Analysis of the treatment of aneurysm involved in vascular Behçet's disease (three cases)

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BACKGROUND: *The involvement of a aneurysm or pseudoaneurysm in vascular Behçet's disease (BD) is rare and difficult to treat. The present study investigates the definitive therapeutic strategy for aneurysm with vascular BD. Case presentation: We report three cases of BD with aneurysm in different arteries. All patients underwent endovascular surgery, but the treatment effects were different. Two patients developed new aneurysms immediately after surgery, and eventually died due to rupture of recurrent aneurysm, while merely one patient survived after endovascular treatment.*
CONCLUSION: *For aneurysms involved in vasculo-Behçet's disease (v-BD), endovascular treatment is suggested during the stable period. At the same time, glucocorticoids and immunosuppressants could be preoperatively and postoperatively used together.*

KEY WORDS: Behçet's disease (BD), Aneurysms, Immunosuppressive therapy

Introduction

Behçet's disease (BD) is a multisystemic chronic vasculitis. The main manifestations are recurrent oral and genital ulcers, uveitis, and skin lesions. When the vascular system is involved in Behçet's disease (BD), including the artery and veins, it is called vasculo-Behçet's disease (v-BD). These venous lesions include thrombosis and superficial phlebitis. These artery lesions may present with, stenosis, and occlusion in the arterial system. The incidence of vascular involvement in patients with BD is 7%-29% [1]. Aneurysm or pseudoaneurysm is the main symptom involved in v-BD.
The present study reports three patients with the involvement of aneurysm or pseudoaneurysm in BD.

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Case Report

CASE N. 1

A 30-year-old female from Hunan province of China was admitted to our hospital due to relapsing oral ulcer for two years, with progressive chest and back pain for four months. In a local hospital, the patient's computed tomography angiography (CTA) revealed pseudoaneurysm of the thoracic aorta. Hence, endovascular repair of the aortic aneurysm was performed. The patient's back pain was not significantly relieved after the operation, and the aorta CTA revealed pseudoaneurysm at the original distal end of the stent-graft, and at the original incision on the femoral artery. Hence, the patient came to our hospital. The patient's pathergy test result was positive, and the laboratory test results revealed an erythrocyte sedimentation rate (ESR) of 45 mm/hr (reference range: 0-15 mm/hour) and a C-reactive protein (CRP) of 110.2 mg/L (reference range: 0-10 mg/L). After rheumatologist consultation, the patient was diagnosed with BD. Hence, methylprednisolone was used at 80 mg per day. After one week, the patient's chest and back pain suddenly aggravated. By CTA, it was inferred that

