Case Report

Endovascular Repair of Supra-Celiac and Abdominal Aortic Pseudo Aneurysms Concomitant with a Right Atrial Mass in a Patient with Behçet's Disease: A Case Report

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Abstract

Behcet's disease is a rare immune mediated systemic vasculitis which besides it's more frequent involvement of eyes and skin, sometimes present with aortic pseudo aneurysm and more rarely cardiac inflammatory masses.A 51-year-old patient with Behçet's Disease presented with two symptomatic aortic pseudoaneurysms concomitant with a right atrial mass. Computed tomography (CT) revealed one supra-celiac and another infrarenal aortic pseudoaneurysms. Echocardiography showed a large mobile mass in the right atrium. Both pseudoaneurysms were successfully excluded simultaneously via endovascular approach with Zenith stent-grafts, and the atrial mass was surgically removed 10 days later. Post-implant CT showed successful exclusion of both pseudo-aneurysms, patency of all relevant arteries, and patient is now asymptomatic and has returned to normal lifestyle. Multiple pseudoaneurysms concomitant with a right atrial mass can be an initial manifestation of Behçet's disease. Endovascular repair can be a good treatment option for the pseudoaneurysms.

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Introduction

Behcet's disease (BD) is a rare immune-mediated small-vessel systemic vasculitis that often presents with mucous membrane ulceration and ocular problems.[1, 2] Since the highest incidence rates of BD are reported in Mediterranean countries, it is also known as the "Silk Road Disease."[1] Cardiovascular involvement in BD has a wide spectrum, affecting different parts of the heart structures and vasculature.[1] Vascular involvement - with an incidence rate of 7-38% - is considered the most common cause of mortality in BD patients.[3] Of this patient group, aortic aneurysms manifest in 2-6%. Of the BD patient group with aortic aneurysms, only 18% present with multiple

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aneurysms.[2] Arterial aneurysms, which may be true or false, may also affect the thoracic aorta, iliac arteries, and/or femoral arteries.[3]

We report a BD case with dual aortic pseudoaneurysms concomitant with a right atrial mass and coronary artery disease.

Case Report

A 51-year-old male, known to have BD with a history of popliteal aneurysm surgery, presented with abdominal pain mostly related to food intake.

In routine echocardiography, a large mobile mass (27mm × 14mm) in the right atrium was reported, which had an origin from the coronary sinus - albeit without any tricuspid dysfunction. CT Angiography of the aorta showed two pseudoaneurysms: a supra-celiacpseudoaneurysm- 66mm in diameter- and an infrarenal pseudoaneurysm- 55m in diameter (Figure 1). Both had a compressive affect on the aorta, and both were leaking. Within the first two days of admission, the hemoglobin level dropped from 13.2 to 8.0 mg/dl and, therefore, two units of pack cells were transfused.

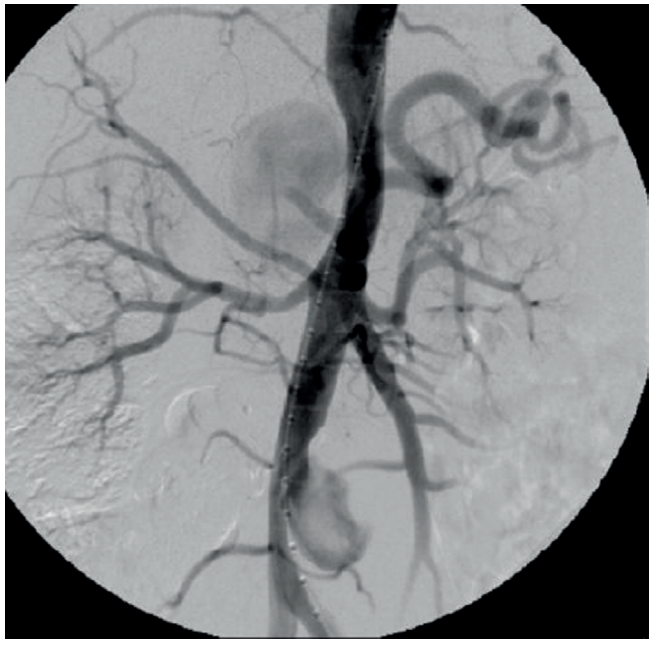


Figure 1. A supra-celiac pseudoaneurysm (66 mm in diameter) and an infrarenal pseudoaneurysm(55 mm in diameter) can be seen in pre-implant digital subtraction angiography (DSA)

