Journal of Korean Medical Science
Vol. 2, No. 2, P 133–136. June 1987

Crohn's Disease

– A Case Report –

Kyung Ja Cho, M.D., Yeon Lim Suh, M.D., Chul Woo Kim, M.D. and Je G. Chi, M.D.

Department of Pathology, Seoul National University Children's Hospital and College of Medicine, Seoul National University

A case of rather typical Crohn's disease in a 10 year old girl is described. She had suffered from intractable abdominal pain, diarrhea and fever for 1 year. Eventual right hemicolectomy revealed diffuse involvement of terminal ileum, cecum and ascending colon by confluent ulcerations and transmural inflammation. Histologically there were numerous well developed non-caseating granulomas scattered transmurally and in regional lymph nodes. Deep penetrating ulcerations were characteristic. Acid fast staining failed to demonstrate any organism. The rarity of Crohn's disease in Korea and this occurrence in pediatric age prompted this report.

Key Words: *Crohn's disease, granulomatous colitis, regional enteritis.*

INTRODUCTION

The rarity of Crohn's disease and prevalence of tuberculous enterocolitis in Korea tend to unnecessarily make it hesitate to diagnose a Crohn's disease, both clinically and histologically. Its differentiation from ulcerative colitis is always a problem, but the distinction from tuberculosis is rather more difficult in Korea, if the lesion is granulomatous. The presenting case was clinically misdiagnosed as tuberculosis and ineffective medication was continued for 1 year. Surgically resected intestine again required a close examination for various diagnostic criteria to prove a Crohn's disease.

CASE REPORT

A 10 year old girl first presented with diarrhea, abdominal pain, vomiting and fever in July 1985. On admission laboratory tests revealed strongly positive C-reative protein, high ASO titer and occult blood in stool. Colonoscopy and colon barium study revealed ulcerative mucosal lesions in ileocecal area and ascending colon, but a biopsy showed nonspecific inflammation. Tuberculin test was negative. But it was interpreted as false negative because she had chickenpox two weeks prior to admission. Anti-tuberculosis medication was started. She appeared slightly improved for a while, but the symptoms such as diarrhea, abdominal pain and fever gradually aggravated, requiring her second admission in October, 1985. Colonoscopy was repeated, and showed inflammatory bowel disease, but the biopsy result was again nonspecific. Anti-tuberculosis medication was continued for another several months. In January 1986, prednisolone therapy was tried followed by marked improvement. After then she was dependent on steroid for months, making it impossible to taper the dosage due to prompt relapse. This intractability eventually necessitated an exploration in July 1986, and she underwent a right hemicolectomy in the end of the long illness.

This paper was presented at the monthly meeting of the Korean Society of Pathologists in September, 1986.
Address for Correspondence: *Je G. Chi, M.C., Department of Pathology, College of Medicine, Seoul National University Children's Hospital, 28 Yunkun-Dong, Chongno-ku, Seoul 110, Korea (02) 7601-3540, 3546.*

Grossly the resected segment of terminal ileum, cecum and ascending colon showed congested and swollen mucosa intersected by diffuse irregular ulcerations, making a cobblestone appearance (Fig. 1). The ulcers were focally serpiginous and intervening normal mucosa formed many pseudopolyps (Fig. 2). Grossly visible fissure, fistula or stenosis were not identifiable and there was an apparently unaffected area separated from the main lesion. The cut surfaces revealed diffuse transmural involvement with mild mural thickening and prominent fat creeping.

Microscopically the lesions were of diffuse chronic inflammation with transmural and focally mucosal or submucosal involvement. Edema and fibrosis of submucosa were also present in diffuse

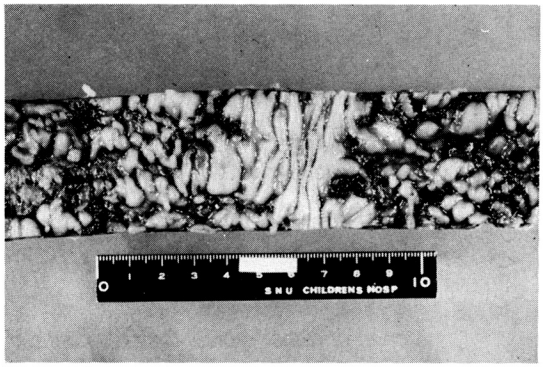

Fig. 1. Resected colonic segment showing multiple ulcerated lesions with a segment of uninvolved mucosa.

