Enteropathy-Associated T-Cell Lymphoma: A Rare Case Presenting as Acute Abdominal Pain

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[ABSTRACT]

Enteropathy-Associated T Cell Lymphoma (EATL) is a rare malignancy with annual incidence of 0.5–1 per million people and typically manifests with symptoms like chronic diarrhea, weight loss, and malabsorption (1). In this case report, we describe an unusual presentation of EATL in a 49-year-old otherwise healthy gentleman who presented with sudden-onset severe abdominal pain and vomiting. Despite the absence of classical symptoms, clinical examination revealed signs of peritonitis, prompting further investigations. Chest X-ray confirmed the presence of free gas under the diaphragm, leading to the necessity of an emergency laparotomy. Intraoperatively, a mobile mass was discovered, and surgical intervention was performed. Histopathological examination confirmed T cell lymphoma, emphasizing the importance of considering rare malignancies in the differential diagnosis of acute abdominal pain.

[KEYWORDS]

Enteropathy-Associated T Cell Lymphoma (EATL), Acute Abdominal Pain, Peritonitis, Gastrointestinal Perforation, Surgical Intervention

[KEY CLNICAL MESSAGE]

Enteropathy-Associated T-Cell Lymphoma (EATL) can present atypically as acute abdominal pain, complicating diagnosis. Clinicians should maintain a high index of suspicion for rare malignancies in emergency settings, even in patients without classical symptoms. Timely surgical intervention is crucial for effective management and improved patient outcomes.

[INTRODUCTION]

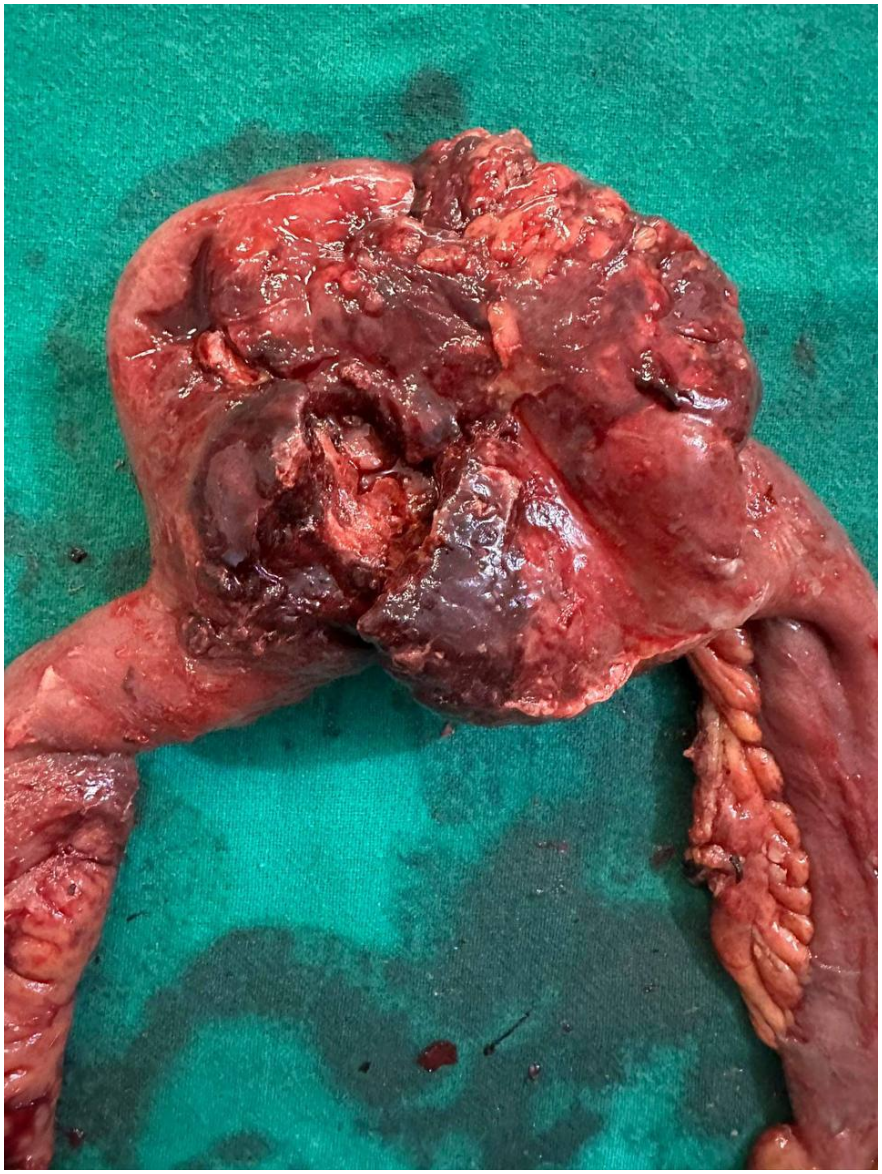
Gastric cancer is the one of the most common and lethal cancer worldwide (2). 95% of Gastric cancers of are adenocarcinomas, followed by primary gastric lymphoma (2). Primary Gastric lymphoma are approximately 5% of gastric neoplasm (3). T-cell lymphomas are types of non-Hodgkin lymphoma (NHL) which can grow in lymphoid tissues such as the lymph nodes and spleen, or outside of lymphoid tissues (i.e., gastrointestinal tract, liver, nasal cavity, skin, and others) (4).

