

**Case
Report**

Multiple Recurrent Pseudoaneurysms after Endovascular Repair of Abdominal Aortic Aneurysm in a Patient with Behçet's Disease

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In Behçet's disease (BD) patients, endovascular repair is a reasonable alternative treatment for aortic aneurysms to avoid postoperative anastomotic pseudoaneurysms. However, there are some complications that may occur after endovascular repair. We herein report the case of a 40-year-old man with active BD developed recurrent aortic pseudoaneurysms at the proximal and distal margins of the stent graft and a femoral puncture site pseudoaneurysm 3 months after endovascular abdominal aortic aneurysm (AAA) repair. The aortic pseudoaneurysms were treated endovascularly, including the use of the chimney technique for the proximal pseudoaneurysm close to the renal arteries and the femoral pseudoaneurysm with surgical excision and reconstruction. Intensive immunosuppressive therapy was initiated immediately after the operation. The patient is in good condition without any complications at 8-month follow-up. This case suggests the utility of the chimney technique and postoperative immediate intensive immunosuppressive therapy in treating recurrent aortic pseudoaneurysms in emergency, active BD patients.

Keywords: Behçet's disease, pseudoaneurysm, endovascular aneurysm repair, chimney technique, immunosuppressive therapy

Introduction

Behçet's disease (BD) is a multisystemic inflammatory disease of unknown etiology, characterized by recurrent oral and genital ulcers, skin lesions, and

uveitis.¹⁾ BD can also affect the cardiovascular, gastrointestinal, and central nervous system. Vascular involvement, known as vasculo-BD, may present as venous thrombosis and arterial pseudoaneurysm formation. Pseudoaneurysms are a major contributing factor to the overall mortality in BD patients and the most common site of aneurysm formation is the abdominal aorta.²⁾ Conventional open repair of abdominal aortic aneurysm (AAA) in BD patients is associated with a high recurrence rate of pseudoaneurysms at anastomotic sites. Endovascular aneurysm repair (EVAR) offers a reasonable alternative for BD patients, as it obviates the need for aortic anastomosis, but there are some typical complications after EVAR. We herein report a case of a patient with BD who developed multiple recurrent pseudoaneurysms after endovascular repair of an AAA. The patient consented to the publication of this case report.

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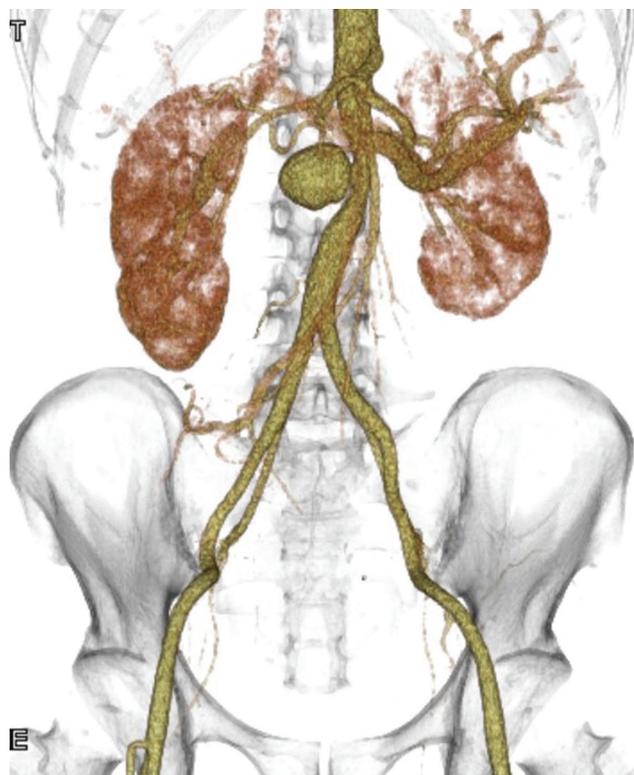


