

CASE REPORT

Intestinal Behcet's disease with esophageal ulcers and colonic longitudinal ulcers

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Abstract

Intestinal Behcet's disease in a 38-year-old woman was diagnosed because of the history of recurrent oral aphthous ulcers, erythema nodosum-like eruptions, genital ulcer, and endoscopic findings of esophageal and ileocolonic punched-out ulcers with colonic longitudinal ulcers. Esophageal lesions and colonic longitudinal ulcers are rarely seen in intestinal Behcet's disease. The ulcers of esophagus and ileocolon healed with 3 wk of treatment with prednisolone and mesalazine without any adverse effect. Mesalazine may decrease the total dose of prednisolone required to treat the disease.

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Key words: Colonic longitudinal ulcer; Esophageal ulcer; Intestinal Behcet's disease; Mesalazine

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INTRODUCTION

In Japan, Behcet's disease, when accompanied by intestinal involvement, is called intestinal Behcet's disease, which primarily affects the terminal ileum, caecum, or ascending colon^[1]. However, esophageal aphthous ulcerations and colonic longitudinal ulcers are rare in intestinal Behcet's

disease. Intestinal lesions in Crohn's disease tend to be longitudinal ulcers with a cobblestone appearance, while those in Behcet's disease are round and oval "punched-out" ulcers. Moreover, epithelioid granuloma is one of the pathological characteristics of Crohn's disease, whereas it is uncommon in intestinal Behcet's disease. Another feature of Behcet's colitis is lymphocyte venulitis, which is a disorder of vasculitis. Despite these differences, it can be difficult on occasions to make a differential diagnosis between these two diseases. We present a case of intestinal Behcet's disease with esophageal and colonic longitudinal ulcers.

CASE REPORT

The patient was a 38-year-old woman who was admitted to our department in December 2003 for epigastralgia and melena that occurred after transient fever. For the previous two days, she was in good health, except for numerous recurrences of aphthous stomatitis. Physical examinations on admission revealed the presence of multiple oral aphthous ulcers, in addition to slight tenderness in the epigastrium. Ophthalmologic and neurologic examinations showed no remarkable findings despite the presence of erythema nodosum-like eruptions in the bilateral inferior limbs and genital ulcers. Negative results were obtained by a prick test of the skin. Clinical laboratory tests showed the following: white blood cells, 7830/ μ L; red blood cells, 360 $\times 10^4$ / μ L; hemoglobin, 9.6 g/dL; hematocrit, 29.7%; platelets, 61 $\times 10^4$ / μ L; C-reactive protein, 10.9 mg/dL; total protein, 8.6 g/dL; AST, 36 IU/L; ALT, 64 IU/L; LDH, 171 IU/L; and total bilirubin, 0.2 mg/dL. Human lymphocyte antigens were negative for B51, but positive for B52. All bacteriologic examinations of blood, urine, and cultured stool showed negative results. Upper gastrointestinal endoscopy demonstrated small oval and discrete ulcers with reddish margin scattered between the middle and lower parts of the esophagus (Figure 1A). Colonoscopy revealed multiple aphthas and erosions scattered in the terminal ileum (Figure 1B) as well as small aphthas on the ileocecal valve (data not shown). However, punched-out ulcers were not observed on ileocecal lesions. Multiple erosions and ulcers including longitudinal irregular-outlined ulcers were extant in the ascending and transverse colon (Figure 1C). Pathological examination of the endoscopic biopsy specimen showed nonspecific ulceration and no evidence of Crohn's disease could be found.



Figure 1 Endoscopic view of discrete ulcers with reddish margin in the middle part of the esophagus (A), multiple aphthas and erosions scattered in the terminal ileum (B), and multiple erosions and ulcers including longitudinal irregular-outlined ulcers extant in the transverse colon (C).

Table 1 Reported cases of Behcet's disease with colonic longitudinal ulcers

n	Age	Gender	Symptoms				Type	Colonic longitudinal ulcer		Other ulcers	Treatment	Outcome
			Oral	Eye	Skin	Genital		Location	Histology			
1	29	Female	+	+	+	+	Complete	Transverse Descending	Vasculitis		Operation Steroid	Healing
2	70	Female	+		+	+	Incomplete	Ascending Transverse Descending	Nonspecific colitis	Ileum	Colchicine Salazopyrin	Died
3	37	Female	+		+	+	Incomplete	Ascending Transverse	Granuloma	Ileum	Steroid 5-Aminosalicylate	Healing
4 ¹	39	Female	+		+	+	Incomplete	Ascending Transverse	Nonspecific colitis	Ileum	Healing Mesalazine	Healing

¹Case reported here.

