[症例報告]Mucosal prolapse syndrome simulating primary limtis plastica carcinoma of the rectum : report of two cases and a review of the literature

Mucosal prolapse syndrome simulating primary linitis plastica carcinoma of the rectum : report of two cases and a review of the literature

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ABSTRACT

Case 1 (a 53-year-old man, postoperative status for hepatocellular carcinoma, follow-up of 1.5 years) developed anal pain on defecation. A barium enema showed a segmental stricture. Colonoscopy revealed a multinodular mass with no obvious ulcer 4 cm from the anal verge. Endoscopic ultrasonography showed a mucosal mass with a preserved layer structure of the rectal wall. A biopsy of the lesion showed the characteristic features of fibromuscular proliferation in the lamina propria. He underwent a colostomy because of rectal stricture. He remained asymptomatic after the colostomy. Case 2 (a 54-year-old man, postgastrectomy status for gastric carcinoma, follow-up of 10 months) presented with thin stool and rectal pain on defecation. A barium enema revealed a segmental stricutre. Colonoscopy showed a multipolypoid mass with no ulceration 5 cm from the anus. A pelvic CT scan showed a mucosal mass with a normal layer structure of the rectal wall. The bioptic findings were identical to those of Case 1. He also underwent a colostomy and showed a good recovery. The two above described patients were both misdiagnosed to have rectal carcinoma, especially linitis plastica on the colonoscopic appearances. A biopsy of the lesion may lead to an accurate diagnosis of mucosal prolapse syndrome. A colostomy is thus considered to be a useful alternative treatment as a temporary strategy for such patients. *Ryukyu Med. J., 19(1)35~38, 1999*

Key words: mucosal prolapse syndrome, linitis plastica, barium -enema, colonoscopy, rectum.

INTRODUCTION

Mucosal prolapse syndrome (MPS) is a rare benign rectal disease which occurs due to mucosal prolapse. MPS can resemble rectal carcinoma in its clinical presentation, endoscopic and radiographic appearances and, in some cases also in the histopathologic findings due to the presence of an atypical proliferating epithelium in fibromuscular tissue. Therefore, patients with MPS are often misdiagnosed to have rectal carcinoma. We herein present two patients with MPS resembling rectal carcinoma, especially linitis plastica carcinoma, and discuss its differential diagnosis from rectal carcinoma.

CASE REPORT

Case 1. A 53-year-old man was admitted to the Ryukyu University Hospital for an evaluation of changes in his bowel habits and painful defecation on May 10, 1997. He had been followed up after undergoing an operation for hepatocellular carcinoma (HCC) for 1.5 years before this admission. He had previously undergone percutaneous transhepatic portal vein embolization for HCC in November 1995 and then underwent a right hepatectomy for HCC in January 1996, followed by an enucleation of a recurrent small HCC 10 months later. On this admission, he appeared mildly malnourished. The physical examination showed hepatectomy scars on the right upper quadrant, and a rectal examination revealed a multipolypoid lesion located 4 cm from the anal verge. The polypoid lesion was tender, firm, and had a somewhat irregular surface. Laboratory data, such as complete blood count, glucose, and serum liver chemistries, were slightly abnormal. A barium enema showed nodularity of the distal mucosa

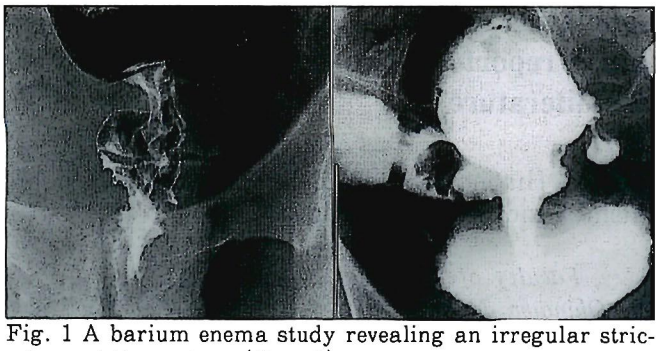

Fig. 1 A barium enema study revealing an irregular stricture of the rectum (Case 1).

