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# An unusual presentation of EATL type 1: Emergency surgery due to life-threatening gastrointestinal bleeding

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ABSTRACT

*INTRODUCTION:* Enteropathy-associated T-cell lymphoma (EATL) is a very rare malignancy. Reasons for hospital admission are variable.
*PRESENTATION OF CASE:* 76 years old man admitted to emergency service with sudden and massive obscure gastrointestinal bleeding. There was no complaints in his history. After initial evaluation, emergency laparatomy had to be done. Bleeding lesion in proximal jejunum was resected. Histopathologically, the muscularis propria had abundant atypical lymphoid infiltrate in diffuse pattern. Atypical lymphoid cells expressed CD3 and CD30. The jejunal mucosa adjacent to the tumor showed effacement of normal villous architecture.
*DISCUSSION:* EATL is known to cause anemia as a result of chronic bleeding. However in this case, the bleeding was abundant, irreplaceable and requiring emergency surgery. To our knowledge it is not reported previously.
*CONCLUSION:* A sudden and massive gastrointestinal bleeding can be the first and unique sign of EATL.


## 1. Introduction

Enteropathy-associated T-cell lymphoma (EATL) is an intestinal tumor arise from intraepithelial T-cells.[1] It is a very rare disease with the annual incidence rate is 0.5 to 1 per million.[2]

EATL is strongly associated with celiac disease (CD).[1,3–6] Although CD is a long standing disease, EATL is more often preceded by a short history of adult CD diagnosed after 50 years of age. In some cases, there is no history of CD, but histopathologic features of it are found in the tissue surrounding the tumor after resection.[1,7] In 2008, two types of EATL are introduced into the World Health Organization (WHO) classification of tumors of hematopoietic and lymphoid tissues.[8] The distinction between two types is made according to morphologic and genetic features. Type 1 EATL is more frequently associated with celiac disease.[1,7–10]

Non-specific signs and symptoms can be seen in patients with EATL. Sometimes occult or overt gastrointestinal bleeding or bowel perforation can be reason of hospital admission. Gastrointestinal bleeding seen in patients with lymphoma is usually insidious and it is manifested by iron deficiency anemia.[1,10,11] To our knowledge, in these patients, sudden and massive gastrointestinal hemorrhage has not reported previously. In this article, we report the very unusual case of EATL which manifested by sudden and life-threatening massive gastrointestinal bleeding, without previously known CD.

## 2. Case report

A 76-years-old male admitted to emergency service with sudden and severe gastrointestinal bleeding and hypovolemic shock. There were massive hematochezia and hematemesis. There is no history of celiac disease in his/family history. Systolic arterial blood pressure was 60 mmHg, diastolic was unmeasurable. The patient was monitored and resuscitation were started immediately. Crystalloids and erythrocyte suspensions were given intravenously. After the initial resuscitation hemodynamic stability was provided and then the patient transferred to Intensive Care Unit (ICU). The hemoglobin value increased from 4 mg/100 mL to 8 mg/100 mL. Emergency upper and lower gastrointestinal endoscopy revealed no lesion or active bleeding. There was blood clots in the colon and stomach. A short time later, second severe bleeding was observed in ICU. Despite the resuscitation and transfusions, the patient re-entered the hypovolemic shock. We decided to emergency laparatomy in doubt.

In exploration there was an adhesion between the transverse mesocolon and the jejunum just after the ligament of Trietz. We divided the adhesion. There was a perforated lesion that had been bleeding, on antimesenteric surface of jejunum (Fig. 1). Perforated

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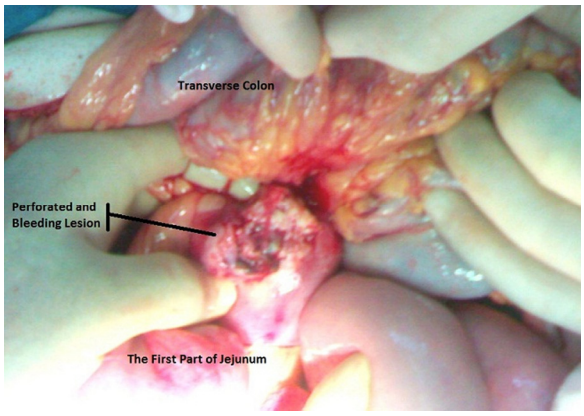


**Fig. 1.** Perforated and actively bleeding tumor on jejunum.

area was attached to adjecent intestine so that there was no blood in peritoneal cavity. We resected the lesion with margins of a few centimeters of healthy jejunum in both sides. End-to-end anastomosis was performed.

In histopathological examination, macroscopically, there were traces of bleeding and almost complete necrosis. Microscopically, intestinal mucosa showed extensive ulceration and necrosis. Extensive necrosis made the diagnosis very hard. But the following findings led us to the diagnosis of EATL type 1. The muscularis propria had abundant atypical lymphoid infiltrate in diffuse pattern (Fig. 2a). Neoplastic lymphocytes were medium to large sized cells with irregular nuclear contours and hyperchromatic nuclei. Some large cells with vesicular nuclei had prominent nucleoli (Fig. 2c). The jejunal mucosa adjacent to the tumor showed effacement of normal villous architecture (villous atrophy, crypt hyperplasia, intraepithelial lymphocytosis, pyloric metaplasia) (Fig. 3a and b). Immunohistochemically, atypical lymphoid cells positive for CD3 (Fig. 2b) and CD30 (Fig. 2d). They were immunonegative for CD56, CD4, CD8, CD20, ALK1. The background population was composed of reactive B and T lymphocytes. The intraepithelial lymphocytes in the adjacent mucosa were positive for CD3 and negative for CD4 and CD8.