An urgent endovascular plan was devised, followed by coronary angiography and digital subtraction angiography of the descending aorta and iliac arteries in a hybrid-suite. In the same session, the supra-celiac pseudoaneurysm was excluded endovascularly using a Zenith TX2 stent-graft extension cuff (TBE-30-80-PF). Also, the infrarenal pseudoaneurysm was excluded using a Zenith Renu aorto-uni-iliac stent graft (AX1-1-28-125) and extended into the right common iliac artery, and the left common iliac artery was occluded with a 14-mm iliac plug (Cook Medical Inc., Bloomington, Indiana). Subsequently, a femoral-femoral bypass was conducted to reestablish the flow to the left leg (Figure 2). An aorto-uni-iliac stent graft was chosen instead of a standard bifurcated endograft because the infrarenal pseudoaneurysm had compressed the diameter of the aorta to a mere 8mm, which would not allow the two iliac legs of the bifurcated device to fully deploy above the aortic bifurcation.

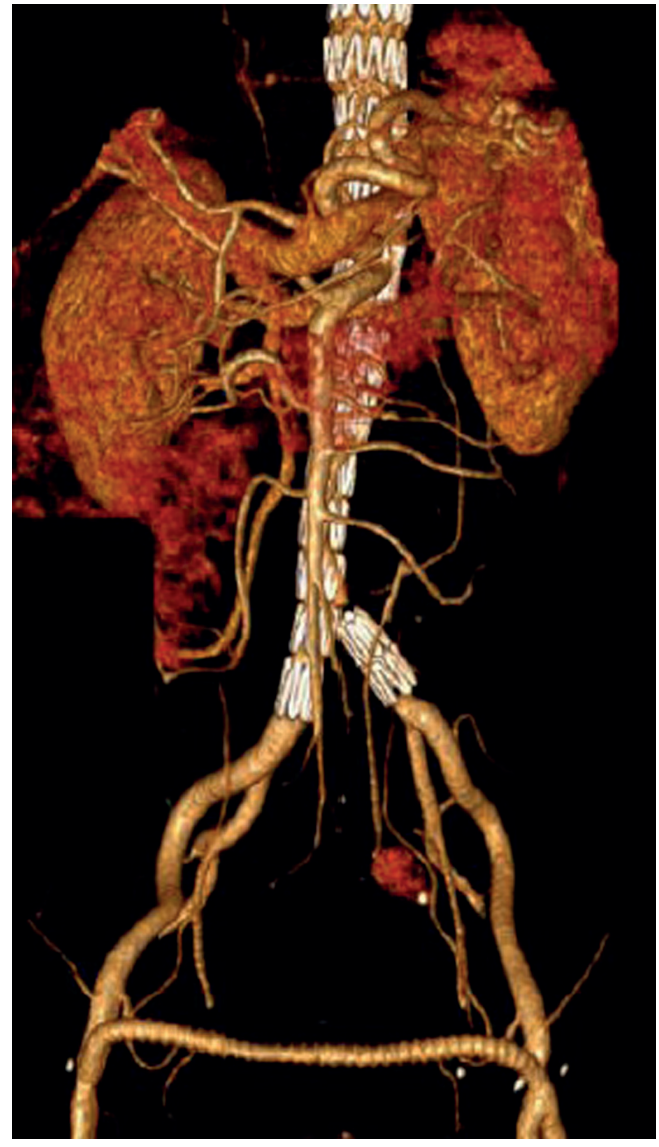
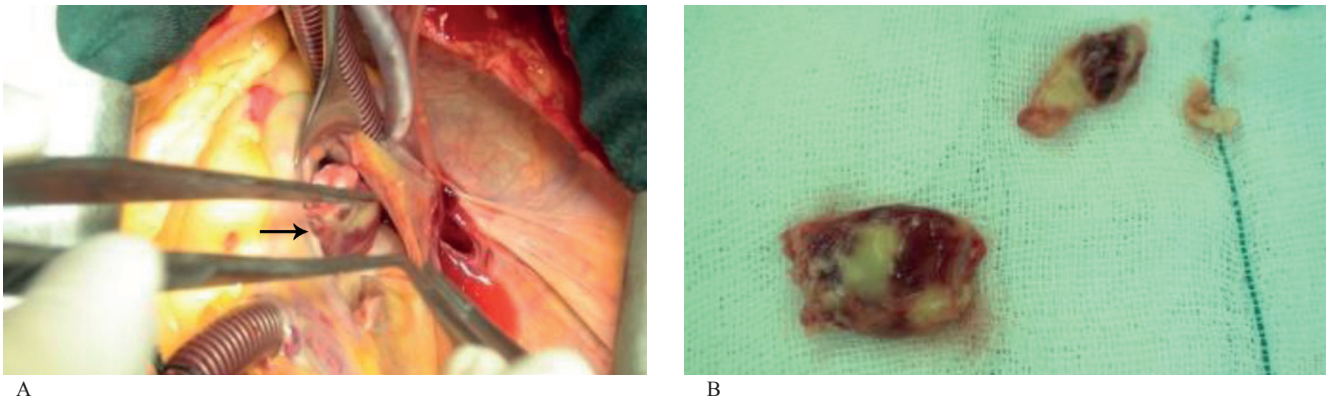
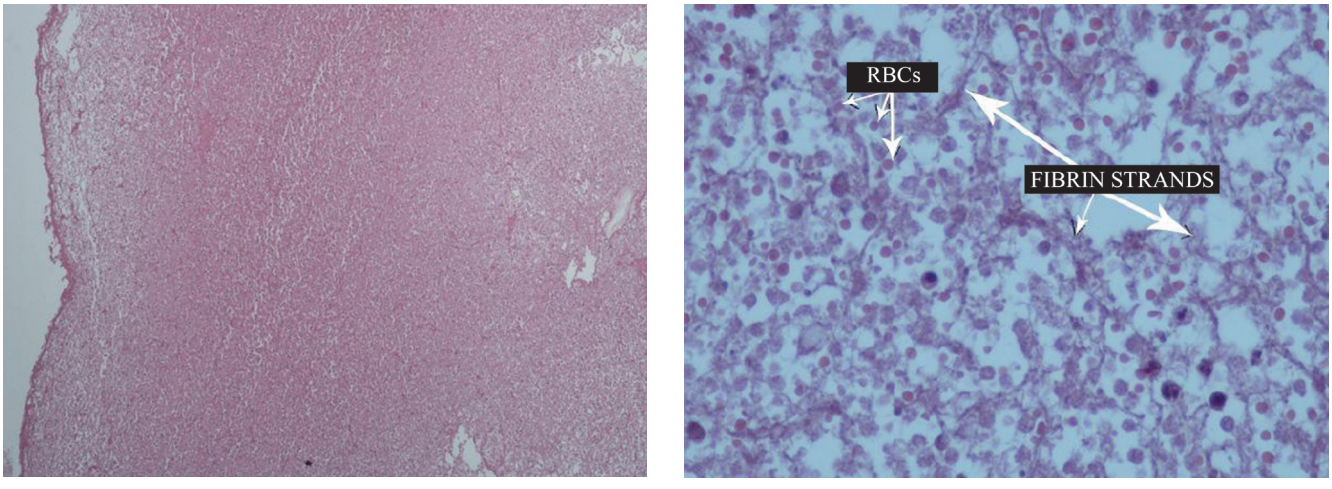


