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Lymphoma - an unusual cause of Ileocolic Intussuception

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IN BRIEF

Introduction - Intussusception is defined as telescoping of one segment of the gastrointestinal tract into an adjacent one. Approximately 5%of peripheral NHL arise in ileum and mostly seen in adults. We report a case of primary NHL of ileum presenting as intussusception in a child. *Case*- A 14 year old boy presented with pain abdomen, vomiting and obstipation of 5 days duration. Per abdomen – tenderness and guarding in RIF with lump and hyperperistaltic bowel sounds. USG abdomen was indicative of intussusception. Emergency laparotomy revealed ileocolocolic intussusception as the cause of obstruction. Patient underwent right hemicolectomy with restoration of the bowel continuity. Histopathology revealed the mass to be a B-cell lymphoma. **Conclusion** - Aim of this reporting is to recognize NHL as a cause of intussusception in child and its correct management.

INTRODUCTION

Intussusception is defined as telescoping of one segment of the gastrointestinal tract into an adjacent one. It is relatively a common entity in childhood, where it can present as an acute abdomen with peak age between 6-9 months during the weaning period. In adults, an underlying cause is present in 80% of cases which may include polyps, lipomas and malignant tumors arising in gut as well as oedema and fibrosis from recent or previous surgery [1]. We report a rare case of childhood intestinal intussusception caused by a primary B-cell non-Hodgkin lymphoma (NHL) of the terminal part of ileum.

The primary NHL found in ileum is rare, it occurs 23 to 30% of gastrointestinal

lymphomas and approximately 5% of peripheral NHL[2] . It differs from gastric MALTomas in presentation, management, and prognosis. Although substantial progress has been achieved in the diagnosis and treatment of gastric lymphomas in recent years [3,4] primary intestinal lymphomas are not well characterized, and standardized concepts for their clinical diagnosis and management are absent.

Patient and case report

A 14 year old boy presented with pain abdomen, vomiting, decreased frequency of passage of stools and flatus since 5 days. Pain was colicky, non-radiating in nature. No history of fever, jaundice, abdominal distension was present. No history of TB, DM, or chronic disease of childhood, previous abdominal surgery was present.

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On examination there was tachycardia of 110/min, BP- 104/70 mm Hg, and the patient was dehydrated and afebrile. Per abdomen – tenderness and guarding in RIF associated with a vague lump, no rebound tenderness and hyper peristaltic bowel sounds were noted. Rest of systems appeared normal.

A second evaluation of abdomen, a few hours later , showed in addition to earlier findings, a small lump of about 4*4 cm in RIF and extending into right lumbar quadrant, which was tender and immobile. A rectal examination revealed no stools and collapsed rectum.

A clinical diagnosis of appendicular lump was made.

Initial management included correction of dehydration, routine blood tests, CXR and X-ray erect abdomen, as well as an emergency USG.

Haemogram revealed a raised TLC of 14,500. Erect abdominal X-ray showed dilated small bowel loops. Emergency USG abdomen revealed a concentric mass of bowel loops suggestive of a ileocolic intussusception.

Patient was put on expectant management initially which included- nasogastric tube insertion, IV fluids and parental antibiotics. An exploratory laparotomy was contemplated when the patient didn't respond to expectant management.

Intra-operatively an Ileocolic intussusception was noted in RIF, with circumferential 4*3 growths involving the caecum. Firm intraluminal masses were palpable in the lumen of ileum 30 cm and 50 cm from ICJ with enlarged multiple mesenteric lymph nodes were present.

A right hemicolectomy with ileo-transverse anastomosis with mesenteric lymph node

Fig. 1: Multiple enlarged mesenteric lymph nodes.

