CASE REPORT

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Jejunocolic Fistula Associated with an Intestinal T Cell Lymphoma

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Malignant fistula of the small bowel to the colon is rare and is most often due to adenocarcinoma. Small bowel lymphoma is unusual, representing less than 1 percent of all gastrointestinal malignancies. We report a case of intestinal lymphoma presenting with diarrhea and abdominal pain. A jejunocolic fistula was discovered during colonoscopy. Celiotomy revealed a large, ulcerated fistula tract between the jejunum and distal transverse colon, and pathology was consistent with peripheral T-cell lymphoma. This is a rare entity in a nonimmunocompromised individual and has not been previously described in Korea. **(Gut Liver 2011;5:387-390)**

Key Words: Peripheral T-cell lymphoma; Jejunocolic fistula; Diarrhea

INTRODUCTION

Enterocolic fistulas are usually caused by inflammatory conditions such as Crohn's disease. It can also be caused by prior surgery, foreign bodies, pancreatitis, diverticulitis, and malignancy.[1-4] Small bowel tumors account for 1% to 3% of all gastrointestinal neoplasms.[5] Malignant lymphoma accounts for up to 20% of all primary malignant tumors of the small bowel,[6] and most primary small intestine lymphomas are of B-cell origin. Lymphomas originating from T-cells are rare.[6,7] In Japan, only 24 cases of T-cell malignant lymphoma of the small intestine have been reported and primary intestinal peripheral T-cell lymphoma (PTCL) is especially rare.[8] Intestinal fistula is known to be a late complication of primary intestinal lymphoma, and so is rarely encountered. This report describes a case-to our knowledge this is first reported case in Korean population-of primary intestinal PTCL causing jejunocolic fistula and perforation.

CASE REPORT

A 56-year-old man presented with an acute diarrheal illness that had begun 14 days prior to first hospital visit. The patient had intermittent fevers, chills, night sweat 1 month previously and a 9 kg loss of body weight in the previous 3 months. The patient was afebrile and had stable hemodynamic vital signs. The patient was cachectic and pale, but appeared nontoxic on physical examination with a soft and flat abdomen, normoactive bowel sounds and no tenderness or palpable lymphadenopathy. The past medical history was significant for pulmonary tuberculosis in the third decade of life and chronic hepatitis B carrier state. There was no prior history of abdominal surgery.

Initial laboratory tests revealed the following results: leukocyte counts 7.2×10^9/L (neutrophile 75.5%), Hemoglobin 11 g/dL, platelets 407×10^9/L, total protein 6.6 g/dL, albumin 3.9 g/dL, total bilirubin 1.0 mg/dL, AST 73 IU/L, ALT 65 IU/L, Alkaline phosphatase 193 IU/L, LDH 212 IU/L, BUN 12.3 mg/dL, creatinine 0.96 mg/dL, sodium 129 mmol/L, potassium 3.3 mmol/L, Cl^- 93 mmol/L, plasma CEA 0.2 ng/mL, with a normal blood cell morphology. The work up for infectious diarrhea was negative. Contrast enhanced computed tomography (CT) of the abdomen showed focal wall thickening in the distal transverse colon with enlarged regional lymph nodes (Fig. 1).

During the colonoscopy, a fistula opening communicating with a small bowel was identified at the distal transverse colon, and a circumferential ulcer was detected in the small bowel side through the fistula opening (Figs 2 and 3). The colonoscope was not advanced through the fistula due to the risk of perforation. Biopsies from the circumferential ulcer of small bowel side showed an aggregation of atypical lymphoid cells suggestive of lymphoma. However, biopsies from the margin of the colonic fistula opening showed chronic inflammation only.

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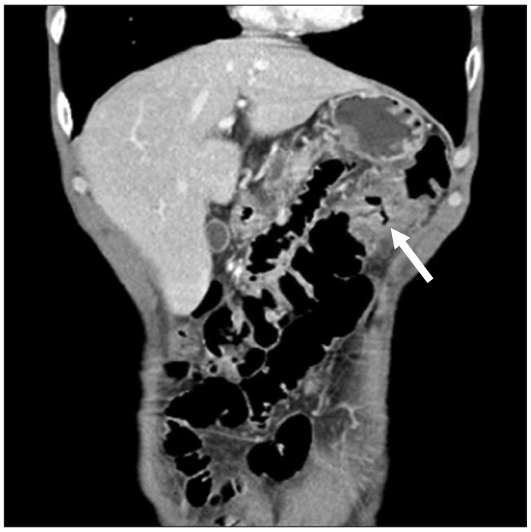

Fig. 1. Contrast enhanced computed tomography of the abdomen shows focal wall thickening in distal transverse colon (white arrow) with enlarged regional lymph nodes.