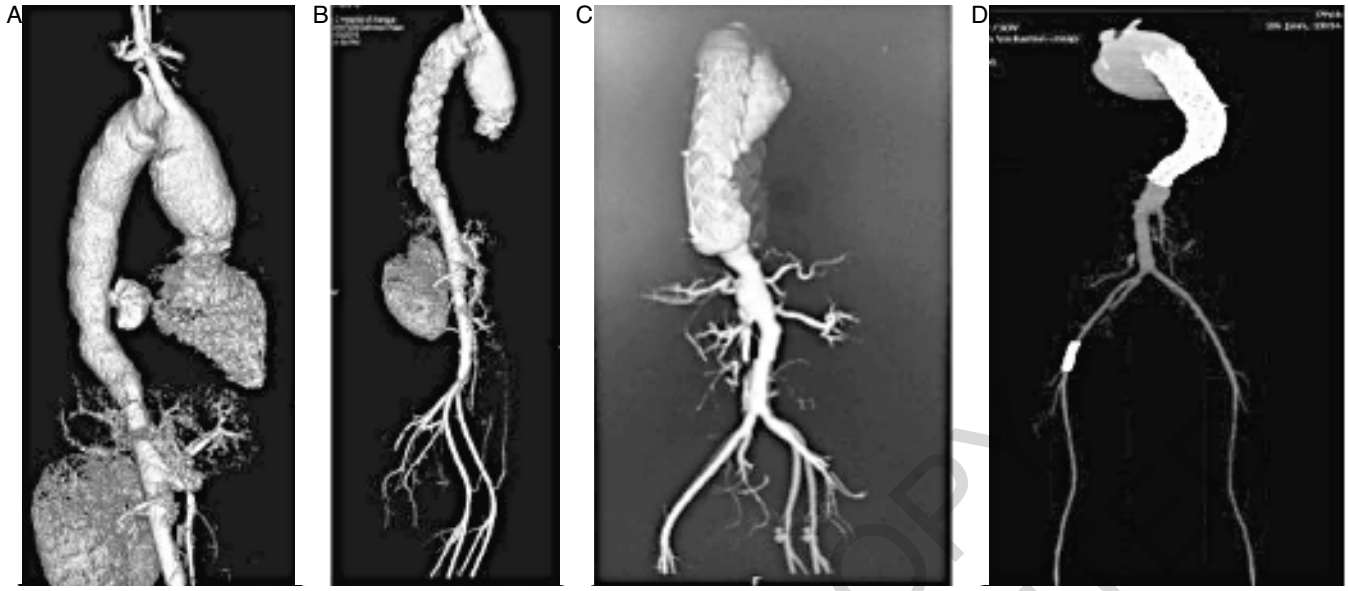

Fig. 1: Case 1 CTA(A) CTA revealed the pseudoaneurysm of the thoracic aorta. (B) The first operation after the endovascular procedure (C). CTA revealed the pseudoaneurysm at the original distal end of the stent-graft, and at the original incision in the femoral artery. (D) The second operation after the endovascular procedure is shown.

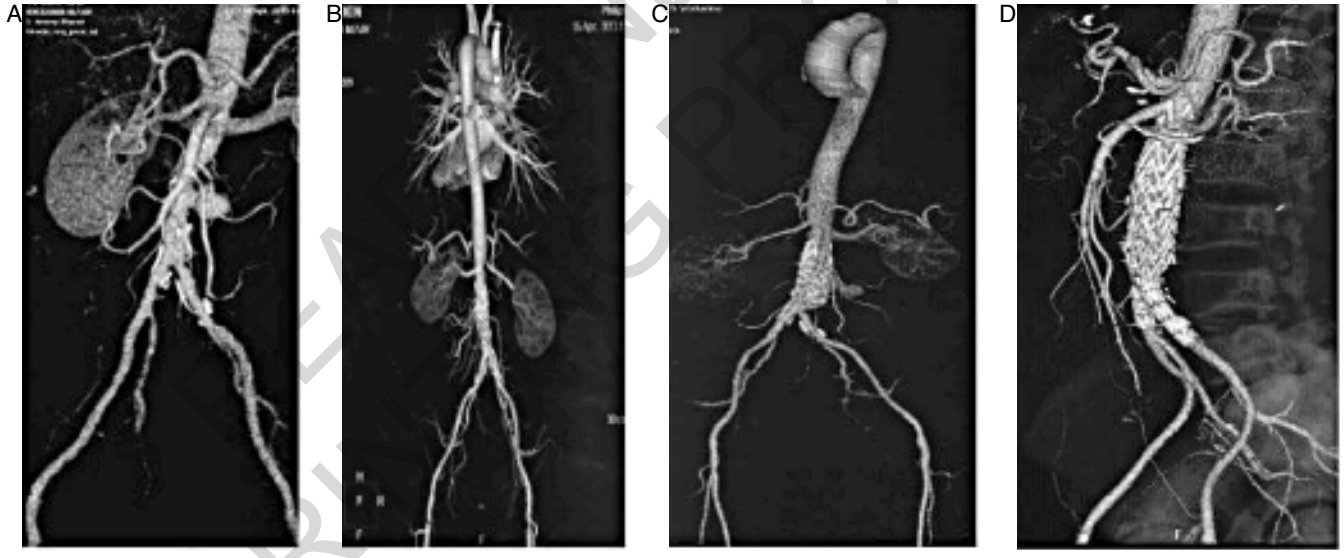

Fig. 2: Case 2 CTA(A) CTA revealed the abdominal aortic aneurysm. (B) The first operation after endovascular procedure is shown. (C) CTA revealed the rupture of the aortic aneurysm at the distal end of stent-graft. (D) The second operation after the endovascular procedure is shown.

the aneurysm ruptured, because the thoracic aortic aneurysm diameter was larger than before. Therefore, the investigators decided to perform emergency surgery. Stents were placed at the distal end of the original stent and at the inferior segment of the right femoral false aneurysm. The patient was given 40 mg of prednisone and 0.5 mg of colchicine daily after the operation. The ESR level was 10 mm/hr and CRP was 16.79 mg/L. Furthermore, the chest and back pain of the patient was also relieved after two weeks. The patient was discharged and continued to orally take 20 mg of prednisone and 0.5 mg of colchicine daily. However, the patient died due to rupture of the recurrent aneurysm after one month in a local hospital (Fig. 1).

Fig. 3: (A) The CTA revealed the aneurysm of the left iliac artery. (B) The situation after the endovascular treatment is shown.

Case n. 2

A 50–year-old male from Shanxi, China had a chief complaint of oral ulcer, which repeatedly occurred for more than one year, intermittent abdominal pain for nine days and hematochezia, which suddenly occurred for one day. The patient had BD for one year and intermittently received methylprednisolone.

The blood test revealed that white blood cell count was 12.95×10^9/L, and ESR and CRP levels were high. The abdominal CTA revealed rupture of the abdominal aortic aneurysm.