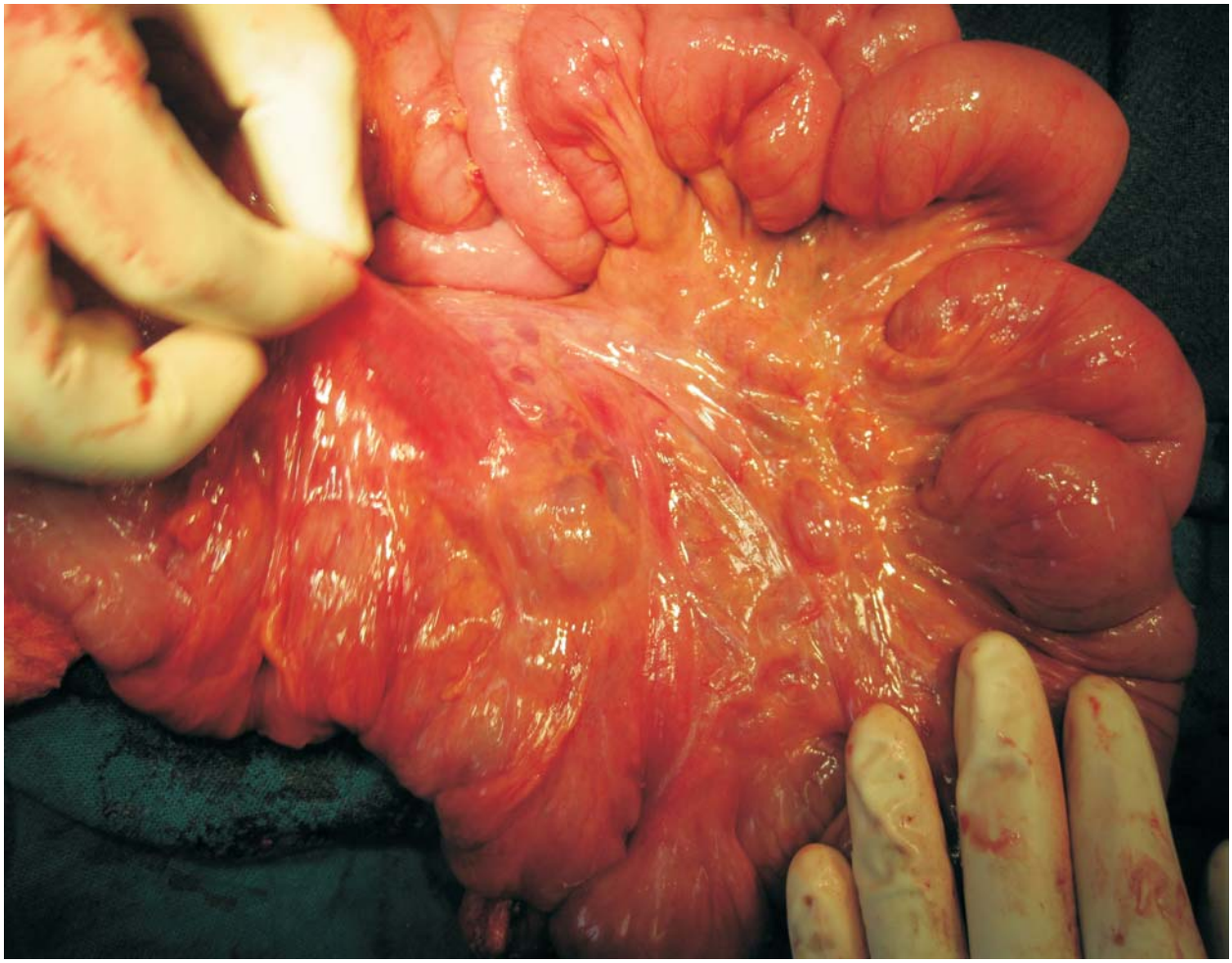
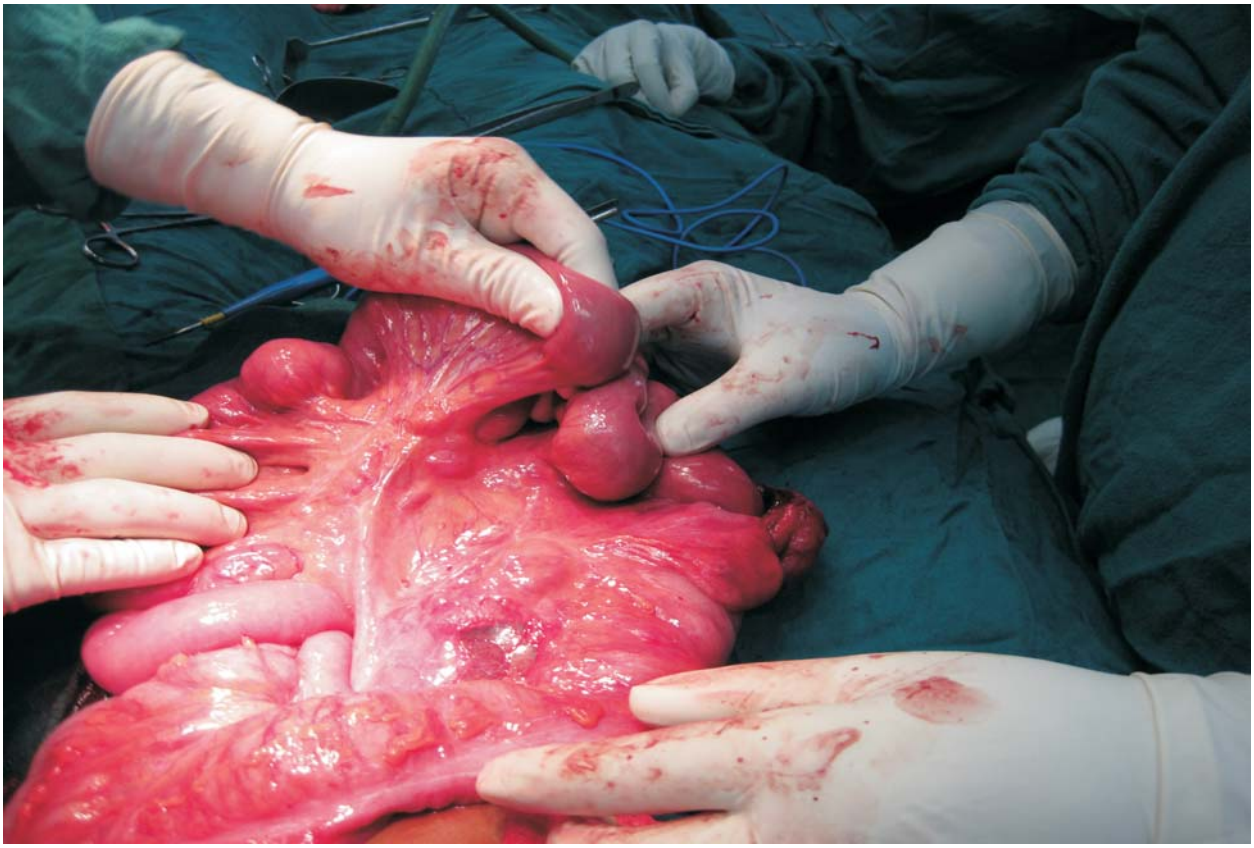
