INTRODUCTION

Behcet’s disease is a systemic vasculitis characterized by systemic organ involvement. Although the gastrointestinal and systemic features of Behcet’s disease and inflammatory bowel disease overlap to a considerable extent, they are generally viewed as two distinct diseases. A 39-yr-old female was diagnosed as having Behcet’s disease. She was admitted to our hospital because of oral and genital ulcer, lower abdominal pain, and frequent diarrhea. Colonoscopy showed diffuse involvement of multiple longitudinal ulcers with inflammatory pseudopolyps with a cobblestone appearance and ano-rectal fistula was suspected. These findings are extremely rare in Behcet’s disease. However, there were no granulomas, the hallmark of Crohn’s colitis. Microscopically, perivasculitis and multiple lymph follicles compatible with Behcet’s disease were seen. Although being rarely encountered, multiple longitudinal ulcers, cobblestone appearance, and ano-rectal fistula can develop in Behcet’s disease, as in Crohn’s colitis. Therefore, Behcet’s disease and Crohn’s disease may be closely related and part of a spectrum of disease.

CASE REPORT

A 39-yr-old female presented with recurrent oro-genital ulcers, erythema nodosum, and arthralgia. She was diagnosed as having Behcet’s disease. She had taken the maintenance therapy with colchicines. Two years later, she was admitted to our hospital because of oral and genital ulcer, lower abdominal pain, and frequent diarrhea. Colonoscopy showed diffuse involvement of multiple longitudinal ulcers with inflammatory pseudopolyps with a cobblestone appearance and ano-rectal fistula was suspected. These findings are extremely rare in Behcet’s disease. However, there were no granulomas, the hallmark of Crohn’s colitis. Microscopically, perivasculitis and multiple lymph follicles compatible with Behcet’s disease were seen. Although being rarely encountered, multiple longitudinal ulcers, cobblestone appearance, and ano-rectal fistula can develop in Behcet’s disease, as in Crohn’s colitis. Therefore, Behcet’s disease and Crohn’s disease may be closely related and part of a spectrum of disease.
A Case of Intestinal Behcet's Disease Similar to Crohn's Colitis

The external genitalia showed a linear to ovoid shaped ulcerating wound at the perineum and vulva area.

Fig. 1. The external genitalia showed a linear to ovoid shaped ulcerating wound at the perineum and vulva area.

DISCUSSION

Behcet's disease was originally described in 1937 and characterized by oral and genital ulceration and ocular inflammation (6). It is now acknowledged that this disorder has a wide spectrum of clinical manifestation. Although many diagnostic criteria have been established, there is no universally accepted definition. The diagnosis is clinical and is now based on criteria suggested by an international study group for Behcet's disease (7). In the present case, oro-genital ulceration and skin lesions led to the diagnosis of Behcet's disease. The etiology remains unknown, but the most widely held hypothesis of pathogenesis is that a profound inflammatory response is triggered by an infectious agent in a genetically susceptible host (6). Other possible mechanism is that Behcet's disease may be autoimmune in origin (8, 9). An inflammatory response to several antigens is found, and generalized aberrant T cell responses results in enhanced nonspecific inflammation. An increased production of interferon gamma (IFN-γ) by T cells has been demonstrated in active Behcet's disease, and circulating T cells have the T-helper phenotype (Th1) predominantly (10).

Oshima et al. (12) reported that over 40% of Behcet's disease patients had gastrointestinal complaints. Symptoms included abdominal pain, diarrhea, nausea, anorexia, and abdominal distension. Although gastrointestinal symptoms are common, the demonstration of gastrointestinal ulcer is rare. This so-called intestinal Behcet's disease accounts for only 1-2% of cases (1, 2). In intestinal Behcet's disease, ulceration of the gastrointestinal tract can be found throughout the intestine, but the most frequent area of involvement is the ileocecal region. Only 15% of intestinal Behcet's disease diffusely involve the colon (13). Behcet's colitis can appear as ulcerative colitis or Crohn's disease when there are skip lesions with rectal sparing. The ulcers are usually large, discrete, punch-out appearing lesions and can extend to the serosa. Formation of fistula, hemorrhage, or perforation occurs in up to 50% of cases involving the intestine. The ulcers are found within normal or minimally inflamed mucosa. Microscopic examination reveals vasculitis involving small and medium sized vessels. The lymphocytes are infiltrated, and dense perivascular infiltrate is frequently seen (3, 4).

There are many extra-intestinal findings of Crohn's disease, such as oral and genital ulcers, erythema nodosum, uveitis and arthritis, resembling the manifestations of Behcet's disease. It is also very difficult to distinguish the intestinal Behcet's disease from that of Crohn's disease in some patients. It is possible that a patient with Crohn's disease meets the criteria for Behcet's disease. There are several reports on the coexistence of Behcet's disease and Crohn's disease (14, 15). In the present case, we made the diagnosis of Behcet's disease as described above. However, the gastrointestinal mani-
The findings in the family reported by Yim and White (16), suggest that inflammatory bowel disease and Behcet's disease may be closely related and part of a spectrum of disease rather than distinct disease entities. In our patient, clinical and pathological findings are characteristic of intestinal Behcet's disease. Patient's bowel symptoms, endoscopic appearance, and the response to medical treatment were compatible with Crohn's colitis. These findings suggest that Behcet's disease may be a part of the spectrum of chronic inflammatory bowel disease.

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Fig. 3. Microscopic examination from ulcers of the colon showed shallow ulcerations with inflammatory infiltration consisting of lymphocytes and plasma cells. There were no granulomas (A: H&E stain, ×100; B: H&E stain, ×400).

Fig. 4. Microscopic examination of the perineal lesion revealed chronic ulcer with acute and chronic inflammatory cell infiltration and increased small blood vessels (A: H&E stain, ×100; B: H&E stain, ×200).
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