Incomplete intestinal obstruction as the possible main complaint in Behcet’s disease after surgery for recurrent abdominal aortic pseudoaneurysms: a case report and literature review

Fang Jiang†, Hui Xiang† and Zhi-Yong Peng*†

Abstract

Background: Behcet’s disease (BD) is a systemic vasculitis characterized by oral and genital aphthosis, and ocular and skin lesions. The disease is involved in vascular, gastrointestinal, and central nervous systems. Vasculitis may exacerbate fatal problems, such as anastomotic pseudoaneurysms. If the mesenteric vessels are involved, severe abdominal symptoms such as intestinal obstruction may occur.

Case presentation: This case report describes a young female patient who suffered from BD with recurrent abdominal aortic pseudoaneurysms, as well as deep venous thrombosis and subsequent complications of incomplete intestinal obstruction. This patient first underwent stent grafting, which was followed by rupture of two newly formed anastomotic pseudoaneurysms within six months. Emergency open surgical repair (OSR) was then performed on the ruptured pseudoaneurysms. Thrombosis and incomplete ileus occurred five months after surgery. This case was unique due to the presence of incomplete intestinal obstruction being the possible main complaint for a patient with Behcet’s disease, and it is the first ever case to be reported.

Conclusion: Intestinal obstruction may present as the possible main complaint in BD. Careful and attentive strategy should be carried out to prevent fatal outcomes.

Keywords: Behcet’s disease, Recurrent abdominal aortic pseudoaneurysm, Incomplete intestinal obstruction

Background

Behcet’s disease (BD) is a disease that involves complex multisystem disorders. However, the exact physiopathologic mechanism of BD remains unclear. Evidence shows that arterial and venous systems are involved in BD. Involvement of the venous systems results in thrombosis and superficial phlebitis. Involvement of the arterial systems result in aneurysm or pseudoaneurysm, stenosis, and occlusion [1]. The pseudoaneurysm, which evolves quickly, is prone to rupture and can become life-threatening. Therefore, it is crucial to diagnose and manipulate this fatal condition early. If the mesenteric vessels are involved, severe abdominal symptoms such as intestinal obstruction may occur [2]. However, such symptoms, especially those presenting as the main complaint, are easily misdiagnosed. Here, we report a rare case of a patient who presented with incomplete intestinal obstruction as the possible main complaint after a second surgery. We believe that an examination of this case will help the field make progress toward developing a common procedural strategy for treating such rare conditions.

Case presentation

A 28-year-old female Chinese patient presented with the onset of acute continuous right abdomen pain, nausea...
and vomiting at the emergency department. On admission, abdominal dynamic computed tomography (CT) with a multislice detector row CT scanner showed several air-fluid levels in the enteric cavity, and the diagnosis was considered to be ileus.

The patient was diagnosed with BD four years ago. She had received medications regularly, including immunosuppressive therapy with oral prednisone (60 mg/day) and cyclophosphamide (100 mg/day).

The patient first presented with abdominal pain at the hospital. A computed tomographic angiography (CTA) was performed, which (Fig. 1) indicated an aneurysm of 5.67 cm*5.28 cm*0.97 cm located in the left junction of the thoracic and abdominal aorta. An approximately 0.8 cm segment was found to block the starting part of the celiac trunk. Subsequently, a graft stent was implanted. Follow-up CTA (Fig. 2) showed no residual aneurysm. However, six months after the intervention, a rapidly growing mass was found in the lower abdomen, and the patient presented with nausea, vomiting, progressive and intermittent pain in the abdomen which radiated to her back. Before being admitted to the intensive care unit (ICU), the patient underwent CTA, which showed that there was a haematoma of approximately 6.0 × 4.8 cm in the abdomen. The extravasation of contrast agent was located in the opening of the renal artery, with a mixed soft tissue mass of 3.7 cm (Fig. 3) in the haemoperitoneum. The haemoglobin concentration decreased to 4.36 g/dL with abrupt hypotension (60/43 mmHg). The critical condition of the patient prompted the cardiac surgeons to perform open surgical repair (OSR) rather than a more conservative treatment. The patient was transferred to the ICU after the operation. Three days later, after her vital signs were stable, she was transferred back to the general ward. No recurrence of pseudoaneurysm was found during a follow-up of 15 days (Fig. 4). She continued receiving immunosuppressive therapy as usual. This course of treatment was decided based on previous studies showing that use of immunosuppressive therapy with cyclophosphamide and corticosteroids before and after surgical intervention could help prevent BD activation [3, 4].