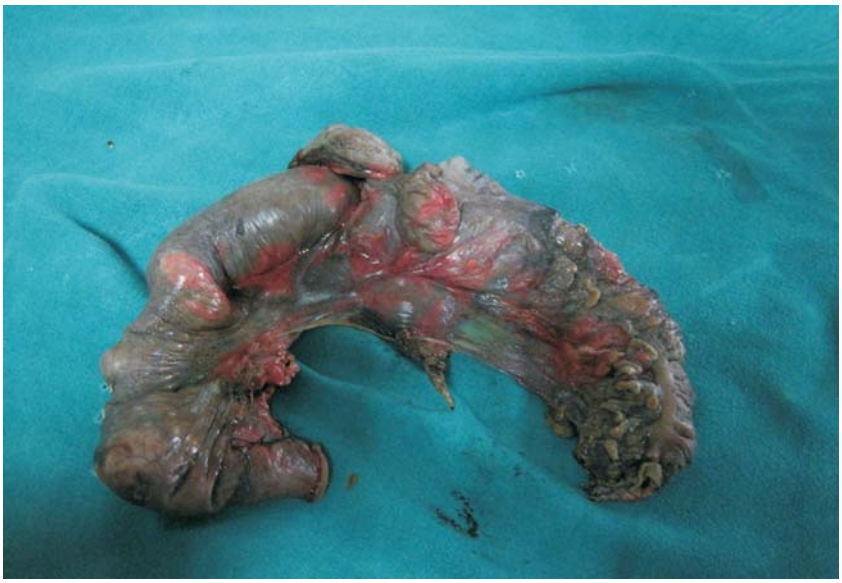