Table 1. The main gross and microscopic characteristics of inflammatory bowel diseases and the present case. (Referring to Kent et al., 1970, Lee et al., 1979, and Tandon et al., 1972)

	Crohn's disease	Chronic ulcerative colitis	Indeterminate colitis	Tuberculous enterocolitis	Present Case
GROSS					
Site	Ileum 70% Ileum & colon 15% Colon 15%	Rectosigmoid 100% Entire colon 40% Ileum 10%	Any level of colon Rectum 50%	Ileocecal area Lower ileum Cecum	Distal ileum, cecum, appendix & ascending colon
Ulcer	Longitudinal Serpiginous	Irregular Broad-based Coalescent	Irregular	Transverse Annular	Irregular Serpiginous, focally
Fissure	Common	Rare	Common (superficial)	Rare	Present
Fistulation	Common	Extremely rare	Rare	Rare	Absent
Skip area	Common	Absent	Common	Common	Present (?)
Wall thickening	Common	Rare	Minimal	Common	Minimal
Fat creeping	Common	Rare	Minimal	Common	Prominent
Pseudopolyp	Rare	Common	Common	Minimal	Present
MICRO					
Thickness of involvement	Transmural	Mucosal and submucosal	Transmural	Transmural	Mucosal and submucosal to transmural
Granuloma	Common	Absent	Absent	Multiple with caseation	Present
Lymphoid aggregate	Common	Rare	Rare	Common	Prominent
Lymphangiectasia	Common	Rare	Rare	Common	Prominent
Penetrating ulcer	Common	Rare	Common	Absent	Present
Edema and fibrosis	Marked	Minimal	Moderate	Moderate	Present
Crypt abscess	Rare	Common	Rare	Common	Present
Neutrophilic infiltrate	Rare	Common	Rare	Common	Minimal

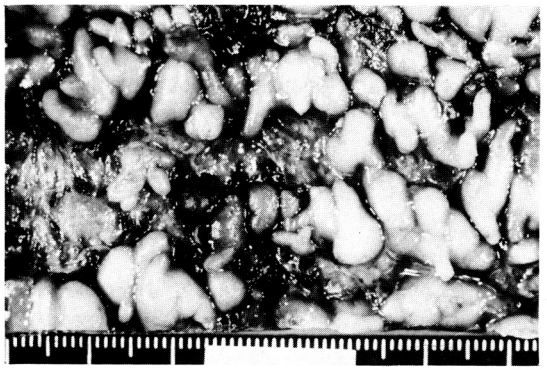

Fig. 2. Close up of Fig 1, showing cobblestone appearance with multiple pseudopolyps.

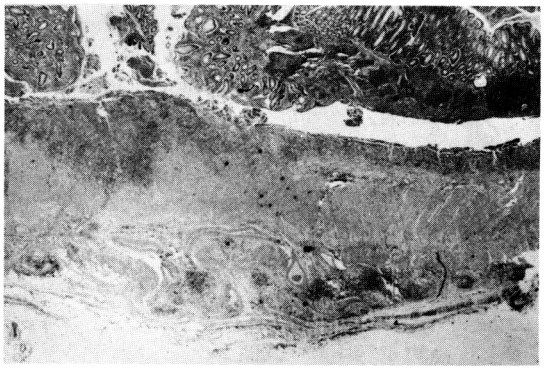

Fig. 3. Photomicrograph showing transmural involvement and a deeply penetrating fissure. H&E x 40.

fashion. The ulcers were well demarcated and characteristically showed fissuring and cracking at the bases which comprised of active granulation tissue and lymphoplasma cell infiltrate (Fig. 3). The adjacent nonulcerated areas showed multiple lymphoid aggregates, lymphangiectasia, telangiectasia and edema in submucosa, often featuring pseudopolyps (Fig. 4). The most striking finding was many noncaseating granulomas scattered transmurally. They were observed at the ulcer base or discretely within the submucosa and muscle coat, and in the subserosa as well (Fig. 5). They were composed of epithelioid histiocytes and Langhans giant cells, surrounded by rim of lymphocytes, and showed no necrosis. Similar granulomas were also found in the subcapsular portion of two pericolic lymph nodes. Carbol-Fuschin and Auramine-Rhodamine stainings for acid fast bacilli revealed no organism.

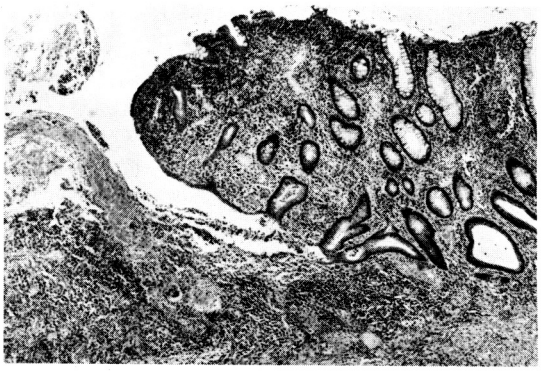

Fig. 4. Microscopic picture showing pseudopolyp and an ulcer resting on granulation tissue base. H&E x 100.