Ten months later, after the implantation of artificial vascularization in the thoracic and abdominal aorta,
doppler-ultrasound indicated deep venous thrombosis in the left popliteal vein (Fig. 5). Therefore, the patient was treated with anticoagulant therapy using hypodermic injections of low-molecular-weight heparin at a daily dose of 4100 U.

One month later, the patient suffered from persistent right abdominal pain with nausea and vomiting accompanied by oral aphtha and genital ulcer. Abdominal CT showed the occurrence of an air-fluid level in part of the ileum and colon (Fig. 6). The diagnosis was determined to be an incomplete intestinal obstruction, which may have been caused by the previous aneurysms. Mesenteric artery angiography showed that the root of the celiac trunk and superior mesenteric artery were stenosed (Fig. 7). Then, conservative treatment was administered, such as fasting, gastrointestinal decompression and

---

Fig. 3 The hematocyst occurred at 6 months after intervention. The size was 6.0*4.8 cm. The extravasation of contrast agent located in the opening of the renal artery, and the thickness of this mixed soft tissue mass was 3.7 cm.

Fig. 4 Implantation of artificial vascular in the thoracic and abdominal aorta. Two transplanted vessels were seen in picture D, one of which was connected to abdominal aorta, the other connected to right common iliac artery.

Fig. 5 The deep venous thrombosis in the left popliteal vein. It was detected by doppler-ultrasound. Arrow pointed to the thrombosis. Jiang et al. BMC Cardiovascular Disorders (2018) 18:238
enema. Approximately 20 days later, the patient recovered well and was discharged from the hospital.

**Discussion and conclusions**

Here we demonstrated a case of a patient with a diagnosis of BD who presented with incomplete intestinal obstruction as the possible main complaint. To the best of our knowledge, this is the first report of the presence of incomplete intestinal obstruction after a second surgery for BD.

Recent studies have reported that HLA-B51 is associated with a more severe disease and is related to familial BD [5]. Other studies have shown that polymorphism in the IL-21, IL-10, and IL-8 genes and in the tumour necrosis factor-(TNF-)alpha-1031C allele are correlated with the pathogenesis of BD [6–11]. The role of the IL-23/IL-17 axis has been found to be associated with the onset of BD [12, 13]. Meanwhile, delayed-type hypersensitivity to infections such as streptococcus species (especially sanguis) has also been proposed to be an important factor in inducing BD [14]. Hematological diseases such as coagulation abnormalities and platelet overactivity may play additional roles in the development of BD [15–17]. Nitric oxide (NO) accumulation after induction by interferon-gamma is also related to the activation of BD [18].

BD can be divided into two categories according to the pathological mechanism: occlusive and lesion. Although venous involvement is the most common vasculo-BD complication, Ketari et al. has been reported that arterial involvement is more common than venous involvement [19]. The most severe complication is aneurysm formation and rupture. Approximately 60% of reported arterial lesions associated with BD are aneurysms [20]. The aorta is the most commonly affected artery followed by the pulmonary artery. Aneurysms occur more frequently in the abdominal aorta than in the thoracic aorta [21–23].

Surgical repair is the standard treatment for aneurysms with dilatation, stenosis, or obstruction, while alternative treatments involve minimally invasive endovascular therapy for aneurysm repair [22, 24, 25].
Shen et al. reported that both open surgery and endovascular repair were safe and effective for treating aortic pseudoaneurysm in BD [26]. Furthermore, Naganuma et al. reported that ulcer lesions in the ileum and colon, which could lead to intestinal obstruction, were found in surgical patients with intestinal BD [27].

In our report, it was challenging to diagnose the patient who presented with initial symptoms of abdominal pain and vomiting. The differential diagnoses included pseudoaneurysm, ileus, new recurrent arterial aneurysm, mesenteric arterial embolism and so on. To obtain an accurate diagnosis, abdominal computed tomography and angiography were performed. CT/CTA results showed that the patient suffered from intestinal obstruction. The cause may be related to chronic vascular lesions, which could finally lead to an insufficient blood supply in the mesenteric artery. Literature review demonstrated that intestinal obstruction presented with BD in three cases. Of them, two cases of BD that presented with abdominal aortic aneurysm were diagnosed with ileus after stent graft interposition [28]. One patient with BD suffered from complete obstruction 6 years after several operations. The operations included an aortobifemoral bypass grafting for an abdominal aortic aneurysm [2] (Table 1). However, incomplete intestine obstruction was not reported as the main complaint for these patients. Our case report is the first to present incomplete intestinal obstruction as the possible main complaint of BD. Unfortunately, there are no certain guidelines for how to prevent and treat intestinal obstruction caused by BD. Thus, prompt differential diagnosis and management are essential for future treatment.