Figure 2. Post-implant 3 dimensional Computed Tomography Angiography: an endograft extension cuff stent-graft is deployed in the supra-celiac aorta, an aorto-uni-iliac stent-graft is deployed in the infrarenal aorta, an iliac vascular plug is placed in the left common iliac artery, and the procedure is finished with a fem-fem-surgical bypass with a vascular supported tube-graft



A B

Figure 3. A) Peri-procedure photograph of the surgical removal of the right atrial mass. B) Surgically removed right atrial mass



A B

Figure 4. A) Fibrinous blood clot in low power field. B) High power field shows entrapped red blood cells (RBCs) in fibrin strands

After endovascular repair, the patient was followed up with daily echocardiography for the right atrial mass and was placed under treatment with heparin and high-dose steroids (60 mg of oral prednisolone daily). Due to the large size of the right atrial mass and its hypermobility, the risk of pulmonary emboli was very high. Additionally, there was no response to the anti-coagulant and immunosuppressive treatments and the severe stenosis of the left anterior descending, left circumflex, and posterior descending artery was seen on angiography. Therefore, ten days later, the surgical removal of the right atrial mass performed, and it was followed immediately by coronary artery bypass graft surgery (CABG) (Figure 3). The pathology report of the right atrial mass was a fibrinous clot (Figure 4).

"One week later, the patient was discharged from the hospital in good clinical condition. Since then, he has been under strict clinical care of rheumatologist. After 6 months, his general condition was good, new CT angiography showed no residual aneurysm, endoleak or any sign of migration or new aneurysm formation and there was no new mass in his cardiac chambers".

Discussion

To the knowledge of the authors, this is the first reported case of dual pseudoaneurysms in the aorta concomitant with a large right atrial mass. Our patient had a supra-renal pseudoaneurysm and an infra-renal pseudoaneurysm, for which we preferred endovascular repair to classic surgery as a treatment option due the high rate of aneurysm recurrence after surgery as reported by Bautista-Hernández et al.[2]

Endovascular repair of the dual pseudoaneurysms simplified the procedure significantly, allowing us to complete the entire endovascular procedure in less than one hour. A risk that we faced with the supra-celiac aneurysm was that the short distal landing zone from the aneurysm to the celiac trunk of less than 2 cm created the possibility of celiac trunk occlusion due to graft migration. However, in our case perioperative angiography - to evaluate the collateral flow from the superior mesenteric artery - and anchoring barbs - positioned incrementally on the extension cuff - reduced the above-noted risks.

In patients with BD, recurrence can be a complication

of endovascular repair due to the chronic irritation of the stent graft as a foreign material.[4] Therefore, the use of immunosuppressive drugs before and after repair is highly recommended.[4] Althoughthe intensity and duration of the treatment still remains controversial, patients' clinical manifestations and disease activity can determine the immunosuppressive protocol.[4]

There are only a few reported cases of a cardiac mass in patients with BD. Marashi et al.[5] reported the case of a 13-year-old male with a large left ventricular mass, for which surgical treatment was selected. Vahedian et al.[6] reported a right ventricular mass in a 17-year-old male; the problem was successfully resolved after 4 months with anticoagulants and immunosuppressive therapy.[6] In our case, the surgical option was chosen based on the large size and hypermobility of the mass as well as the great risk of emboli and valvular destruction, which did not respond well to intensive immunosuppressive therapy. Due to the apparent risks to valvular integrity, distal embolization, and concomitant coronary artery disease, we decided to combine the two surgical procedures of right atrial mass removal and CABG into one session.

Conclusion

Multiple pseudoaneurysms concomitant with a right atrial mass can be an initial manifestation of BD. In patients with this unusual presentation, one should assume BD as a possible diagnosis. Endovascular repair can be a good treatment option of the pseudoaneurysms. Although a cardiac mass usually consists of thrombus and inflammatory components –which in most patients can solely be treated with medical therapy - in our view, surgery may be necessary for high-risk patients.

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