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CASE REPORT

분출성 동맥출혈과 내강 협착을 동반한 회장 말트림프종 1예

표정의, 이범재, 이현주, 김지원, 김경진, 박종재, 김재선, 박영태, 장유진[1]
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A Case of Ileal Mucosa-associated Lymphoid Tissue Lymphoma Accompanied by Luminal Stricture and Arterial Spurting

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Primary small intestinal lymphoma is relatively uncommon. Small bowel tumors are difficult to diagnose, because they are usually asymptomatic in the initial phase, and they are not easily detected by traditional methods of investigating the small intestine. This case shows a successfully detected and treated gastrointestinal bleeding from rare ileal mucosa-associated lymphoid tissue lymphoma, using double balloon endoscopy. **(Korean J Gastroenterol 2013;62:365-369)**

Key Words: Mucosa-associated lymphoid tissue lymphoma; Gastrointestinal bleeding; Small intestine; Double balloon endoscopy

INTRODUCTION

Although the gastrointestinal tract is one of the most frequent sites of extranodal malignant lymphoma, the occurrence of primary small intestinal lymphoma is relatively uncommon, accounting for 19-38% of small bowel malignancies,[1] and 20-30% of primary gut lymphoma.[2] Since the first description in 1983,[3] the concept of mucosa-associated lymphoid tissue (MALT) lymphoma has been widely accepted, and has also been incorporated into the new histologic classification for extranodal lymphomas, including primary gut lymphoma.[4] Previous studies have analyzed the detailed clinicopathologic features of MALT lymphoma of the stomach,[5] but little remains known regarding the MALT lymphoma of the small bowel. To date, only a few cases of small bowel MALT lymphoma have been reported. We report a case of small bowel MALT lymphoma complicated with annular stricture and active bleeding, which was successfully detected and treated, using double balloon endoscopy (DBE).

CASE REPORT

A 67-year-old man was referred to an outside hospital for the treatment of hematochezia. Upper gastrointestinal endoscopy and total colonoscopy were performed, but failed to localize the bleeding site. Abdominal-pelvic CT scan did not show abnormal findings. Video capsule endoscopy (VCE) showed fresh blood in the middle ileum, but also failed to localize the bleeding site (Fig. 1). The patient was referred to our center, for enteroscopy. The patient reported a 3-day history of intermittent hematochezia, and a past history of recurrent abdominal pain for the past 3 years; but endoscopic

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examination and abdominal pelvic CT findings performed at previous centers did not detect any abnormalities. He had been taking aspirin for 17 months. On admission, his vital signs were blood pressure 90/60 mmHg, heart rate 96 beats/min, body temperature 36.6°C, and respiratory rate 18 breaths/min. On physical exam, the patient was alert and not pale, and digital rectal examination revealed hematochezia. Laboratory findings were as follows; hemoglobin 9.6 g/dL, white blood cells count 7,200/μL (neutrophile 60%, lymphocyte 31.5%, eosinophil 3.3%), platelet 112/μL, PT 14.2 sec (INR 1.09), aPTT 34.9 sec, protein 5.7 g/dL, albumin 3.6 g/dL, AST/ALT/ALP 126/28/103 IU/L, total bilirunbin 1.06 mg/dL. Considering the examinations done in the previous hospital, we performed enteroscopy for hemostasis.

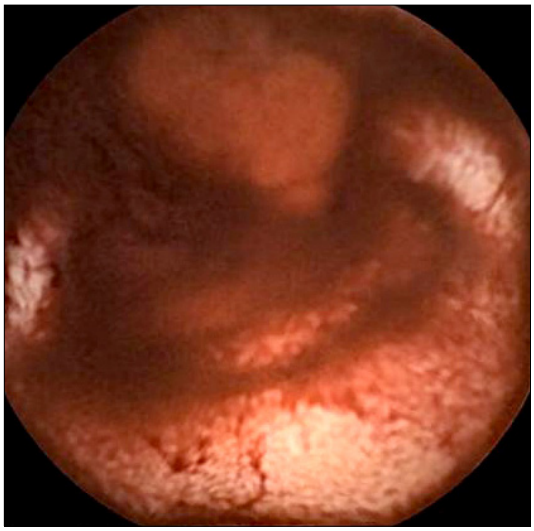

Fig. 1. Video capsule endoscopy, showing fresh blood in the middle ileum.

