The Uncommon Acute Abdomen: Enteropathy-Associated T-Cell Lymphoma Revealed by Jejunal Perforation



INTRODUCTION

- Long-standing Celiac Disease (CD) that is untreated or refractory to gluten-free diet can have dire consequences.
- Enteropathy-associated T-cell Lymphoma is a rare (<1%) form of non-Hodgkin's intestinal lymphoma that carries a poor prognosis with high mortality.
- EATL may manifest as **complicated small bowel disease** such as **obstruction or perforation**.
- We highlight a Celiac Disease patient presenting with an acute abdomen found to have jejunal perforation due to EATL.

CASE PRESENTATION

- 51-year-old male with history of CD non-compliant with glutenfree diet and peptic ulcer disease presented with acute abdomen.
- Abdominal CT revealed closed loop obstruction and pneumatosis with hollow viscus perforation. Urgent ex-lap discovered large volume purulent fluid and a perforation in the mid-jejunum which was resected.
- Biopsy of the resected jejunum revealed malignant lymphoid infiltrate in a background of mild villous blunting, increased intraepithelial lymphocytes, and ulceration. This pathology was consistent with EATL.

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Jejunal perforation (yellow arrows) see on CT A/P in Coronal and Axial views.



CASE PRESENTATION (cont.)

- chemotherapy.
- transplant.
- failure. Transitioned to comfort care.

 - abdomen.





• T-cell receptor rearrangement study was **negative** and bone marrow biopsy showed **no involvement**.

• Initiated on **BV-CHOP** therapy by Oncology. PET/CT showed complete remission of lymphoma after 6 cycles of

After induction chemotherapy, patient opted for consolidation (BEAM) chemotherapy followed by autologous bone marrow

• Unfortunately, treatment course was complicated by neutropenic fever leading to septic shock and multi-organ

DISCUSSION

Strict gluten-free diet (GFD) is the **primary** prevention of EATL in CD patients. Non-compliance significantly increases risk of developing EATL.

Small bowel perforation is a rare, but welldocumented complication of EATL that **must be** recognized in any CD patient presenting with acute

 Current treatment of EATL with chemotherapy and autologous bone marrow transplant has shown promising increase in survival rates.