Enteropathy-associated T-cell lymphoma (EATL) is a rare primary extra nodal T-cell lymphoma primarily affecting the small intestine (5,6). It is an aggressive T-cell lymphoma. EATL develops most frequently in the middle part of the small intestine, called the jejunum (7). The cause of EATL is not fully understood yet by definition is related to celiac disease (5,6). Classically presenting with chronic diarrhea, weight loss, and malabsorption, EATL poses a diagnostic challenge when presenting with atypical symptoms such as acute abdominal pain without the typical constitutional manifestations.

[CASE HISTORY AND EXAMINATION]

A 49-year-old otherwise healthy gentleman presented to the emergency department with sudden-onset severe abdominal pain, nausea and vomiting. He had no history of fever, anorexia, weight loss, abdominal distension or jaundice. Vitals were stable. Per abdominal examination revealed localized tenderness, signs of peritonitis, prompting further investigations. Bowel sounds were heard and no additional observations were noted in the systemic examinations. On Laboratory investigations full blood count, renal function test, liver function test, serum amylase, lipase, PT-INR were all within normal limits.

Chest X-ray was done which showed the presence of free gas under the diaphragm suggestive of gastrointestinal perforation, necessitating emergency laparotomy. Bowel resection with anastomosis was performed and a significant finding was identified intraoperatively. A mobile mass, measuring approximately 10 by 10 centimeters in size, was discovered within the abdominal cavity which was situated 250 centimeters proximal to the ileocecal junction and was found to be covered by omentum (Figure 1).