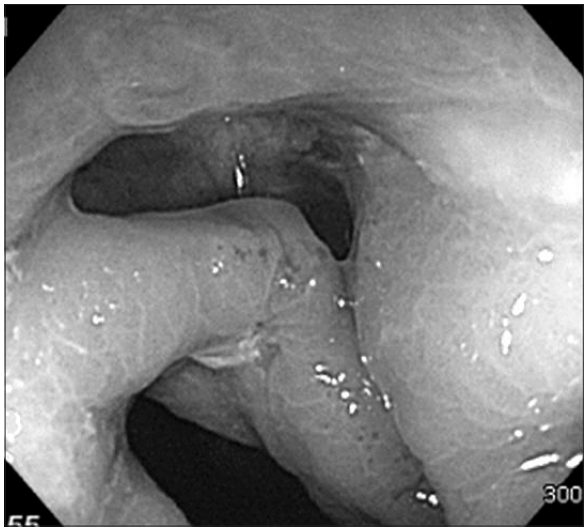

Fig. 2. Colonoscopic examination reveals a fistula opening at the distal transverse colon.

Patient refused to admit and further evaluation until histologic diagnosis of the lesion is determined. Therefore, the further studies including CT enterography and follow-up colonoscopy for additional biopsy were delayed.

Two weeks after the colonoscopy, at his 3rd visit to outpatient clinic for admission, the patient presented with acute abdominal distress. An emergency laparotomy was performed, which revealed a large inflammatory mass-like lesion covered with omentum. After adhesiolysis, jejunocolic fistula and jejunojejunal adhesion were noted. A perforation site was found in the jejunocolic fistula. An extended right hemicolectomy and segmental jejunectomy were performed with dissection of regional mesenteric lymph nodes.

Pathology revealed a 7.0×5.5 cm-sized ulcerated fistula tract between the jejunum and the distal transverse colon (Fig. 4).

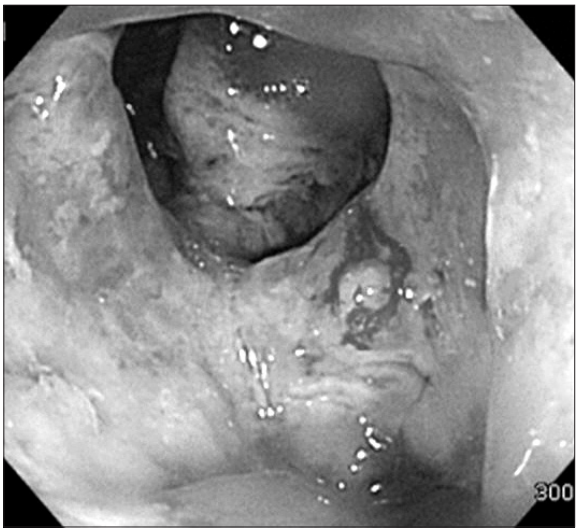

Fig. 3. Colonoscopic examination reveals circumferential ulcer and fistula tract at the small bowel.

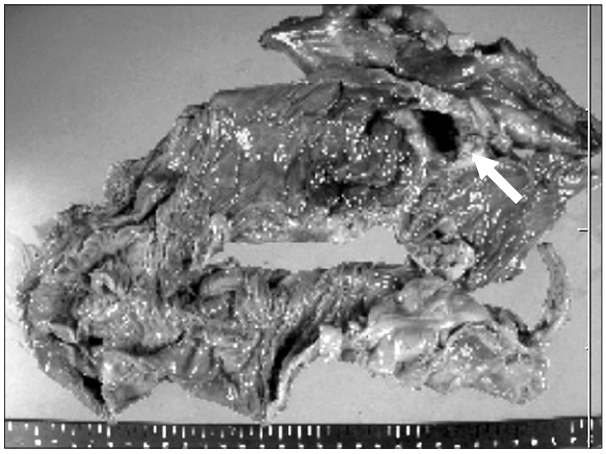

Fig. 4. Pathology reveals a 7.0×5.5 cm-sized ulcerated fistula tract opening (white arrow) between the jejunum and the distal transverse colon.

