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# Intestinal Behçet's Disease in a Child

## – A Case Report –

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***Behçet disease is relatively rare in pediatric age group. And the bowel involvement is seen in only a small portion of Behçet disease. However, once the bowel is involved it is potentially life threatening event. We report a 15 year old boy with intestinal Behçet's disease who had a history of recurrent oral and genital ulcers for several years. He underwent right hemicolectomy under the impression of intestinal tumor. Pathologically the lesion was a large sharply delineated ulcer in the cecum. The ulcer was round and deep with elevating margin, and was associated with thickening of affected intestinal wall. Microscopically, the ulcer base consisted of granulation tissue with fissurings and underminings. Characteristic phlebitis and occlusive arterial lesion were seen in intestinal wall. The inflammatory lesion was most pronounced around the ulcer but could be recognizable throughout the resected specimen.***

**Key Words:** Behçet disease, entero-Behçet disease, intestinal Behçet disease.

## INTRODUCTION

Behçet disease is a disease of increasing interest as the etiology and pathogenesis are still unknown. This disease is known to be prevalent in Japan as well as mid-east and mediterranean countries. In Korea, although it's presence was known for a long time the exact figures on the incidence are not available (Kim et al., 1962, Kim et al, 1981, Rhim et al., 1980, Choi et al., 1983). Although over hundred cases of Behçet disease were reported (Eun et al., 1984), documented cases of entero-Behçet are very rare, particularly in pediatric age group. There is only one previous case of entero-Behçet in childhood in Korean literature (Ko et al, 1982).

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This report presents a description of a 15 year old patient who had intestinal Behçet disease that was diagnosed by pathological examination of resected bowel.

## CASE REPORT

This 14 10/12 year old boy was admitted to Department of Pediatric Surgery because of a palpable mass in the right lower quadrant. He had suffered from fatigue, anorexia, excessive night sweating and nervousness for one year. Eight months prior to this admission he developed intermittent vomiting, for which he underwent an upper gastrointestinal series at other hospital. It revealed no abnormal findings. This time he came to Seoul Natonal University Children's Hospital for abdominal pain and mass. After surgery he was found to have recurrent oral ulcer since his time of elementary school days and also genital ulcer for the last one year. A few days prior to admission knee joint pain developed in the left side. Physical examination on admission

showed a 5 x 7.5cm sized mass in the right lower abdomen, which was round, movable and firm. The mass was tender. Laboratory findings remained within normal limits except for occult blood in the stool and increased level of C-reactive protein. He underwent a right hemicolectomy under the impression of intestinal tumor.

Pathology: Resected specimen consisted of terminal ileum, cecum and ascending colon. Opening the lumen there was a sharply delineated ulcer, 3 x 3cm, in the cecum, partly involving ileocecal valve. The ulcer was round and deeply excavated reach-

Microscopically the ulcer consisted of necrosis resting on granulation tissue, with areas of undering muscularis externa, associated with marginal elevation and thickening of intestinal wall. Ulcer base was rather smooth and fibrotic. Submucosal edema around the ulcer extended toward distal ileum and ascending colon (Fig. 1, 2).

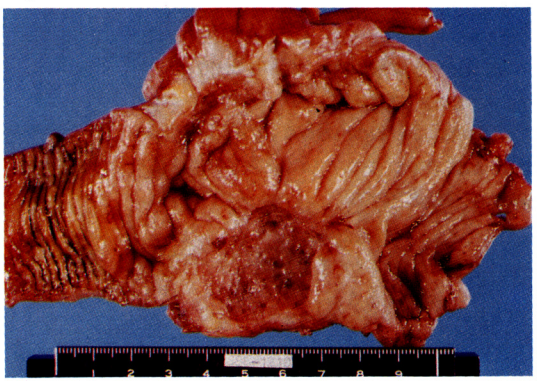

Fig. 1. Right hemicoletomy specimen showing a sharply delineated round ulcer and marked submucosal edema in the cecum.

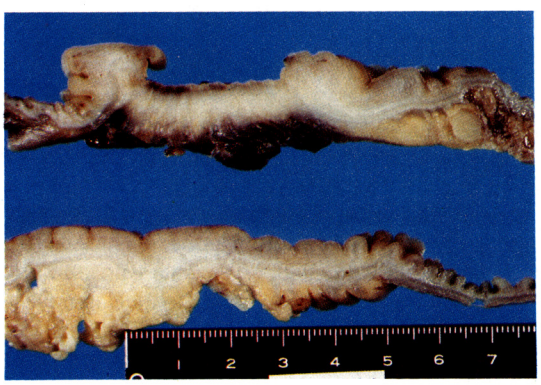

Fig. 2. The ulcer is deeply excavated reaching muscularis externa, associated with marginal elevation of mucular layer and thickening of intestinal wall (upper). Note a marked submucosal edema around the ulcer (lower).