The patient was discharged after surgical healing and referred to hematology clinic in eighth day postoperatively. The patient received combined chemotherapy regimen composed of bleomycin, doxorubicin, cyclophosphamide, vincristine, and prednisone. Since then he has been followed up by 18F-fluorodeoxyglucose positron emission tomography. The patient is alive in twenty-fourth month after diagnosis, there were no

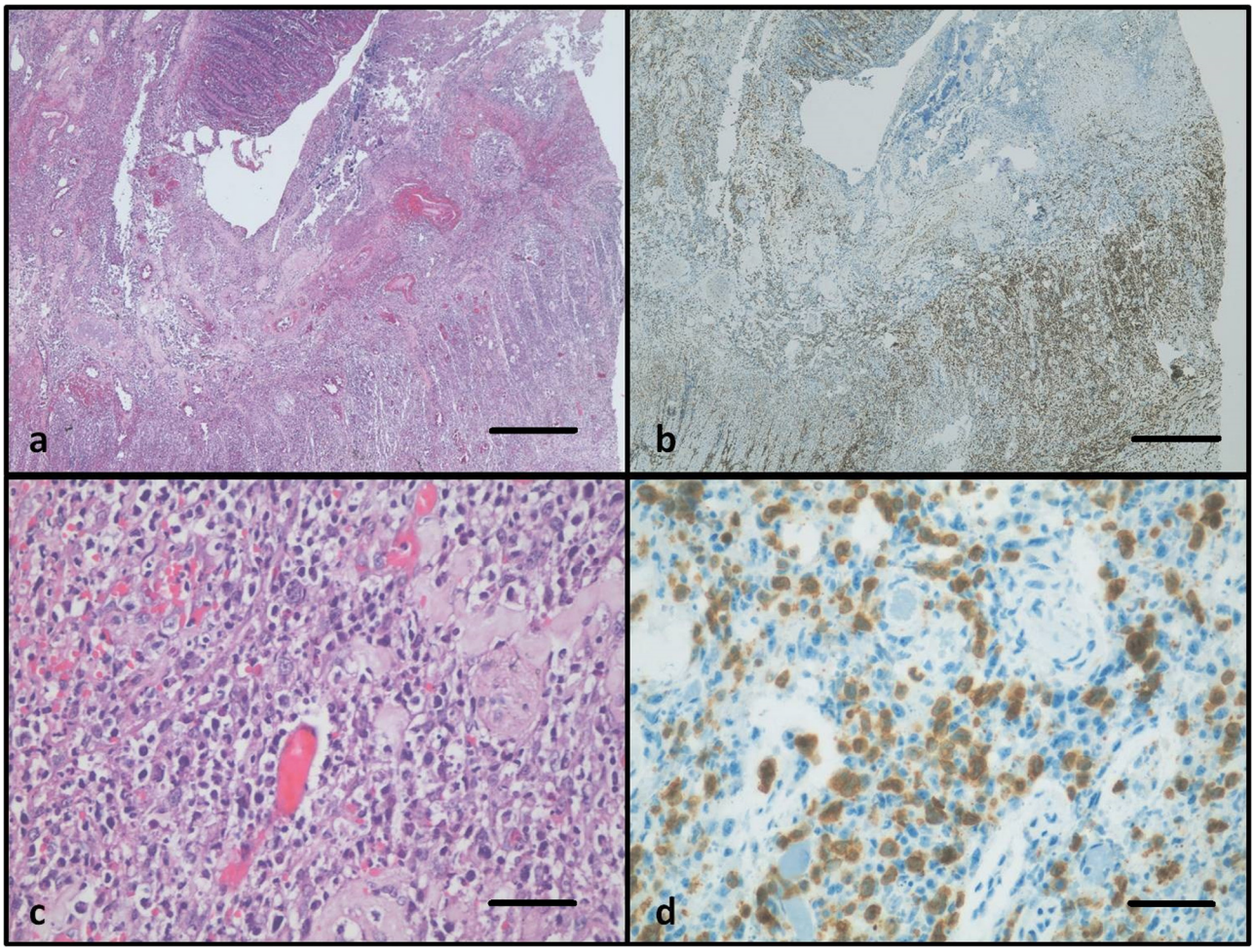


**Fig. 2.** (a) Extensive mucosal ulceration and diffuse lymphoid infiltration in submucosal and muscular layer of jejunal wall (hematoxylin-eosin, original magnification 20×). (b) Immunohistochemically, tumor cells are reactive for CD3 (immunoperoxidase, original magnification 20×). (c) At high power magnification, variably sized atypical pleomorphic lymphocytes within a necrotic background (hematoxylin-eosin, original magnification 400×). (d) The neoplastic cells commonly express CD30 (immunoperoxidase, original magnification 400×). Scale bar = 100 μm.