Journal of Korean Medical Science
Vol 7, No. 1, 76-78 March 1992

Localized Form of Colitis Cystica Profunda

— A Case of Occurrence in the Descending Colon —

Woo Ho Kim, M.D., Ghee Young Choe, M.D., Yong Il Kim, M.D. and Jin Pok Kim*, M.D.

Department of Pathology and Surgery Seoul National University College of Medicine, Seoul, Korea*

An unusual localization of localized colitis cystica profunda in a 31-year-old man is described. The patient presented as anal bleeding and a protruding mass at the descending colon; the mass was polypoid and was made up of papillary epithelial hyperplasia with downward herniation of glands into the submucosa. Only one similar case involving a descending colon has been reported in the world literature.

Key Words: *Colitis cystica profunda, Colitis cystica polyposa, Solitary rectal ulcer, Descending colon*

INTRODUCTION

Colitis cystica profunda (CCP) is a rare colonic pseudoneoplastic disorder characterized by a polypoid colonic mass with epithelial-lined cysts in the submucosa or proper muscle coat. While the diffuse form usually involves several segments of the colon, the localized form tends to be confined to the rectum, and thus its relationship with solitary rectal ulcer syndrome has been stressed (Wayte and Helwig, 1967; Madigan and Morson, 1969). To the best of our knowledge, only a single case of localized CCP involving the colon other than rectum has been described (Fechner, 1967).

CASE REPORT

A 31-year-old salesman was admitted to Seoul National University Hospital for the excision of a "colonic tumor". The patient had remained healthy until two years previously when he first noted fresh anal bleeding that stopped spontaneously but reappeared three months before this hospitalization. The barium study revealed a protruding mass at the descending colon. On colonofiberscopy, a polypoid, soft, lobulated and sessile mass was found 55cm above the anal verge. Biopsy from the mass showed only a reactive mucosal hyperplasia. Physical examination and routine laboratory tests were within normal limits. A segmental resection with end-to-end anastomosis of the descending colon was done.

The resected segment contained a 2.7×2.1cm sized, sessile, polypoid and lobulated mass, involving half of the circumference. Microscopically, the polypoid lesion was made up of hyperplastic surface epithelium in papillary configuration with herniation of the glands into the submucosa. The muscularis mucosae was irregularly thickened. Glandular epithelium showed complex branching and depletion of the goblet cells. Submucosal lymphoid aggregates were prominent and

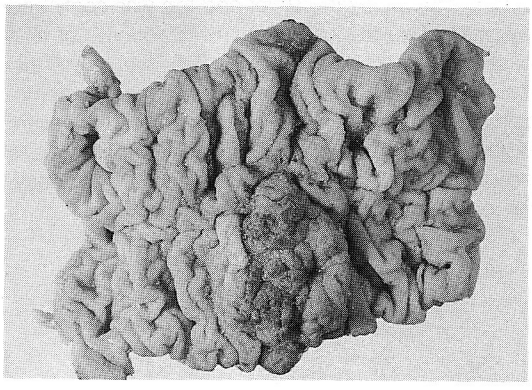

Fig. 1. The resected segment of the descending colon reveals a 2.7×2.1cm sized, sessile, polypoid and lobulated mass involving half of the circumference.

Address for correspondence: *Woo Ho Kim, Department of Pathology, Seoul National University College of Medicine, 28 Yongon-Dong, Chongno-gu, Seoul 110-460, Korea. (Tel: 02-760-3390)*

Supported by a grant (1988) from the Seoul National University Hospital Research Fund.