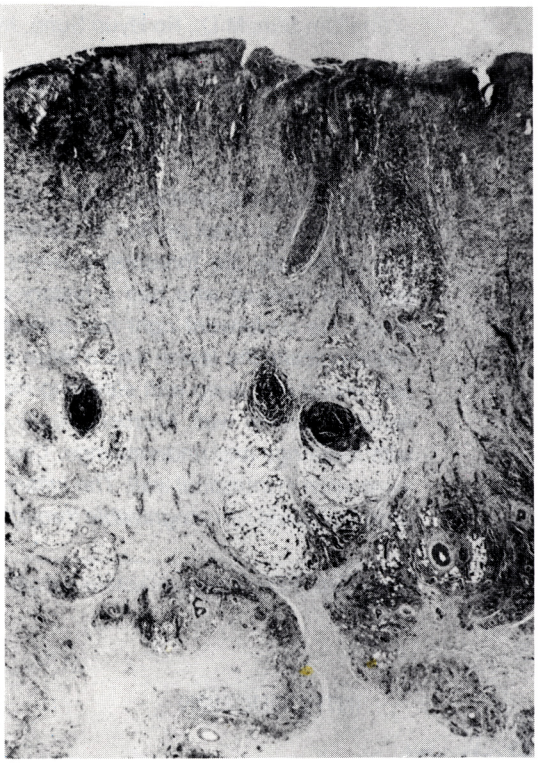

Fig. 3. The ulcer base showing transmural involvement of nonspecific chronic inflammation, fibrosis and scattered lymphoid follicles (H & E, x10).

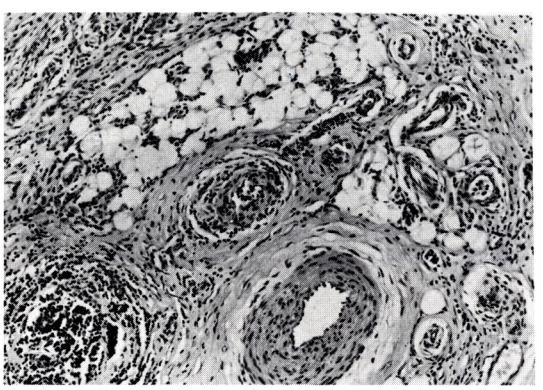

Fig. 4. Note the phlebits, lymphocytic infiltration around capillaries and venules, and an artery with intimal thickening in the serosa of colon (H & E, x 40).

mining and fissuring. The ulcer base showed a nonspecific chronic inflammatory infiltration with lymphocytes, and a few eosinophils and plasma cells were seen in the fibrotic intestinal wall, penetrating into the subserosal fat in some areas. Nodular collections of lymphoid cells simulating lymphoid follicle were also scattered throughout the thickened intestinal wall (Fig. 3). There were prominent vascular changes throughout the entire thickness of bowel wall in the ulcer base and the surrounding areas of the ulcer. The vascular changes were characterized by lymphocytic infiltration in and around the slightly fibrotic vascular wall. It was more pronounced in the venules and capillaries in the serosa. Arteries and arterioles were unaffected although small arteries were often found stenotic or occluded due to subintimal fibrosis (Fig. 4). There was no evidence of fibrinoid necrosis in the vascular wall. Submucosal edema was prominent, associated with dilatation of lymphatic vessels.

## DISCUSSION

Behçet's syndrome is initially described only as a peculiar triple symptom complex of recurrent oral and genital ulcerations and relapsing iritis (Behçet, 1940). But it is soon recognized as a systemic disease of unknown etiology. The clinical features of Behçet's disease have been well characterized, and various diagnostic criteria have been formulated. However, the pathologic features of the Behçet's disease has been fragmentarily described. About one half of the patients presents some of gastrointestinal symptoms (Shimizu et al., 1975). Behçet's disease associated with ulcerative change of intestine is called as intestinal or entero-Behçet's disease in Japan (Baba et al., 1976, Tsukada et al., 1964). It was first reported by Bøe et al. in 1958. Recently, the number of case reports has increased in Japan (Seki et al, 1972, Shimizu et al., 1968, Kasahara et al., 1981). Pathologic features were described in five resected cases of adult intestinal Behçet's disease in Korea (Kim et al., 1985). One of us (JGC) has reported a case of Behçet's disease with skin and intestinal involvement in a 14 year old girl who had had multiple jejunal perforation due to Behçet disease (Ko et al., 1982). Fukuda reported nine resected cases of intestinal Behçet's disease and divided the intestinal ulcers into three types (Fukuda et al., 1979); necrotic, granulomatous and combined. The ulcerating region of necrotic type is characterized by necrosis as the main change, without elevating muscular layer or thickening of intestinal wall. The granulomatous type is composed of granulation tissue and shows obvious elevating muscular layer and thickening of intestinal wall. The ulcerating lesion of combined type shows characteristic findings of both necrotic and granulomatous types. Intestinal ulcer of this case fits for combined type according to the above histologic classification.

