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Fatal Expanding Thoracoabdominal Aneurysm in Known but Mistreated Behcet's Disease in a Young Patient

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Abstract

Introduction: Arterial involvement in Behcet's disease has been previously described. We report a rare case of a large and long-segment thoracoabdominal aneurysm, which was associated with Behcet's disease and had an unfavorable evolution.
Case Presentation: A 23-year-old man was diagnosed with Behcet's disease, as revealed by a carotid aneurysm, and was treated with a prosthetic graft reconstruction as well as immunosuppressive therapy. The patient was lost to follow-up. He stopped the medications of his own will 1 month after his discharge from the hospital. Two years later, he presented with chest pain of 1 week's duration. A large aneurysm involving a long aorta segment from the sinus of Valsalva to the abdominal aorta above the renal arteries was identified by computed tomography angiography. Unfortunately, the patient died despite immunosuppressive therapy and before any surgical or endovascular intervention could be performed.
Conclusions: This rare observation supports the role of immunosuppressive therapy in preventing the recurrence of life-threatening vascular lesions in the management of Behcet's disease.

Keywords: Behcet's, Vascular Disease, Thoracoabdominal Aneurysm

1. Introduction

Behcet's disease (BD) is a chronic inflammatory condition that can involve the cardiovascular system, hence the term vasculo-Behcet's (1). The incidence of vascular involvement in BD found in the literature is about 29% (2). Thoracoabdominal aortic aneurysms in a patient with BD are rare. We describe a patient with a 2-year history of vasculo-Behcet's, who developed a fatal voluminous aortic aneurysm extending from the sinus of Valsalva to the suprarenal abdominal aorta after he had dropped off prescribed medications of his own will. We highlight the importance of long-term adherence to immunosuppressive therapy in BD with arterial involvement.

2. Case Presentation

A 23-year-old man presented with a rapidly growing neck swelling, which had appeared in the preceding month before his referral to the hospital in the absence of any neurological manifestations. Angiography showed a fusiform aneurysm in the left common carotid artery with a transverse diameter of 45 mm and a narrow neck communicating with the main artery of 7 mm diameter (Figure 1). Clinical investigations revealed recurrent oral and genital ulcerations and relapsing ocular symptoms. The patient was diagnosed with BD according to the O'Duffy criteria. C-reactive protein (CRP) levels were elevated. Imaging investigations were conducted to exclude aneurysms of the other arteries (The diameters of the different aortic segments were normal). The carotid bifurcation and intracranial vessels were patent.

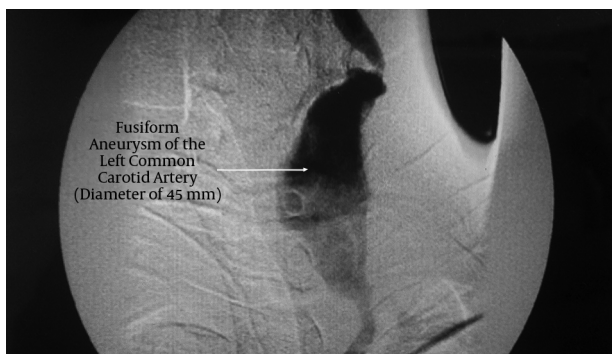

Figure 1. Carotid Angiography Shows a Fusiform Aneurysm in the Left Common Carotid Artery

Due to the rapid enlargement of the aneurysm, the patient was treated surgically shortly after starting immunosuppressive medications and before full control of systemic inflammation and decline in CRP. The treatment consisted of excision and prosthetic graft reconstruction. Dissection of the common carotid artery distal to the aneurysm was attempted to reach a healthy segment of the artery with no adherence to the surrounding tissues or thickening of the artery wall. Anastomotic suture lines were wrapped with a felt strip. The postoperative course was uneventful, and control computed tomography angiography showed an intact repair. The pathological specimen of the aneurysmal wall revealed an adventitial lymphocyte infiltration, which was consistent with the diagnosis of BD. Microbiological cultures grew no organisms.

Immunosuppressive medications were administered before and after surgery with high-dose corticosteroid therapy (methylprednisolone 1 g/d for 3 d), followed by prednisolone and colchicine in tandem with oral cyclophosphamide as a steroid-sparing immunosuppressive drug.