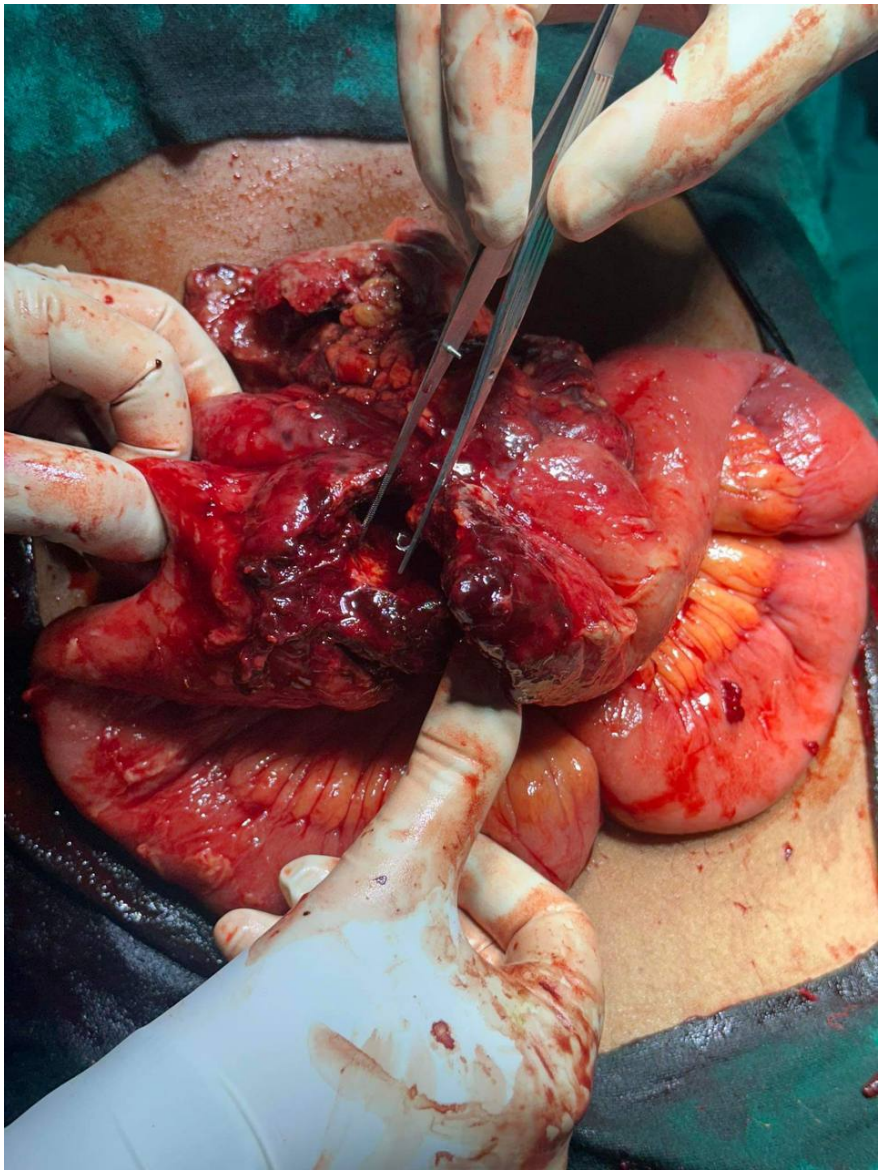
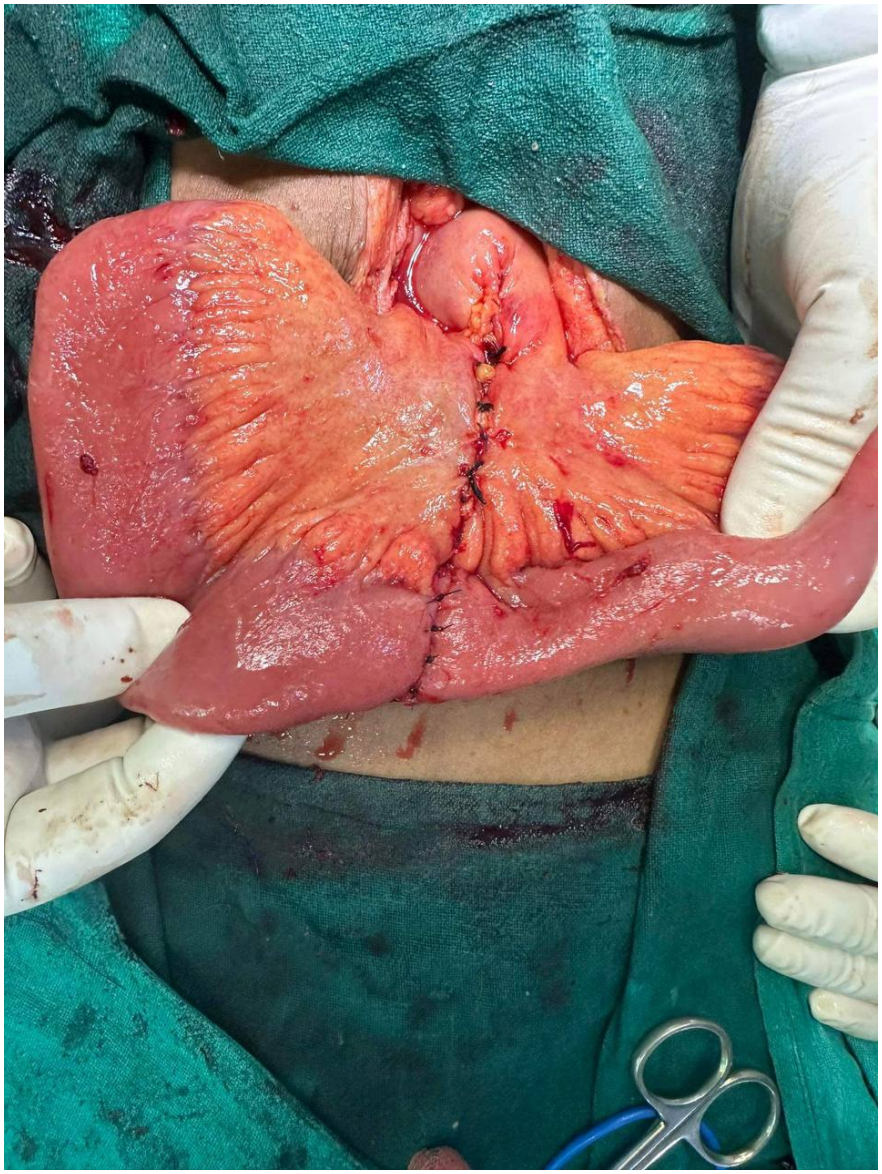
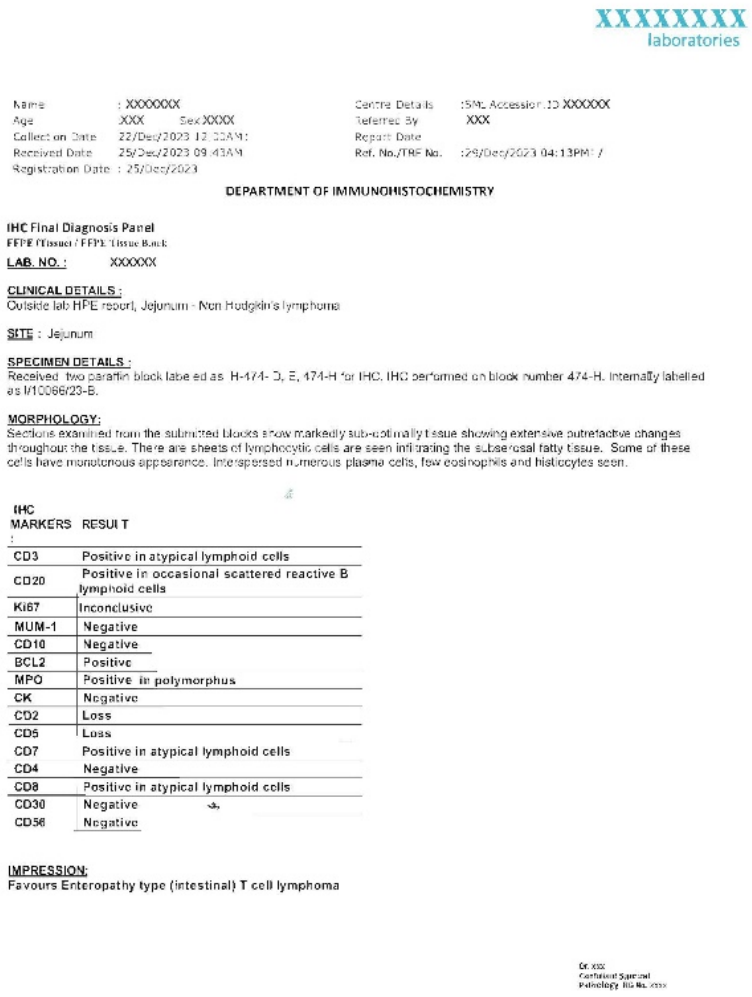