Considering the paucity of data available, the management of patients with intestinal obstruction should be individualized, and a multidisciplinary approach should be taken. Differential diagnosis and management are essential when addressing this fatal problem.

Technologies such as abdominal computed tomography and angiography are recommended resources when determining differential diagnosis. Finally, if intestinal obstruction is confirmed with the diagnosis, appropriate measures (surgical or nonsurgical treatment) to treat this disease should be taken.

### Table 1
Characteristics of lesion, treatment and outcome for patients complicated with intestinal obstruction from literature review

| Patient | Location     | Symptom | Treatment (graft size & type) | Complication                      | Immuno-suppressant | Recurrence | Follow up | Result |
|---------|--------------|---------|-------------------------------|-----------------------------------|---------------------|------------|-----------|--------|
| 38 yrs. male | Infrarenal | C. Rupture | Interposition (Polytetrafluoroethylene) 16 × 8, bifurcated | Incomplete Intestinal Obstruction | Prednisolone + Colchicines | None       | 98 M      | Alive  |
| 31 yrs. male | Suprarenal  | Rupture  | Patch (Polytetrafluoroethylene) | Incomplete Intestinal Obstruction | Colchicines + Azathioprine | 18 M       | 43 M      | Alive  |
| 50 yrs. male | Suprarenal  | vascular | Stent graft | Complete Intestinal Obstruction | Not clear            | 72 M       | 5 M       | Alive  |

### Abbreviations
BD: Behcet’s disease; CT: Computed tomography; CTA: Computed tomographic angiogram; ICU: Intensive care unit; NO: Nitric oxide; OSR: Open surgical repair

### Acknowledgements
We would like to express our sincere appreciation to Dr. Jinping Zhao and Dr. Ming Xu of the Department of Cardiothoracic Surgery for their valuable comments.

### Funding
This study was supported by the National Natural Science Foundation of China (Z.P. 81560131) and Hubei Province Key Projects (Z.P. WJ2017Z008).

### Availability of data and materials
All relevant data supporting the conclusions are contained within the article.

### Authors’ contributions
ZP designed this study and revised the manuscript. FJ & HX analyzed and interpreted the patient data regarding Behcet’s disease. FJ & ZP conducted the manuscript writing. ZP, FJ & HX treated the patient in the ICU. All authors read and approved the final manuscript.

### Ethics approval and consent to participate
Not applicable.

### Consent for publication
Written consent was obtained from the patient’s father for the publication of this case report, because the patient was critically ill and her father was authorized to act as her guardian.

### Competing interests
The authors declare that they have no competing interests.

### Publisher’s Note
Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

Received: 12 March 2018 Accepted: 6 December 2018
Published online: 17 December 2018

### References
1. Nair JR, Moote RJ. Behcet’s disease. Clin Med (Lond). 2017;17(1):71–7.
2. Bayar S, Unal E, Esen S, Kurt T, Ensari C, Alacayir I. Aortobifemoral graft causing complete intestinal obstruction in a patient with Behcet’s disease: report of a case. Surg Today. 2004;34(2):185–7.
3. Tuzun H, Seyahi E, Arslan C, Hamuryudan V, Besirli K, Yazici H. Management and prognosis of nonpulmonary large arterial disease in patients with Behcet disease. J Vasc Surg. 2012;55(1):157–63.
4. Barnes CG. Treatment of Behcet’s syndrome. Rheumatology (Oxford). 2006;45(3):246–7.
5. Freisko I, Soy M, Hamuryudan V, Yurdakul S, Yavuz S, Turner Z, Yazici H. Genetic anticipation in Behcet’s syndrome. Ann Rheum Dis. 1998;57(1):45–8.
6. Gori G, Terrier B, Rosenzweig M, Wechsler B, Touzot M, Selhieman D, Tran TA, Bodaghi B, Musset L, Soumelis V, et al. Critical role of IL-21 in modulating TH17 and regulatory T cells in Behcet disease. J Allergy Clin Immunol. 2011;128(3):655–64.
7. Touma Z, Farra C, Hamdan A, Shamseddeen W, Uthman I, Hourani H, Arayssi T. TNF polymorphisms in patients with Behcet disease: a meta-analysis. Arch Med Res. 2010;41(2):142–6.