Intestinal Behçet's disease occurring in clinically incomplete type of Behçet's disease is very difficult to differentiate from Crohn's disease associated with oral and genital uclers. Morphological features observed in this case can be compared with those of Crohn's disease described in textbook (Robbins et al., 1984). Important features that help to diff-disease are; (1) the ulcers of intestinal Behçet's disease are more round to geographic and deeply excavated with sharp demarcation and marginal elevation, (2) the ulcers locate in the ileo-cecum, more on cecal side and at the opposite side of mesentery, (3) there is no skip lesion or cobble stone mucosa as in Crohn's disease, (4) histologically, vasculitis or perivascular lymphocytic infiltration is observed, and (5) neither sarcoid-like granuloma is present nor the fissure extends significantly into the muscular layer.

Clinically or radiologically this case was thought to be intestinal lymphoma before its removal. Because the patient was presented as an abdominal mass and radiologically the mass appeared to be intramural and partly extrinsic in nature. Considering the facts that abdominal pain is the most common symptom of intestinal Behçet's disease and that the high incidence in the fourth and fifth decades of life, this case is rather exceptional. Although intestinal Behçet's disease is reltively rare entity particularly in childhood, it still needs to be considered as a differential diagnosis of intestinal ulcer in children.

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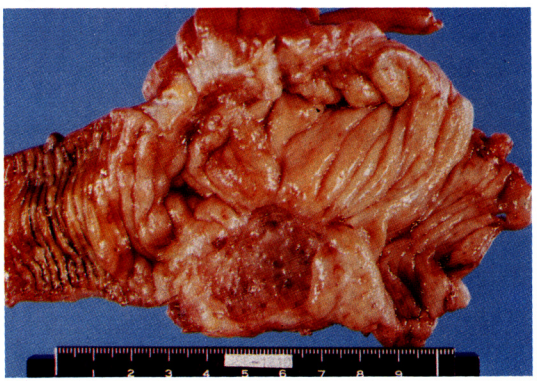

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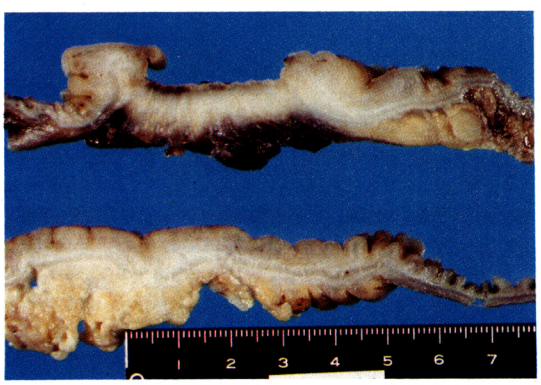

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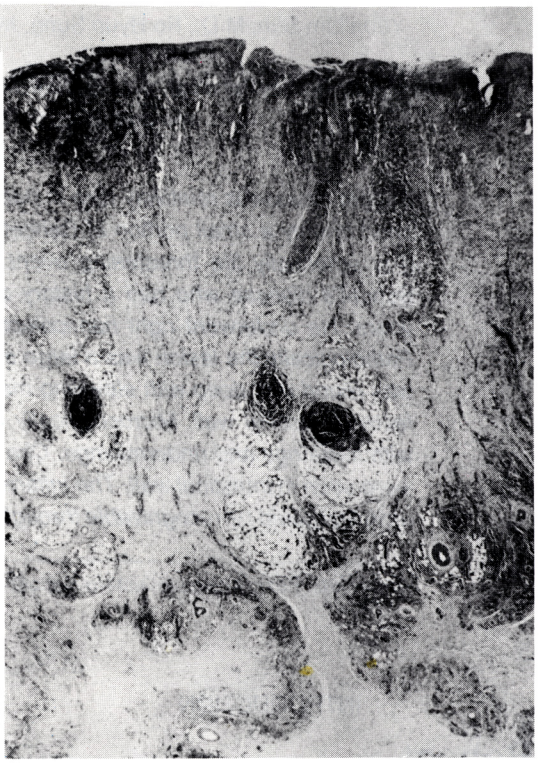

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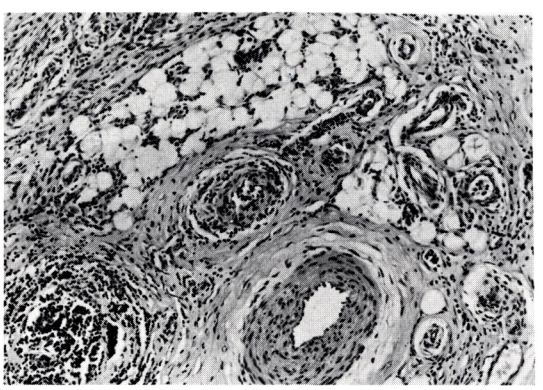

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