Antegrade DBE (EN450T5; Fujinon, Saitama, Japan) revealed arterial spurting from an irregular ulcerative lesion, with severe luminal stricture at the middle ileum. The scope could not pass through this lesion. We used epinephrine injection and argon plasma coagulation for bleeding control, and hemostasis was successfully achieved (Fig. 2). Retrograde DBE was performed, but no other synchronous lesions were found. The whole procedure took about 150 min. DBE guided biopsy revealed an active ulcer. Since he had suffered persistent abdominal pain for 3 years, and the endoscopic appearance was highly suggestive of malignancy, he subsequently underwent small bowel resection. The surgical pathology revealed extranodal marginal zone B-cell lymphoma of MALT type (also known as MALT lymphoma), characterized by the appearance of poorly defined follicular areas, which were composed of monocytoid B cells that featured enlarged nuclei and lymphoepithelial lesions (Fig. 3). Post-operative staging work-ups showed no evidence of disseminated disease. Urea breath test for *Helicobacter pylori* was negative. The patient has been followed up regularly for the last 9 months, and no signs of recurrence have been found.

DISCUSSION

Small bowel tumors are relatively rare disorders, comprising less than 10% of all gastrointestinal tumors. The development of capsule endoscopy and balloon-assisted endoscopy launched a new era for the diagnosis and management of small bowel diseases.[6] Several studies have been published on the role of DBE in the diagnosis and treatment of small bowel tumors. Dinesen et al.[7] prospectively assessed the di-

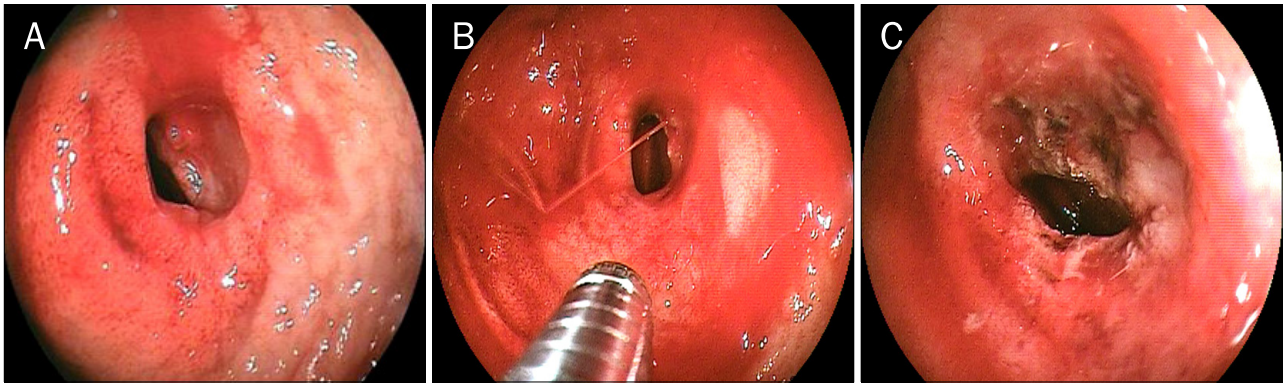

Fig. 2. (A, B) Double balloon endoscopy shows active ulcer with luminal stricture. Arterial spurting was noted at the ulcer base. (C) Argon plasma coagulation with ephinephrine-normal saline mixture injection was used to achieve hemostasis.

