A Rare Case of MALT Lymphoma Underlying Ileocecal Intussusception

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Intussusception is an extremely rare diagnosis in adults, with an etiologic process identified in the majority of cases. The authors describe an unusual case of mucosa-associated lymphoid tissue (MALT) lymphoma as the underlying cause of ileocecal intussusception in an elderly woman. The patient presented with complaints of abdominal pain of variable intensity that had increased in severity over the past several months. A contrast-enhanced computed tomographic scan revealed evidence of ileocecal intussusception, and a subsequent exploratory laparotomy revealed high-grade bowel obstruction. Early recognition of intussusception is critical to appropriate management and resolution.

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First described in 1983, B-cell lymphoma of mucosa-associated lymphoid tissue (MALT) is a rare cause of primary gastrointestinal (GI) lymphoma.[1] As defined by Jayabackthan et al, "MALT lymphoma is defined as an extranodal marginal zone B-cell lymphoma of MALT type."[2(p119)] Mucosa-associated lymphoid tissue can be found throughout the body, with the highest amount found in the GI tract. When this tissue undergoes abnormal cell proliferation, lymphoma can develop. Most commonly in the GI tract, MALT lymphoma comprises up to 50% of cases.[3(p158)] Within the GI tract, the stomach is the most common location (85%), followed by the small intestine and colon.[3(p158)]

Intussusception in adults accounts for 5% of all cases of intussusception and 1% to 5% of all cases of intestinal obstructions.[4(p407)] The symptoms of MALT lymphoma are nonspecific, which makes diagnosis difficult. An underlying pathologic condition is identified as the cause of almost 90% of cases of intussusception in adults.[4(p407)] We present a case of MALT lymphoma underlying as intussusception in an adult who complained of abdominal pain.

Report of Case

An 83-year-old woman presented to the emergency department with several months of abdominal pain of variable intensity that had increased in severity over the past 2 days. Her pain was intensified by oral intake of food and was accompanied by diarrhea. Physical examination revealed abdominal distention, well-healed scars from previous open cholecystectomy and open appendectomy, and a nonpulsatile palpable mass in the right lower quadrant. The patient was admitted to the hospital, and a computed tomographic (CT) scan was ordered. The CT scan of the abdomen and pelvis with oral and intravenous contrast revealed evidence of intussusception of the terminal ileum into the colon with high-grade small bowel obstruction (*Figure 1* and *Figure 2*). The patient underwent an exploratory laparotomy and a subsequent ileocolectomy on the day of admission. Gross

examination during laparotomy consisted of the cecum (9 cm in length by 8.5 cm internal circumference, with an average wall thickness of 0.5 cm) and the terminal ileum (28 cm in length by 9 cm internal circumference with an average wall thickness of 0.5 cm) without an apparent appendix. Focal areas of duskiness, hemorrhage, and erosion were evident in the distal ilium. A firm nodular tan mucosa (3.5 × 3 × 3 cm area) was underlying the puckered serosa, where the appendix was previously located. It was located within the cecum, 2.5 cm distal to the ileocecal valve, and was dissected intraoperatively. Tissue samples were resected for laboratory evaluation.

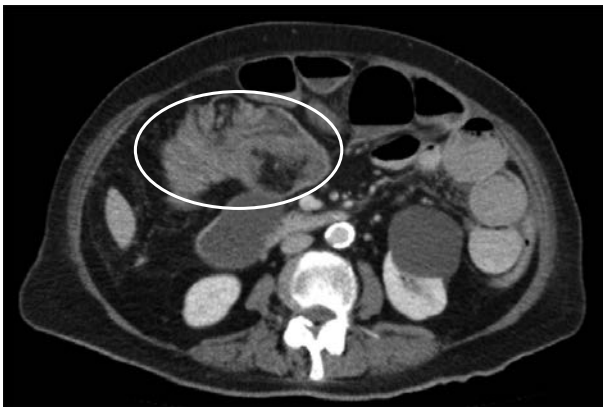

Figure 1.
Computed tomographic image revealing invagination of the terminal ileum into the cecum with visible mass (circle) in an elderly woman presenting with abdominal pain.

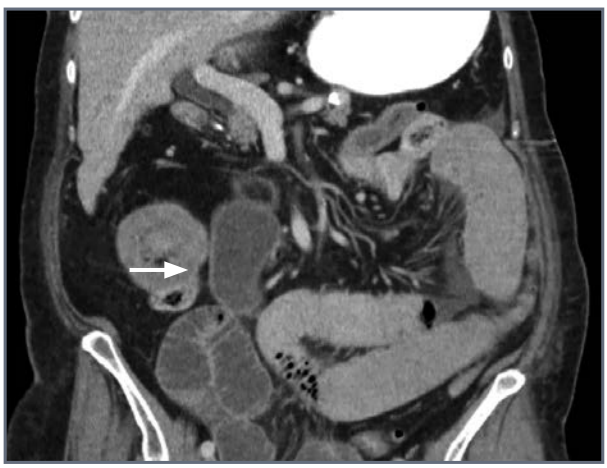

Figure 2.
Contrast-enhanced computed tomographic image demonstrating a classic target pattern (arrow), revealing the telescoping of the terminal ileum into the cecum, in an elderly woman presenting with abdominal pain.