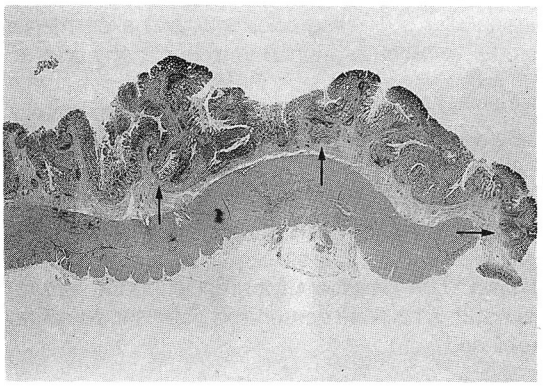

Fig. 2. The polypoid lesion is microscopically composed of hyperplastic surface epithelia in papillary configuration with herniation of the glands into the submucosa (arrow). (×3.7)

Fig. 3. A submucosal herniation is noted. The muscularis mucosae are irregularly thickened and lymphocytes and plasma cells are heavily infiltrated in the lamina propria. (×40)

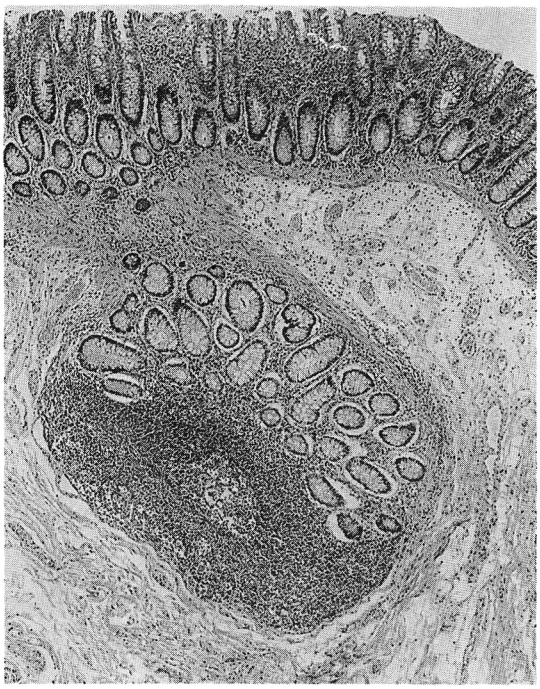

Fig. 4. Mucosa-associated lymphoid tissue (MALT) is prominent and it is especially located along the mucosal herniation. (×100)

they were located along the mucosal herniation. Heavy lympho-plasma cell infiltration in the lamina propria was noted. The lamina propria was replaced with disoriented collagen and smooth muscle fibers. Between the proper muscle coats, the myenteric plexus and ganglion cells were increased.

DISCUSSION

The CCP is a rare colo-rectal polypoid lesion, being characterized by the presence of epithelial-lined mucous cysts in the muscularis mucosae or submucosa. Although gross appearance of this case is similar to neoplastic polyp, deep downward herniations of hyperplastic epithelial glands provide easy diagnosis.

Colitis cystica profunda was first described in 1766 by Stark. In 1863, Virchow designated this lesion as colitis cystica polyposa (Wayte and Helwig, 1967). The superficial and diffuse lesions associated with pellagra were separately named as colitis cystica superficialis thereafter (Denton, 1925). In contrast, CCP extends below the muscularis mucosae. CCP is not common and no more than one hundred and fifty cases have been reported in the world literature (Guest and Rexnick, 1989). Herman and Nabseth (1973) have classified CCP into three forms according to distribution; diffuse, segmental and localized forms. In the diffuse form, the lesions are scattered throughout the colon. Bentlery et al. (1985) collected nine cases of diffuse CCP from the world literature; six of them were associated with ulcerative colitis, one with salmonellosis and the remaining two cases with multiple colonic polyps. The segmental form presenting as multiple polypoid lesions involves one or more segments of the large bowel, and seven such cases have been reported (Goodal and Sinclair, 1957; Allen, 1966; Castleman, 1966; Epstein et al., 1966; Herman and Nabesth, 1973). They were mostly localized in the rectosigmoid area, and one case was associated with ulcerative colitis (Barner, 1967). The localized form, the most common variant, presents with a single mass or

polyp, and is usually localized to the rectum within 5-12cm from the anal verge. Our case is an example of the localized form, but unusually located in the descending colon, and only a single case of CCP involving the descending colon has been reported in the literature (Fechner, 1967).