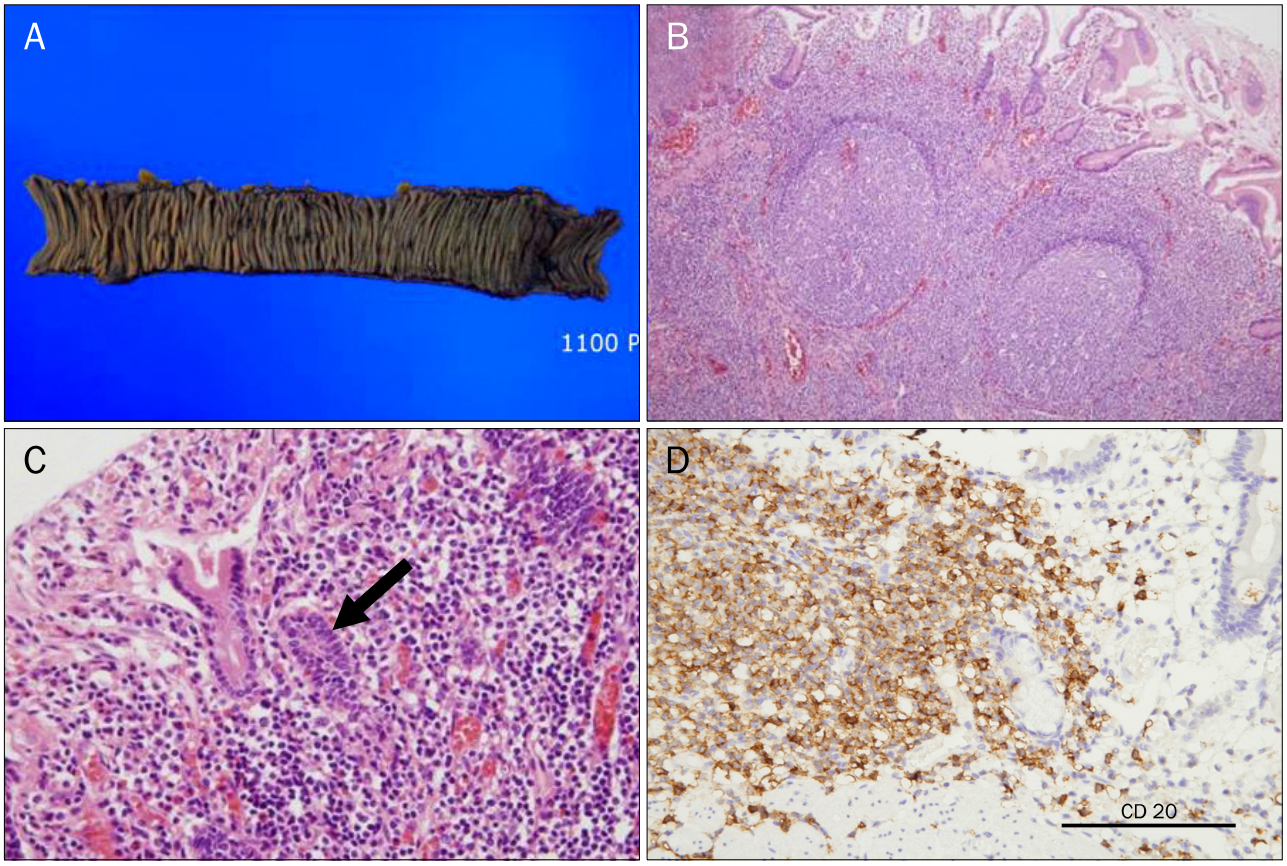

Fig. 3. Histopathologic findings of mucosa-associated lymphoid tissue lymphoma. (A) The tumor measured 1.5×1.3 cm across. (B, C) Microscopic examination shows the poorly defined follicular areas that are composed of monocytoid B cells that feature enlarged nuclei (H&E, ×100), and infiltration and distortion of epithelial structures by aggregates of neoplastic lymphoid cells (H&E, ×400). (D) Immunohistochemistry shows strong positive activity for CD 20 (×400).