In this patient, ulcers were detected in the esophagus, terminal ileum, and ascending and transverse colon, in addition to erythema nodosum-like eruptions in the bilateral inferior limbs and genital ulcers. Based on these findings and the history of recurrent oral aphthous ulcers, the patient was diagnosed as having incomplete type intestinal Behcet's disease.^[1] The treatment for this patient was initiated by oral administration of prednisolone (50 g) and mesalazine (1 500 mg daily). Consequently, her symptoms disappeared rapidly, and the results of all clinical laboratory tests were normalized. Upper gastrointestinal endoscopy and colonoscopy performed 3 wk after admission revealed the disappearance of ulcers. Mesalazine was maintained at the same dose, but the dose of prednisolone was gradually decreased to 2.5 mg daily, which was maintained thereafter. However, remission was not obtained until 8 mo.

DISCUSSION

Behcet's disease is characterized by repeated eye, skin and visible mucosal lesions. The prevalence of this disease differs widely among races; the rate is 0.3/100 000 in the United States, but 1/10 000 in Japan^[2]. Abdominal complaints also differ among races. Shimizu *et al* reported abdominal pain in 75% of patients with Behcet's disease^[3], as noted in the present patient showing epigastralgia.

In Japan, diagnostic criteria for Behcet's disease have

been established by the Behcet's Disease Research Committee^[1]. Based on these criteria, a diagnosis was made in the present case of incomplete type intestinal Behcet's disease manifests mainly in the terminal ileum, cecum, and ascending colon, although esophageal lesions are rare and colonic longitudinal ulcers are very rare rather than esophageal lesions. To the best of our knowledge, there are only three existing reports describing intestinal Behcet's disease with longitudinal ulcers (Table 1)^[4-6]. Although Lee presumed the cause of the longitudinal ulcers to be multifocal vasculitis, in our case vasculitis was not significant. However, we cannot conclude that the patient had Crohn's disease, because microscopic characteristics of Crohn's disease - that is, chronic inflammation involving all layers of intestinal wall or granulomas - were absent.

In addition, only nine cases including our case of intestinal Behcet's disease with both esophageal and ileocolonic ulcers were reported in English literature^[7-13]. One case was of the complete type; 6, the incomplete type; and 2, the suspected type. Three patients had strictures; and 2 had perforation. The treatment of intestinal Behcet's disease is controversial. Surgical treatment was not effective in 1 of the 9 patients (Table 2). However, the esophageal ulcers healed in 6 of 7 patients with medical treatment, including corticosteroids or acid suppressive drugs. Corticosteroids, the major therapeutic agent in this disease, were effective in 3 of the 4 patients. However, they can have serious adverse effects, and their use may be

Table 2 Reported cases of Behcet's disease with esophageal and ileocolonic ulcers

n	Age	Gender	Symptoms				Type	Esophageal ulcer		Other ulcers	Treatment	Outcome
			Oral	Eye	Skin	Genital		Number	Complication			
1	52	Female	+	+	+		Incomplete	1	Perforation	Stomach, Duodenum Jejunum, Ileum	?	?
2	21	Female	+			+	Suspected	3		Colon	Antacid Transfer factor	Healing
3	16	Female	+	+		+	Incomplete	1	Perforation	Colon	Steroid	Healing
4	12	Female	+	+	+	+	Complete	1	Stenosis	Ileum	Steroid Operaton	No healing
5	50	Male	+		+	+	Incomplete	1	Stenosis	Ileum, Colon	?	?
6	52	Male	+	+		+	Incomplete	Diffuse	Stenosis	Ileum	Healing	Healing
7	19	Female	+				Suspected	A few		Ileum	Healing Mesalazine	Healing
8	33	Male	+			+	Incomplete	1		Ileum	Healing	Healing
9 ¹	39	Female	+		+	+	Incomplete	8		Ileum, Colon	Healing Mesalazine	Healing

¹Case reported here.

associated with colonic perforation.

Sonta *et al* have suggested that mesalazine is effective for treatment of intestinal Behcet's disease^[13]. It may decrease the total dose of a corticosteroid required to treat the disease. The esophageal and ileocolonic ulcers in the present case healed 3 wk of treatment with prednisolone and mesalazine without any adverse effect. However, it was reported that the recurrence rate even with medical treatment was 90% in patients with Behcet's disease^[14]. The present case has not relapsed for 8 mo with treatment of 2.5 mg prednisolone and 1500 mg mesalazine. Thus, the post-treatment course of this patient should be followed up carefully.

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