Figure 1: Intra-operative image showing tumor with perforation at the center

Figure 2: Resected tumor segment with 10 cm margin both proximally and distally

Further examination revealed a rent, or tear, leading to a moderate accumulation of fluid within the abdomen, indicative of a potential bowel perforation. To address this issue, the affected segment of the bowel loop was resected, ensuring a 10-centimeter margin both proximally and distally to ensure removal of compromised tissue (Figure 2). Following the resection, the remaining healthy ends of the bowel were carefully joined together using a hand-sewn anastomosis technique, creating a new connection between the two segments of bowel aiming to restore bowel continuity and minimizing the risk of postoperative complications such as leakage or strictures (Figure 3).





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DEPARTMENT OF IMMUNOHISTOCHEMISTRY

IHC Final Diagnosis Panel
FFPE (Tissue / FFPE Tissue Block

LAB. NO. : XXXXXX

CLINICAL DETAILS :
Outside lab HPE report, Jejunum - Non Hodgkin's lymphoma

SITE : Jejunum

SPECIMEN DETAILS :
Received two paraffin block labe ed as H-474- D, E, 474-H for IHC. IHC performed on block number 474-H. Internally labelled as M10066/23-B.

MORPHOLOGY:
Sections examined from the submitted blocks show markedly sub-optimally tissue showing extensive putrefactive changes throughout the tissue. There are sheets of lymphocytic cells are seen infiltrating the subserosal fatty tissue. Some of these cells have monotonous appearance. Interspersed numerous plasma cells, few eosinophils and histiocytes seen.

