

Primary Non-Hodgkin Lymphoma of large bowel presenting as ileocolocolic intussusception in a child - A rare clinical presentation

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Citation

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Abstract

Primary Non-Hodgkin Lymphoma presenting as ileocolocolic intussusception in paediatric age group is a very rare clinical entity. A ten year old male child presented in emergency department with bowel obstruction. Emergency laparotomy revealed ileocolocolic intussusception as the cause of obstruction. Patient underwent right hemicolectomy with restoration of the bowel continuity. Pathologist diagnosed it as diffuse type (B-cell) Non-Hodgkin Lymphoma. Patient is well two months after surgery. He is on chemotherapy.

INTRODUCTION

Intussusception is invagination of a bowel segment, usually proximal, into distal bowel segment. It is a common cause of bowel obstruction in children with a peak incidence at age 3 - 9 months. Between 70% - 95% cases are idiopathic. Change of climate and weaning are related with intussusception in children.

Ileocolic is the commonest variety in children while colocolic variety is the commonest in adults. Most intussusceptions in adults are pathological and malignancy is the commonest cause. Most are diagnosed intraoperatively.

Primary Non-Hodgkin lymphoma of bowel may present as mass, obstruction or perforation. Here we report a case of a ten year old child who presented with clinical features of acute bowel obstruction with mass in right hypochondrium that was found to be ileocolocolic intussusception on exploration. Biopsy and further investigation showed primary Non-Hodgkin Lymphoma (B-cell variety) of large bowel (caecum and ascending colon) and terminal ileum. On extensive review of indexed literature only a single case of Non Hodgkin Lymphoma presenting as ileocecolic intussusception could be found. However in this reported case the affected part of the bowel with lymphomatous involvement was terminal ileum only^[1]. To the best of our knowledge this is the first case of its kind to be reported.

CASE REPORT

A ten year old male child presented to surgery emergency with history of severe pain in abdomen, vomiting, fever, inability to pass faeces and flatus for last three days. On clinical examination patient had fever (101°F), high pulse rate (110/min), normal blood pressure (106/70 mm of Hg), and respiratory rate (20/min.). There was no icterus, cyanosis or peripheral lymphadenopathy.

Respiratory and cardiac systems were normal. A hard tender mass 12x11 cm was palpable in right hypochondrium. Right iliac fossa was empty. There was no free fluid in abdomen and digital rectal examination was unremarkable. Bowel sounds were exaggerated.

Lab investigations revealed TLC 20,000/cmm with normal renal and liver function tests. Chest X-ray was normal. CECT abdomen revealed a large heterogeneously enhancing phlegmonous mass extending from right iliac fossa to right hypochondrium with extensive fat stranding in the surrounding mesentery, thickening of the fascial planes and loculated collection surrounding the mass with moderate right hydronephrosis. Terminal ileum, caecum and ascending colon were not seen separate from the mass. A streak of air density was seen extending into the mass. Possibility of acute ileocolic intussusception or an inflammatory mass was suspected (Fig. 1, 2).

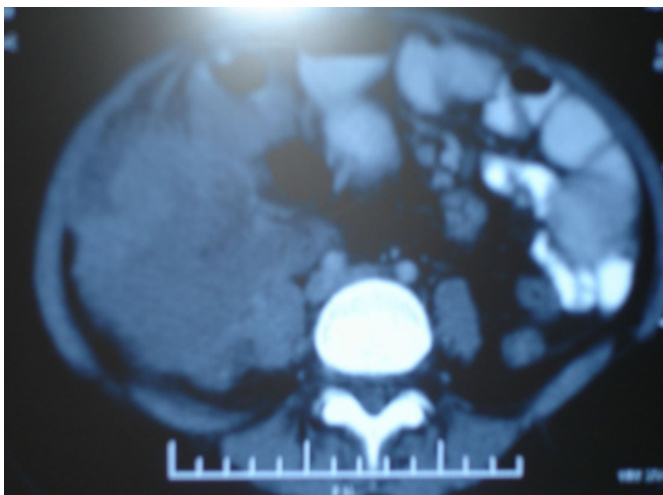
Figure 1

Figure 1: Coronal section of contrast CT abdomen showing phlegmonous mass in right hypochondrium



Figure 2

Figure 2: Axial section of contrast CT abdomen showing a heterogeneously enhancing large mass with fat stranding and enlarged mesenteric nodes



At laparotomy, a 12x12x8cm hard mass was found in right hypochondrium reaching upto the hepatic flexure. Minimal ascites was present. Mesenteric nodes were enlarged. Right ureter was entangled in the mass and was difficult to separate. Right hemicolectomy was performed; the involved ureteric segment was excised en-block and end-to-end

anastomosis was done over a D-J stent.