At the time of his discharge, on the 7th postoperative day, the patient was given colchicine (1 mg/d), prednisolone (15 mg/d), and cyclophosphamide (100 mg/d). CRP had decreased to a normal range. The patient was lost to follow-up; he skipped outpatient controls probably owing to a low socioeconomic background. The communication with the patient or his local district hospital was not possible. It later transpired that he stopped medication of his own volition 1 month after his discharge from the hospital.

Two years later, the patient presented with 1 week's history of chest pain. Skin and eye lesions and genital ulcerations were absent. Chest radiography showed abnormal aortic shadows (Figure 2). Blood examination showed CRP of 6.0 mg/dL (normal < 1.0 mg/dL), erythrocyte sedimentation rate of 78 mm/1 hour, and white blood cell count of 9900/mL.

A large fusiform aneurysm with a maximum transverse diameter of 104 mm involving the aorta from the sinus of Valsalva to the abdominal segment and extending to the level of the renal arteries with a total thrombosis of the carotid prosthetic graft and a moderate ectasia in the origin of the left subclavian artery (19 mm in diameter) were identified by computed tomography angiography (Figures 3 - 5). A partial wall thrombus in the abdominal portion of the aneurysm was also observed (Figure 6).

Because of the high risk of rupture, we scheduled the patient for an urgent operation. High-dose steroid intravenous therapy was started because of a high systemic inflammation. Unfortunately, the patient died suddenly on the 3rd day of his hospitalization, probably due to the rupture of the aneurysm. Autopsy was not performed, but a postmortem ultrasound showed a large retroperitoneal effusion, suggesting a rupture of the abdominal segment of the aorta aneurysm.

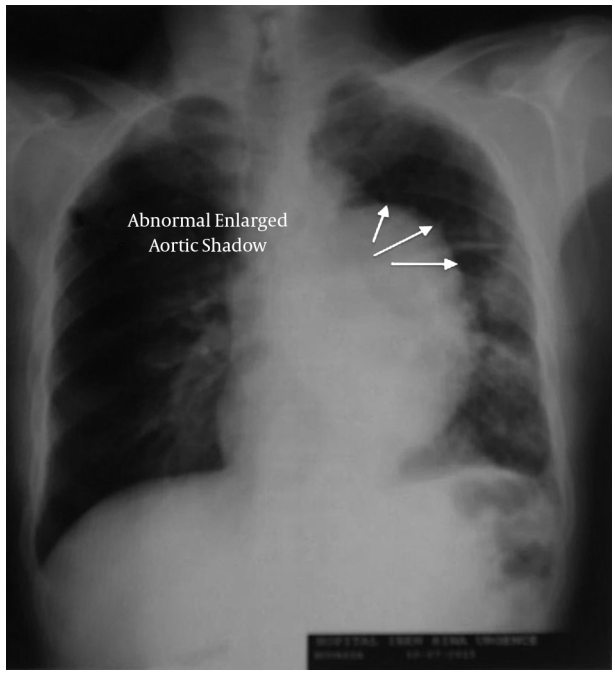

Figure 2. Chest Radiography Shows Abnormal Aortic Shadows

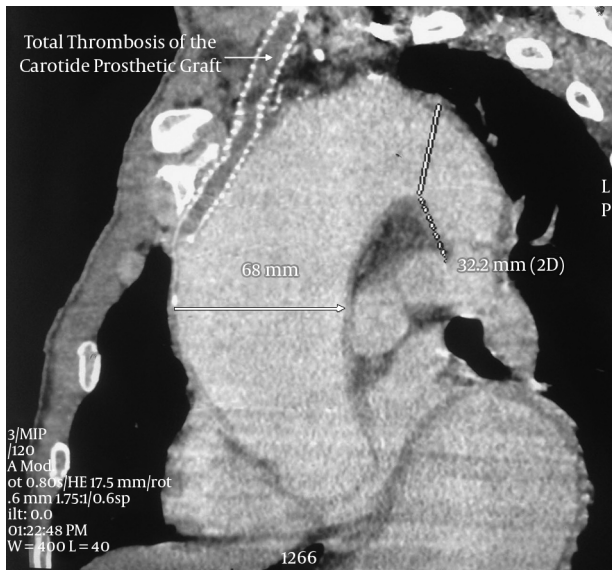

Figure 3. Computed Tomography Angiography Demonstrates an Ascending, Transverse, and Descending Thoracic Aortic Aneurysm

3. Discussion

Arterial involvement of BD is rare, but nearly every major artery has been reported to be involved. Among BD patients with vascular involvement, arterial aneurysms occur in 4.5% to 30% (3, 4). Large-vessel aneurysm formation is the most frequent observed lesion in these cases. Multiple aneurysms with BD are relatively common. A Japanese meta-analysis reported that aneurysms were multiple in 36% of patients with BD aneurysms (5). Cases of extended aneurysms involving thoracic and abdominal aortas in BD are exceptional (6, 7). A literature survey by Hamza et al. (8) reported 67 patients with BD aneurysms; only 1 patient had a thoracoabdominal aortic aneurysm.