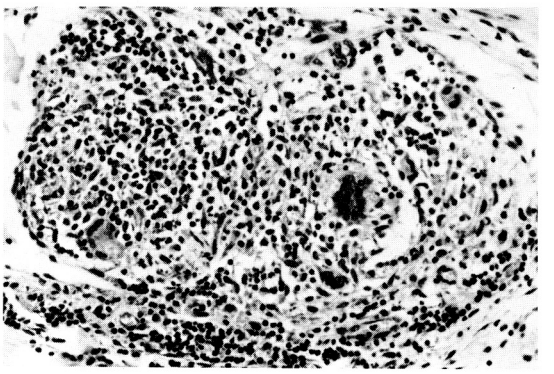

Fig. 5. Photomicrograph of a subserosal epithelioid cell granuloma. H&E x 200.

DISCUSSION

Although there have been a few reports on clinical cases of Crohn's disease in past (Bae et al, 1975; Kim et al, 1964), definite cases of Crohn's disease that were thoroughly examined pathologically and satisfied all the criteria in making diagnosis are extremely rare in Korea. At Seoul Nat'l University Hospital we had 3 cases of Crohn's disease for the last 25 years. One of three cases was reported and it was granulomatous colitis in a 17 year old female (Moon et al, 1979). Its incidence in childhood is not conclusively estimated and vary in various reports, but it is now thought that most of the disease have their onset in the second to third decade of life and are common between the ages of

ulcer or fistulation seen in Crohn's disease is generally absent (Tandon et al, 1972; Suh and Kim, 1986). These gross differences, and microscopic

absence of caseation necrosis and acid-fast bacilli in the granulomas in both bowel and lymph nodes give sufficient clues to exclude tuberculosis. Clinical intractability to anti-tuberculous therapy and good response to steroid therapy in this patient also support the diagnosis of Crohn's disease.

Emerging cases of Crohn's disease in Korea are worth paying attention, and we need more serious consideration of Crohn's disease as a differential diagnosis of chronic inflammatory bowel disease, particularly in young age.

10 and 15 years (Rubin et al, 1967).

The diagnosis of Crohn's disease is always harboring difficulties in differentiation from other inflammatory bowel disease. The gross and microscopic characteristics of various inflammatory bowel diseases and those of the present case are summarized in Table 1.

When large intestinal involvement is extensive and granuloma is deficient, it must be differentiated from ulcerative colitis. The gross appearance of the present case was more close to ulcerative colitis, showing diffuse colonic involvement, irregular diffuse ulcerations without definite skip area, minimal mural thickening and many pseudopolyps, but definite transmural involvement with fat creeping and fissure formation favored Crohn's disease. Numerous well developed epithelioid granulomas seen on microscopic examination helped us to exclude ulcerative colitis and indeterminate form (Kent et al, 1970; Price, 1978; Lee, 1979), but required a differentiation from intestiinal tuberculosis which is common in Korea. In tuberculous enteritis, transverse annular ulceration associated with luminal stenosis is characteristic, but deeply penetrating

REFERENCES

Bae SW, Kim CK, Kim KY, Min JS, Kim CK: *Clinicopathological study of Crohn's disease. J Kor Surg Soc 17: 209-218, 1975.*

Kent TH, Ammon RK, DenBesten L: *Differentiation of ulcerative colitis and regional enteritis of colon. Arch Pathol 89: 20-29, 1970.*

Kim KY, Suh SH, Chang MW, Min KS: *Crohn's disease. Case report and article review. J Kor Surg Soc 6: 121-131, 1964.*

Lee KS, Medline A, Shockey S: *Indeterminate colitis in the spectrum of inflammatory bowel disease. Arch Pathol Lab Med 103: 173-176, 1979.*

Moon JH, Yoo IH, Park YH, Koh CS, Kim MS, Lee HS, Chi JG: *Crohn's colitis. A case report. J Kor Surg Soc 21: 971-980, 1979.*

Price AB: *Overlap in the spectrum of non-specific inflammatory bowel disease-'colitis indeterminate'. J Clin Pathol 31: 567-577, 1978.*

Rubin S, Lambie RW, Chapman J: *Regional ileitis in childhood. Am J Dis Child 114: 106-110, 1967.*

Suh YL, Kim YI: *Pathology of tuberculous enterocolitis. Analysis of 10 cases in special reference to macroscopic features. Kor J Pathol 20: 42-48, 1986.*

Tandon HD, Prakash A: *Pathology of intestinal tuberculosis and its distinction from Crohn's disease. Gut 13: 260-269, 1972.*