IHC MARKERS	RESULT
CD3	Positive in atypical lymphoid cells
CD20	Positive in occasional scattered reactive B lymphoid cells
Ki67	Inconclusive
MUM-1	Negative
CD10	Negative
BCL2	Positive
MPO	Positive in polymorphus
CK	Negative
CD2	Loss
CD5	Loss
CD7	Positive in atypical lymphoid cells
CD4	Negative
CD8	Positive in atypical lymphoid cells
CD30	Negative
CD56	Negative

IMPRESSION:
Favours Enteropathy type (intestinal) T cell lymphoma

Dr. xxx
Consultant Specialist
Pathology Reg No. xxxx

Figure 3: Resection anastomosis of the perforated bowel segment.

Figure 4: Immunohistochemistry report

[METHODS]

The resected segment was sent for histopathological examination. On gross examination the specimen showed brown growth-like area measuring 10*9 cm. Microscopic examination of the segment showed sheets of lymphocytic cells infiltrating the subserosal fatty tissue where some of the cells had monotonous appearance with interspersed numerous plasma cells, few eosinophils and histiocytes (Figure 5).

Differential of Non-Hodgkin's Lymphoma was made and advised for Immunohistochemistry. Immunohistochemistry was conclusive of Enteropathy Type T-cell Lymphoma with strong presence of CD3 marker positive in atypical lymphoid cells, CD20 marker positive in occasional scattered reactive B lymphoid cells,

BCL2 positive, MPO positive in polymorphus, CD7 and CD8 positive in atypical lymphoid cells and negative for CD4, CD10, CD30, CD56, MUM-1, CK, CD2, CD5 (Figure 4).

The post-operative course was uneventful, and the patient was discharged on the 7th postoperative day.

The patient was subsequently referred to an oncologist for further management.

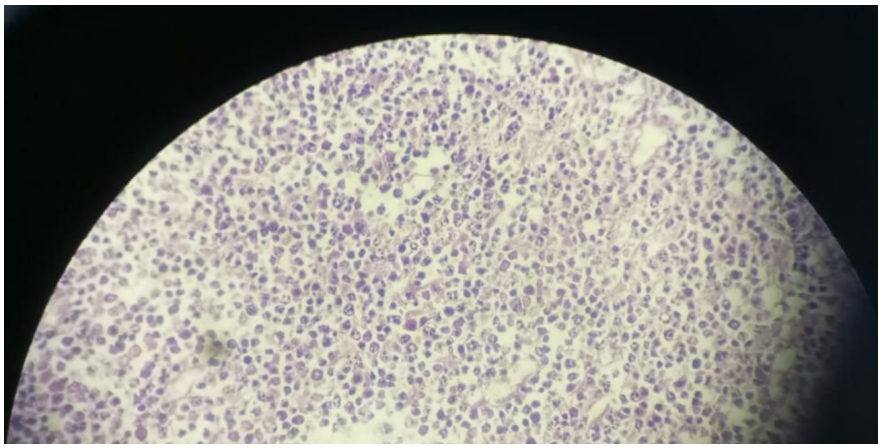

Figure 5: High power view of monotonous population of lymphoid cells suggestive of Non-Hodgkin Lymphoma.

[CONCLUSION]

This Case Report aids as a reminder of the diverse presentations of rare malignancies like Enteropathy-Associated T Cell Lymphoma. Early recognition and prompt surgical intervention played a fundamental role in optimizing outcomes for this patient. Clinicians should remain cautious to atypical presentations, ensuring timely and appropriate management for improved patient outcomes. Increased awareness of rare diseases with diverse clinical manifestations is essential for providing comprehensive and effective healthcare.

[DISCUSSION]

Enteropathy-associated T-cell lymphoma (EATL) is a rare form of intraepithelial T-cell intestinal lymphoma (1). The annual incidence rate is 0.5–1 per million people in Western countries and this is a rare form of malignancy, covering approximately 35% of all small bowel lymphomas (1). The pathogenesis of EATL is multifactorial, involving chronic inflammation and immune dysregulation within the small intestine, particularly in individuals predisposed to celiac disease. Most commonly affected area is small intestine (jejunum> duodenum/ileum) (7). Even though this kind of lymphoma is uncommon, it is one of the leading causes of mortality for adults with celiac disease (CD) (8).

In 1937, Farley and Mackie initially described the association between intestinal lymphoma and malabsorption (9). O'Farrelly coined the term 'Enteropathy associated T cell lymphoma' in 1986, establishing the close relationship between this lymphoma with villous atrophy of the jejunal mucosa adjacent to EATL (10).

