Clinical Case Report

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Ulcerative colitis followed by the development of typical intestinal Behçet disease

A case report

Zhenhua Zhu, MD, PhD, Xu Shu, MD, PhD, Shunhua Long, MD, PhD, Xiaozhen Jiang, MD, Nonghua Lu, MD, Xuan Zhu, MD*, Wangdi Liao, MD, PhD*

Abstract

Rationale: Intestinal Behçet disease (intestinal BD) and inflammatory bowel disease (IBD) share a lot of characteristics, including genetic background, clinical manifestations, and therapeutic strategies, especially the extraintestinal manifestations, such as oral ulcers, arthralgia, eye lesions, skin lesions, etc, but the coexistence of these 2 diseases are uncommon. Behçet disease with gastrointestinal involvement in ulcerative colitis (UC) patient has been reported in just 1 previous case report, but, which can not be diagnosed as definite intestinal BD based on Korean novel diagnositic criteria due to lacking the typical ileocecal ulcer.

Patient concerns: We present a 23-year-old woman with ulcerative disease who developed typical intestinal BD, which is the first case report of patient with coexisting UC and typical intestinal BD.

Diagnoses: This patient was diagnosed as coexistence of intestinal BD and UC base on the clinical manifestations, extra intestinal manifestations and typical colonoscopic findings.

Interventions: Steroid and methotrexate were administered.

Outcomes: This patient achieved clinical remission and mucosal healing.

Lessons: Coexistence of intestinal BD and UC is uncommon, and the combination with steroid, methotrexate, and 5-aminosalicylic acids is an effective therapy.

Abbreviations: 5-ASA = 5-aminosalicylic acids, BD = Behçet disease, CMV = cytomegalovirus, CRP = C-reactive protein, EBV = Epstein-Barr virus, ESR = erythrocyte sedimentation rate, IBD = inflammatory bowel disease, intestinal BD = intestinal Behçet disease, ISG = International Study Group, UC = ulcerative colitis.

Keywords: intestinal Behçet disease, treatment, ulcerative colitis

1. Introduction

Behçet's disease (BD) is regarded as a chronic relapsing disease with multiple organ system involvement characterized clinically by oral and genital aphthae, cutaneous lesions, and ophthalmological, neurological, and/or gastrointestinal manifestations.[1,2] Although intestinal lesions associated with BD may cause serious complications, such as perforation, and decreased quality of life, the diagnosis, and management of intestinal BD lesions has not been standardized. According to the Korean novel diagnostic criteria for intestinal BD, systemic BD patients with typical ileocecal ulcers, a few giant oval-shaped deep punched-out ulcer in the ileocecal area, should be diagnosed as having "definite intestinal BD." Coexistence is uncommon between inflammatory bowel disease (IBD) and intestinal Behçet disease (intestinal BD) in spite of the similarities among their clinical features.[3] Behçet disease with gastrointestinal involvement in ulcerative colitis (UC) patient has been reported in just 1 previous case report, but, which can not be diagnosed as definite intestinal BD based on Korean novel diagnositic criteria due to lacking the typical ileocecal ulcer. Here, we present a patient diagnosed with "definite intestinal BD" during clinical follow-up for UC. Informed consent was obtained from the patient.

2. Case report

A 23-year-old woman was admitted to our gastroenterology outpatient clinic with abdominal pain and bloody stools 10 to 20 times per day. She had a history of UC for 6 years and was treated with oral and topical 5-aminosalicylic acids (5-ASA) (colonosopy for the last relapse and remission are shown in Fig. 1). Two years after the 5-ASA discontinue by herself, the patient was hospitalized with the diagnosis of UC reactivation. On physical examination at admission, her temperature was 38.0 °C. Abdomen tenderness at the left lower quadrant was detected

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ZZ and XS have contributed equally to this work.

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Department of Gastroenterology, The First Affiliated Hospital, Nanchang University, Nanchang, Jiangxi, China.

