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Ileocecal Ulcer with a Cecocecal Fistula in Behcet's Disease

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We describe a case of Behcet's disease (BD) which showed the ileocecal ulcer and cecocecal fistula. This 38-year-old man had appendectomy six years ago because of colicky pain in the right lower abdomen (RLA). There are some reports on fistula formation in BD. In those, some are related to surgery and others are not. BD with cecocecal fistula, possibly associated with a past operation, has not been reported in the literature.

Key Words : *Behcet's disease, Cecocecal fistula, Ileocecal ulcer, Appendectomy*

INTRODUCTION

Though gastrointestinal symptoms are relatively common in BD, ulcerative change of the intestine is not that frequent. The most common sites for intestinal BD are the terminal ileum and the cecum. There are reports of fistula formation in BD. These include rectovaginal fistula[1], vesicovaginal fistula[2], aortoatrial fistula[3] and postoperative complication such as aortoenteric fistula[4] and enterocutaneus fistula[5]. Our patient, who presented with massive, bloody diarrhea and severe pain in RLA, had a large, deep cavitating ileocecal ulcer along with a cecocecal fistula around the previous appendectomy site.

CASE

A 38-year-old man presented with massive, bloody diarrhea and colicky pain in RLA. He had been having recurrent oral ulcer, genital ulcer and erythema nodosum-like lesion for the past ten years. Six years ago, he had an appendectomy done at a primary clinic. At that time, the multiple ulcers were noted in the ileocecal region and histology showed minimal inflammation in the appendix. He was referred to the university medical center and diagnosed as having an intestinal BD. He had been treated with low-dose prednisolone and sulfasalazine. After surgery, intermittent, severe abdominal pain occurred while he had been on medication on and off.

On examination, he had a regular pulse of 120 beats/min, blood pressure of 70/40 mm Hg and body temperature at 36.5℃. His conjunctiva was mildly pale. The lungs were clear to auscultation. Though his abdomen was soft, the abdominal examination revealed tenderness and rebound tenderness in RLA. He had a hematocrit of 33.5%, a white blood cell count of 11.8 109/mm^3 and a platelet count of 185 109/mm^3. The antinuclear antibody, rheumatoid factor and antineutrophil cytoplasmic antibody were negative. Plain abdomen showed no specific findings except for the focal ileus in RLA.

Even with enough saline and transfusion of packed red blood cells, he had persisting hypotension and severe abdominal pain. Emergency ileocecal segmental resection was performed. Pathologic examination revealed large cavitating ulcers(figure 1), lymphocyte aggregates and lymphocytic vasculitis (figure 2) in the ileocecal region. There was cecocecal fistula (figures 3 & 4), presumably around the previous appendectomy site.

He was started on oral prednisolone 10 mg/day, sulfasalazine 2 g/day and cyclophosphamide 100 mg/day

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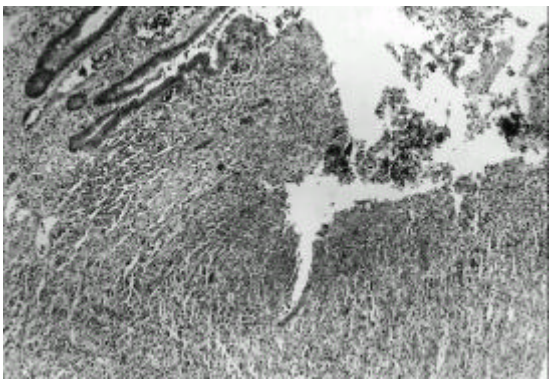

Figure 1. Histopathologic examination of the ulcer showing necroinflammation on the right side of the photograph (H&E stain, original magnification×40).

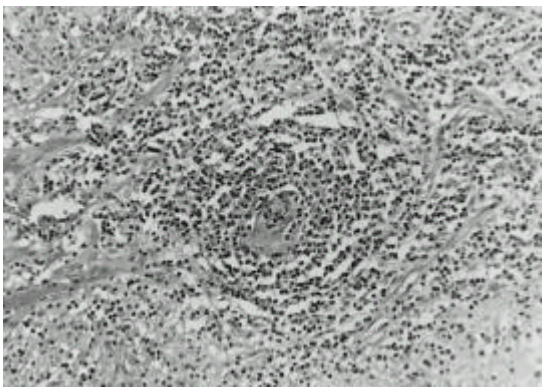

Figure 2. Marked perivascular lymphocytic infiltration noted, and some in the wall of this arteriole as well (H&E stain, original magnification×100).

in the sixth postoperative day. His postoperative course was uneventful.

