DOI: 10.4274/gulhane.galenos.2024.02259
Gulhane Med J 2025;67(1):51-54

Acute abdomen with perforated viscus: A case of intestinal T-cell lymphoma, not otherwise specified

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Cite this article as: Lung WW, Zakaria AD, Yusoff MZM, Hussain FA. Acute abdomen with perforated viscus: a case of intestinal T-cell lymphoma, not otherwise specified. Gulhane Med J. 2025;67(1):51-54.

Date submitted:
30.11.2024
Date accepted:
08.12.2024
Epub:
30.01.2025
Publication Date:
20.02.2025

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Keywords: Small intestine, neoplasm, lymphoma, T-cell

ABSTRACT

Small bowel lymphoma is a rare disease with nonspecific presentation. Intestinal T-cell lymphoma, not otherwise specified, is an aggressive lymphoma type with a poor prognosis and may be widespread upon presentation. We report a case of a 72-year-old man who presented with abdominal pain and was diagnosed with a small bowel tumor. Small bowel resection with primary anastomosis was performed. We highlighted the importance of high clinical suspicion when diagnosing such disease and tumor aggressiveness.

Introduction

Primary gastrointestinal (GI) lymphoma accounts for about 1-4% of all GI malignancies (1). Approximately 90% of GI lymphoma cases are of the B-cell lineage, with the rest indicating Hodgkin lymphoma and T-cell lymphoma (2). World Health Organization 2022 classification classifies intestinal T-cell lymphomas as (I) enteropathy-associated T-cell lymphoma (EATL), (II) monomorphic epitheliotropic intestinal T-cell lymphoma, (III) indolent T-cell lymphoproliferative disorder of the GI tract, (IV) intestinal T-cell lymphoma, not otherwise specified (ITCL-NOS), and (V) indolent natural killer cell lymphoproliferative disorder of the GI tract. The prognosis

is different in each lymphoma type; therefore, prompt diagnosis and treatment are crucial to achieve a cure (3). Therapeutic management requires a multidisciplinary approach, and surgery is necessary in patients with complications like perforation or hemorrhage.

Case Presentation

A 72-year-old man with a past diagnosis of hypertension was admitted with fever and worsening abdominal pain for 2 days. The pain was colicky, localized on the left side, with no radiation. He had frequent colicky abdominal pain for 1 month, loss of appetite, and weight loss. He reported no recurrent fever or other GI symptoms. He was of medium build, had pallor, and was mildly dehydrated. The abdomen showed tenderness in the left hypochondrium with a vague palpable mass. The blood tests showed leucocytosis ($23x10^9$/L), anemia (8.3 g/dL), increased ALP (476 U/L), lactate dehydrogenase (LDH) (557 U/L, tumor markers like carcinoembryonic antigen, was normal (1.0 ng/mL). Computed tomography (CT) scan showed a heterogenously enhanced small bowel mass with multiple enlarged mesenteric lymph nodes (Figure 1). He was pre-diagnosed with a sealed perforated small bowel tumor.

The patient underwent exploratory laparotomy, which showed a small bowel tumor, a small perforation covered with slough, and multiple enlarged mesenteric nodes 30 cm from the duodenojejunal junction. Another intraluminal mass was noted 20 cm distally from the terminal ileum. Segmental bowel resection was performed on both tumor sides, with primary anastomosis; however, proximal enlarged mesenteric nodes could not be completely resected because they were fixed and close to the mesenteric vessels (Figure 2).

Post-operatively, his recovery was complicated by cardiac events and sepsis caused by hospital-acquired pneumonia. During approximately 1 month of hospital stay, he developed intestinal obstruction, and the CT scan showed an enlarged mesenteric node with tumor progression and obstruction in the small bowel. Unfortunately, he succumbed to illness 40 days after the initial surgery.

Histopathological assessment showed diffuse tumor infiltration within the submucosa, extending into the serosa and causing perforation in the jejunum. Both tumors were reported as aggressive intestinal T-cell lymphoma, not otherwise specified (ITCL-NOS). Tumor cells were positive for CD3, CD2, CD4, and CD8, perforin with high Ki67, and negative to CD20 and CD56 (Figure 3).

Consent to participate in this case report was provided by the patient's next of kin.