Light microscopy examination of the involved small intestinal mucosa showed diffuse infiltration of predominantly medium-sized atypical lymphoid cells in the lamina propria (H&E stain, ×400), but findings of normal small bowel mucosa showed normal villi without atrophic change. Immunohistochemical staining was positive for leukocyte common antigen, CD30, CD3, and Galectin-3; weakly positive for CD4; and negative for CD56, anaplastic lymphoma kinase, and CD8 (Fig. 5). Thirty of 65 lymph nodes were also involved by atypical lymphoid cells. Thus, the final pathology was confirmed as unspecified primary small-intestinal PTCL.

After surgery, the patient's condition continued to deteriorate and he died 2 months after surgery due to peritonitis and sepsis.

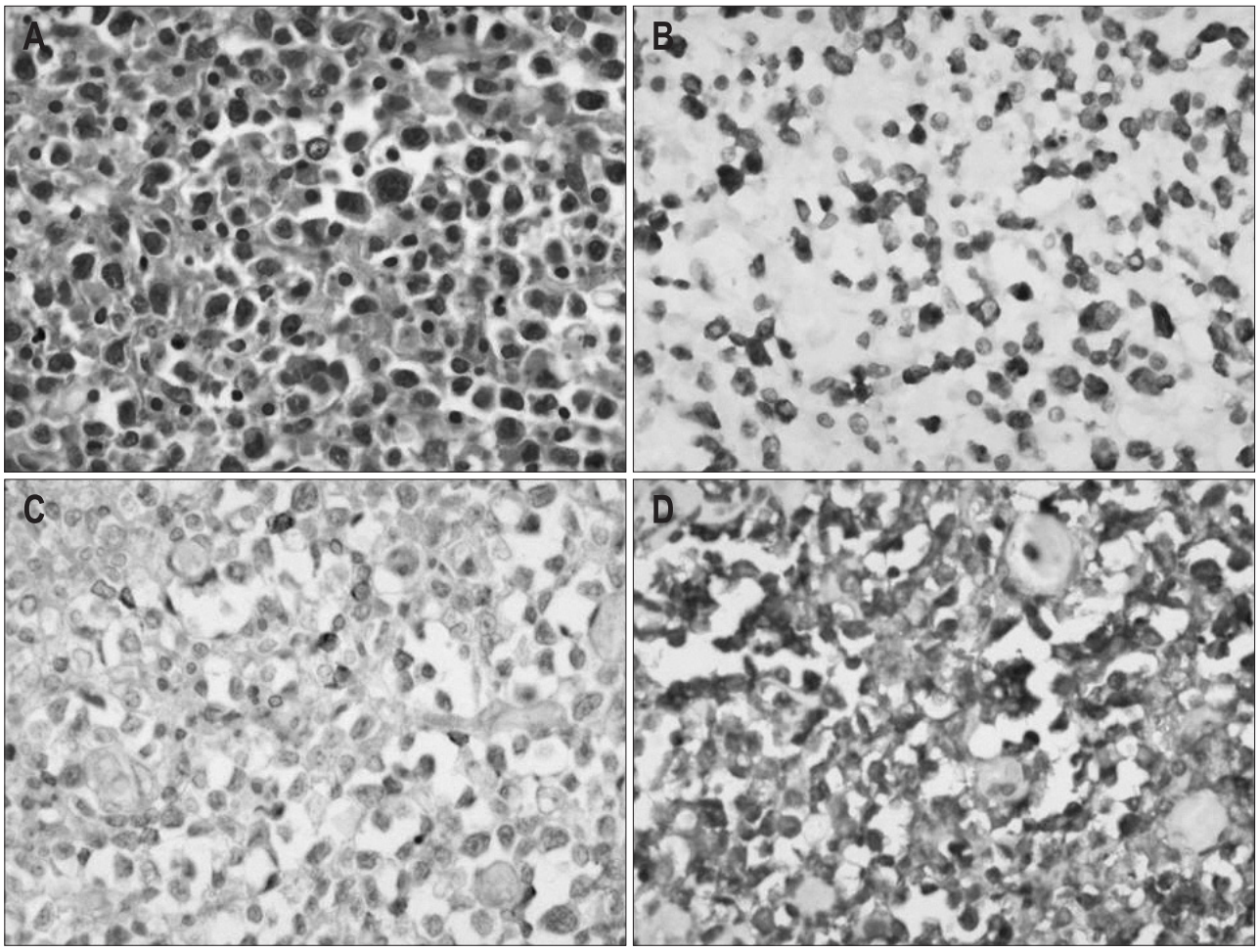

Fig. 5. Results of H&E staining of tumor cells (×400). Diffuse infiltration of medium to large lymphoid cells with pleomorphic and irregular nuclei is noted (A). Tumor cells are strongly positive for CD3 (B), weakly positive for CD4 (C), and strongly positive for LCA (D).

DISCUSSION

The gastrointestinal tract is the most frequent site of extranodal involvement in malignant lymphoma. However, primary gastrointestinal lymphoma remains relatively rare. Gastrointestinal lymphomas represent 1% to 10% of all gastrointestinal malignancies,[9,10] up to 20% of all small bowel malignancies[5,6] and only 0.2% to 0.4% of large bowel malignancies.[11] Most primary small intestine lymphomas originate from B-cells, with T-cell origin being rare.[6,7] The most common location of PTCL is the jejunum[12] and common symptoms include abdominal pain, nausea, vomiting, diarrhea, fatigue, weight loss and fever. Unfortunately, these symptoms are not disease specific. Therefore, the diagnosis of lymphoma of the small intestine is often made after complications such as hemorrhage, obstruction or perforation develop.[6,13]

Enterocolic fistula is a rare complication in patients with intestinal lymphoma. It has been noted that most gastrointestinal malignant fistulas arise in the presence of locally invading adenocarcinoma.[14] In the natural course of PTCL, enterocolic fistula is likely to represent a late complication and suggests an advanced disease.[15]