Endovascular aortic aneurysm repair was practiced in emergency. However, after three months, the patient was admitted to the hospital due to left hip and left thigh pain. The CTA revealed rupture of the aortic aneurysm at the distal end of the stent-graft. Hence, endovascular treatment was performed again. Two stent-grafts were used. The patient also received prednisone after the operation. Finally, the patient died due to rupture of aneurysm after one year (Fig. 2).

Case n. 3

A 38–year-old male, who lived in Beijing, China, was admitted to our hospital due to swelling and pain of his left lower limb, with intermittent claudication for more than one month. The patient was diagnosed with BD for six months, and had been regularly receiving methylprednisolone. The CTA revealed aneurysm of the left iliac artery. The laboratory test results revealed that ESR was 30 mm/hr and CRP was 18.3 mg/L. The patient received methylprednisolone and cyclophosphamide daily. When the level of ESR and CRP returned to normal, the patient underwent iliac aneurysm endovascular exclusion. The stents have been used. After the operation, the patient continued to received methylprednisolone and cyclophosphamide. The pain of his left lower limb was relieved significantly. He could walk regularly without any pain. Two year after the operation, the patient is still alive (Fig. 3).

Discussion

The vascular involvement in BD includes the vein and artery. Aneurysm and pseudoaneurysm are mostly observed in the arterial system, and thrombosis occurs in the venous system. Many artery locations could be involved, such as the pulmonary artery, femoral artery 2, abdominal aorta, and intracranial artery [3,4]. The most dangerous event of aneurysm is high risk of rupture [5]. The treatment for aneurysm due to BD is surgery. Open surgical therapy is the treatment option for these aneurysms. However, this approach often results in the development of pseudoaneurysm or leakage at the anastomotic sites, or graft occlusions during the postoperative period [6,7]. Furthermore, the pro-coagulative state and inflammatory activity in patients with BD might lead to

vessel wall thickening and occlusions in both native arteries and implanted grafts [8]. In order to avoid the recurrence rate of anastomotic pseudoaneurysm, immunosuppressive therapy (glucocorticoid and cyclophosphamide) is performed before surgery [9].

The first endovascular treatment in BD to treat aortoiliac aneurysms was performed in 1998 [10]. Many investigators have attempted this surgical approach to treat aneurysms due to BD [11,12]. Although this is less invasive than open surgical therapy, aneurysms after the operation can recur. In the present study, two of the three cases had recurrence of aneurysms after each operation, and finally died. These two patients had BD before the operation, but they all did not regularly receive immunosuppressive drugs. Moreover, these two patients performed endovascular treatment in emergency and in the active inflammation stage of BD. The third patient took methylprednisolone and cyclophosphamide at pre-operation and post-operation. Hence, endovascular repair and adjunctive immunosuppressive therapy were necessary to reduce the risk of recurrence of aneurysms due to BD [13]. Endovascular procedures are less invasive than open surgery, stents or stent grafts may provoke inflammation, and mechanical irritation could contribute to the recurrence of psedoaneurysm after endovascular treatment. Pre- and post-operative immunosuppressive therapy is use for BD patients to modulate the inflammation, and thereby reduce the risk of complications. It is suggested that the operation should be practiced until the inflammatory markers diminishes. The application of post-operative immunosuppressive therapy for at least two years is effective [13]. The perioperative administration of corticosteroids with immunosuppressants is also effective in decreasing the incidence of recurrent aneurysm [14].

In conclusion, it is suggested that endovascular treatment and adjunctive immunosuppressive therapy should be necessity when treating aneurysms of v-BD v-BD.

Meanwhile, an operation should be performed when the patient's condition is at the stable phase of BD. Furthermore, glucocorticoids and immunosuppressive drugs need to be continuously used after the operation. In addition, all patients should undergo long-term observation and follow-ups. Since samples are rare, multicenter and prospective studies are needed.

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Riassunto

Il coinvolgimento di un aneurisma o di uno pseudoaneurisma nella malattia vascolare di Behçet (BD) è raro e difficile da trattare. Il presente studio indaga i risultati di una strategia terapeutica conservativa per aneurisma con BD vascolare. Vengono presentati tre casi di BD con aneurisma in diverse arterie. Tutti i pazienti sono stati sottoposti a chirurgia endovascolare, ma gli effetti del trattamento sono stati diversi. Due pazienti hanno sviluppato nuovi aneurismi subito dopo l'intervento chirurgico e alla fine sono morti a causa della rottura dell'aneurisma ricorrente, mentre solo un paziente è sopravvissuto dopo il trattamento endovascolare.

In conclusione, per gli aneurismi in corso di malattia di vasculo-Behçet (v-BD), si suggerisce un trattamento endovascolare durante il periodo di satbilizzazione. Allo stesso tempo, i glucocorticoidi e gli immunosoppressori potrebbero essere usati in associazione prima e dopo l'intervento.

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