Mucosa-associated lymphoid tissue lymphoma presenting as intestinal obstruction: a case report

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ABSTRACT

Background: Mucosa-associated lymphoid tissue (MALT) lymphoma is a rare variant of extranodal marginal zone B cell lymphoma occurring at mucosal sites. Gastric MALT lymphoma is well-described, but non-gastric MALT lymphomas, particularly small intestinal, ceacal, and appendiceal MALT lymphoma, are rare and present diagnostic problems. Intestinal MALT lymphoma is frequently diagnosed late and can be associated with nonspecific symptoms in patients.

Case presentation: A 69-year-old female with diabetes mellitus, hypertension, and hypothyroidism, admitted to the emergency room for 24 hours with colicky lower abdominal pain and vomiting, without bowel movements. Abdominal distension and right iliac fossa tenderness were noted on clinical examination. Imaging, however, suggested a closed-loop obstruction of a twisted segment of the ileum. A laparoscopic right hemicolectomy with resection and anastomosis was carried out on the patient. Extranodal marginal zone lymphoma of MALT type arising from the terminal ileum, cecum, and appendix with ischemic changes was identified by histopathological analysis of the resected specimen.

Conclusion: The importance of considering MALT lymphoma in the elderly with bowel obstruction, without an established chronic inflammatory or autoimmune condition, is emphasized in this case. Mechanical obstruction requires surgical resection, but systemic therapy is the treatment of choice for disseminated disease. Future research should address the role of molecular diagnostics and targeted therapies in non-gastric MALT lymphoma to enhance early detection and management programs.

Keywords: Extranodal marginal zone lymphoma, gastrointestinal lymphoma, intestinal obstruction, malt lymphoma, case report.

Type of Article: CASE REPORT **Specialty:** General Surgery

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Introduction

Mucosa-associated lymphoid tissue (MALT) lymphoma is a subtype of non-Hodgkin lymphoma in which B-cells develop from the MALTs' marginal zone. In 2016, 7,460 new cases were estimated in the US, representing approximately 7% of all mature non-Hodgkin lymphomas [1]. Most patients with MALT lymphoma are aged between 50 and 60 years, and the frequency is higher in patients aged over 40 years [2].

The most common MALT lymphoma site of involvement is the stomach, affecting about half of cases [3]. Other affected sites include the skin (11%), lung (14%), thyroid gland (4%), ocular adnexa (12%), salivary gland (6%), and neck and head regions (14%) [4]. However, MALT lymphoma can rarely occur in the small intestine, though it is much less common than the sites mentioned. Primary small intestinal MALT lymphoma accounts for <5% of cases (likely even rarer than thyroid or salivary gland involvement) [4]. Non-gastric MALT lymphomas are also common; notably, the ocular adnexa is the most common extranodal site (34%) [5].

The pathogenesis of MALT lymphoma is closely related to chronic antigenic stimulation by persistent infection or autoimmune disorders. For example, gastric MALT lymphoma is strongly associated with chronic infection with the gastric bacterium Helicobacter pylori, leading to lymphoid tissue proliferation and the potential for malignant transformation [6]. Infections with Chlamydophila psittaci have also been reported to cause ocular adnexal MALT lymphomas [7].

The clinical manifestation of MALT lymphomas depends on the anatomical site involved. Most often, gastric MALT lymphoma presents with nonspecific gastrointestinal symptoms, such as dyspepsia, epigastric pain, or bleeding [2]. Respiratory symptoms like cough and dyspnea might be present in pulmonary MALT lymphoma with or without being asymptomatic [8].

Painless orbital mass or swelling typically indicates the presentation of ocular adnexal MALT lymphoma [9].

In rare cases, MALT lymphoma can present as acute diseases, like gastric MALT lymphoma with multi-organ involvement has been described in 5.2% of cases and indicates that these patients should be carefully examined [10].

In general, the prognosis for MALT lymphoma is favorable. But prognosis depends on the primary site of involvement, stage of diagnosis, and presence of certain genetic abnormalities. Pulmonary MALT lymphoma, for example, has a higher frequency of disseminated disease compared to gastric MALT lymphoma, where disseminated disease is present in <25% of cases [11]. This case report details related to the presentation, diagnosis, and management of a patient with MALT lymphoma of the terminal ileum presenting with intestinal obstruction.

