A Case of Enteropathy-Type T-Cell Lymphoma Diagnosed by Small Bowel Enteroscopy: A Perspective on Imaging-Enhanced Endoscopy

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Enteropathy-type T-cell lymphoma (ETL) or enteropathy-associated T-cell lymphoma is a very rare malignant intestinal tumor. ETL is usually diagnosed by surgery. Endoscopic findings of ETL are not well known, and there are few reports of findings from endoscopy that has been performed only using white light. Additionally, there are no definite treatment guidelines for ETL. Therefore, we report a case of ETL diagnosed by enteroscopy with imaging-enhanced endoscopy and also review recently developed treatment options. (Gut Liver 2012;6:516-519)

Key Words: Enteropathy-type T-cell lymphoma; Imaging-enhanced endoscopy; Narrow band image

INTRODUCTION

Enteropathy-type T-cell lymphoma (ETL) is primary extranodal T-cell lymphoma arising in the intestine originating from intraepithelial T-cells. ETL is very rare and accounts for fewer than 5% of all gastrointestinal tract lymphomas. The clinical course of ETL is very aggressive. Five-year survival rate is 20% to 25%. The treatment options are chemotherapy with or without radiotherapy and surgery. Currently, there are no definite treatment guidelines for ETL. Diagnosis of ETL was very difficult and usually made by surgery because of complications of ETL at presenting time, such as intestinal perforation or obstruction, or lack of endoscopic experiences. There were few articles about endoscopic findings of ETL with only white light and capsule endoscopy.

CASE REPORT

A 70-year-old malnourished male patient visited because of anemia and hematochezia. Hemoglobin was 8.9 g/dL and albumin was 2.9 g/dL. There was no definite bleeding focus found on gastroscopy and colonoscopy. Relatively long segmental wall thickening of jejunum with mild dilatation, fluid collection and suspicious mesenteric fat infiltration was noted on abdominal computed tomography. Increased fluorodeoxyglucose (FDG) uptake (SUV, 6.0) was also noted on 18-FDG positron emission tomography (Fig. 1). To evaluate for obscure gastrointestinal

Fig. 1. Abdominal computed tomography (CT) and positron emission tomography (PET) CT. (A) Diffuse enhancing wall thickening and mild dilatation with fluid collection of jejunum is noted on abdominal CT. (B) Long segmental hypermetabolism (SUV, 6.0) is noted on 18-fluorodeoxyglucose PET.
bleeding and small intestinal mass, double-balloon enteroscopy (EN-450T5; Fujinon, Saitama, Japan) was performed via oral route and showed multiple small shallow ulcerations, edematous villi and fusion of villi (Fig. 2). Push enteroscopy with conventional colonoscopy via oral route (CF-H260AL; Olympus, Tokyo, Japan) showed mosaic and fissured like villi and more prominent nodularity mucosal pattern with narrow band imaging (NBI) and indigocarmin spray (Fig. 3). The biopsy result was mature T-cell lymphoma. Immunohistochemical staining showed CD3 (+), CD8 (+), CD56 (+), TLA-1 (+), CD4 (-), CD5 (-), CD20 (-), and CD30 (-) (Fig. 4). On bone marrow examination, there was no abnormality.

The patient was treated with rituximab, cyclophosphamide, adriamycin, vincristine, and prednisone (R-CHOP) chemotherapy and radiotherapy. Six months after the initial diagnosis, the patient underwent an operation because of intestinal obstruction with perforation and died from sepsis.

**DISCUSSION**

Mean age of diagnosed ETL was 60 years old and sex ratio of ETL was about 1:1 (male:female). ETL commonly arises in the proximal jejunum. But, less frequently, ETL can arise anywhere in the small intestine, stomach and colon. About 40% of ETL patients have acute clinical manifestations such as acute peritonitis due to perforation and intestinal obstruction which needs emergent operation. Other clinical manifestations are abdominal pain, weight loss and protein losing enteropathy. In early disease, ETL is localized in gastrointestinal tract or mesenteric lymph node. As progression of disease, it can spread to liver, spleen, skin, or other organs. Prognosis of ETL is very poor. Five-year survival rate is 20% to 25%.

Few endoscopic findings of ETL were reported by enteroscopy and capsule endoscopy. Typical endoscopic findings of ETL are multiple shallow ulcerations and diffuse thickening of mucosa with innumerable coarse or fine granular elevations (mosaic mucosal pattern). No articles have been published about NBI of ETL. In our case of ETL, more prominent mucosal nodularity and scalloping was observed with NBI and indigocarmin spray. The specimen of ETL endoscopic mucosal biopsy shows that mucosal edema and villous distortion by tumor cell infiltration (Fig. 3). This is a reason of nodularity with NBI and indigocarmin spray.

There is no definite treatment guideline of ETL. The surgery for ETL has poor outcomes and its role is limited to debulking and/or treatment of ETL complications such as perforation and obstruction. Radiation therapy has been indicated for bulky disease or incomplete resection. Chemotherapy such as vincristine,
doxorubicin, methotrexate, and prednisolone (VAMP); vincristine, doxorubicin, and prednisolone (VAP); CHOP; procarbazine, etoposide, prednisolone, and doxorubicin (PEPA); cyclophosphamide, etoposide, vincristine, and prednisone (CEOP); prednisolone, cytarabine, lomustine, etoposide, and thioguanine-bleomycin, vincristine, and methotrexate (PEACE-BOM); and surgery are options for treatment of ETL. But, more than half were unable to complete their planned chemotherapy courses and early treatment failure is frequently occurred because of malnourished state and complications of disease or chemotherapy. Overall response rate of treatment is 58%. But 16% of patients died after first cycle of chemotherapy. Mean recurrence time after chemotherapy or surgery was 6 months.

Recently, some clinical trials for feasibility and activity of high-dose chemotherapy supported by autologous stem cells transplantation (ASCT) as therapeutic option for ETL in small series and retrospective studies. In these clinical trials, ASCT could increase overall survival rate, even up to 60%. There are two reports of chemotherapy with ASCT about prevention of ETL and reduction in aberrant T-cells in patients with refractory celiac disease.

Alemtuzumab, humanized anti-CD52 monoclonal antibody currently used in treatment of chronic lymphocytic leukemia or T-cell lymphoma, can be another option for ETL. Alemtuzumab can treat ETL successfully with chemotherapy. But, all ETL cases who were treated by alemtuzumab were only two. There is the report about increased risk for ETL with using this monoclonal antibody for treatment of refractory celiac disease. So, more clinical studies are needed for alemtuzumab.

In conclusion, ETL is a very rare intestinal tumor. The endoscopic characteristics are edematous villi, fusion of villi, and multiple shallow ulcerations. In chromoendoscopy, indigocarmine and NBI, prominent mucosal nodularity, fissured or mosaic patterns are the characteristics of ETL. There is no treatment guideline for ETL. But, recently, molecular target agent such as alemtuzumab and ASCT can be the treatment options for ETL instead of conventional chemotherapy with poor outcomes.

CONFLICTS OF INTEREST

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

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