A Case of Behcet’s Disease with Bowel Perforation as the First Presentation and Bowel Resection and Literature Review

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Abstract

Behcet's disease is a chronic recurrent idiopathic inflammatory disease that occurs in the gastrointestinal tract and involves many systems. The most commonly involved site is the ileocecal region. Bowel ulcers caused by Behcet's disease can cause perforation. A 19-year-old male patient was admitted with a diagnosis of Behcet's disease. He underwent surgery at our clinic, and during the laparotomy, it was determined that the terminal ileum had multiple perforations. Part of the ileum was excised. The patient was discharged from the hospital after surgery without any problems. The patient's ileostomy was closed at the second month after the first surgery. There were no clinically significant problems during the 16-month follow-up period. In this article, Behcet's disease Multiple perforation of the ileum is a rare complication at the time of diagnosis and is discussed with the knowledge of published cases. This article summarized the clinical data of 1 case of enterobechase disease with intestinal perforation as the first clinical manifestation in our hospital and reviewed the literature, discussed the clinical characteristics, diagnosis and the latest treatment progress of enterobechase disease, in order to improve the understanding of this disease.

Keywords

Behcet's disease, Bowel behcet's disease, Diagnosis, Treatment

Behcet's disease, also known as Behcet's disease BD, oral-eye-genital triad, etc. is a chronic, inflammatory, relapsing immune-mediated disease of unknown etiology involving multiple organs, characterized by recurrent oral and genital ulcers, eye disorders, gastrointestinal ulcers, vascular disorders and skin lesions. According to the damage to the visceral system, BD can be divided into vascular, neurological and gastrointestinal types. When BD is involved in the gastrointestinal system, serious complications will occur, including massive bleeding, intestinal perforation and fistula, with the incidence of 10%-50% [1], most commonly occurs in young and middle-aged people aged between 20 and 50, with a male/female incidence ratio of 1.4:1. However, there is no special clinical manifestation of bowel behcet disease, and some patients even take gastrointestinal symptoms as the first manifestation. The diagnosis of bowel Behcet disease lacks specific laboratory examination and pathological diagnosis, which is mainly based on the comprehensive analysis of the clinical manifestations, symptoms and signs of the patients. In order to improve the understanding and early diagnosis and treatment of this disease, a case of enterobehcasse disease with intestinal perforation as the first clinical manifestation admitted to our hospital was reviewed and relevant literature was used to discuss the diagnosis, treatment and management of intestinal BD, as reported below.
1. Case Profile

1.1 Case summary

A 47-year-old male patient, 2022-02-15, was diagnosed with "abdominal pain accompanied by abnormal stool characteristics 10+Day, aggravated 2 days" in the department of gastroenterology. Patient 10+Days ago there was no obvious cause of lower abdominal pain with abnormal stool characteristics, stool for water, 2 days ago abdominal pain symptoms aggravated, the pain degree is tolerable, nothing to do with the change of position, there is still water stool. Then the outpatient department to "cause of abnormal defecation" income hospital. Past 1+(2020-11-26) due to "sudden lower abdominal pain 18 years ago+She was diagnosed as perforation of the small intestine and multiple penetrating ulcers of the colon" in the emergency department of our hospital. Laparotomy was performed under static aspiration combined with general anesthesia: intestinal adhesion release, abdominal abscess removal, partial resection of the small intestine, resection of the right half colon, and enterostomy. Intraoperative examination showed pathological findings: high hyperemia and edema of intestinal wall tissue, chronic inflammation of intestinal mucosa with mucosal exfoliation. He was discharged after a good recovery. Patient's private complaint 1+Before perforated bowel resection, the patient had repeated ulcers in the oral cavity and external genitalia, but they were not paid attention to. Now the patient came to our hospital for systematic diagnosis and treatment of the above discomfort.

