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 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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Dear Editor,

The treatment of Hodgkin lymphoma with Adriamycin, Bleomycin, Vinblastine, Dacarbazine (ABVD) regimen is considered as one of the most efficacious and least toxic modalities and is the standard of care today. Delays due to hematological toxicity do occur occasionally, but severe toxicity requiring treatment modifications are rare and rarer still are the incidences of neurotoxicity. A 20-year-old lady, with no known comorbidities presented to us with B symptoms and chest pain of 6 months duration. A contrast computed tomography (CT) chest showed a 10.9 × 9.5 × 9.2 cm large heterogeneously enhancing anterior mediastinal mass lesion without any focal lung lesion or any pleural effusion [Figure 1]. Trucut biopsy of the mediastinal mass showed features suggestive of Nodular Sclerosing Hodgkin lymphoma. Contrast CT of abdomen, bronchoscopy, and bone marrow examination were normal. She was diagnosed as Hodgkin lymphoma Stage IIBx and managed with three cycles of ABVD (Adriamycin 45 mg/Bleomycin 17 mg/Vinblastine 10 mg/Dacarbazine 640 mg) based chemotherapy to which she responded well. She was a healthy young lady with a body surface area of 1.72 m² and she did not require any growth factor supplements during these three cycles. She also denied intake of any alternative medicines. However, after two cycles she developed difficulty in walking and after third cycle developed frank bilateral foot drop. Clinically, there was no evidence of any other neurological deficit. Nerve conduction studies were suggestive of mixed sensory-motor neuropathy, predominantly axonal motor neuropathy involving the lower limbs [Figures 2 and 3]. An interim positron emission tomography (PET) scan done showed complete metabolic response and also decrease in size of the mass. Workup for preexisting causes of neuropathy and diseases like megaloblastic anemia, diabetes mellitus, hypothyroidism, syphilis, and connective tissue disorders like rheumatoid arthritis and systemic lupus erythematosus was negative. Pending a decision on how best to modify further chemotherapy we treated her with 30 Gy/15# of involved field radiotherapy to mediastinum.

We did an extensive literature search on incidence of foot drop or severe neurological toxicity secondary to vinblastine in ABVD therapy and were surprised to note that no significant data existed. [1,2] The major toxicity reported with vinblastine was hematological and there were not even case reports on severe neurotoxicity. [1] Literature also did not support the omission of vinblastine in further therapy.

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