** Correspondence: Xuan Zhu and Wangdi Liao, Department of Gastroenterology, The First Affiliated Hospital, Nanchang University, Nanchang, Jiangxi, 330006, China (e-mails: jyyfyzx@163.com; 25062224@qq.com).*

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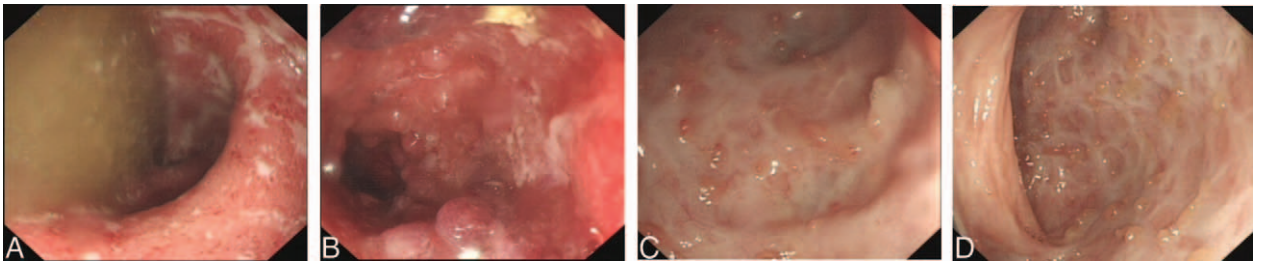

Figure 1. Colonoscopic appearance of last time relapse (A. rectum, B. sigmoid colon) and remission (C. rectum, D. sigmoid colon).

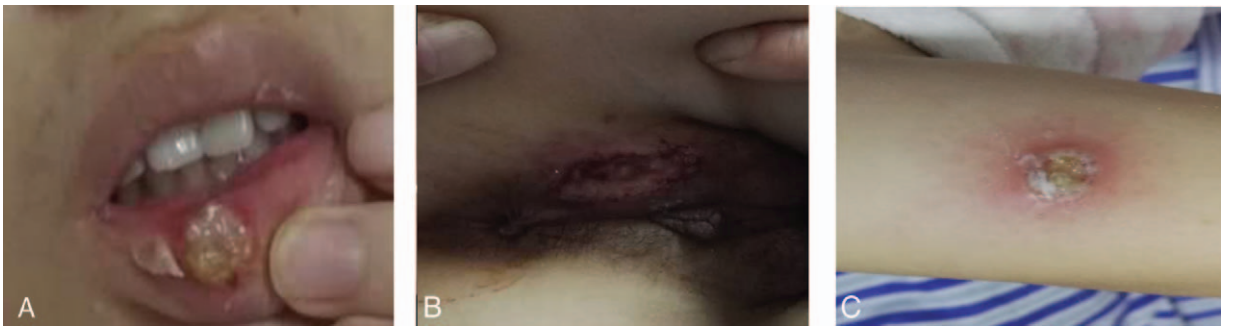

Figure 2. Oral ulcers (A), genital ulcer (B), pyoderma gangrenosum (C).

and she had several mucosal ulcers on his lower lip, one painful genital ulcer around vulva, an pyoderma gangrenosum-like lesion on her left forearm (Fig. 2). Laboratory findings showed as follows: white blood cell count: 8.77/mm^3, hemoglobin: 9.2 g/dL, platelet count 485,000/mm^3, and normal blood urea nitrogen, creatinine, glucose, and liver function tests. Elevations in C-reactive protein (CRP, 14.60 mg/dL) and erythrocyte sedimentation rate (ESR, 34 mm/h) were observed; no infectious pathogens was discovered in the stool sample. CMV-DNA and EBV-DNA were negative. Subsequently, the patient was treated with intravenous methylprednisolone (60 mg/d) and 5-ASA (Etiasa) 3000 mg/d. Then the abdominal pain and bloody stools were improved 1 day after steroid injection. One week later, bloody stools was decreased to 2 to 4 times per day without abdominal pain and the temperature returned to normal, CRP (14.60 mg/dL) and ESR (34 mm/h) were decreased to 5.6 mg/dL and 24 mm/h, respectively. Then, Colonoscopy was performed and revealed continuous inflammation and deep ulcers from rectum to transverse colon with clear demarcation, and mucosa between transverse colon to ileocecum was normal except 2 well-demarcated, large, deep ulcers with exudate at the ileocecum (Fig. 3), 1 ulcer is at the ascending colon near the ileocecum valve and another is opposite. Colonic biopsies at the ileocecum showed ulcer with a necroinflammatory exudate (Fig. 4). Finally she was diagnosed as UC with intestinal BD according to the clinical symptoms and examination. Therefore, methotrexate (20 mg/wk) was added to treat intestinal BD and steroid was slowly tapered for 3 months. A follow-up colonoscopy after 4 months demonstrated that the ulcers at the ileocecum were replaced by scar and normal mucosa (Fig. 5) with complete mucosal healing. Oral ulcers, genital ulcer, and a pyoderma gangrenosum-like lesion were replaced by scars (Fig. 6).