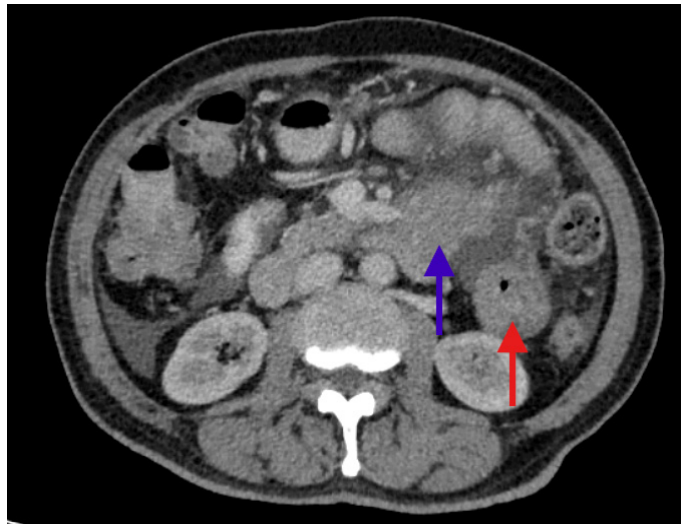

Figure 1. Preoperative contrast-enhanced computed tomography scan of the abdomen. The blue arrow denotes enlarged mesenteric nodes. The red arrow denotes an intraluminal heterogeneous mass at the proximal small bowel

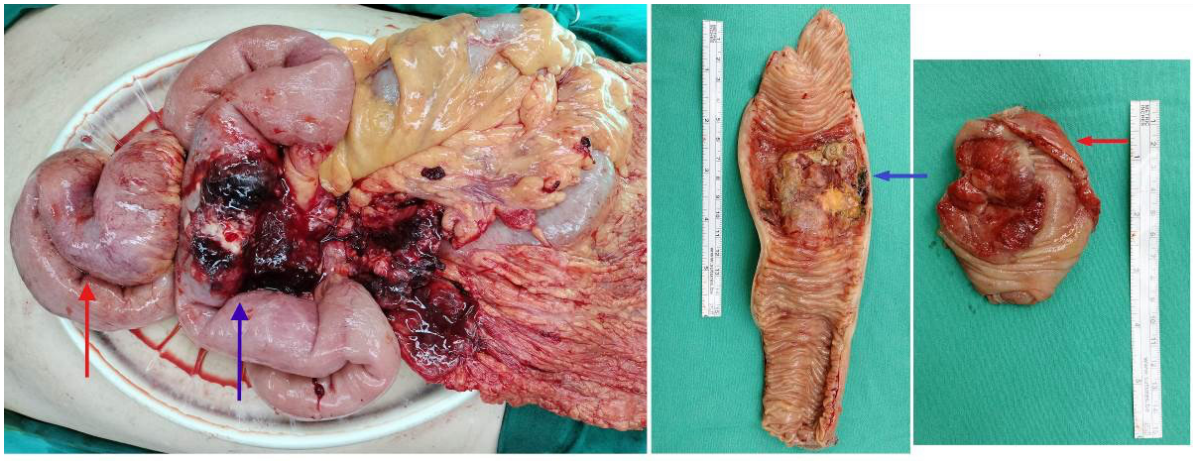

Figure 2. Left: Intraoperative findings during exploratory laparotomy. The blue arrow denotes the jejunal tumor site with multiple enlarged mesenteric nodes. The red arrow denotes the site of the intraluminal mass felt at the distal ileum. Right: Bivalved specimen showing an ulceroinfiltrative mass at the intraluminal bowel, Blue arrow: Jejunal specimen, Red arrow: Terminal ileum specimen