Case Presentation

A 69-year-old female with known diabetes mellitus, hypertension, and hypothyroidism, and left-eye blindness was admitted with a 1-day history of colicky lower abdominal pain, mildly relieved by antispasmodics. She experienced two episodes of vomiting and had no bowel movements or flatus for 24 hours. She reported altered bowel habits for several months, including constipation and the passage of hard stool, and anorexia. Upon physical examination, a reducible periumbilical hernia and pain in the right iliac fossa were found along with an enlarged abdomen. Bowel sounds were audible and normotonic. Digital rectal examination was unremarkable.

Initial laboratory investigations revealed normal values for white blood cell count, hemoglobin, Lipase, Amylase, sodium (Na), potassium (K), and Troponin.

A laparoscopic right hemicolectomy with resection and anastomosis was performed on the patient. Pathological examination of the resected specimen revealed a MALT lymphoma involving the terminal ileum, cecum, and appendix (Figures 1 and 2).

The specimen was taken to histopathology. The analysis identified atypical B-cell proliferation in the terminal ileum, raising suspicion for extranodal marginal zone lymphoma of MALT. Additionally, ischemic changes were noted in the terminal ileum and cecum, indicative of early-stage ischemic damage. A tubular adenoma was identified in the cecum, and the appendix exhibited fibrous obliteration. Assessment of the resection margins revealed ischemic changes in the proximal margin, while the distal margin remained viable.

Discussion

Gastrointestinal MALT lymphomas often present nonspecific symptoms, making early diagnosis challenging. Common manifestations include weight loss, altered bowel habits, abdominal pain, and, occasionally, gastrointestinal bleeding [12]. However, acute presentations like intestinal obstruction are uncommon. In this case, the patient's acute colicky lower abdominal pain, vomiting, and absence of bowel movements were indicative of a mechanical obstruction. Imaging studies suggested a closed-loop obstruction due to a twisted segment of the ileum, leading to surgical intervention. This aligns with reports where MALT lymphoma was incidentally discovered during emergency surgeries for bowel obstruction without prior chronic inflammatory or autoimmune conditions [13].

In another case, a 67-year-old man with jejunal MALT lymphoma showed up with almost complete intestinal lumen blockage, highlighting the potential for MALT lymphoma to cause significant luminal narrowing and obstruction [14]. In a case involving chronic bowel obstruction, a patient presented with increased peristalsis and an abdominal mass. MALT lymphoma was histopathologically confirmed, demonstrating variability in clinical presentations [15].

The involvement of the appendix is also exceedingly rare, with only very few cases described in the literature. Most patients with appendiceal MALT lymphoma present with symptoms that mimic acute appendicitis and, therefore, are initially misdiagnosed [16]. For example, a case report by Nureta et al. [17] involved a 22-year-old man with recurrent abdominal pain initially suspected to be chronic appendicitis that was later found to be primary appendiceal MALT lymphoma.

In this case, histopathological examination was consistent with extranodal marginal zone lymphoma of MALT, with atypical B-cell proliferation in the terminal ileum. Early ischemic changes were seen in the terminal ileum and cecum, tubular adenoma of the cecum, and fibrous obliteration of the appendix. MALT lymphoma, however, very rarely involves multiple contiguous sites (terminal ileum, cecum, and appendix).

For example, a MALT lymphoma presented as a semi-pedunculated polyp in the sigmoid colon, demonstrating the varied appearance of this lymphoma in the gastrointestinal tract [18]. Ischemic changes in the bowel and MALT lymphoma coexist rarely. Ischemic colitis is due to inadequate blood flow to the colon and results in inflammation and even necrosis in some cases [19]. Lymphoma can cause bowel wall thickening and compromise vascular supply, resulting in ischemia, but the relationship between MALT lymphoma and ischemic changes is still unclear. Inban et al. [19] reported a case of Epstein-Barr virus B cell lymphoma in an 80-year-old patient with ischemic colitis, hinting at a possible association with ischemic bowel disease and lymphoproliferative disorders.

The histopathology of MALT lymphoma under high-power (400×) Hematoxylin and Eosin stained section showed a dense lymphoid infiltrate with characteristic lymphoepithelial lesions (arrow). The infiltrate is composed of small to medium-sized lymphoid cells with irregular nuclear contours, consistent with extranodal marginal zone B-cell lymphoma of MALT type [20] (Figure 3).