Cut section of the specimen showed intussusception of terminal ileum into the caecum which was intussuscepting into the ascending colon. There was patchy necrosis of caecum (Fig. 3).

Figure 3

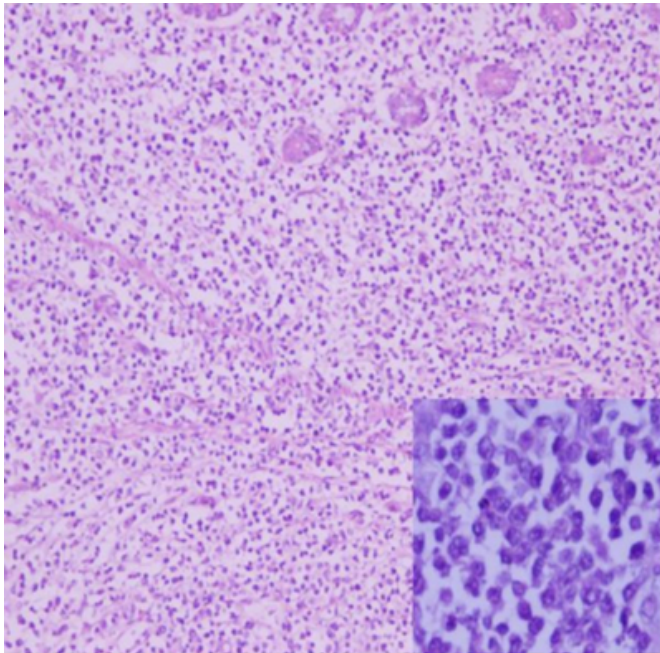
Figure 3: Photograph showing the resected specimen exhibiting intussusception of terminal ileum into the large bowel



Histopathological examination revealed diffuse type (B-cell variety, intermediate grade, Working formulation) Non-Hodgkin Lymphoma of colon and terminal ileum with transmural involvement and infiltration into surrounding mesenteric fat and lymph nodes (Fig. 4).

Figure 4

Figure 4: Microscopic photograph showing lymphoma cells infiltrating the mucosa and submucosa of the colon. Inset: higher magnification of lymphoma cells (H&E, 400x)



On immunohistochemistry CD-20 count was positive. Postoperatively patient made an uneventful recovery. In view of histopathological findings patient was subjected to CECT of thorax, and bone-marrow biopsy; both were normal. PET-CT scan showed minimal residual disease in right iliac fossa.

Patient is receiving Vincristine, Adriamycin, Cyclophosphamide & Prednisolone based chemotherapy at department of Oncology and is well two months postoperatively.

DISCUSSION

Lymphomas of the gastrointestinal tract are the most common type of primary extra-nodal lymphomas^[2] accounting for 5 to 10% of all non-Hodgkin's lymphomas. Primary lymphoma of the colon is rare and comprises less than 1% of large bowel malignancies.^[3] The peak age for gastrointestinal NHL in children is 5-15 years with male sex preponderance 1.8-2.5 times that of females . Tumor is considered primary when:

Intestinal lymphoma is confirmed on histology,

Palpable peripheral lymphadenopathy and hepatosplenomegaly are absent

There is no evidence of lymphoma on chest x-ray or chest CT scan.

Peripheral blood smear and bone-marrow biopsy are normal.^[4]

The predominant GIT malignant tumor in children is NHL of the distal small bowel and the caecum.^[5] According to the WHO histological classification of NHL in children, B-cell immunophenotypes (Burkitt, Burkitt-like, large B-cell) most commonly predilect the abdomen as a primary site of presentation. Burkitt's NHL is the most common.^[6]

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

The most frequent symptoms are abdominal pain, nausea, vomiting and weight loss. Most of these patients present clinically as an abdominal mass, bowel obstruction, perforation, bleeding or intussusception. A number of diagnostic tools such as ultrasonography, 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.^[7]

In majority of children the diagnosis is made at laparotomy Surgery plays a pivotal role in the management.^[8,10] Spontaneous bowel perforation from the lymphoma or during surgical manipulation increases the risk of perioperative mortality. If patient survives long term survival is significantly reduced due to delay in instituting chemotherapy.^[9] 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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