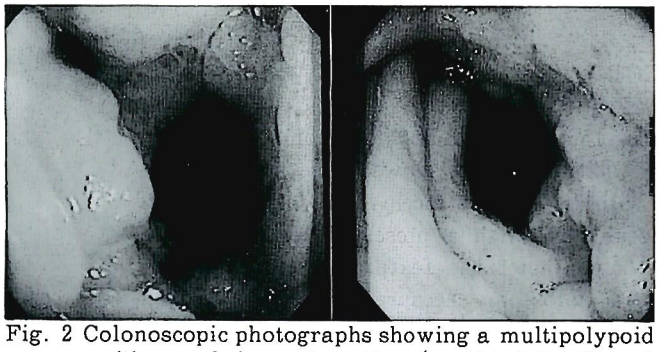

Fig. 2 Colonoscopic photographs showing a multipolypoid mass with no obvious ulceration (on the left, Case 1 and on the right, Case 2).

and stricture in the last 3 cm of the rectum (Fig. 1). Colonoscopy revealed a posterior multipolypoid mass and the absence of any obvious ulcers 4 cm from the anus (Fig. 2). Histologically, the lesion showed characteristic fibromuscular proliferation within the lamina propria (Fig. 4). Treatment with steroid enemas produced very little improvement in his condition, and he continued to have pain on defecation. Subsequently, he underwent a colostomy in August 1997 which has to date produced a favorable improvement. On colonoscopy, the polypoid masses had mostly disappeared, and only a few were observed to remain.

Case 2. A 54-year-old man presented with thin stool and anal pain on defecation. A barium enema showed a rectal stricture measuring 4 cm in length at a location 5 cm from the anal verge. The patient was referred to Ryukyu University Hospital for a further evaluation of rectal stricture on June 2, 1998. His past medical history revealed a previous total gastrectomy for diffusely infiltrating type of gastric carcinoma (Roux-en-Y reconstruction, stage IVa, signet-ring cell carcinoma) in October 1997. The curability of the gastric carcinoma was eveluated as "Curability B". Consequently, he received adjuvant chemotherapy (CDDP 5mg+5-FU 250 mg on Days 1 to 5 of each cycle). The chemotherapy cycles were repeated every 4 weeks for 4 cycles. At this admission, the patient appeared ill with

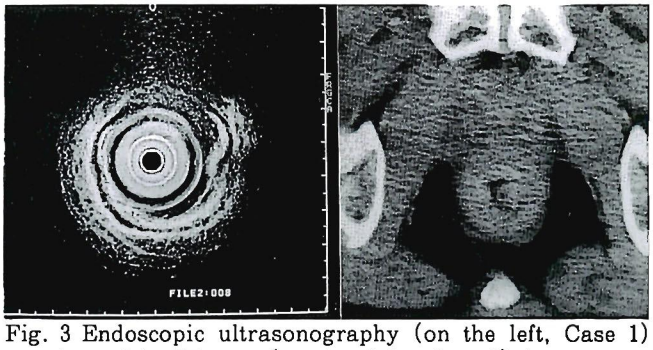

Fig. 3 Endoscopic ultrasonography (on the left, Case 1) and a pelvic CT scan (on the right, Case 2) demonstrating a mucosal mass with the preservation of the layer structure of the rectal wall.

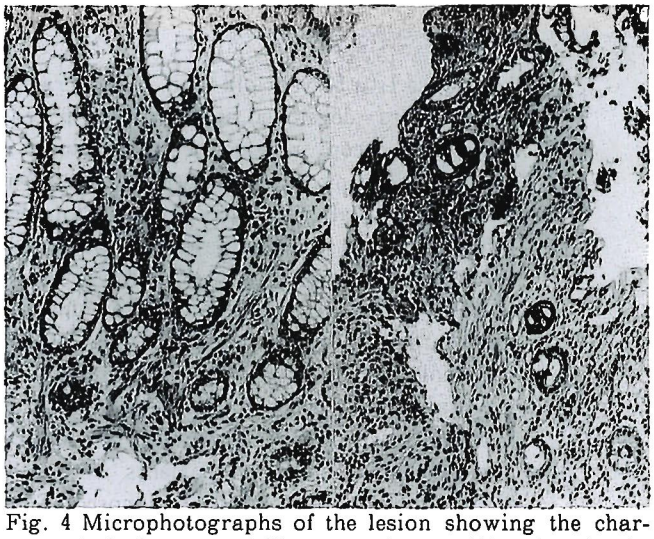

Fig. 4 Microphotographs of the lesion showing the characteristic features of fibromuscular proliferation in the lamina propria (on the left, Case 1 and right, Case 2) (HE, X50).