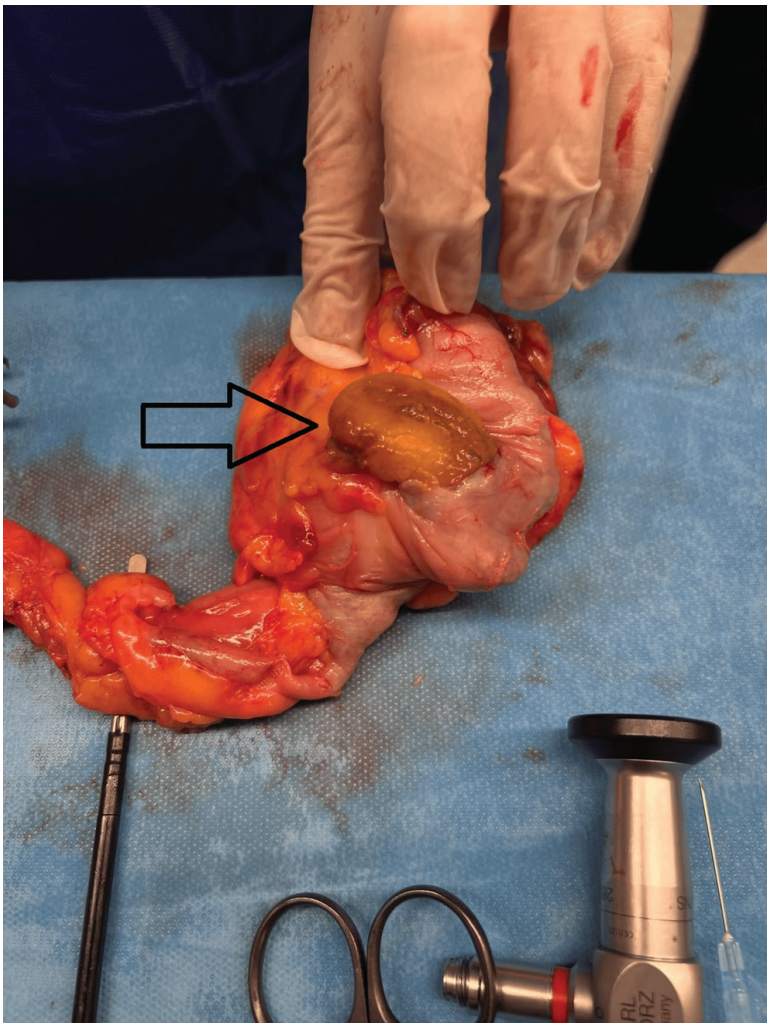

Figure 1. *Gross findings of MALT lymphoma.*

The immunohistochemistry in MALT lymphoma under high-power (400×) immunohistochemical stain showed a prominent lymphoepithelial lesion (arrow) characteristic of MALT lymphoma. The surrounding neoplastic B-cells demonstrated strong membranous positivity, consistent with CD20 immunostaining, highlighting the dense infiltrate and its interaction with epithelial structures [20] (Figure 4).

Intestinal MALT lymphoma has a nonspecific clinical presentation, and the diagnosis is often incidental or delayed. Mucosal abnormalities ranging from erosions, ulcerations, or polypoid lesions might be seen on endoscopic evaluations, but these are not pathognomonic [2]. The cornerstone of diagnosis is histopathological examination, which typically shows diffuse infiltration of small to medium-sized lymphocytes with irregular nuclei and lymphoepithelial lesions [21]. To differentiate MALT lymphoma from other lymphoproliferative disorders, immunohistochemical staining is essential [12].