agnostic and therapeutic impact of DBE, in patients with suspected or documented small bowel neoplasia, seen on VCE or CT scan. Over a 6-year period, a total of 580 DBE procedures were carried out. A total of 48 patients were found to have neoplastic disease/masses, and 6% were pathologically proven to be lymphomas. Imaoka et al.[8] conducted a retrospective chart review in 227 patients who had undergone DBE, and found that small bowel tumor groups contained more symptomatic patients than the non-small bowel tumor group (90% *vs.* 49%, $p < 0.05$), with a significantly higher rate of gastrointestinal symptoms at presentation (72% *vs.* 33%, $p < 0.05$). In a Korean multi-center study, among the 112 patients with small bowel tumors who received DBE, benign polyp was the most common (n=38, 33.9%), and lymphomas were found in 16.1% (n=18). Small bowel lymphomas were located in the jejunum (50.0%), ileum (38.9%), and duodenum (5.6%), evenly distributed throughout the entire small bowel.[6]

Because small bowel lymphomas are uncommon, the majority of studies on endoscopic findings have been performed in small bowel tumors, and studies on the endoscopic findings of small bowel lymphomas are limited. DBE improved the endoscopic approach to the small bowel, and enabled preoperative histopatholgic investigations and minimally invasive surgery, in small bowel tumors, including lymphomas. In this case, endoscopic treatment was performed immediately, for active arterial spurting. The endoscopic appearances of small bowel lymphoma on DBE included stenosis, ulceration, and a mass or ulcerated mass lesion.[8] Endoscopy commonly demonstrates MALT lymphoma of the small intestine, presented as either the ulcerative or polypoid type, and was indistinguishable from other high grade B-cell or T-cell lymphomas.[9] In a Japanese retrospective study,[8] among the 5 patients with small bowel lymphoma who had undergone DBE, the endoscopic findings of 2 cases of MALT lymphoma were multiple whitish nodules.

Clinical manifestations may vary from each patient, who may present with any one, or a combination of any, of the following: dyspepsia, epigastric pain, abdominal pain, nausea, vomiting, diarrhea, weight loss, malabsorption, obstruction, anemia, and to a lesser extent ulceration, perforation, and intussusceptions.[10-12] The most frequent symptoms for small bowel tumors were obscure gastrointestinal bleeding (57%), and chronic abdominal pain/diarrhea/obstruction (15%).[6] Conversely, a few patients were reportedly asymptomatic.[13] In our case, the patients had abdominal pain that persisted for 3 years, and hematochezia. Endoscopic finding showed arterial spurting in the ileum, with annular stricture. To date, only a few cases of MALT lymphoma of the small bowel have shown annular stricture. Yanai et al.[14] reported a rare case of MALT lymphoma of the small bowel with annular stricture, presumably induced by NSAID.

MALT lymphomas display unusually indolent behavior, remaining localized to their site of origin for long periods of time without disseminating, a feature which has made them uniquely amenable to cure by local therapy, such as surgical excision, with or without radiation therapy.[15] Gastric MALT lymphomas, which represent up to 48% of all primary gastric lymphomas, are associated with *H. pylori* infection, and the eradication of *H. pylori* correlates with tumor regression.[16] In contrast to gastric MALT lymphoma, the relationship between small bowel MALT lymphoma and *H. pylori* infection has not been established; and whether *H. pylori* eradication may, or may not lead to lymphoma regression, has also not been established.[16,17] *H. pylori* test showed negative in our case. Relapses involving the small bowel are rare, unlike gastric MALT lymphoma, where, following complete remission after the eradication of *H. pylori*, the risk of relapse justifies life long follow-up examinations.[18]

In conclusion, small bowel MALT lymphoma is a relatively rare form of gastrointestinal tumor. To date, only a few cases of small bowel MALT lymphoma have been reported, and to our knowledge, this is the first case to report small bowel MALT lymphoma complicated with annular stricture and active bleeding. Small bowel tumors are difficult to diagnose, because they are usually asymptomatic in the initial phase; and traditional methods of investigating the small intestine, such as small bowel follow-through and CT scanning, have low yield for tumors.[19] This case represents the successful detection and treatment of arterial bleeding from rare ileal MALT lymphoma accompanied by annular stricture, using DBE.

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