Fig. 2: Right hemicolectomy specimen cut open to show tumor masses in ileum.





biopsy was done. The resected part was sent for HPE.

Histopathological examination revealed B-cell Non-Hodgkin lymphoma which was of diffuse large B cell type. Immunohistochemistry showed atypical cells with strong membrane positivity for LCA and CD 20. The resected margins were free of tumor.

The disease was staged as primary ileal Stage 1 disease according to the Ann-Arbor classification. A postoperative chemotherapy (CHOP-R) was recommended but refused by the patient.

Post –operative course and follow up-

Patient regained bowel movements on post operative day 3 and oral sips were allowed on the same day with gradual reversion to normal diet. Patient was discharged on day 6 Patient was followed up after 15 days and 2 months after discharge was found be symptomless.

DISCUSSION

Intussusception is mainly a disease of childhood, whereas in adults its rare but associated with organic lesions in 80% cases in ileo-colic region (benign and malignant); with child to adult ratio being 20:1[5].

More rare are intussusceptions caused by lymphoma's involving of the ileum. Only a few cases are reported in the literature [6]. Small bowel lymphomas are rare due to several reasons [7]; the incidence of this disease has been rising in recent years particularly among immunocompromised patients [8,9].

Lymphomas of the gastrointestinal tract are the most common type of primary extra-nodal lymphomas [10] accounting for 5 to 10% of all non-Hodgkin's lymphomas. Majority of these tumors arise from small bowel and colon [11]. Age group maximally affected is 5-15 years with male to female ratio of 1.8-2.5

Tumor is considered primary when:

Histopathology is positive, Palpable peripheral lymphadenopathy and no hepatosplenomegaly. There is no evidence of lymphoma on chest x-ray or CT scan. Peripheral blood smear and bone-marrow are negative [12].

Malignant NHL in children originates in the distal small bowel and the caecum [13]. According to the WHO histological classification of NHL in children, B-cell immunophenotypes (Burkitt, Burkitt-like, large B-cell) most commonly arise in abdomen with Burkitt's lymphoma being the most common [14].

Lymphoma arises in the lymphoid follicles of the submucosa of the bowel from where it can either project into lumen as a large mass or a polypoid lesion or may then invade the serosa to mesentery and beyond.

The common symptoms are pain abdomen, nausea, and vomiting and weight loss. Presentation may vary clinically as an abdominal mass, bowel obstruction, perforation, bleeding or intussusception. Many diagnostic tools such as ultrasonography [sausage shaped mass, coiled spring appearance], contrast CT scan, barium studies, angiography and radionuclide scan are available but characteristic 'target mass' appearance on CECT of abdomen is the most sensitive test[15].

The diagnosis is made at laparotomy in children and surgery plays a pivotal role in the management [16,17].Spontaneous bowel perforation from the lymphoma or during surgical manipulation increases the risk of perioperative mortality. Patient should be started on chemotherapy in the immediate post-operative period as it affects the long term survival significantly [17]. Chemotherapy represents a cornerstone in the treatment of these patients and offers an excellent chance for long term disease free survival. Localized disease, low stage disease and complete resection favours survival in lymphoma.

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