Gastrointestinal MALT lymphoma is multifaceted, most commonly undergoing a sequence of surgical, medical, and occasionally radiotherapeutic management. Also, eradication of Helicobacter pylori has been effective in inducing remission of localized MALT lymphomas of the stomach [22]. However, for non-gastric MALT lymphomas, such as those in the small intestine or colon, the role of antibiotics is less clear, and treatment often involves surgery, radiotherapy, or immunochemotherapy. A review of 52 cases of small intestinal MALT lymphoma by Markopoulos et al. [23] indicated that surgery was the primary diagnostic and therapeutic modality, with clinical remission achieved in 82% of cases. During screening colonoscopy, a 2-cm semi-pedunculated polyp in the sigmoid colon was discovered in a 54-year-old male. The endoscopic mucosal excision was used to remove the polyp, and histologic examination revealed MALT lymphoma, demonstrating that endoscopic resection can be both diagnostic and therapeutic in certain cases [18]. Anti-CD20 monoclonal antibody, rituximab, has been proven effective in treating MALT lymphoma, either alone or in combination with chemotherapy [24]. Systemic treatment with rituximab and chemotherapy regimens such as CHOP (cyclophosphamide, doxorubicin, vincristine, and

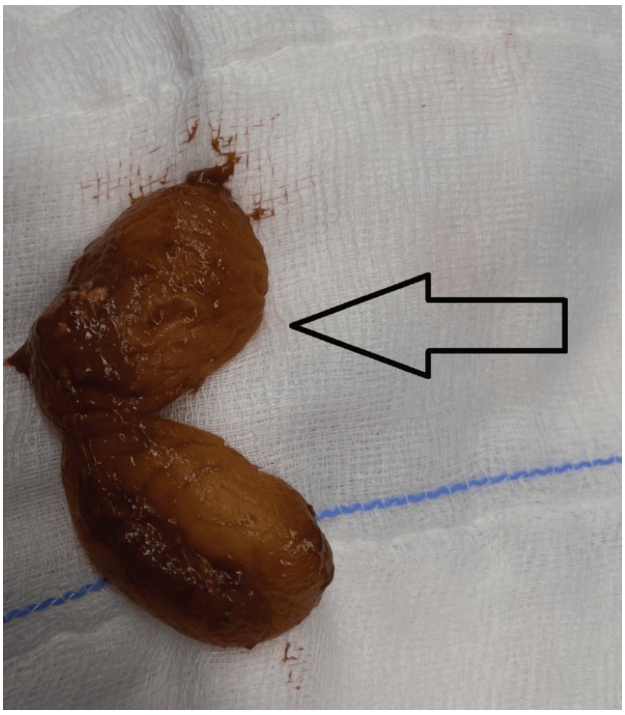

Figure 2. *The specimen was taken to histopathology.*

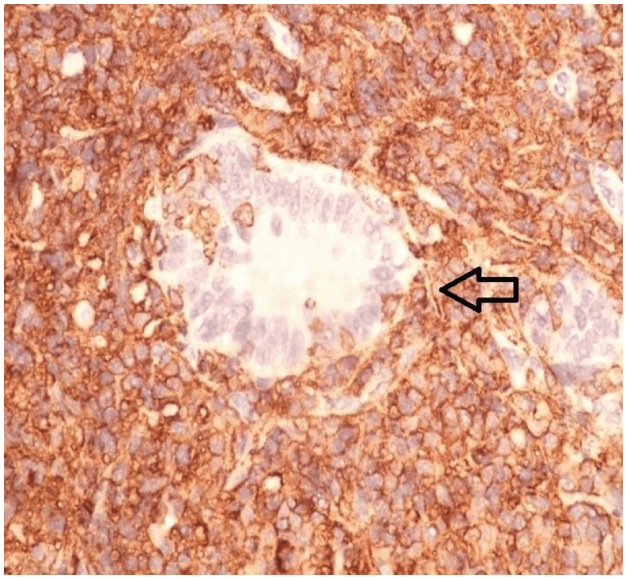

Figure 3. *Histopathology image of MALT Lymphoma.*

Immunostain CD20 shows strong positivity in lymphoma cells.
Stain: Hematoxylin and Eosin (H&E)
Magnification: 400x (40x objective with 10x eyepiece)
Source: International Scientific
Information, Inc. /Hafsa Abbas, Masooma Niazi, Jasbir Makker
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prednisone) has been used in cases where surgical resection is not feasible or in advanced-stage disease [25].