Microscopic evaluation results obtained 5 days after the surgical specimen was removed revealed terminal ileitis with ulceration and microabscess formation. Thirteen lymph nodes were identified but were found to be uninvolved. A histologic diagnosis of cecal MALT lymphoma was reported and supported by immunotyping results (*Figure 3*, *Figure 4*, and *Figure 5*), which were positive for B-cell markers CD20 (*Figure 4*) and Pax-5, focal positive for Bcl-2, and negative for CD3, CD5, CD43, CD10, and Cyclin-D1. The tumor had a low proliferative index, with Ki-67 less than 10%. The patient recovered postoperatively without complications and was referred to the Oncology Department for further treatment.

Discussion

As defined by Marinis et al, "Intussusception of the bowel is the telescoping of a proximal segment of the bowel within the lumen of the adjacent segment."[4(p407)] Most commonly seen in children, intussusception in adults is exceedingly rare.[4(p407)] Unlike the classic triad of symptoms seen in children presenting with intussusception—abdominal pain, stools reminiscent of red currant jelly, and a tender abdominal mass—symptoms in adults vary considerably. Nausea, vomiting, bleeding, changes in bowel habits, constipation, and abdominal distention have been identified as nonspecific symptoms associated with intussusception in adults.[4(p408)]

Abdominal CT is the most sensitive imaging modality for the diagnosis of intussusception.[4(p410)] When the CT beam is perpendicular to the longitudinal axis of the intussusception, it can be visualized as a classic target-like lesion (*Figure 2*, arrow). Most cases of adult intussusception are caused by an

underlying pathologic condition, such as malignant processes, lipomas, polyps, diverticula, strictures, or benign neoplasms.[4(p407),5] In MALT lymphomas, the GI tract is the most commonly involved site, with about 50% of all MALT lymphomas occurring there.[6(p215)] Within the GI tract, the stomach is the most common location. As in the present case, the most common site of MALT lymphoma in the large intestine is the cecum and rectum.[2(p119)] Other sites of MALT lymphoma may include the breast, lung, ovary, thyroid, prostate, parotid glands, salivary glands, and other head and neck regions.[6(p222)]

Owing to the association of adult intussusception with underlying malignant organic lesions, surgical intervention is often required.[4(p410)] Management typically includes an exploratory laparotomy or laparoscopy followed by resection of lead point masses or ischemia.[7(p80)] Hematopathologic review with immunotyping of resected materials is essential to diagnosis. Recommended B-cell markers to identify MALT lymphoma include CD20, CD3, CD5, CD10, CD21 or CD23, kappa/lambda, CCND1, and BCL2 with flow cytometry markers CD19, CD20, CD5, CD23, and CD10.[6(p222)] The characteristic immunophenotype for MALT lymphoma is $CD5^{-}$, $CD10^{-}$, $CD20^{+}$, $CD23^{-/+}$, $CD43^{-/+}$, cyclin $D1^{-}$, and $BCL2^{-}$.[6(p222)] In the present case, MALT lymphoma was identified by the monocytoid appearance (*Figure 3*), arranged in a somewhat irregular crowding nodular pattern, and involved the full thickness of the cecal wall with mucosa erosion. The diagnosis was supported by immunophenotyping results, which revealed atypical cells diffusely positive for B-cell marker CD20 (*Figure 4*) and prominent lymphoepithelial lesions (*Figure 5*).

After diagnosis, the workup of nongastric MALT lymphoma is similar to other non-Hodgkin lymphomas. Workup should include a comprehensive physical examination; laboratory evaluation involving complete blood cell count with differentials, comprehensive metabolic panel, and measurement of lactate dehydrogenase; and contrast-enhanced CT of the chest, abdomen, and pelvis.[6(p222)]

Conclusion

Mucosa-associated lymphoid tissue lymphoma is a rare cause of intussusception in adults. Because of the known role of *Helicobacter pylori* and the response to antibiotic therapy, management of gastric lymphoma is well established.[8(p572)] However, the best treatment modality for MALT lymphoma at other locations has not been identified. The combination of surgery and chemotherapy is believed to be superior to other approaches. It is important to understand this disease process to aid in the evaluation of patients and the diagnosis and management of this condition.

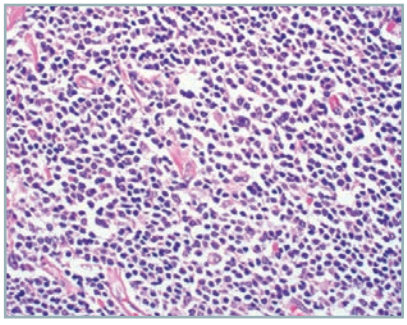

Figure 3.
Pale cytoplasm with a monocytoid appearance (hematoxylin-eosin).

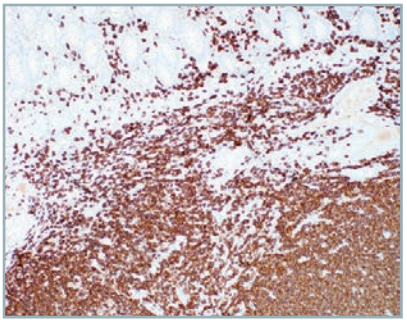

Figure 4.
Atypical cells diffusely positive for B-cell marker CD20 (immunohistochemistry CD20, original magnification ×10).

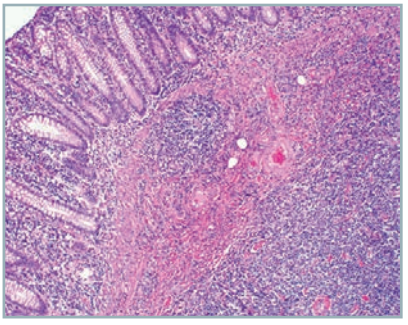

Figure 5.
Atypical lymphoid cells forming lymphoepithelial lesions. (hematoxylin-eosin, original magnification ×10).

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