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Case Report

Enteropathy-Type Intestinal T-Cell Lymphoma Showing Jejunoileal Fistula: Report of a Case

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Jejunoileal fistula is an extremely rare complication in patients with intestinal lymphoma. Here, we report a Japanese male patient with enteropathy-type intestinal T-cell lymphoma presenting abdominal pain and weight loss. A jejunoileal fistula was discovered during colonoscopy and pathological diagnosis was performed preoperatively by forceps biopsy. After elective surgery for partial resections of jejunum, ileum, and sigmoid colon, eight cycles of cyclophosphamide, doxorubicin, vincristine and prednisolone chemotherapy led complete remission of the disease.

Keywords: Jejunoileal fistula; T-cell lymphoma; Enteropathy; Intestine

Introduction

Primary gastrointestinal lymphoma is a relatively rare disease constituting 5-10% of all gastrointestinal tumors.1 Most cases are derived from B cells, whereas T-cell lymphomas are rare and are often related to celiac disease.2-3 One characteristic of intestinal T-cell lymphoma is intestinal perforation, which occurs in 30-50% of patients.1,2,3 Jejunoileal fistula is an extremely rare complication in patients with intestinal lymphomas, and is most often caused by Crohn's disease and less frequently by diverticular disease. It is often seen in association with malnutrition and diarrhea in patients with abdominal pain. Here, we present a case of enteropathy-type intestinal T-cell lymphoma showing jejunoileal fistula.

Case report

In May 2004, a 62-year-old Japanese male presented with lower abdominal pain, which had been developing for three years. His body weight had decreased by 10 kg in a single year. In 1999, he experienced polyradiculoneuropathy, but he had no history of celiac disease. Upper gastrointestinal endoscopy and pathological diagnosis was performed preoperatively by forceps biopsy. After elective surgery for partial resections of jejunum, ileum, and sigmoid colon, eight cycles of cyclophosphamide, doxorubicin, vincristine and prednisolone chemotherapy led complete remission of the disease.

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performed. The specimen exhibited diffuse proliferation of atypical medium to large lymphoid cells that were positive for TIA-1, CD8 and CD3, but negative for CD20, CD4 and CD30. Enteropathy-type intestinal T-cell lymphoma was considered. Small bowel X-ray (Figure 2) showed a large cavitary lesion forming an ileo-jejunal fistula. Partial ileectomy, partial jejunectomy and partial sigmoidectomy were performed on June 15, 2004. The tumor measured 60 x 70 mm and penetrated the jejunum, adhering to the mesocolon of the Sigmoid colon. The tumor comprised diffuse atypical large lymphoma cells (Figure 3). Immunohistochemistry of frozen sections confirmed that the lymphoma cells were positive for CD2, CD3, CD8 and TCRγδ, but negative for CD4, CD16, CD56, CD57, CD103 and TCRδ. On Southern blot analysis, tumor tissue was positive for TCR Cβ1 and TCR Jγ. Based on the histopathological, immunophenotypic and genetic findings, a diagnosis of enteropathy-type intestinal T-cell lymphoma was made.

Because remnants of the tumor were suspected to persist in this case, CHOP (cyclophosphamide, doxorubicin, vincristine and prednisolone) was initiated. The patient tolerated this therapy well and had received eight full cycles of chemotherapy in February 2005. At the last follow-up examination, four years after initial diagnosis, the patient was in good condition with no evidence of recurrence of the disease.

Discussion

We described a case of enteropathy-type intestinal T-cell lymphoma showing jejunoileal fistula diagnosed preoperatively by clinical and histological examination. The present patient showed severe abdominal pain and was not previously diagnosed with either celiac disease or enteropathy. Because intestinal T-cell lymphoma often arises prior to celiac disease-like symptoms, the term "enteropathy-type intestinal T-cell lymphoma", as specified by the World Health Organization Classification, is now widely accepted. Gale et al. demonstrated that only 12 of 31 patients with a diagnosis of enteropathy-type intestinal T-cell lymphoma have a documented clinical history of celiac disease and important clinical features are weight loss, abdominal pain and diarrhea. Katoh et al. demonstrated that only 7 of 18 patients with intestinal T-cell lymphoma present with enteropathy. Enteropathy-type intestinal T-cell lymphoma does not always occur with enteropathy.
Malignant lymphoma is apparently inclined to form fistulas, and since Nadal et al. first reported gastroduodenal fistula caused by malignant lymphoma, numerous cases describing tracheoesophageal, gastroporeal, gastroplenic, jejunoileal, ileocolic and choledodhoduodenal fistulas have been reported. Most of the organs involved and involved in fistula formation are fixed and located in the retroperitoneum. A search of the 1966-2008 MEDLINE databases failed to find reports describing jejunoileal fistula caused by malignant lymphoma. Jejunoileal fistula is an extremely rare complication in patients with intestinal lymphomas.

The prognosis for enteropathy-type intestinal T-cell lymphoma is extremely poor. A multicenter prospective clinical study of 65 patients with primary intestinal lymphoma demonstrated that the two-year survival rate in patients with intestinal T-cell lymphoma is 28%, which is markedly less than in patients with intestinal B cell lymphoma (94%). Two large retrospective analyses in Western populations indicated that the 1-year survival rate for enteropathy-type intestinal T-cell lymphoma is 31% to 39%. The poor clinical outcome, in part, reflects emergency surgery for perforation and late diagnosis. The present case has shown favorable prognosis to date because we were able to preoperatively diagnose the condition and perform elective surgery for enteral fistula. Because the small bowel is sometimes overlooked as the cause of abdominal pain and is examined only indirectly through barium radiography, diagnosis of intestinal lymphoma tends to be delayed. It is important to examine the terminal ileum when colonoscopy is performed during examination of abdominal pain and celiac disease. Capsule endoscopy was recently developed to investigate diseases of the small intestine. Joyce et al. performed capsule endoscopy to assess the extent of enteropathy-type intestinal T-cell lymphoma after chemotherapy and reported that the findings of endoscopy were correlated with those of barium radiography. Capsule endoscopy may thus be a useful modality in patients with celiac disease.

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References