



## Enteropathy-Associated T-cell Lymphoma: A diagnostically challenging complication of celiac disease

Ahmet Faruk Ağan<sup>1</sup>, Ahmet Yılmaz<sup>2</sup>, Cem Cahit Barışık<sup>3</sup>

### Dear Editor;

Enteropathy-associated T-cell Lymphoma (EATL) is a rare, but fatal complication of celiac disease. EATL may occasionally present simultaneously with celiac disease. In most cases, the clinical features of EATL are very similar to celiac disease in the initial stages, which usually leads to a late diagnosis. EATL is primarily observed during the sixth decade of life in patients with celiac disease who have no diet restrictions. In this article, we describe the diagnostic features of a case of EATL that was identified in a patient with celiac disease.

A 72-year-old woman, presenting with a 7-month history of abdominal pain, vomiting, and weight loss, was diagnosed with celiac disease. She was unable to maintain a state of wellbeing, despite following a strict diet. Physical examination revealed cachexia. Hemoglobin level was 11 g/dL. The findings of gastro-duodenal biopsy, abdominal ultrasonography, and computed tomography (CT) appeared normal. The findings of small bowel barium-graphy (SBBG) were non-specific. However, CT enteroclysis (CTE) showed a narrow 10-mm segment at the distal ileum, with few adjacent micro-lymph nodes. Laparoscopy indicated 6 foci of lymphoma, including 3 in the jejunum and 3 in the ileum.

Approximately 40% of EATL cases are diagnosed in the emergency room and present with perforation, bleeding, or ileus (1). Early diagnosis is crucial for improving survival chances to a few months, and the diagnostic method used is also critical.

Positron emission tomography (PET) and double balloon endoscopy have been suggested as the most reliable diagnostic methods, obviating the need for a laparotomy in the last years. PET can identify the lymphoma in the attached segments (2). Double balloon enteroscopy is the optimal non-surgical modality for the diagnosis of EATL, with the advantage of being able to obtain a biopsy specimen (3); however, the major complications

with this technique include acute pancreatitis and perforation.

Capsule endoscopy may also be used for the diagnosis of EATL; however, the major disadvantages include capsule impaction and inability to obtain biopsy, whereas a high false-negative result rate of this procedure is another disadvantage (4). CTE may enable the detection of tumors as small as 5 mm, with a sensitivity and a specificity of 95-100%. Magnetic resonance imaging (MRI) has a limited role in the diagnosis of EATL. EATL may not be identified with duodenal endoscopic biopsy, CT, ultrasonography, and SBBG (5).

In this case, the clinical presentation of EATL was not apparent, and a diagnosis was therefore difficult. The difference between the CTE and laparotomy findings reflects the diagnostic challenge this condition presents. We believe that the most effective diagnostic modalities available should be employed in such cases, to achieve the best possible chance of survival.

### REFERENCES

1. Yang Y, Batth SS, Chen M, et al. Enteropathy-associated T cell lymphoma presenting with acute abdominal syndrome: a case report and review of literature. *J Gastrointest Surg* 2012;16:1446-9.
2. Chandesris MO, Malamut G, Verkarre V, et al. Enteropathy-associated T-cell lymphoma: A review on clinical presentation, diagnosis, therapeutic strategies and perspectives. *Gastroenterol Clin Biol* 2010;34:590-605
3. Hadithi M, Peña AS. Current methods to diagnose the unresponsive and complicated forms of coeliac disease. *Eur J Intern Med* 2010;21:247-53
4. Lewis BS, Eisen GM, Friedman S. A pooled analysis to evaluate results of capsule endoscopy trials. *Endoscopy* 2005;37:960-5
5. Malamut G, Chandesris O, Verkarre V, et al. Enteropathy-associated T cell lymphoma in celiac disease: A large retrospective study. *Dig Liver Dis* 2013;45:377-84

Istanbul Medipol University, Medical School, Departments of Gastroenterology<sup>1</sup>, General Surgery<sup>2</sup> and Radiology<sup>3</sup>, Istanbul, Turkey

Received: 14.05.2015, Accepted: 07.07.2015

Correspondence: Ahmet Faruk Ağan Haydarpaşa-Harem Yolu, Kadıköy 34718 Istanbul. Tel : 02165446630 Fax: 02163394444 E-mail: afagan@medipol.edu.tr