World health organization(WHO) divided enteropathy associated t cell lymphoma(EATL) into two subtypes in 2008, into EATL type I and EATL Type II (11). EATL type I is typically associated with refractory celiac disease, which accounts for 80-90% of cases, and often presents with large-cell or pleomorphic cytology, with infrequent expression of CD8 and CD56 (12). While EATL type II, constituting 10-20% of cases, is sporadic, less commonly associated with celiac disease, and characterized by monomorphic cytology with frequent expression of CD8 and CD56 (13). In 2016 WHO redefined two diseases as, EATL Type I as EATL, and EATL type II as Monomorphic epithelia-tropic intestinal t cell lymphoma(MEITL) (14). EATL is 5-10 times more common than MEITL (15).

The common age group for EATL is 60's and 70's with similar prevalence in both genders (16). EATL most commonly develops in the jejunum but it can also develop in other parts of small intestine and colon (17). It may also spread to spleen, stomach, lymph node, liver and gall bladder (18). Despite its primary localization in the small intestine, EATL manifesting in the stomach, poses diagnostic challenges as it may mimic other gastric lesions such as peptic ulcers, leading to diagnostic confusion. Accurate diagnosis is crucial as EATL requires distinct management strategies compared to other gastric malignancies. Affected patient can present with similar symptoms of celiac disease like abdominal pain/distention, malabsorption, weight loss, night sweats, chronic recurrent diarrhea. Sometimes it can suddenly develop with serious symptoms of bowel perforation and/or bowel obstruction (16,19).

The atypical presentation of EATL as acute abdominal pain without classical symptoms underscores the need for a comprehensive differential diagnosis in emergency settings. While gastrointestinal perforation is an uncommon manifestation of lymphomas (20), this case emphasizes the importance of a timely surgical approach for both diagnosis and therapeutic intervention. Despite advancements in diagnostic techniques and treatment modalities, EATL remains a challenging entity to manage due to its rarity and heterogeneous clinical presentation. Therefore, increased awareness among healthcare professionals regarding the association between celiac disease and EATL is paramount for timely diagnosis and appropriate management. The exact mechanisms underlying the development of EATL remain poorly understood, highlighting the need for further research in this.

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[NOTES]

Conflict of interest

All authors declare that they have no potential conflicts of interest.

Patient consent

The patient involved in this study has granted consent for the publication of this case report.