1.2 Physical examination

Physical examination Body temperature: 36.4℃, pulse: 108 beats/min, respiration: 20 beats/min, blood pressure: 98/73mmHg. The sclera of the skin was not yellow and superficial lymph nodes were not swollen. No abnormality was found in cardiopulmonary examination. The abdomen was flat and soft, no gastrointestinal type or peristaltic wave was observed, no varicose veins in the abdominal wall, no mass was touched, light tenderness in the lower abdomen, no tenderness in the remaining abdomen, no rebound pain, no muscle tension, the liver and spleen were not touched, Murphy's Sign was negative, the mobility voiced voice was negative, the liver voiced voice boundary existed, and the bowel ringing was normal.

1.3 Ancillary examination

After admission, colonoscopy was performed. Under the microscope, multiple large ulcers with a diameter of 0.3-1.5cm deep were observed at the anastomosis on the side of the small intestine, the bottom was covered with thick white moss, the surrounding mucosa was congested, the biopsy quality was tough, and the small intestinal cavity was narrow. Multiple large ulcers with a diameter of 0.5-1.0cm were observed 20cm and 30cm away from the anus, and the bottom was covered with thick white moss. There was a fissure ulcer about 0.2×0.5cm in size in the sigmoid colon 28cm from the anus with bleeding at the margin. Endoscopic diagnosis: 1. Postoperative colon perforation with anastomotic ulcer and anastomotic nail residue; 2.2. Crohn's disease? Behcet's disease? (Please combine pathology). Pathological results showed moderate chronic mucosal inflammation with ulcer formation, activity and focal lymphoid tissue hyperplasia. Laboratory examination: Blood routine showed a white blood cell count of 9.04×10^9/L, neutrophil percentage 70.80%, lymphocyte percentage 19.20%, hemoglobin 122.00g/L, platelet count 383.00×10^9/L; Fecal OB weak positive; Esr 54mm/h; C- reactive protein 43.110mg/L; ANA antibody profile showed negative titer of anti-nuclear antibody 1 and weak positive titer of anti-SSA antibody. Behcet disease related gene test (HLA-B51) positive; Immunoglobulin, rheumatoid factor, anti- streptococcal hemolysin O, cardiolipin antibody, anti-neutrophil cytoplasmic antibody, tuberculosis infection T cell detection were not abnormal. Imaging examination: enhanced CT of small intestine: 1. Thickened wall of the greater curvature of the stomach, rectum and left upper abdomen, increased number of abdominal and retroperitoneal lymph nodes with slightly larger parts, please combine with endoscopic examination. 2. Postoperative changes of digestive tract perforation should be combined with medical history.

1.4 Diagnosis

Diagnosis 1. Behcet's disease complicated with gastrointestinal bleeding; 2. After small intestinal perforation resection.

1.5 Treatment

After the diagnosis was confirmed, he was given prednisone acetate 20mg/d orally, mesalazine 4g/d orally, supplemented with PPI drugs to suppress acid and protect the stomach, confuxin, probiotics, etc. After the above
treatment, his symptoms improved and no abdominal pain or blood in the stool was found, so he was discharged from the hospital. After discharge, he was instructed to take prednisone acetate 20mg/d orally. Calcium carbonate D3 tablets, Esomeprazole magnesium enteric-soluble capsules, Clostridium enterococcus triad viable bacteria tablets, Kangfuxin solution, L-glutamine sodium gualenate granules. During the monitoring of stool condition, every 2 weeks I outpatient visit. On 2022-04-27, the patient presented with increased abdominal pain accompanied by unformed stool and was hospitalized again. After the diagnosis was confirmed and conjunctivized, the biological agent infliximab was added for targeted therapy on 2022-05-03 with mesalazine sustained-release granules 4g/d and prednisone acetate 40g/d. The possibility of pulmonary tuberculosis could not be ruled out due to chest CT indicating double pulmonary nodules. After bronchoscopy and alveolar lavage were performed to detect rifampicin resistance genes of mycobacterium tuberculosis, no abnormalities were found. After consultation, isoniazid 0.3g/d and rifampicin tablets 0.45g/d were recommended for anti-tuberculosis treatment in the Department of Respiratory and Critical Care Medicine. After the treatment, the abdominal pain improved and the patient was discharged from the hospital. The patient was asked to return to the hospital on time (week 2, week 6, and then received an injection every 6-8 weeks) for another infusion of infliximab targeted therapy. After regular follow-up and planned infusion of infliximab targeted therapy, the condition was stable.