8. Bonyadi M, Jahanafrooz Z, Esmaeili M, Kolahi S, Khabazi A, Ebrahimi AA, Hajialoo M, Dastgiri S, Khabazi A, Ebrahimi AA, Haji-Alli M. TNF-alpha gene polymorphisms in Iranian Azeri Turkish patients with Behcet’s disease. Hum Immunol. 2009;70(7):549–54.

9. Shah A, Alspoy E, Kosara M, Balar M, Yavuzer U, Alspoy E, Yegin O. TNF-alpha gene 1031 T/C polymorphism in Turkish patients with Behcet’s disease. J Biomed Sci. 2009;16(2):285–9.

10. Kamoun M, Chellbi H, Houman MH, Lahou B, Hamzaoui K. Tumor necrosis factor gene polymorphisms in Tunisian patients with Behcet’s disease. Hum Immunol. 2007;68(9):201–5.

11. Akman A, Sallakci N, Kacaroglu H, Tosun O, Yavuzer U, Alpsoy E, Yegin O. Relationship between periodontal findings and the TNF-alpha gene 1031 T/C polymorphism in Turkish patients with Behcet’s disease. J Eur Acad Dermatol Venereol. 2008;22(8):950–7.

12. Leng RX, Chen GM, Pan HF, Ye DQ. The role of IL-23/IL-17 axis in the etiopathogenesis of Behcet’s disease. Clin Rheumatol. 2010;29(10):1209.

13. Dincle B, Alspoy E, Karakas AA, Yilmaz SB, Yegin O. Homocysteine in vascular Behcet disease: a meta-analysis. Arterioscler Thromb Vasc Biol. 2010;30(10):2067.

14. Shanker J, Gasparian AY, Kitas GD, Kakkar W. Platelet function and antithrombotic therapy in cardiovascular disease: implications of genetic polymorphisms. Curr Vasc Pharmacol. 2011;9(4):479–89.

15. Gasparian AY, Ayazyan L, Mikhailidis DP, Kitas GD. Mean platelet volume: a link between thrombosis and inflammation? Curr Pharm Des. 2011;17(1):147–58.

16. Belouzard M, Descoyenne D, Lahmar K, Ahmedi I, Medjebre O, Carton D, Lahou-Boukoffa O, Touil-Boukoffa C. Interferon-gamma and nitric oxide production during Behcet uveitis: immunomodulatory effect of interferon-gamma. J Interf Cytokine Res. 2011;31(9):643–51.

17. Kim WH, Choi D, Kim JS, Ko YG, Jang Y, Shim WH. Effectiveness and safety of endovascular aneurysm repair in Behcet disease complicated by recurrent thoracoabdominal aortic aneurysms. Vasc Endovasc Surg. 2010;44(2):146–9.

18. Akin U, Sarmak C, Turan T, Ipekci A, Sargin N, Karabulut H. Aortic arch aneurysm in Behcet disease. Int J Radiat Oncol Biol Phys. 2010;76(1):216–21.

19. Yamada T, Yamamoto I, Ohtsuki Y, Katsumura T, Tsuchida T, Kishi H, Tsuchida Y, Nakamura Y. Characteristics of Streptococcus suis isolated from patients with Behcet’s disease. Microbiol Immunol. 1995;39(9):729–32.

20. Kim WH, Choi D, Kim JS, Ko YG, Jang Y, Shim WH. Effectiveness and safety of endovascular aneurysm repair in Behcet disease complicated by recurrent thoracoabdominal aortic aneurysms. Vasc Endovasc Surg. 2010;44(2):146–9.

21. Calguneri M, Kiraz S, Erenli I, Benlik M, Karacsil Y, Celik I. The effect of prophylactic penicillin treatment on the course of arthritis episodes in patients with Behcet’s disease. A randomized clinical trial. Arthritis Rheum. 1996;39(12):2062–5.

22. Naganuma M, Iwao Y, Inoue N, Hisamatsu T, Imai T, Ishii H, Kanai T, Watanabe M, Hibi T. Analysis of clinical course and long-term prognosis of surgical and nonsurgical patients with intestinal Behçet’s disease. Am J Gastroenterol. 2000;95(1):631–8.