Fig. 1 A CT image showing an abdominal aortic aneurysm. CT: computed tomography

Case Report

A 40-year-old man was referred to our hospital for further evaluation of a fever and recurrent abdominal pain. His medical history was significant, including a 20-year history of recurrent oral ulcers and several attacks of erythema nodosum on the lower limbs. The etiology of the erythema nodosum had not been identified at the time of the attacks. Three months prior, he was diagnosed with AAA (**Fig. 1**) during an evaluation of abdominal pain, and he underwent EVAR using an iliac leg stent graft (Gore Excluder PXC161000J; W.L. Gore & Associates Inc., Flagstaff, AZ, USA) at another hospital. The device was introduced through a 14-Fr sheath via percutaneous right femoral access and deployed above the aortic bifurcation. The right femoral puncture site was closed using two suture-mediated closure devices (Perclose Proglide; Abbott Vascular Inc., Redwood City, CA, USA). About 1 month after the endovascular treatment, postoperative computed tomography (CT) confirmed exclusion of the aneurysm. Several days before admission to our hospital, he developed a fever and recurrent abdominal pain. On a physical examination, he had a fever of 38.7°C, aphthous ulcers on the

inside of the lower lip, and a pulsatile mass in the right groin. Laboratory data showed an erythrocyte sedimentation rate (ESR) of 117 mm/h and a C-reactive protein (CRP) level of 15.56 mg/dL. Enhanced CT at admission revealed aortic pseudoaneurysms at the proximal and distal margins of the previously inserted stent graft and another pseudoaneurysm at the right femoral access site (**Figs. 2A** and **2B**). A diagnosis of BD was made from his total course, according to the International Criteria for BD,¹⁾ but he had never received any immunosuppressive therapy. Emergency repair was indicated although his inflammatory markers were markedly elevated because the patient had abdominal pain, which was a sign of potential impending rupture. We decided to perform endovascular repair of the aortic pseudoaneurysms and simultaneous repair of the puncture site pseudoaneurysm. Due to the short proximal neck, we decided to adopt the chimney technique for the proximal aortic pseudoaneurysm. Under general anesthesia, a vertical skin incision was made over the right common femoral artery. Resection of the puncture site pseudoaneurysm and reconstruction with a great saphenous vein interposition graft were performed. The graft was anastomosed to the normal artery away from the aneurysmal segment. We then performed endovascular procedures. Right femoral access was obtained proximal to the interposition graft, and left femoral and left brachial artery access were also obtained by surgical cut-down. The left renal artery, which had a lower origin from the aorta than the right, was cannulated via left brachial access. A 5 mm × 19 mm balloon-expandable stent (Express SD; Boston Scientific Corp., Natick, MA, USA) was advanced into the proximal portion of the left renal artery and left undeployed. A 23 mm × 33 mm Excluder aortic cuff (PXA230300J; W.L. Gore & Associates Inc.) was advanced via left femoral access and deployed proximally to the previously inserted stent graft with appropriate overlap. An additional aortic cuff was deployed just below the right renal artery. The left renal chimney stent was then deployed with inflation of the balloon simultaneously with gentle balloon molding of the aortic cuffs. Finally, to exclude the distal aortic pseudoaneurysm, an AFX bifurcated stent graft (BA22-90/I16-30; Endologix Inc., Irvine, CA, USA) was positioned at the aortic bifurcation, overlapping the previous straight stent graft, and deployed. A completion angiogram demonstrated successful exclusion of the aortic pseudoaneurysms and bilateral renal perfusion (**Figs. 3A** and **3B**). We closed all three access sites with surgical sutures.

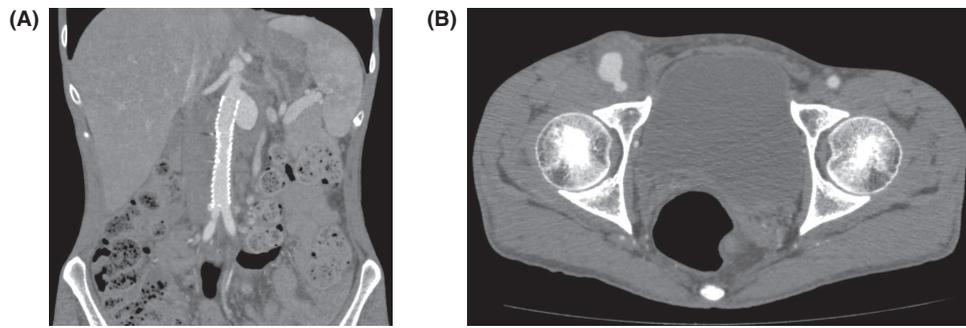


Fig. 2 (A) Aortic pseudoaneurysms at the proximal and distal margins of the stent graft and (B) a pseudoaneurysm at the right femoral puncture site.