3. Discussion

Intestinal BD and IBD share a lot of characteristics, including genetic background, clinical manifestations, and therapeutic strategies, especially the extraintestinal manifestations, such as

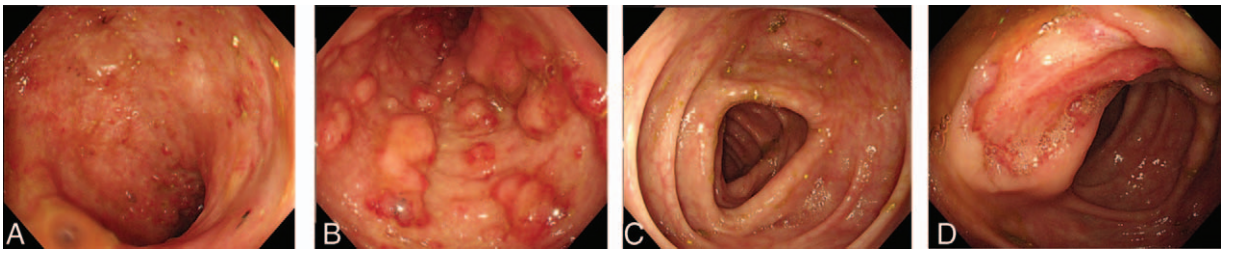

Figure 3. Colonoscopy on admission shows continuous inflammation and deep ulcers from rectum to transverse colon with clear demarcation (A. rectum, B. sigmoid colon), and mucosa between transverse colon to ileocecum was normal (C. transverse colon), except 2 well-demarcated, large, deep ulcers with exudate at the ileocecum (D. ileocecum).

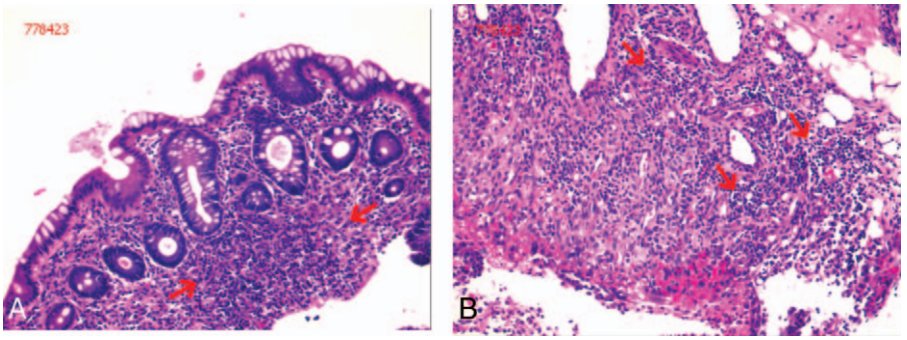

Figure 4. Biopsy from the ileocecum showing ulcers and marked inflammatory cell infiltration, consisted of neutrophils and lymphocytes.

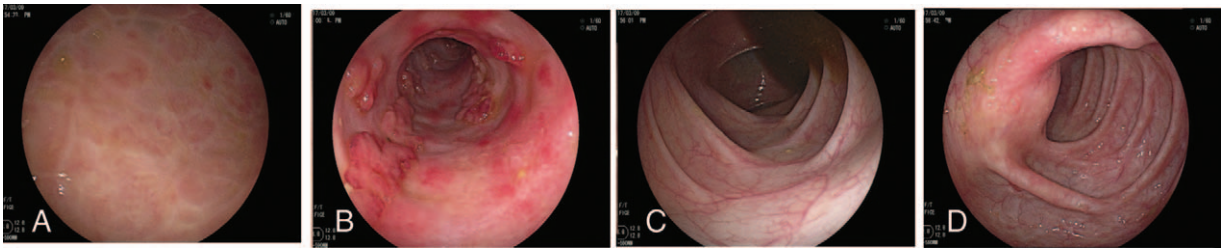

Figure 5. On follow-up colonoscopy at 4 months, continuous inflammation and deep ulcers was replaced by normal mucosa, except mucosa erythema, and inflammatory polyp at the sigmoid colon.

oral ulcers, arthralgia, eye lesions, skin lesions, etc., but the coexistence of these 2 diseases is uncommon.[1] We present a 23-year-old woman with ulcerative disease who developed typical intestinal BD, which is the first case report of patient with coexisting UC and typical intestinal BD.