The pathogenesis of Vasculo-Behcet's is thought to be the consequence of vasa vasorum vasculitis, and the development of the aneurysmal dilatation seems to be due to the destruction of the arterial media.

The reported survival rate of patients with BD complicated by arterial lesions is > 66% at 15 years (9). In a large cohort of BD patients with a long median follow-up, the main cause of death was major vessel disease with mostly arterial aneurysms. In that study, the incidence of arterial complications was 3 times higher among the patients who died and there was an increased mortality among the youngest patients. Also, male gender and a high number of BD flares were independently associated with the risk of death (10). Mortality in BD patients with aneurysm rupture has been found to be as high as 60% (11).

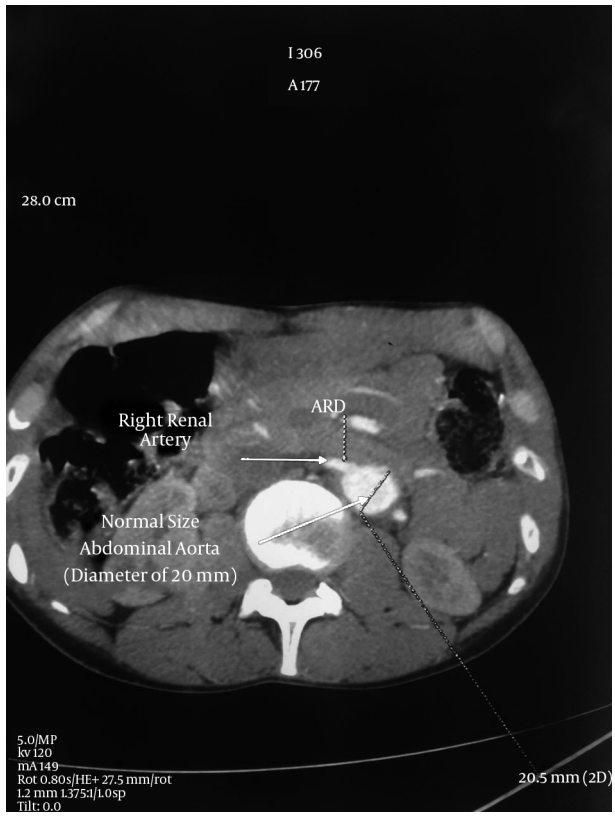

Figure 5. Computed Tomography Angiography Shows a Normal Abdominal Aorta Size at the Level of the Renal Arteries

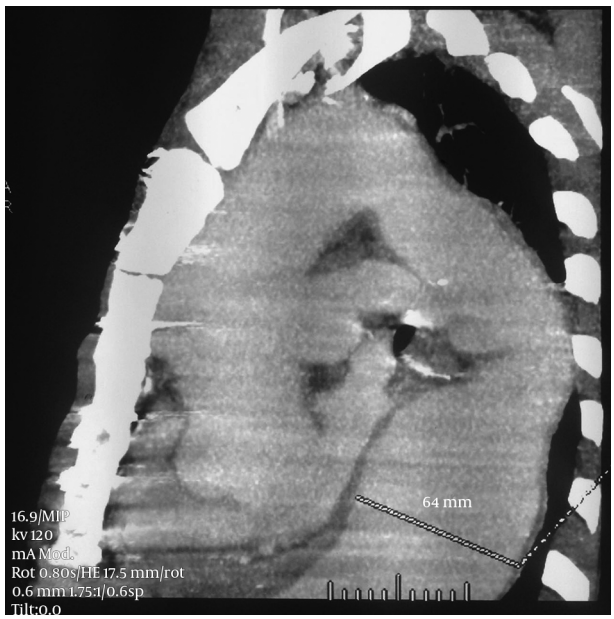

Figure 4. Computed Tomography Angiography Demonstrates an Ascending, Transverse, and Descending Thoracic Aortic Aneurysm