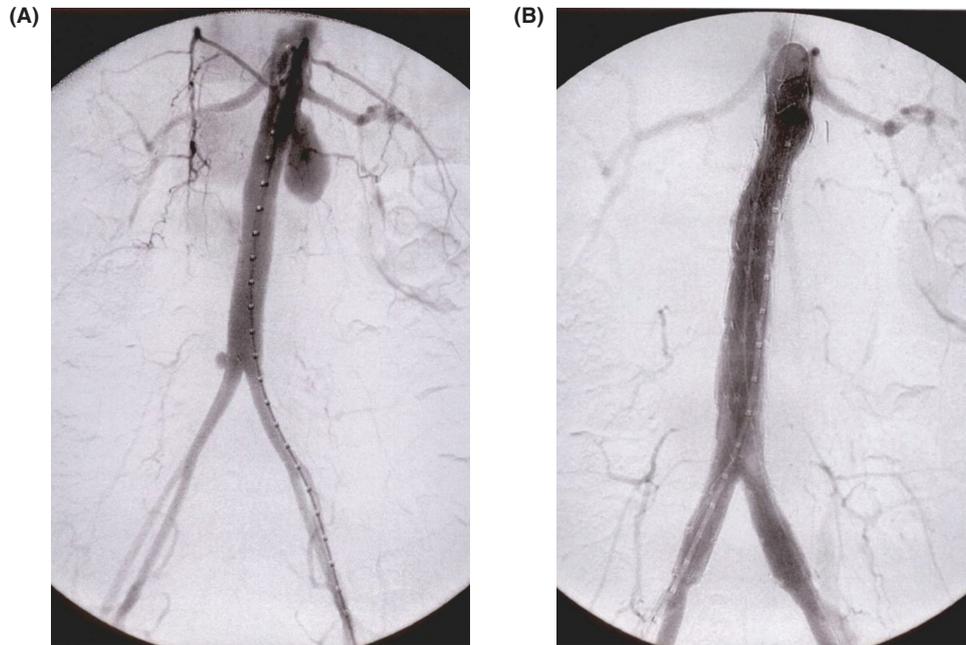


Fig. 3 (A) Intraoperative angiogram showing pseudoaneurysms at the both margins of the stent graft. (B) A completion angiogram showing successful exclusion of the aortic pseudoaneurysms and bilateral renal perfusion.

He received immunosuppressive therapy with oral prednisolone (50 mg) preoperatively and intravenous methylprednisolone (1000 mg/day) for the first 3 postoperative days. Thereafter, oral prednisolone, methotrexate, and colchicine were continued and adjusted according to the disease activity. Infliximab (5 mg/kg) was also added on the third and seventeenth postoperative day. The levels of ESR and CRP dropped to normal ranges within 10 days after the operation, and the patient was discharged on postoperative day 31.

At 8-month follow-up, he was in good condition without any symptoms or signs of aneurysm recurrence. He was taking prednisolone 6 mg/day along with methotrexate 10 mg/week at the most recent visit.

Discussion

Vascular involvement is reported to occur in 7%–38% of BD patients.³⁾ The abdominal aorta is the most common site of aneurysm formation in BD patients. Conventional open repair of AAA in BD patients is associated with a high morbidity. Anastomotic pseudoaneurysm is the most common serious complication after open repair, occurring in 30%–50% of patients.⁴⁾ Over the past 15 years, several reports have described experiences of EVAR in BD as an alternative to open surgical repair. EVAR may be the superior treatment option from the viewpoint of prevention of aortic anastomotic pseudoaneurysms. Liu et al.⁵⁾ observed no

immediate complications after endovascular repair of AAAs, but 2 of 10 patients developed recurrent pseudoaneurysms at the margins of the stent graft, and one patient died due to rupture of a recurrent pseudoaneurysm. In a report by Kim et al.,⁴⁾ 2 of 10 patients experienced recurrence of an aneurysm: one patient with a pseudoaneurysm at the distal margin of the stent graft, and the other with a new pseudoaneurysm of the left subclavian artery.

