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Case Report

Extranodal marginal zone lymphoma of mucosa associated lymphoid tissue: a rare cause of acute abdomen

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ABSTRACT

Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma) is a low-grade B-cell lymphoproliferative disorder that most commonly involves the stomach. Involvement of the small intestine is rare, accounting for a small proportion of gastrointestinal cases, and clinical presentation is often nonspecific. Acute abdomen resulting from intestinal perforation is an exceptional manifestation. Case presentation: We report the case of a 33-year-old male with no significant past medical history who presented with a two-week history of progressive diffuse abdominal pain, fever, and anorexia. Physical examination revealed signs of peritonitis and a palpable inflammatory mass in the right lower quadrant. Given the clinical diagnosis of acute abdomen, an emergency exploratory laparotomy was performed, revealing a small bowel inflammatory mass with intestinal perforation and purulent peritonitis, located approximately 50 cm from the ileocecal valve. Segmental small bowel resection with primary anastomosis and appendectomy were performed. Histopathological and immunohistochemical analysis of the resected specimen confirmed extranodal marginal zone lymphoma of MALT type. The postoperative course was uneventful, and the patient was discharged without complications. Small intestinal extranodal marginal zone lymphoma is a rare entity with an indolent course and nonspecific clinical manifestations, which may delay diagnosis. In exceptional cases, it may present as an acute abdomen due to complications such as intestinal perforation, requiring urgent surgical intervention. This case underscores the importance of maintaining a high index of suspicion and highlights the role of a multidisciplinary approach in diagnosis and management.

Keywords: Extranodal marginal zone lymphoma, MALT lymphoma, Acute abdomen, Small intestine, Intestinal perforation

INTRODUCTION

Extranodal marginal zone lymphoma (EMZL) is a low-grade lymphoproliferative neoplasm associated with mucosa-associated lymphoid tissue (MALT) of the gastrointestinal tract. Its localization in the small intestine is rare, accounting for approximately 5-8% of cases.[1-4] Due to its indolent course and nonspecific clinical manifestations, early diagnosis is challenging. In atypical presentations, EMZL may manifest with intestinal perforation, placing the patient in a life-threatening condition that requires surgical management. We present an atypical case that aims to broaden the understanding of the clinical presentation and surgical management of this entity.

CASE REPORT

A 33-year-old male with no significant medical or surgical history presented to the emergency department

after being referred from another institution due to a two-week history of progressive, diffuse abdominal pain predominantly involving the hypogastrium and right flank, accompanied by fever and anorexia. He had been treated with ceftriaxone, chloramphenicol, and analgesics without clinical improvement.

On admission, the patient appeared pale, diaphoretic, tachycardic, febrile, and adopted an antalgic position. Abdominal examination revealed distension, decreased bowel sounds, generalized muscular guarding, and a palpable inflammatory mass in the right iliac fossa with marked tenderness. Laboratory tests showed mildly elevated C-reactive protein levels and bandemia, without leukocytosis.

Given the diagnosis of acute abdomen, we performed an emergency exploratory laparotomy. Intraoperative findings included an inflammatory mass of the small intestine located approximately 50 cm from the ileocecal valve, associated with intestinal perforation, purulent peritonitis, and signs of periappendicitis. Segmental small bowel resection with primary anastomosis and appendectomy were performed (Figure 1).

The postoperative course was uneventful, and the patient was discharged on postoperative day seven. During follow-up, persistent but less intense abdominal pain prompted a contrast-enhanced computed tomography scan, which demonstrated marked thickening of a small bowel segment without intra-abdominal collections (Figure 2). Histopathological examination of the resected specimen revealed extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue.

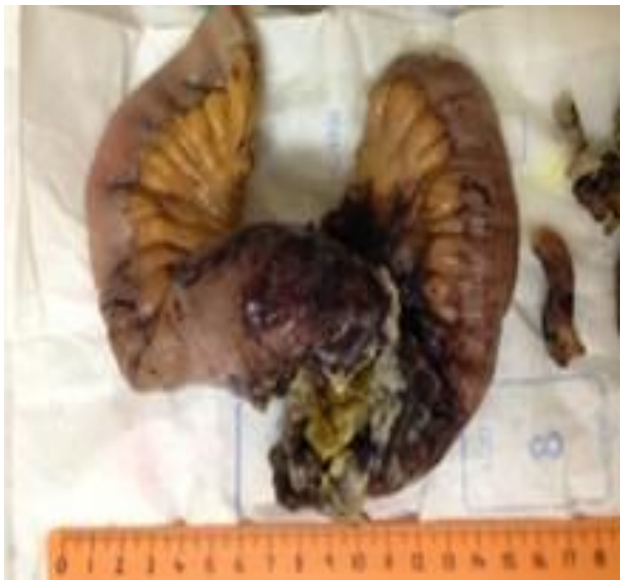

Figure 1: Gross specimen of the small intestine resected during emergency laparotomy, demonstrating marked wall thickening, transmural involvement, and an area of perforation associated with inflammatory changes.

