



Recurrent intestinal ischemia related to Behçet's Disease

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ABSTRACT

Behçet's disease is a systemic inflammatory disease that may affect multiple organs. However, intraabdominal complications requiring surgical intervention are rare in the natural course of the disease. A 32-year-old male patient with Behçet's disease who had been followed for 5 years with a diagnosis of acute abdominal syndrome was operated on twice in 18 days. Intestinal ischemia was identified in different segments of the jejunum during each operation. Recurrent segmental intestinal ischemia within a short time interval is rare, although the gastrointestinal involvement can be seen in the normal course of Behçet's disease.

Keywords: Behçet's disease, intestinal Behçet's disease, vasculitis, small bowel ischemia

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INTRODUCTION

Behçet's disease (BD) is a systemic vasculitis characterized by ocular inflammation (e.g., uveitis) and recurrent oral or genital ulceration. Various other organs and systems, including the locomotor system, central nervous system, and gastrointestinal system (GIS), may also be affected during the disease's natural course. The etiology of BD is unclear.

Behçet's disease with GIS involvement is referred to as 'intestinal Behçet's disease' or 'entero-Behçet's disease' in the literature. The prevalence of this condition varies according to different geographic regions, but it has been noted in 1%–2% of all BD patients (1). Abdominal pain, vomiting, and loss of appetite are major symptoms of the GIS involvement; however, these are commonly encountered symptoms in emergency rooms. Although rare, massive gastrointestinal bleeding and hollow organ perforation may also be encountered in BD (2).

Medical treatment is the preferred option for BD in the absence of complications (3, 4). In this paper, a BD patient who was operated on twice within a short time interval is described.

CASE PRESENTATION

A 32-year-old male patient was admitted via the emergency room with a 2-day history of abdominal pain, fever (38.5°C), nausea, and vomiting. The patient had been admitted to another clinic 5 years earlier with complaints of left lower extremity swelling and painful oral and genital lesions; he was diagnosed with BD (HLA B51⁺) based on his medical history. Medical treatment (sodium warfarin, 5 mg/day; and colchicine, 1.5 mg/day) was started due to left-thigh deep venous thrombosis (DVT). One year later, a steroid (prednisolone 40 mg/day) was added to the anti-coagulant and colchicine regimen due to polyarthritis. This treatment regimen was followed regularly by the patient; however, 3 years later, the patient was diagnosed with the right-lower extremity DVT and was treated medically. After this condition had resolved, the patient stopped all medications and was lost to follow-up for 1 year.

On admission, diffuse abdominal tenderness and peritoneal irritation signs were detected during a physical examination. All laboratory findings were normal, except leukocytosis ($12.6 \times 10^3/\text{mm}^3$). The patient's air–fluid levels on abdominal radiography suggested intestinal obstruction. Intraabdominal free fluid and localized dilatation in the proximal small bowel segment were detected on the unenhanced (due to contrast allergy) abdominal computed tomography (CT; Figure 1). A laparoscopic exploration was planned with acute abdominal syndrome as the preliminary diagnosis. During the exploration, segmental small bowel ischemia was found in the proximal jejunum (30 cm in length, 40 cm away from the Treitz ligament). A laparoscopic segmental small bowel resection was performed. The patient was discharged uneventfully on postoperative Day 6 with steroid therapy (40 mg/day). After 7 days, the patient was readmitted with complaints of abdominal pain, nausea, and vomiting. Similar findings to the first admission were detected on a physical examination (diffuse abdominal tenderness and peritoneal irritation signs) and on the abdominal CT (intraabdominal diffuse free fluid and localized dilatation in the

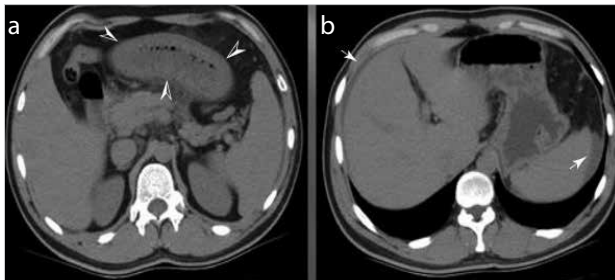


Figure 1. a, b. Abdominal CT. Wall thickening at the proximal jejunum (image A, arrow heads) and intraabdominal free fluid in the perihepatic–perisplenic areas (image B, arrows)

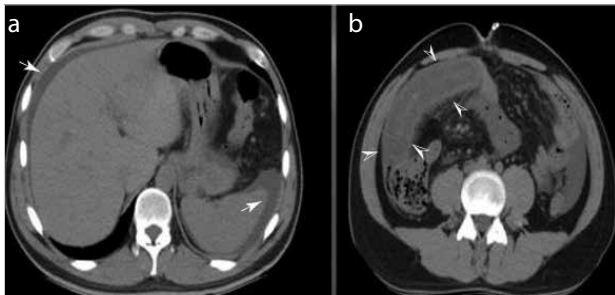


Figure 2. a, b. Abdominal CT. Similar findings to the first CT are seen: intraabdominal free fluid in the perihepatic and perisplenic areas (image A, arrows) and small bowel wall thickening (image B, arrow heads)

proximal small bowel segments) (Figure 2). Given the patient's history of the small bowel resection, we decided to reexplore with a preliminary diagnosis of possible surgical complications or recurrent ischemia. During the exploration, recurrent ischemia was detected in two different small bowel segments (first: 50 cm in length, within the jejunum, starting at the end of the previous anastomosis; second: 50 cm in length, within the ileum, located 100 cm away from the ileocecal valve) (Figure 3), and the previous anastomosis was apparently normal. The ischemic segments were resected and two end-to-end anastomoses were performed separately. The patient was discharged uneventfully on postoperative Day 7. Findings of vasculitis, compatible with BD, and ischemic changes (superficial epithelial necrosis in villous structures, extensive hemorrhage in the lamina propria, vascular congestion, thrombosis, and extensive edema and hemorrhage in the submucosa) were obtained by histopathological examinations (Figure 4). The patient has been followed uneventfully for 38 months with regular rheumatology consultations.