In case of an aortic aneurysm, an aggressive approach with surgical repair is indicated regardless of long-term complications because of the high incidence of rupture or dissection (12, 13). However, if possible, surgery should be postponed until the active inflammation phase has subsided. Omitting surgery in the initial phase may avoid acute exacerbation of inflammation and formation of new aneurysms at the site of the suture line. Indeed, in patients with Vasculo-Behcet's, a high recurrence rate of aneurysms after open surgery has been reported (14, 15). For the prevention of postoperative anastomotic aneurysms, the use of the healthy aortic wall for suturing and end-to-end anastomosis are very important. Full-thickness anastomosis at sites sufficiently distant from inflammatory vascular involvement is required (16). Some authors have proposed an extra-anatomic vascular reconstruction at a site distant from the lesions of the aorta (17), whereas some others have advised not to use a foreign material (6). Recently, advances in endovascular therapies have led to the endoluminal exclusion of aortic aneurysms as an alternative

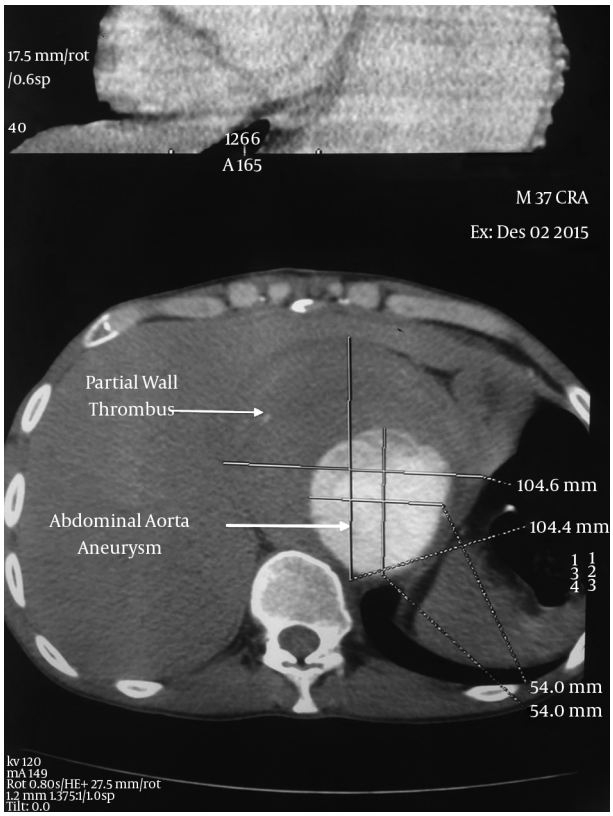

Figure 6. Computed Tomography Angiography Shows a 10-cm Abdominal Aorta Aneurysm With a Partial Wall Thrombus

to open surgery. This sutureless technique seems to be a reasonable approach (18, 19). Nonetheless, the recurrence of pseudoaneurysms after endovascular treatments at the margins of the stent is possible, and experience with this technique in BD is still limited (20). In addition, further studies with long-term follow-up of small cohorts are required to provide more data on the safety and effectiveness of this technique (21).

Screening for other aneurysms as well as long-term follow-up is extremely crucial to detecting the recurrence and development of new aneurysms, especially if the systemic inflammation is not well-controlled. In studies on small cohorts, Kwon Koo et al. (21) and Liu et al. (19) reported that without immunosuppressive therapy, the recurrence rate of aneurysms was high. The unfavorable evolution of the thoracoabdominal aneurysm in our patient can suggest an uncontrolled inflammation of the vessel wall after the discontinuation of immunosuppressive therapy. Nevertheless, we cannot conclude about the role of immunosuppressive therapy in preventing recurrence based on only 1 case report.

3.1. Conclusions

Early recognition and treatment of patients with Vasculo-Behcet's is important because of the increased risk of the recurrence of vascular lesions. Postoperative immunosuppressive regimen might be helpful. It is important to maintain the immunosuppressive therapy and a regular follow-up to prevent these complications.

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Footnotes

Authors' Contribution: Bakkali Tarik and Hamzi Mohamed Amine contributed equally to this work. All authors were involved in drafting the article and approved the final version for publication.

Conflict of Interest: The authors declare that no conflict of interest exists.

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