BD is a multisystemic inflammatory disorder of an unknown etiology and shows a chronic recurrent clinical course. Several diagnostic criteria for BD have been proposed. International Study Group (ISG) is the widely used diagnostic criteria for Behçet disease including recurrent oral ulcer plus at least 2 of the following 4 factors—recurrent genital ulcers, eye lesions, skin lesions, and positive pathergy test.

Intestinal BD occurs in 3% to 60% of patients with BD.[3–5] According to the Korean novel diagnostic criteria for intestinal BD, a few giant oval-shaped deep punched-out ulcer in the ileocecal area is the typical gastrointestinal lesion. These ulcers are characterized by nonspecific inflammation and edematous swelling with craters around the ulcer margin. However, the ulcer shape and distribution pattern including frequent right colonic predominance, discontinuous involvement, and often rectal sparing, is clearly different from that of ulcerative colitis.[6,7] In our case, the patient have a history of UC with typical clinical manifestation, colonoscopic and histopathologic findings, and the treatment with 5-ASA is effective. So, we think that the diagnosis of UC is definite. Two years after the 5-ASA discontinue by herself, the patient was hospitalized for UC reactivation with abdominal pain and bloody stools 10 to 20 times per day. We routinely add intravenous methylprednisolone (40 mg/d) on the basis of mesalazine for severe UC patients, if the effect is not good, the dose will be increased to 60 mg/d. At the same time, the

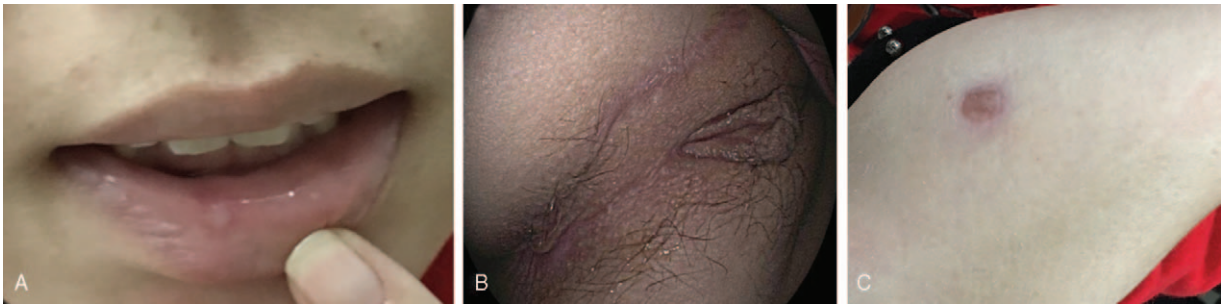

Figure 6. Oral ulcers (A), genital ulcer (B), pyoderma gangrenosum (C) were replaced by scar at 4 months.

patient developed extraintestinal manifestations, such as oral ulcers, genital ulcer, and pyoderma gangrenosum, and the colonoscopy shows 2 giant oval-shaped deep punched-out ulcer in the ileocecal area. Genital ulcer is common in intestinal BD, but rare in IBD, especially in UC.[2] Due to the different therapy between intestinal BD and UC, we should determine whether the patient coexisting with intestinal BD and UC. According to the history of UC, typical intestinal ulcer, genital ulcer, etc., we consider that the patient have both intestinal BD and UC based on the ISD criteria and Korean criteria. Additionally, CMV colitis mimicks an acute exacerbation of UC, so we excluded Cytomegalovirus infection by CMV-DNA detection through PCR and histopathology. Due to coexistence of the 2 diseases, the therapy should cover them. Disease activity index for intestinal BD (DAIBD) of this patient scoring 125 indicating a severe intestinal BD case. As we know, 5-ASA can be primarily administered in mild disease of intestinal BD and younger age (<35 years), higher CRP level, and higher disease activity were associated with a poor response to 5-ASA, whereas corticosteroids and/or immunomodulators can be administered in moderate-to severe cases.[8] Due to the coexistence of UC, we added methylprednisolone and methotrexate to treat this patient on the basis of the 5-ASA. This is the first case that we have treated patient using this combination and obtained a remarkable therapeutic efficacy eventually.

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