To the best of our knowledge, only six cases enterocolic fistula associated with lymphoma have previously reported in the English literatures. Of these, four were B-cell lymphoma[15-18] and two cases were T-cell lymphoma.[9,19] In all six cases, diarrhea was an initial symptom. There has been no other case of intestinal lymphoma causing alimentary tract fistula reported in Korea.

The present patient also presented with watery diarrhea with nonspecific and vague abdominal pain. Although the patient already had intestinal adhesion and enterocolic fistula at the time of the first hospital visit, physical examination revealed no abdominal tenderness or palpable abdominal mass. Fever, night sweat and extensive weight loss were the only clues to suspect a malignant disease. The patient's diarrhea may be explained by short bowel syndrome and small bowel bacterial overgrowth syndrome caused by refluxed colonic bacteria.[20,21] In summary of the symptoms above, fever and extensive weight loss were clinical clues to the diagnosis of malignant disease. Subacute diarrhea that was intractable with the conventional antidiarrheal agent and negative stool studies were clues to presume an in-

testinal fistula. Barium studies, endoscopy and CT enterography are useful tools for demonstrating a fistulous tract.

The treatment options for intestinal lymphoma are varied and not standardized. Resection of tumor and regional lymph nodes constitutes the initial treatment of small bowel lymphoma. After surgery, combination chemotherapy is the preferred option.[22] In comparison with B-cell lymphoma, PTCL is a heterogeneous group of neoplasms presenting as advanced disease, and is characterized by widespread dissemination, aggressive behavior and a very poor outcome.[23] The therapeutic outcome of PTCL is usually worse than that of diffuse large B-cell lymphoma.[24]

In conclusion, we report a male patient with primary intestinal PTCL complicated with enterocolic fistula and small bowel peforation. Although intestinal PTCL is an extremely rare disease, particular attention should be paid its nonspecific clinical manifestation for early diagnosis and treatment of this aggressive disease.

CONFLICTS OF INTEREST

No potential conflict of interest relevant to this article was reported.

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