DISCUSSION

BD is a multisystemic disorder characterized by oral ulcer, genital ulcer, uveitis and skin lesions, most likely occurring with the underlying vasculitis. There is other organ involvement of joints, heart and lungs, as well as neurologic and gastrointestinal involvement. The etiology remains unclear. Genetic and environmental factors probably have a role in the pathogenesis. There are no specific diagnostic or laboratory tests for BD. Diagnosis depends upon the proper history and clinical manifestations. Our case fulfilled the diagnostic criteria of

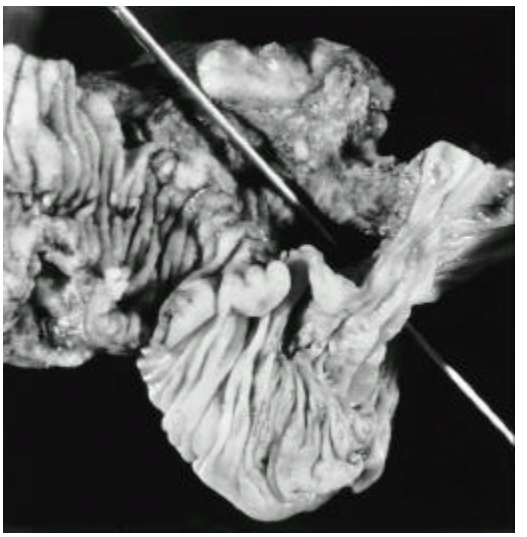

Figure 3. Gross photograph of this ileocecectomy specimen, revealing cecocecal fistula demonstrated by the probe. Ileocecal valve is destroyed with deep ulceration. The probe is passing throuch the cavitating ulcer in the most proximal cecum on the left upper to more distal non-ulcerated portion of the cecum on the right portion of the photograph.

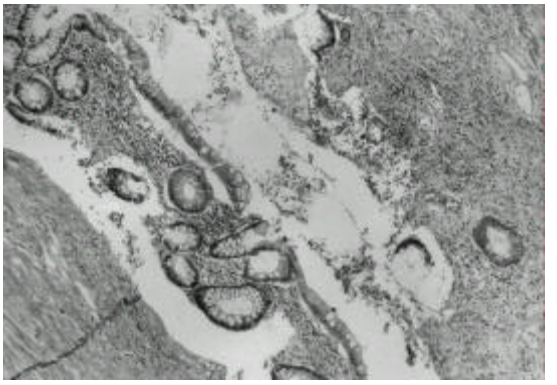

Figure 4. Photomicrograph showing fistulous track partly lined by the colonic epithelial tissue (H&E stain, original magnification×40).

the Intestinal Study Group for Behcet's disease[6].

Many patients complained of gastrointestinal symptoms such as nausea, vomiting and abdominal pain, but the ulcerative changes in the intestine were found in 1% or less of all patients with BD[7]. The commonest sites for the ulcerative changes of intestinal BD were terminal ileum in 44%, followed by the ileocecal region in 34% and the

cecum in 12%[5]. The clinical manifestations of intestinal BD are similar to inflammatory bowel disease, especially Crohn's disease. But lymphoid aggregates, submucosal fibrosis, no granuloma[8], and deep penetrating, easily perforating ulcer[9] in Behcet's colitis help to distinguish it from Crohn's disease. Histology in our case revealed lymphoid aggregates, no granuloma and deep penetrating ulcers in the ileocecal region. About 22% of the patients with intestinal BD developed symptoms mimicking appendicitis during the clinical course[10]. Because the most common sites of intestinal BD are the terminal ileum and the ileocecal region, some cases can be diagnosed as an appendicitis. Our case had a past history of erroneous diagnosis with an appendectomy. The opening of the cecocecal fistula was around the site of the appendectomy. The exact mechanism of fistula formation in BD is unclear. We could speculate that vasculitis and ulcer necrosis, along with pathergy reaction by surgical trauma in our case, might have contributed to the fistula formation.

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