DISCUSSION

Behçet's disease, a systemic disease characterized by recurrent genital–oral ulcers and ocular inflammation, was first described by Hulusi Behçet in 1937 (2, 3). The prevalence of the GIS involvement in BD, which usually affects adults aged 20–40 years, is 1%–2% (1). Despite its unclear etiology, BD is thought to occur after the exposure to an environmental agent in people with a genetic predisposition. The prevalence of BD varies by geographic regions; this finding has been used to support environmental and genetic causes of the disease (1, 5–7). Human leukocyte antigen (HLA) B-51 is the most powerful positive antigen in genetic studies (5). In our case, the



Figure 3. Intraoperative view during the second operation: small bowel ischemia

age range and HLA B-51 positivity of the patient were compatible with BD.

Recurrent oral ulcers (99%), genital ulcers (75%), and uveitis are classical first symptoms of BD. The central nervous system and middle–large vessels are usually affected years later, and such involvement can have serious results. In our case, large vessel involvement (left lower extremity DVT) was present at the first diagnosis of BD 5 years earlier.

The symptoms of BD are usually non-specific (diarrhea, nausea, vomiting, weight loss, and recurrent abdominal pain) in the presence of the GIS involvement; for this reason, making a diagnosis is nearly impossible based on these symptoms in a patient who is not already known to have BD (4–9). The GIS involvement may occur at any location, and intestinal–extraintestinal symptoms may coexist with intestinal BD-like inflammatory bowel disease (IBD) (2). While the ileocecal area is the most commonly affected area, the rectum is the least commonly affected area (1%) in BD (10, 11). In fact, intestinal complications requiring surgical intervention (e.g., an intestinal fistula, perforation, or bleeding) are more common in BD than in IBD, but they are rarely encountered with BD in daily practice (9). Such complications usually occur 4–6 years after

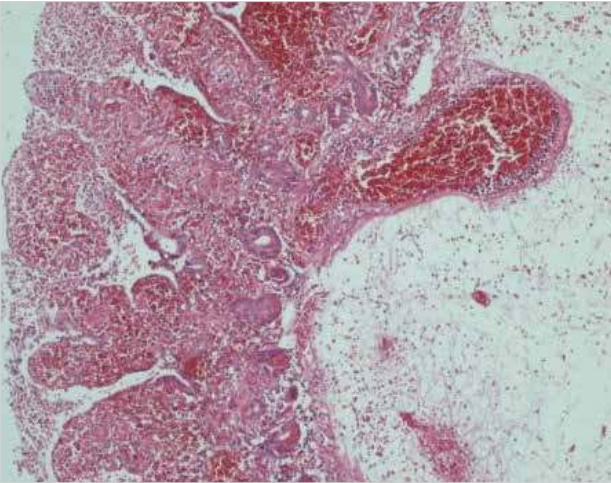


Figure 4. Histopathological view: superficial epithelial necrosis in villous structures, extensive hemorrhage in the lamina propria, vascular congestion, thrombosis and extensive edema, and hemorrhage in the submucosa (hematoxylin and eosin, $\times 40$)

the detection of oral ulcers. In our case, BD was diagnosed because of 'classical' BD symptoms that occurred 5 years earlier and a 2-year history of recurrent abdominal pain and mild weight loss (3–4 kg in the last year).

Despite being extremely rare, BD may present with intestinal ischemia due to mesenteric vascular thrombosis because it is a vasculitis in origin. Generalized peritonitis, which has high mortality and morbidity rates, may occur depending on post-ischemic necrosis and perforation. Despite consensus in the literature about the need for surgical intervention when these complications develop, there is no consensus about the details, including the length of the resection and management of intestinal continuity (12). Because the complication rate is high after anastomosis, the dominant opinion in the literature is that the intestinal continuity should be provided with a stoma instead of anastomosis (4, 10). Although a high recurrence rate (40%–60%) is expected after surgery in patients with complex BD, patients who require a second operation usually have an inadequate postoperative medical treatment history (2, 4, 9, 12). In contrast to previous reports, we chose primary anastomosis after resection for intestinal continuity. If we had chosen a stoma, we would have been confronted with short bowel syndrome because the affected intestinal segments were proximal segments in both operations.

CONCLUSION

Even though it is extremely rare, BD may present with acute abdominal syndrome due to intestinal involvement. A combination of gastrointestinal complaints and BD should be taken seriously, and the possibility of intestinal complications requiring surgery must be kept in mind. We believe that primary anastomosis after the resection is a safe option if the affected segment

is short and there are no known poor prognostic criteria (e.g., intraabdominal intense contamination, an intraabdominal abscess, and inadequate blood supply) for anastomosis.

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