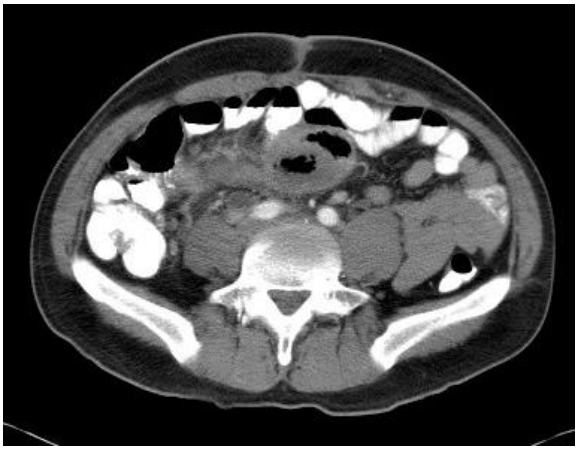

Figure 2: Axial CT image showing persistent mural thickening of the affected small bowel segment without evidence of well-defined intra-abdominal collections.

DISCUSSION

Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue is a low-grade lymphoproliferative neoplasm composed of small B lymphocytes. Owing to the abundance of MALT within the gastrointestinal tract, this region represents one of the most commonly involved sites. Nevertheless, EMZL may also arise in salivary glands, thyroid, lungs, bladder, and skin.[1,2] Involvement of the small intestine is distinctly uncommon, which partly explains the limited characterization of this entity in the literature.[3]

From an epidemiological standpoint, lymphomas account for approximately 1–8% of all gastrointestinal neoplasms. Among these, nearly half correspond to MALT lymphomas; however, only 5–8% involve the small intestine.[2] Reported risk factors include a family history of EMZL, chronic inflammatory conditions, and infection with Campylobacter jejuni. In contrast, Helicobacter pylori infection has not demonstrated a significant association with small intestinal EMZL.[1,2,4]

The pathogenesis of EMZL remains incompletely understood. Chronic antigenic stimulation, secondary to persistent infection or inflammation, is thought to induce immune dysregulation.[1,5] Additionally, recurrent genetic abnormalities such as t (11;18) (q21; q21), t (1;14) (p22; q32), t (14;18) (q32; q21), and t (3;14) (p14; q32) translocations have been identified. These alterations promote activation of the NF-κB pathway or dysregulation of NOTCH signaling, thereby favoring sustained inflammation and resistance to apoptosis.[2,6]

Clinical presentation is typically nonspecific and largely dependent on tumor location, which frequently results in delayed diagnosis. Common symptoms include general malaise, weight loss, and low-grade fever.[1,6] In cases involving the small intestine, patients may present with abdominal pain, iron-deficiency anemia due to chronic

bleeding, or intestinal obstruction. Rare and atypical presentations, such as ileal intussusception and intestinal perforation, have been reported.[5] In a cohort of 1,062 patients with gastrointestinal MALT lymphoma, only 17 cases involved the small intestine, and merely two presented with perforation, underscoring the exceptional nature of this complication.[5,7]

Diagnostic evaluation includes complete blood count, metabolic panel, and lactate dehydrogenase assessment.[6] Contrast-enhanced computed tomography of the chest, abdomen, and pelvis is recommended to evaluate disease extension, spleen size, and lymph node involvement, making it the preferred modality for staging and treatment planning.[8] Magnetic resonance imaging has limited diagnostic utility in intestinal disease, and the role of PET-CT remains controversial.[2]

Endoscopy allows tissue sampling and macroscopic classification of lesions as superficial, ulcerative, or polypoid. However, submucosal involvement without mucosal alteration may limit diagnostic yield. Endoscopic ultrasound has not consistently demonstrated additional diagnostic value.[3]

Histopathological evaluation must be comprehensive, with adequate tissue sampling. Immunohistochemistry is essential for diagnosis and typically demonstrates positivity for CD20, CD79a, and BCL-2. Morphological analysis, flow cytometry, and genetic studies are recommended to confirm the diagnosis and exclude other lymphoproliferative disorders.[5,9]

Therapeutic strategies for small intestinal EMZL are not well established. Some authors advocate initial H. pylori eradication therapy, although responses are less favorable than those observed in gastric EMZL. In cases of deep invasion, systemic chemotherapy or radiotherapy may be required. Oral cyclophosphamide (100 mg/day for 18 months) has been proposed when disease is confined to the mucosa and submucosa. Surgical management is reserved for complications such as hemorrhage, obstruction, or perforation, as in the present case, or when comprehensive diagnostic or therapeutic approaches are not feasible.[1,3,9,10]

CONCLUSION

Small intestinal extranodal marginal zone lymphoma is a rare entity characterized by an indolent, nonspecific, and frequently underrecognized clinical course. In exceptional cases, it may present as acute abdomen due to complications such as intestinal obstruction, intussusception, or intestinal perforation, requiring urgent surgical intervention. Maintaining a high index of clinical suspicion and adopting a multidisciplinary approach are essential, particularly given the limited number of cases reported in the literature. Further studies are needed to refine diagnostic strategies and establish optimal treatment protocols.

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