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Mucosa-associated lymphoid tissue lymphoma of the ileum in a child presenting as intussusception

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Dear Editor,

Mucosa-associated lymphoid tissue (MALT) lymphoma is the third most common non-Hodgkin's lymphoma subtype. Clinical presentation is often insidious as a low-grade lesion and disease tends to remain localized for a long period of time. The gastrointestinal tract, in particular the stomach, is by far the most common extranodal site. MALT lymphoma of the ileum, however, is rare.[1] Ileal intussusception caused by MALT lymphoma as a cause of acute abdomen is extremely rare, with symptoms that often mislead and make diagnosis more difficult.[2] Herein, we present a rare case of ileal intussusception caused by MALT lymphoma in a 14-year-old male child.

A 14-year-old boy presented in the surgical emergency clinic with acute abdominal pain, nausea, vomiting and problems with defecation and flatulence. On physical examination, a distended, diffusely tender and painful abdomen with lower abdominal rebound tenderness was revealed. Abdominal exploration revealed an ileoileal intussusception induced by a 5 cm diameter tumor. A segmental ileal resection was performed.

On gross examination, there was a 5 cm × 4 cm × 3 cm, polypoidal, grey white exophytic tumor arising from the resected ileal segment [Figure 1]. On microscopic examination, diffuse infiltrate of uniform small-to-medium-sized lymphocytes with irregular nuclear contours and abundant cytoplasm resembling centrocytes or monocytoid lymphoid cells was seen. These lymphoid cells were infiltrating all the layers of the ileum with surface ulceration and complete destruction of the intestinal glands [Figure 2]. Scattered transformed blasts and plasma cell differentiation was also seen. Occasional lymphoepithelial lesions were also seen. Immunohistochemical stains showed that these lymphoid cells were positive for CD20 and negative for CD3 and CD5.

In contrast to adult non-Hodgkin's lymphoma, the non-Hodgkin's lymphoma occurring in children is usually found extranodally. The most common site is the abdomen, including the gastrointestinal tract, kidney and pancreas, and the next most common location is the head and neck region.[3] Gastrointestinal non-Hodgkin's lymphoma in adults is usually considered to be MALT lymphoma, but the occurrence of this variety in the small bowel of children is relatively rare, and that occurring in the ileum is still rarer.[4,5]

Intussusception is one of the most common causes of intestinal obstruction in infancy and childhood. However, intussusception confined to the small bowel accounts for less than 10% of all cases of childhood intussusceptions.[5] Ileal maltoma as a cause of ileal intussusception in children

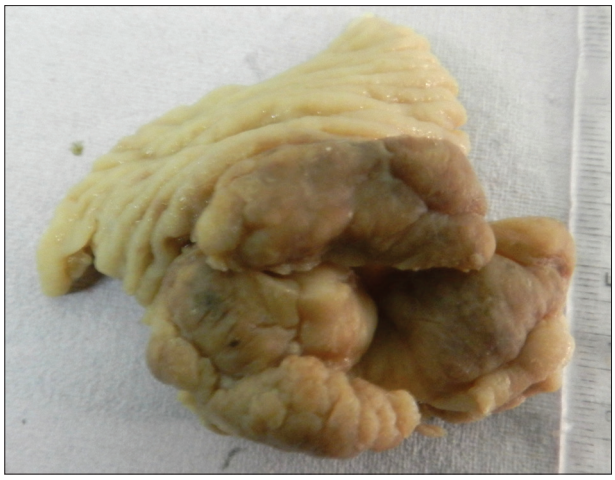

Figure 1: Gross specimen of resected ileal segment revealing grey white polypoidal exophytic growth

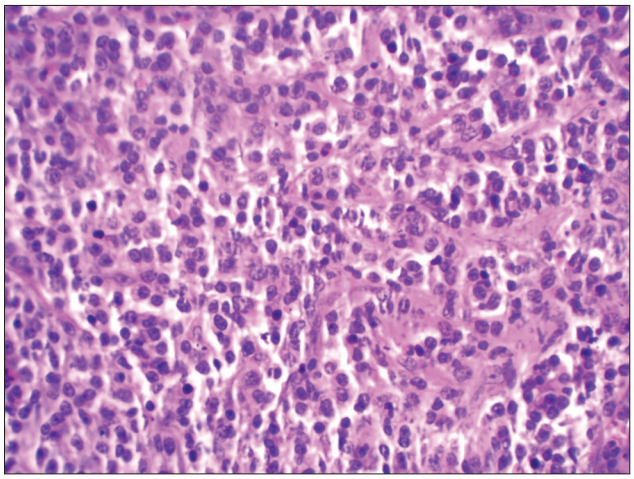

Figure 2: Photomicrograph showing sheets of atypical lymphoid cells diffusely infiltrating the layers of the ileum (hematoxylin and eosin, x20)

is extremely rare and only two cases have been reported in the literature.[4]

Not uncommonly, the diagnosis is only made intraoperatively for intestinal obstruction because of the fact that a palpable abdominal mass is also rare in this type of intussusception. An awareness of this entity among pediatricians is of paramount importance to obviate delays in diagnosis.

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