MALT lymphoma is usually considered an indolent lymphoma with a good prognosis. Eradication therapy for Helicobacter pylori is often effective in the treatment of stomach MALT lymphoma. Intestinal MALT lymphomas, however, especially those with complicating sequelae,

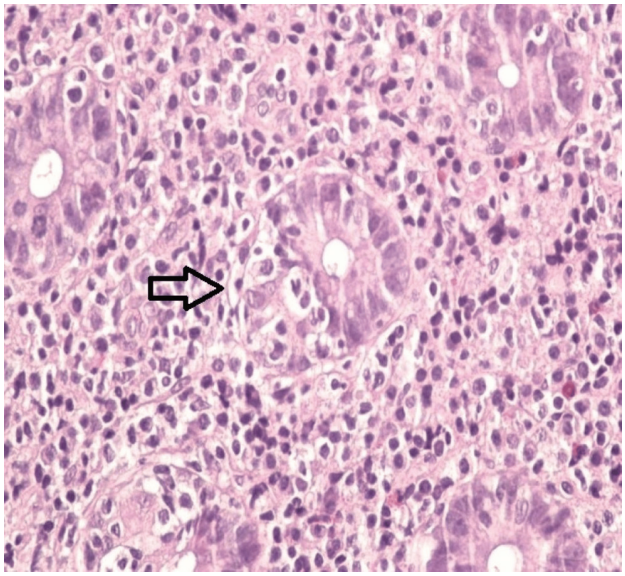

Figure 4. *Histopathology image of MALT Lymphoma.*

MALT lymphoma involving the colon.There is a diffuse infiltrate comprised of small to medium-sized lymphocytes with monocytoid features and plasmacytoid cells.
Stain: CD20 Immunohistochemistry
Magnification: 400x (40x objective with 10x eyepiece)
Source: International Scientific
Information, Inc./ Hafsa Abbas, Masooma Niazi, Jasbir Makker
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such as obstruction, may need more aggressive treatment involving surgical resection, sometimes with adjuvant therapies [22]. The concern regarding recurrence risk of MALT lymphoma is particularly high for patients with multifocal disease or residual microscopic involvement. In a study of MALT extranodal marginal zone lymphoma at an early stage, the survival rate was 89% and the relapse-free survival rate was 60% at 5 years [26].

Gastrointestinal tract MALT lymphoma is rare and its presentation is varied, requiring a high index of suspicion and a multi-disciplinary approach for the accurate diagnosis and management. In chronic inflammation, such as gastric cases with infection by Helicobacter pylori, MALT lymphoma often arises. Molecular diagnostics with the ability to detect specific genetic aberrations may substitute early detection and personalized treatment approaches. Consequently, such infections might be investigated as possible triggers of MALT lymphomas in non-gastric sites and would lead to less invasive treatment options.

Conclusion

MALT lymphoma, though predominantly gastric in origin, can rarely involve the small intestine, often presenting with nonspecific or acute obstructive symptoms that mimic common gastrointestinal disorders. This case highlighted the diagnostic challenges of intestinal MALT lymphoma, particularly in elderly patients without prior chronic inflammatory conditions, where clinical suspicion was low. Surgical intervention might be necessary for complications

like bowel obstruction, but systemic therapies, including rituximab-based regimens, remain crucial for disseminated disease. Given the indolent yet variable nature of MALT lymphoma, a multidisciplinary approach, combining histopathology, immunohistochemistry, and advanced imaging, is essential for accurate diagnosis and tailored management. Future research should explore molecular diagnostics and targeted therapies to improve early detection and treatment outcomes, particularly for non-gastric MALT lymphomas. Enhanced awareness of its diverse presentations would aid clinicians in recognizing this rare entity promptly, optimizing patient care and long-term prognosis.

List of abbreviations

Mucosa-associated lymphoid tissue (MALT)
Potassium (K)
Sodium (Na)

Conflict of interests

The authors declare that there is no conflict of interest regarding the publication of this article.

Funding

None.

Consent for publication

Permission was obtained from the patient/guardians of the patient to publish the case and the accompanying images.

Ethical approval

Ethical approval is not required at our institution to publish an anonymous case report or series.

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Summary of the case

1	Patient (gender, age)	69-year-old female
2	Final Diagnosis	Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue
3	Symptoms	Sltered bowel habits for several months, including constipation and the passage of hard stool, and anorexia
4	Medications	N/A
5	Clinical Procedure	A laparoscopic right hemicolectomy with resection and anastomosis was performed on the patient.
6	Specialty	General Surgery