2. Discussion

Behcet's disease, also known as Behcet's disease BD, mouth - eye - genital triad, etc., was first reported by Behcet in 1937 [2], is a chronic, inflammatory, relapsing immune-mediated disease of unknown etiology that accumulates multiple systems and is characterized by oral and vulvar aphthous ulcers, ophthalmitis, skin lesions, and other clinical manifestations. Its basic pathological changes are multi-vascular involvement of non-specific vasculitis, large, medium and small blood vessels, arteries, veins and capillaries can accumulate throughout the body. BD epidemiology was distributed along the ancient Silk Road. In the Middle East, East Asia and the Mediterranean. The disease occurs in 25-35 years old young adults, male patients slightly more than female patients. The diagnosis of bowel behcet's disease is based on the gastrointestinal symptoms of BD patients and the typical intestinal ulcerative lesions recorded by objective measures [3]. When BD gastrointestinal involvement, serious complications will occur, including massive bleeding, intestinal perforation and fistula, with the incidence of 10%-50% [4].

At present, there is no definite basis for the diagnosis of bowel Behcet's disease, which usually occurs 4-5 years after the diagnosis of Behcet's disease. Due to its diverse clinical manifestations and lack of specificity, the diagnosis is usually made according to the gastrointestinal symptoms and gastroenteroscopy findings of patients. Paraphylaxis of abdominal pain is the most common symptom of BD. If a patient has no typical manifestations of BD, it is easily confused with inflammatory bowel disease, such as ulcerative colitis and Crohn's disease, according to the gastroenteroscopy findings. Therefore, detailed medical history, physical examination, gastroenteroscopy findings and immune-related examinations are helpful for the diagnosis of BD.

Currently, there are no evidence-based medical guidelines that can be recommended for the management of intestinal BD, therefore, the expert opinion is based on Crohn's treatment guidelines for intestinal BD [5]. According to the Japanese Consensus on intestinal BD published in 2007, 5-aminosalicylic acid, corticosteroids, immunomodulator, enteral nutrition, total parenteral nutrition and surgical treatment are included in the standard treatment of intestinal BD [6]. In patients with BD, the number of γδ cells producing TNF-α in the body increases, leading to increased levels of TNF-α and its receptor in blood, and the expression of TNF-α increases with the progression of the disease [7]. Combined with the intestinal BD consensus statement of the 2nd edition published in 2014, it is recommended that anti-TNF-α drugs such as adalimumab and infliximab be used as standardized treatment for intestinal BD [8]. Therefore, the importance of anti-TNF-α drugs is self-evident, and they are also commonly used in the treatment of rheumatoid arthritis, ankylosing spondylitis and other autoimmune diseases. There have been several literatures at home and abroad reporting the efficacy of induction and maintenance of infliximab in the treatment of intestinal BD, which has been widely accepted. However, there is still insufficient evidence on whether the combination of immunomodulator therapy can be used in the postoperative treatment of patients with intestinal resection. Perhaps, close follow-up of this patient can provide valuable data for this kind of study.

To sum up, Behcet's disease is a rare autoimmune disease with unknown etiology, which is called intestinal Behcet's disease or intestinal BD when it accumulates in the digestive system. Its diagnosis is difficult and it lacks specific clinical manifestations, and its endoscopic manifestations are often confused with inflammatory bowel disease. Although intestinal BD is not common, severe complications such as intestinal perforation and bleeding increase the fatality rate. Proper treatment and management of patients is an urgent issue.5-aminosalicylic acid, corticosteroids, immunomodulator and anti-TNF agents have been recommended as standard treatment for intestinal BD.
However, due to its rarity and lack of clinical data, there are currently no established drug treatment guidelines.

References