, which showed a left ventricular apical mural thrombus that necessitated anticoagulation therapy postoperatively. The measured levels of inflammatory markers supported the diagnosis of BD, and immunotherapy (prednisolone and cyclophosphamide) was initiated.

The histopathology report of the resected aortic tissue indicated nonspecific vasculitic changes with neutrophil and lymphocyte infiltration, and the bacterial culture was negative. At 6 months follow-up, CTA showed patent abdominal aorta, SMA and both renal arteries with no signs of a pseudoaneurysm (Fig. 3). Follow-up echocardiography showed normal valvular function, normal left ventricular function, and remission of the previous cardiac thrombus. The erythrocyte sedimentation rate and C-reactive protein level were both normal.

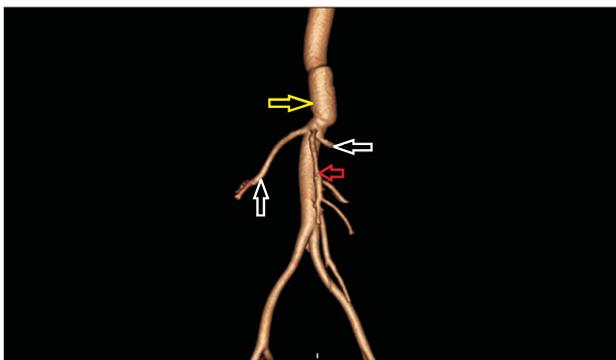


Fig. 3. Postoperative 6 months follow-up three-dimensional reconstruction of the computed tomographic angiogram, showing patent bypass (yellow arrow), superior mesenteric artery (red arrow), and left and right renal arteries (white arrows).

DISCUSSION

BD is a chronic autoimmune inflammatory disease of an unknown etiology. When BD affects the cardiovascular system (vasculo-BD), other manifestations such as oral ulcers, genital ulcers, and skin and neurological symptoms are often neglected.

In vasculo-BD, four different vascular complications have been described: arterial occlusion, arterial aneurysm or pseudoaneurysm, venous thrombosis, and variceal formation [10,11]. The pathogenesis of aneurysms in BD is believed to be obliteration of the vasa vasorum caused by the inflammatory process, resulting in disruption of the nutrient flow to the aortic wall [12]. The intimal layer is thickened by fibroblast and smooth muscle cell proliferation, whereas the media and adventitia layers are both disrupted. Destruction of the media seems to be responsible for the development of a saccular aneurysmal dilatation. Pseudoaneurysms are more common than arterial occlusion/stenosis or true aneurysm in BD, and the rupture of a pseudoaneurysm is a major cause of death [13].

Vascular involvement in BD is reported to occur in up to 38% of patients, with a male predominance [6,10]. There is a high predilection for venous involvement, ranging from 25% to 85%, with deep vein thrombosis being the most common pathology and the lower limbs being the most frequently affected site. The incidence of arterial involvement ranges from 7% to 10%, with pseudoaneurysms being the most common presentation and the aorta being the most frequent site [6,7,14].

The diagnosis of vasculo-BD is usually made through noninvasive methods such as color Doppler ultrasound, CTA, and magnetic resonance imaging. These modalities are preferred because any invasive puncture in the vessel may induce thrombosis or false aneurysm formation [15]. In our case, diagnosis was made using ultrasound and CTA.

The treatment of vasculo-BD involves medical treatment with immunotherapy to suppress the disease activity, as any intervention is better avoided in the active phase of the disease to minimize possible complications. Either surgical or endovascular treatment is an acceptable modality for the definitive treatment of arterial complications in BD.

The endovascular option in the treatment of vasculo-BD is preferred by surgeons when feasible, especially in high-risk patients. Its advantages are lesser operating time and blood loss, shorter hospital stay, and avoidance of late complications such as recurrence and anastomotic disruption. The literature reports that the success rate of endovascular treatment ranges from 80% to 90%, although there is no clinical evidence suggesting that the endovascular option results in better prognosis for patients with BD with

pseudoaneurysms [16,17]. Nevertheless, the endovascular approach was not the best choice in this case because the ruptured false aneurysm involved the visceral segment of the abdominal aorta. In such cases, a fenestrated endograft is deemed necessary; however, it is not readily available and needs time to be customized and delivered from overseas.

Open repair of aneurysms in vasculo-BD is often challenging because the sutures tend to cut through the friable artery, the healing process is usually prolonged, and the graft itself may be affected by new anastomotic pseudoaneurysms and obstruction. A recurrent pseudoaneurysm is reported in 30% to 50% of cases [17,18]. To avoid these complications, anastomoses are performed in disease-free segments, ligating the aneurysm when possible and reinforcing the anastomoses with pledgets [18,19]. The use of synthetic grafts is preferable over autografts in BD because the veins are also usually involved in the disease process [18]. In this case, a synthetic interposition graft was used with both ends anastomosed on macroscopically disease-free segments of the aorta.

Postoperatively, it is advisable to continue steroidal treatment and immunotherapy with regular follow-up investigations including testing for inflammatory markers and CTA scans [5,10,16]. In the case of our patient, the follow-up CTA scan at 6 months was normal (Fig. 3).

In conclusion, when dealing with young patients with

arterial aneurysmal disease, vasculitis such as BD should be suspected as a cause. In such cases, aggressive medical therapy with steroids in combination with immunotherapy should be initiated as soon as possible. Other modalities of treatment, whether surgical or endovascular, once deemed necessary, should be carefully planned according to the urgency of the situation, general condition of the patient, and other associated pathologies. Routine regular follow-ups are always considered necessary in such patients.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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