abdominal distension. The physical examination showed mild abdominal distension and the rectal examination revealed a circumferential multipolypoid mass with stricture and tenderness. Colonoscopy revealed a multipolypoid mass and no evidence of an obvious ulceration (Fig. 2). Endoscopic ultrasonography (EUS) and pelvic CT scan revealed a nodular thickening of the inner layers, preserving the layer structure of the rectal wall and the perirectal fat (Fig. 3). The endoscopic bioptic findings were characteristic of MPS and, these finding were identical to those of Case 1 (Fig. 4). Because of bowel obstruction, he underwent an emergency colostomy. At laparotomy, there was no apparent evidence of any pelvic peritoneal dissemination of gastric or rectal carcinoma. The colostomy successfully improved his condition with a favorable colonoscopic findings.

DISCUSSION

Reports of MPS from Japan are rare[1-4]. The rar-

ity of MPS may be related to its lack of recognition, clinical and pathological underdiagnosis, and also the rare occurrence of this condition in this country. Unfortunately, up to the present the precise reason for this remains unclear. MPS[5] is a rare benign rectal condition due to mucosal prolapse, including solitary rectal ulcer syndrome[6,7], and colitis cystica profunda[8], multiple hamartoma syndrome[9], and the margin of an ileostomy or colostomy[10]. In spite of its name, this condition also includes solitary rectal ulcer syndrome, and its characteristic macroscopic and histologic features do not include ulceration[11]. The endoscopic findings include polypoid or nodular mucosal thickening or a mass lesion with or without ulceration[12-14]. Moreover, the presence of histopathologically erosion and ulceration tend to vary greatly[6]. The pathogenesis of MPS remains to be elucidated. The combination of constipation, excessive straining during defecation, and self-inflicted rectal trauma have been proposed[6,11]. These conditions lead to ischemia due to the internal prolapse and subsequent fibromuscular obliteration of the rectal mucosa. Many MPS studies in the literature have mainly focused on its recognition while little has been reported regarding its differential diagnosis. Two major categories of rectal disease play a role in the differential diagnosis of MPS. Inflammatory bowel disease and rectal carcinoma must be differentiated from MPS because of different therapeutic strategies[7,15,16]. Inflammatory bowel disease, besides having its own clinical and histopathologic features, does not show the characteristic fibromuscular proliferation of the lamina propria. A second crucial dif ferential diagnosis is invasive carcinoma of the rectum for which MPS can also be mistaken, as previously described in the literature[5,6,11]. In our cases, endoscopic features were circumferencial multipolypoid mass and radiographic features were nodularity. Bioptic features were characteristic fibromuscular proliferation. These revealed MPS. But conservative treatment produced very little improvement in his condition (Case 1). Bowel obstruction occurred because of rectal multipolipoid lesion (Case 2). And two cases had past history of malignant disease. Therefore we suspected rectal carcinoma, especially linitis plastica carcinoma because of no epithelial neoplastic change. It is certain that the importance of a rectal biopsy is clear. The presence of a fibromuscular replacement of the lamina is characteristic, and the absence of epithelial neoplastic change indicates the benign nature of MPS. However, a recent study has also described another diagnostic pitfall of MPS[17]. This study indicated that the histopathology of MPS may occasionally represent a characteristic but nonspecific mucosal reactive change to a deeper seated malignancy. Certain clinical features in patients with MPS can serve as a clue for the inflammatory bowel disease and its underlying maligancy. We found diagnostic studies such as endoscopic ultrasonography (EUS) and pelvic CT, to be helpful in assessing the status of the rectal mass. The multipolypoid lesions in ordinary MPS are usually confined to the mucosa while the layer structure of the rectal wall is preserved. In contrast malignant lesions show a solid mass, a transmural invasive lesion or perirectal soft tissue involvement[17-20]. Regarding the therapeutic strategies for this disease, no satisfactory treatment has yet been established for this benign but chronic disorder[11]. In a recent report[21], a patient with a transanal removal of a mass lesion remained asymptomatic and free of recurrence during a 31-month follow-up. The patients in this report both underwent a colostomy for rectal stricture with MPS as a temporary treatment and both have remained asymptomatic with an improvement in the lesion during a follow-up ranging from 3 to 12 months. A colostomy may thus be the optimal treatment as a temporary stategy. A long term follow-up of MPS may allow a clearer picture of its natural history and thereby a more definitive treatment should thus eventually emerge.

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