

## Enteropathy-associated T-cell lymphoma (EATL) without coeliac disorder: a challenging diagnosis of a poor prognosis disease

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### Background

Enteropathy-associated T-cell lymphoma (EATL) is a rare gastrointestinal non-Hodgkin's lymphoma, which is often related to gluten sensitive enteropathy. In patients without a prior history of coeliac disease this is a very rare disorder, and the diagnosis in such cases is often difficult and delayed due to the non-specific nature of the symptoms and a very low index of clinical suspicion. Unfortunately EATL has generally poor prognosis due to high risk of perforation with high mortality rate. The most common presenting symptoms are abdominal pain, weight loss and diarrhoea. Patients may also present with complications such as small bowel perforation or obstruction.

### Case History

We report a case of a 58-year-old male with symptoms of persistent diarrhea, rectorrhagia and abdominal pain for the past 6 months with evening low-grade fever. He revealed a history of previous strangulated hernia with small bowel obstruction occurred twenty seven years before. One month before hospitalization a colonoscopy was performed, showing normal colonoscopic findings. He had no history of coeliac disorder.

At time of our observation, he had ill-looking appearance with alert mental status. The vital signs were normal, with increased bowel sound and diffuse abdominal tenderness.

The laboratory findings revealed white blood cell count 3000/mm<sup>3</sup>, Hb 7,2 g/dL, and Plt 249×10<sup>3</sup>/mm<sup>3</sup> with peripheral blood morphology, showing normocytic normochromic anemia. In liver chemistry, aspartate aminotransferase was increased to 74 IU/L, and C-reactive protein 2,83 mg/dL. Laboratory findings for autoimmune disease and viral screening were normal. Serological testing for *immunoglobulin A tissue transglutaminase (IgA-based*

*TTG)* was normal.

Abdomen CT scan was performed and showed segmental wall thickening of ileal tract and hydropneumoperitoneum indicating bowel micro-perforation. Multiple mesenteric lymphadenopathies were found. The patient underwent emergency laparotomy, which showed small bowel perforation and tenacious adhesions. A 30cm long ileum resection was performed. Histologic findings showed high-grade T-cell Non-Hodgkin's lymphoma of the small bowel: EATL. Post-operatively, the patient's recovery was complicated by steady deterioration. Despite broad spectrum antibiotic therapy the patient continued to decline with further weight loss, and high temperatures. Unfortunately the patient expired two weeks after the operation.

### Discussion

Intestinal T-cell lymphomas affect less than 1 per million people per year. It is associated with coeliac disease, but it can also occur in patients with no prior history of coeliac disorder as in this case, making diagnosis more difficult. However, even with prompt diagnosis, outcomes for this condition remain poor due to deteriorated performance status of the patients at the time of presentation.

### References

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2. Wali GN (2015) A rare but potentially fatal cause of diarrhoea and weight loss: enteropathy-associated T-cell lymphoma. *BMJ Case Rep* 7.