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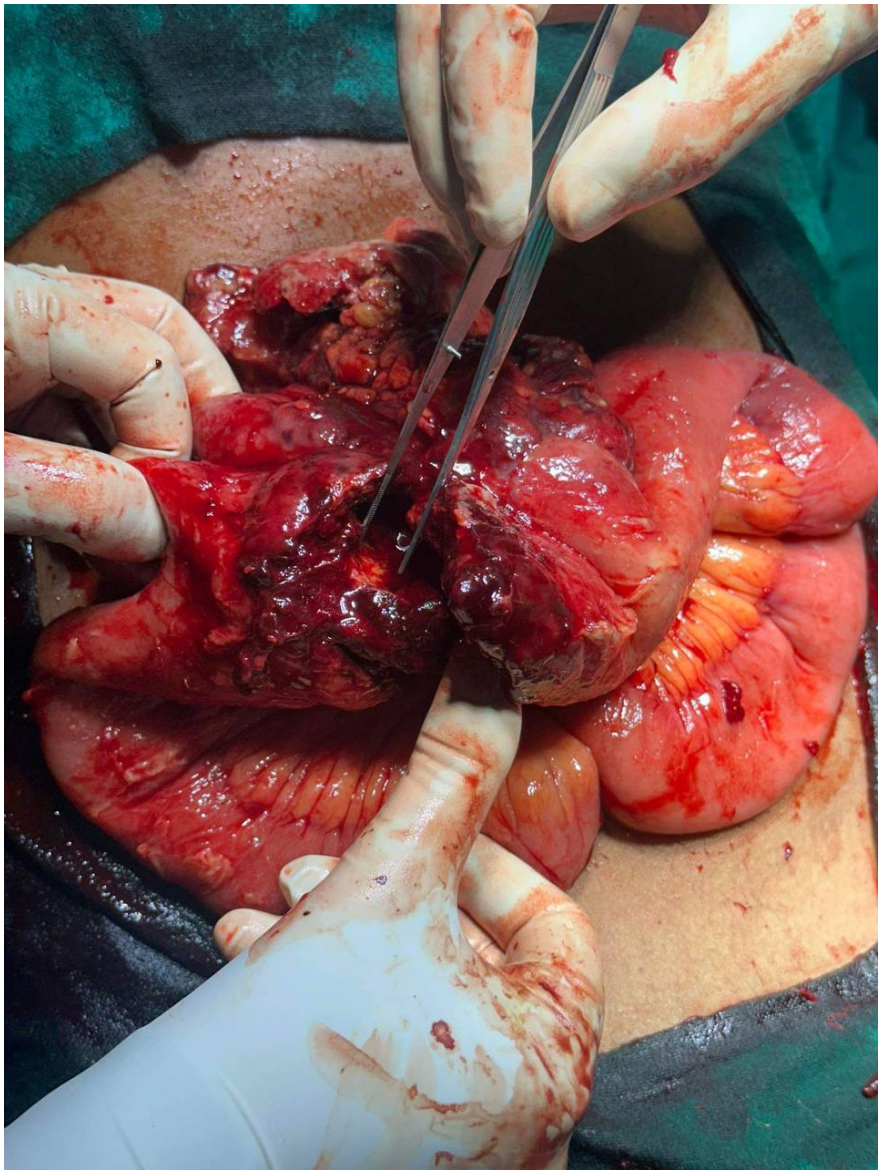
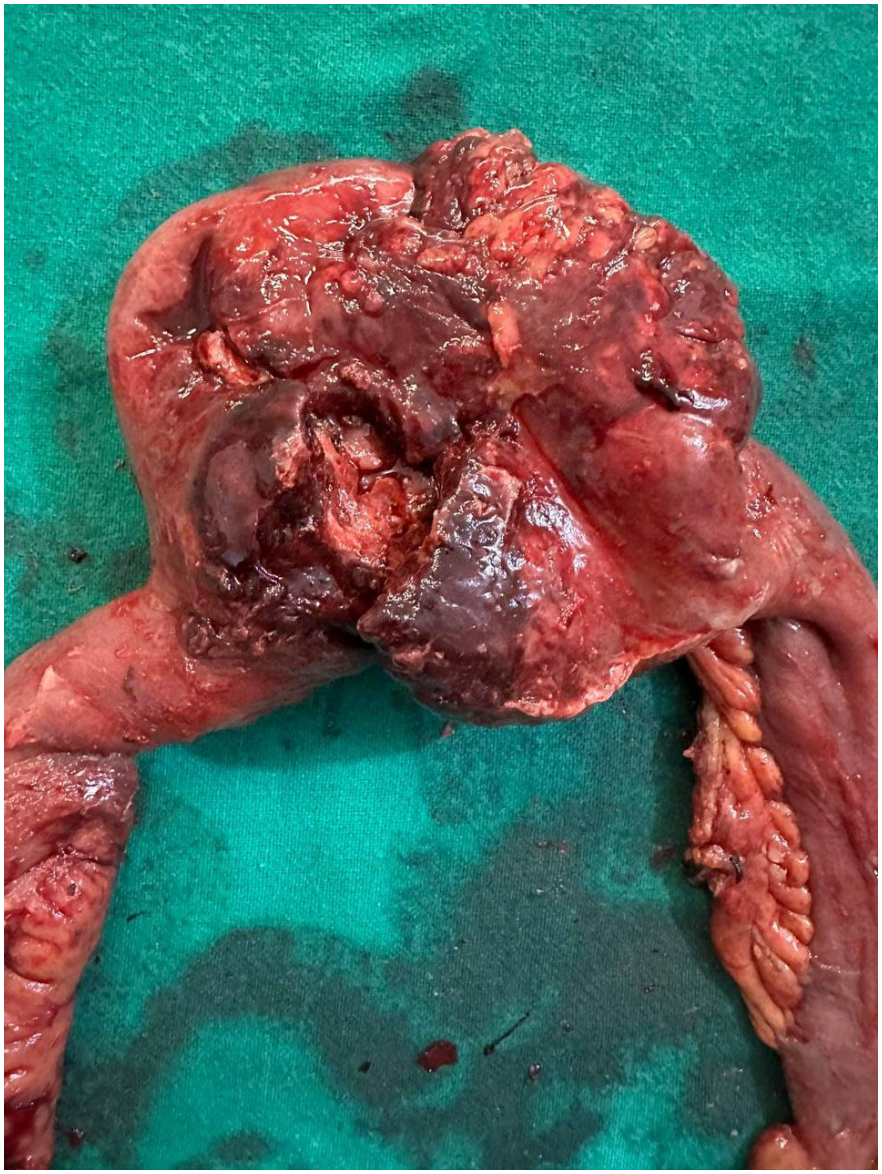
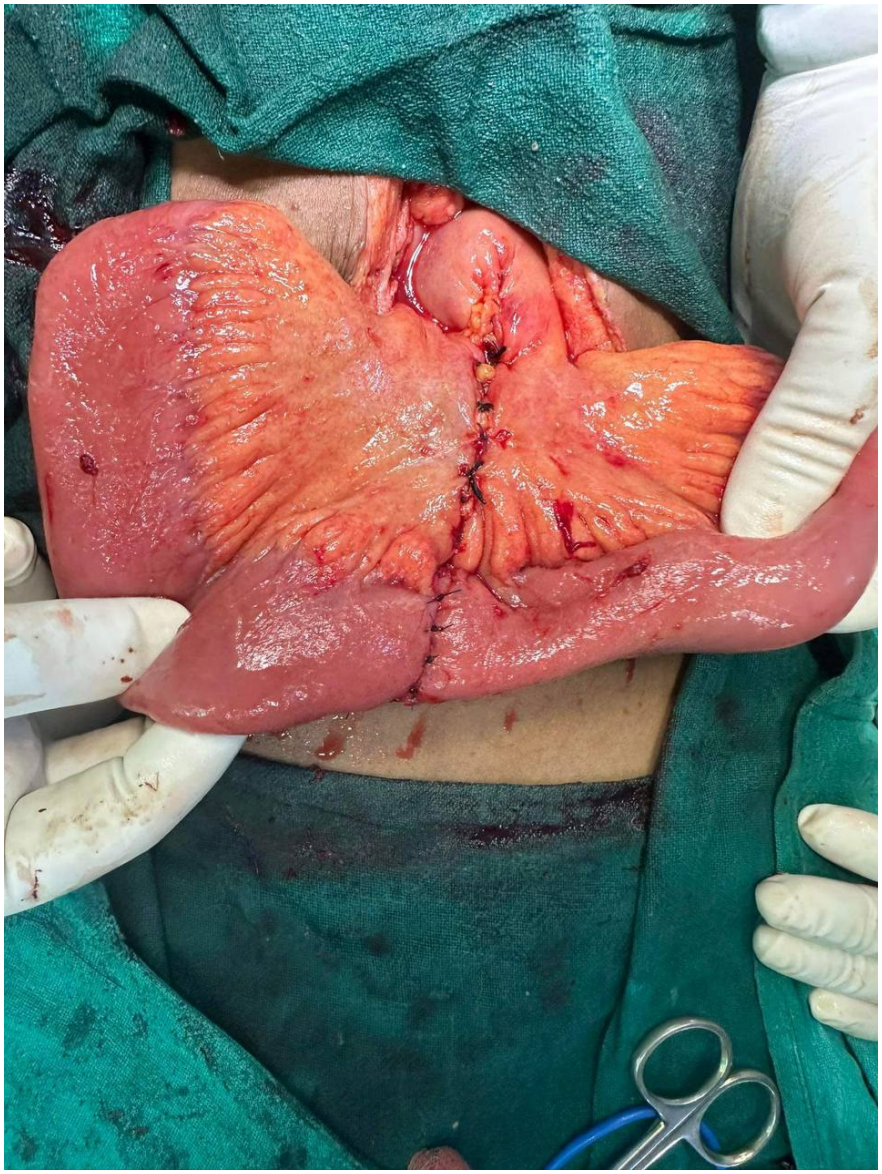
2. **Rupak Subedi**Data curation, Formal analysis, Methodology, Project administration, Software, Writing - original draft, Writing - review & editing
3. **Nischal Khanal** Conceptualization, Supervision, Writing - review & editing
4. **Nirajan Shrestha** Formal analysis, Resources, Validation
5. **Amardip Mandal** Project administration, Resources, Validation, Writing - review & editing
6. **Prajwal Sapkota** Methodology, Software, Visualization, Writing - review & editing
7. **Arjun Khadka** Funding acquisition, Supervision, Writing - review & editing
8. **Rasmi Tamang**Funding acquisition, Resources, Supervision, Writing - review & editing







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DEPARTMENT OF IMMUNOHISTOCHEMISTRY

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Favours Enteropathy type (intestinal) T cell lymphoma

Dr. xxx
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Pathology RG No. xxxx