The pathogenesis of CCP is still obscure. It may be a consequence of an unusual reparative response (Bentley et al., 1985). Among 19 cases collected by Wayte and Helwig (1967) all of the localized forms were confined to the rectum. They noted the accompanying proctitis, and two cases were actually associated with ulcerative colitis. They suggested solitary rectal ulcer as a precursor lesion of the localized CCP, because both diseases share several clinical characteristics; they involve the rectum about 10cm above the anal verge, occur in either sex equally, develop in the 3rd and 4th decades, and produce vague rectal symptoms. The histologic features are also similar; granulation tissue with normal adjoining mucosa and a varying degree of inflammatory reaction. Goodal and Sinclair (1957) explained that the granulation tissue tracts passing through the muscularis mucosae may represent the way, through which the surface mucosa is herniated into the submucosa. In contrast, Madigan and Morson (1967) believed that CCP is hamartomatous and manifested as a precursor of solitary rectal ulcer syndrome. Our case is against both hypotheses in that there was no inflammatory or ulcerative process in the rectum and the lesion was apart from the rectum.

A congenital origin could be supported by embryologic studies, pediatric case reports and association of related disorders with genetic disease (Peutz-Jeghers syndrome). Similar benign inclusion of the intestinal epithelium beneath the submucosa and proper muscle coat is found in Peutz-Jeghers polyps and was named as enteritis cystica profunda (Kyriakos and Stanley, 1978). According to Shepherd et al. (1987), such a pseudocarcinomatous invasion accounts for about 10% of the Peutz-Jeghers polyps. However, the pathogenesis of the mucosal herniation is best explained as a repeated mechanical injury in hamartomatous polyp (Kim et al., 1984). Moreover, absence of CCP in pediatric autopsy series, association of the disorder with conditions involving irritation of the bowel wall, the regression of the lesion after a diverting colostomy, and experimental studies implicating an inflammatory etiology point to an acquired pathogenesis. Guest and Reznick (1989) concluded that this disorder is acquired, and is probably a part of the spectrum of a disease associated with internal prolapse and solitary rectal ulcer.

This case is most unusual in that it is a single lesion and locates at the descending colon, 55cm from the anal verge. Although the localized from of CCP is known to be almost always restricted to the rectum, its occurrence in the descending colon should not be excluded.

REFERENCES

Barner JL: *Colitis cystica profunda. Radiology 89:435-437, 1967.*

Bentley E, Chandrasoma P, Cohen H, Radin R, Ray M: *Colitis cystica profunda: Presenting with complete intestinal obstruction and recurrence. Gastroenterology 89:1157-1161, 1985.*

Castleman B: *Case records of the Massachusetts General Hospital. N Engl J Med 275:608-614, 1966.*

Denton J: *The pathology of pellagra. Amer J Trop Med 5:173-210, 1925.*

Epstein SE, Ascari WQ, Ablow RC, Seaman WB, Lattes R: *Colitis cystica profunda. Amer J Clin Pathol 45:186-201, 1966.*

Guest CB, Reznick RK: *Colitis cystica profunda. Review of the literature. Dis Colon Rectum 32:983-988, 1989.*

Fechner RE: *Polyp of the colon possessing features of colitis cystica profunda. Dis Colon Rectum 10:359-364, 1967.*

Goodall HB, Sinclair ISR: *Colitis cystica profunda. J Path Bact 73:33-42, 1957.*

Herman A, Nabseth D: *Colitis cystica profunda. Arch Surg 73:337-341, 1973.*

Kim YI, Kim WH, Suh DY, Cha KH, Ahn ST, Park CO: *Pseudocarcinomatous invasion in Peutz-Jeghers polyposis. Report of a case. Kor J Pathol 18:447-451, 1984.*

Kyriakos M, Stainley CC: *Enteritis cystica profunda. Amer J Clin Pathol 69:77-85, 1978.*

Madigan MR, Morson BC: *Solitary ulcer of the rectum. Gut 10:871-881, 1969.*

Shepherd NA, Bussey HJR, Jass JR: *Epithelial misplacement in Peutz-Jeghers polyps. A diagnostic pitfall. Amer J Surg Pathol 11:743-749, 1987.*

Wayte DM, Helwig EB: *Colitis cystica profunda. Amer J Clin Pathol 48:159-169, 1967.*