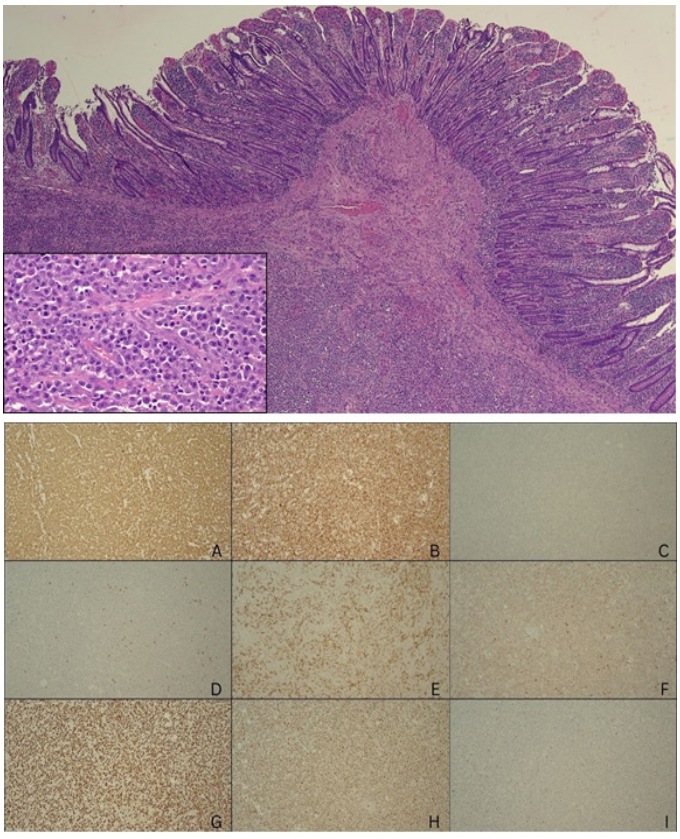

Figure 3. Above: The small bowel with intact mucosa. The tumor cells appear to push the muscularis mucosae without infiltration into the lamina propria of the villi in this field but not in other areas (H and E, 40x) that resulted in perforation (not shown). Inset: Tumor cells are pleomorphic, medium to large in size, irregular vesicular nuclei and large nucleoli, and moderate in amount of cytoplasm. Mitoses are easily seen (H and E, 400x). Below: Immunohistochemistry (x100): A-CD3 positive; B-CD2 positive; C-CD20 negative; D-CD5 negative; E-CD4 positive; F-CD8 positive; G-Ki67 (about 80-90%); H-Perforin positive; I-CD56 negative

Discussion

About 30-40% of all extranodal non-Hodgkin lymphomas occur in the GI tract (4). The most frequently affected sites are the stomach (50-60%), followed by the small bowel (20-30%) (4). Small bowel lymphoma is commonly discovered incidentally in asymptomatic patients, whereas others may present with intestinal obstruction, GI bleeding, perforation, and intussusception (4). Our patient presented with nonspecific abdominal symptoms like pain for a month before the symptoms worsened due to perforation.

Certain tumor subtypes display proclivity toward specific sites, such as mucosa-associated lymphoid tissue lymphoma in the stomach, mantle cell lymphoma (MCL) in the terminal ileum, jejunum, and colon; EATL in the jejunum; and follicular lymphoma in the duodenum (2). Small bowel lymphoma behaves more aggressively than gastric lymphoma (1). ITCL preferentially involves the jejunum with a higher tendency to cause perforation than others (2). In our patient, the tumor involved the jejunum and distal ileum, with complications of perforation at the proximal tumor site.

CT can be used to diagnose small intestinal lymphomas; however, it has a low specificity for small intestinal lesions (1). In our patient, the CT scan that was performed to evaluate the abdominal pain was able to detect a small bowel mass. However, there exist no pathognomonic features on CT to diagnose lymphoma based on imaging alone.

Surgical intervention for gastric and colon lymphoma should be delayed until tumor-related complications (5). The management of GI lymphoma requires a multidisciplinary approach of a radiologist, pathologist, once-hematologist, radiotherapist, and gastroenterologist (6). A previous study reported better prognosis in B-cell subtypes of non-Hodgkin lymphoma and those who underwent systemic chemotherapy and surgical resection compared with surgery alone (1). Hong et al. (5) reported higher morbidity and mortality rates in patients who underwent emergency surgery, but the current study showed no significant differences in early mortality or severe surgery-related complications.

Gross specimens of B-cell lymphoma tend to be fungating or ulcerofungating, whereas T-cell lymphoma tends to appear more ulcerative or ulceroinfiltrative (7). ITCL-NOS is a diagnosis of exclusion and refers to a heterogeneous group of T-cell lymphoma without specific morphologic or phenotypic criteria of other entities (3). They are usually aggressive and may present as widespread disease (3). In immunohistochemistry, tumor cells were positive for CD8 and negative for CD5 (7). As in our case, an aggressive ITCL-NOS diagnosis was made based on histopathological assessment, and other T-cell lymphoma subtypes were ruled out. In a series by Tian et al. (8), patients with prevalent B-cell pathological types had better long-term survival after surgery, whereas T-cell lymphoma and MCL had poor prognosis regardless of whether surgery before chemotherapy or emergency surgery was performed. Risk factors associated with survival included poor performance status, advanced disease stage, bulky disease, extranodal involvement, elevated LDH level, and higher Ki67 proliferative index, where their presence confers lower overall survival (7).

Conclusion

Small bowel lymphoma is rare and requires high suspicion because of a nonspecific presentation and symptoms. Presentation as an acute surgical complication is beneficial in localized disease. The present patient posed diagnostic challenges in small bowel lymphoma because of its rarity and nonspecific presentation.

Ethics

Informed Consent: Consent to participate was granted by the patient's next of kin.

Acknowledgments

We would like to thank Associate Professor Dr. Maya Mazuwin Binti Yahya, Head of the Department of Surgery, Hospital Universiti Sains Malaysia for her support and advice in completing this manuscript.

Footnotes

Authorship Contributions

Surgical and Medical Practices: W.W.L., A.D.Z., M.Z.M.Y., F.A.H., Literature Search: W.W.L., Writing: W.W.L., A.D.Z., M.Z.M.Y., F.A.H.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support.

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