Although EVAR does offer certain advantages over open repair, recurrent pseudoaneurysm is still a serious problem in BD patients. It has been suggested that vessel wall injuries or mechanical stress at the edge of the stent graft may trigger tissue inflammation, leading to aneurysm formation.

Recurrent pseudoaneurysms at the margin of the stent graft generally require timely intervention. This is usually done with additional stent graft components. However, pararenal recurrent pseudoaneurysms after infrarenal EVAR pose a therapeutic challenge because of the insufficient infrarenal neck for additional stent graft. We found six previously reported cases of a recurrent pseudoaneurysm at the proximal margin of an infrarenal stent graft in BD patients. Among them, two patients underwent open surgical repair,^{6,7)} one patient received careful follow-up observation without additional intervention,⁸⁾ and three patients died due to rupture of the pseudoaneurysm.^{5,8,9)} Although the technique was an off-label use of existing devices, we achieved good results with endovascular repair using the chimney technique in the present patient. We used the bare stent for the chimney technique because covered stents are not covered by the national insurance system in Japan. As seen in our patient, aortic pseudoaneurysms in BD are typically of the saccular type with a narrow neck, which means that they originate from a small segment of the normal-sized aorta. Considering this morphologic feature, extension of the proximal landing zone using the chimney technique is effective for achieving better stent graft fixation.

BD patients also tend to develop pseudoaneurysm at puncture sites.¹⁰⁾ In the present case, a pseudoaneurysm developed at the puncture site even after the use of suture-mediated devices. There is no consensus on the appropriate management of puncture sites and peripheral artery pseudoaneurysms in BD patients.^{10,11)} Whether endovascular repair or open surgical repair should be performed and the optimal graft material for arterial reconstruction remain controversial. In the present case,

resection of the aneurysm and interposition grafting was indicated because the lesion was unsuitable for endovascular repair. We used the great saphenous vein for grafting, as it was of good quality.

In addition to appropriate surgical and endovascular interventions, adjunctive immunosuppressive therapy is essential in the treatment of arterial lesions in BD patients. Previous studies have indicated that discontinuing immunosuppressive therapy⁵⁾ or the onset of uncontrolled inflammation despite administering immunosuppressive therapy^{4,6)} are associated with a greater risk of recurrent pseudoaneurysms after EVAR. Our patient had not been diagnosed with BD at the time of initial detection of the AAA, so immunosuppressive therapy was not administered after primary EVAR. His recurrent pseudoaneurysms were strongly suspected to be associated with the postoperative uncontrolled vasculitis. Furthermore, to reduce the risk of postoperative complications, several authors have advocated administering immunosuppressive therapy preoperatively and performing surgical or endovascular intervention when the disease is in remission.^{2,12)} For urgency reasons, we performed surgical and endovascular repair of the recurrent pseudoaneurysms without waiting for remission and initiated intensive immunosuppressive therapy, including intravenous methylprednisolone pulse therapy and infliximab, immediately after the operation. The patient responded well to postoperative immunosuppressive therapy; the levels of ESR and CRP dropped rapidly, and no postoperative complications have been observed to date. ESR and CRP are the most prominent indicators for assessing the disease activity and response to immunosuppressive therapy, but there are no specific biomarkers for BD.⁹⁾ Some authors have reported unpredictable recurrence of pseudoaneurysms in patients with normal ESR and CRP levels.^{10,11)} As such, despite the satisfactory early results in our patient, further careful follow-up remains mandatory.

Conclusion

Recurrent pseudoaneurysms remain a possible complication after EVAR in BD patients. Our experience with the present patient suggests the effectiveness of endovascular repair using the chimney technique and postoperative immediate intensive immunosuppressive therapy for treating recurrent aortic pseudoaneurysms in patients with BD, particularly for emergency, active patients.

Disclosure Statement

All authors have no conflicts of interest.

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