

Non-Hodgkin's lymphoma of the intestine presenting as intussusception in three children – A Case report

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Abstract: Primary Non Hodgkin's lymphoma of the intestine is the most common site of extra-nodal lymphoma. Lymphoma is the most common malignant tumor of intestine in children. We are presenting three cases of Non-Hodgkin's lymphoma of ileocaecal region presenting as intussusception. Intra-operatively we found a nodular mass in ileocaecal region presenting as intussusceptum in all three cases. Involved segment of bowel was resected. All patients were started chemotherapy [Protocol 842] postoperatively. Post chemotherapy FDG PET scan were done to show treatment response evaluation. Management of this condition is discussed and relevant literature is reviewed.

Keywords: Gastrointestinal lymphoma; Intussusception; Intestinal obstruction; Non Hodgkin's lymphoma

INTRODUCTION:

Intussusception is a common cause of bowel obstruction in children with a peak incidence at age 3 - 9 months. Between 70% - 95% cases are idiopathic. In the older children true pathological lead point may be encountered, that may include lymphomas, meckel's diverticulum, hamartomatous polyp, hemangiomas.

Primary Non-Hodgkin lymphoma of bowel may present as mass, obstruction or perforation. Here we report three children who presented with clinical features of acute bowel obstruction, diagnosed as ileocolic intussusception on ultrasonography. Herewith we discuss management of this condition and relevant literature is reviewed.

Case 1:

A 4 1/2 years old male child presented to the emergency department with acute pain abdomen since 3 days. The pain was colicky in nature with relief for 15 to 20 minutes. There was 1 episode of passing of bloody stool. Parents gave history that patient was complaining of pain in abdomen since 2 months. Pain was episodic, more in umbilical region and subsided with medications. Severity of pain increased since three days. On general examination, the child was afebrile

with diffuse tender abdomen with mobile tender mass in right lumbar region. Per rectal examination revealed bloody stool. There was no generalized lymphadenopathy or hepatosplenomegaly. Total leucocyte count was raised. Abdomen x-ray showed air-fluid levels. Ultrasonography of the abdomen revealed lumen in lumen sign (target sign) suggesting acute intestinal obstruction due to ileocolic variety of intussusception. Patient was taken up for emergency laparotomy. Manual reduction of intussusception failed; so involved segment of bowel consisting of ileocecal junction was resected. On cutting open the specimen a nodular mass was found [fig 1]. Histopathology of the lesion revealed mass composed of monotonous population of immature lymphoid cells infiltrating entire wall. Immunohistochemistry showed tumor cells positive for CD 20, CD 79a, and Negative for CD 3, Tdt and Bcl2. These features were suggestive of high grade Non Hodgkin's lymphoma Burkitt's type. Mesenteric lymph nodes showed reactive hyperplasia. Resected margins were clear. Patient was started chemotherapy [Protocol 842]. Post chemotherapy FDG PET scan were done to show treatment response evaluation. No abnormal tracer uptake noted in bowel mucosa. Liver, spleen, bone marrow were unremarkable.



Fig 1: Nodular mass in resected bowel, cut opened

Case 2:

A 3 years old male child presented to us with acute pain abdomen since previous night. Patient had one episode of vomiting. Parents gave history that patient was complaining of pain in abdomen since 1 month. Pain was episodic, more in umbilical region and subsided with medications. On general examination, the child was afebrile with tenderness in umbilical region. Per rectal examination was normal. There was no generalized lymphadenopathy or hepatosplenomegaly. Total leucocyte count was raised. Abdomen x-ray was normal. Ultrasonography of the abdomen revealed

lumen in lumen sign (target sign) suggesting ileocolic variety of intussusception. Hydrostatic reduction under USG guidance attempted but was unsuccessful. Patient was taken up for emergency laparotomy. Manual reduction of intussusception failed; so involved segment of bowel consisting of ileocaecal junction was resected. A polypoid mass [fig 2] was found which was found to be high grade Non Hodgkin's lymphoma Burkitt's type by histopathology and immunohistochemistry. Patient was given chemotherapy [Protocol 842] and further evaluated by FDG PET scan.



Fig 2: Polypoid mass in resected bowel

Case 3:

A 6 years old female child presented to us with increasing pain in abdomen since six days. Patient had history of vomiting. Parents gave history that patient was complaining of pain in abdomen since 2 months.

On general examination, patient had tachycardia and tachypnoea. A lump was palpable in right iliac fossa region. Per rectal examination was normal. There was no generalized lymphadenopathy or hepatosplenomegaly. Ultrasonography of the abdomen

revealed intestinal obstruction due to ileocolic variety of intussusception. Patient was taken up for emergency laparotomy. Manual reduction of intussusception failed; so involved segment of bowel consisting of ileocaecal junction was resected. An ileocaecal mass was found

which was found to be high grade Non Hodgkin's lymphoma Burkitt's type by histopathology and immunohistochemistry.(fig 3) Patient was given same chemotherapy and further evaluated by FDG PET scan.



Fig 3: Ileocaecal mass

DISCUSSION

Primary lymphoma of the small bowel accounts for less than 2% of the gastrointestinal malignancies and 10-20% of bowel malignancies [1]. They arise from the lymphoid tissue in the small bowel, and are more common in the ileum owing to the presence of lymphoid Payer's patches. 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. The peak age for gastrointestinal NHL in children is 5-15 years with male sex preponderance 1.8-2.5 times that of females. Dawson has given criteria for the small bowel lymphoma to be described as primary [3]. It includes no mediastinal or peripheral lymphadenopathy, no spleen and hepatic involvement, normal white blood cell and differential counts, predominant intestinal involvement. Our three cases discussed meet all these criteria, hence described as primary lymphoma. Lymphoma arises in the lymphoid follicles of the sub mucosa 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, and vomiting and weight loss. Our patients had abdominal pain as most common symptom. Most of these patients present clinically as an abdominal mass, bowel obstruction, perforation,

bleeding or intussusceptions. All three cases in present report had intussusception as presentation. In review of the literature, the outcome of primary Non Hodgkin's lymphoma presenting as intussusception in children, two authors opine indifferent ways. In Ein SH et al. study only 3 out of 10 children were long term survivors [4]. On the contrary Pure P. *et al.*; reported only one death in entire series of 292 children with primary lymphoma of the intestine presenting as intussusception [5]. For localized disease surgery is the main modality of treatment, followed by adjuvant radiotherapy or chemotherapy. Complete resection of the involved segment along with enlarged mesenteric lymph node is important in long term survival as shown in all three cases in the present report. Complete resection of the bowel has the main advantage of reducing complication like bowel perforation, bleeding or the tumour lysis syndrome during chemotherapy phase [6].

CONCLUSION

Whenever a child between 3-12 years age group presents with acute abdomen due to intussusception high index of suspicion should be there for the presence of primary lymphoma of the intestine. Complete resection of the localized disease along with involved mesenteric lymph nodes followed by adjuvant

systemic chemotherapy is the main management of primary Non Hodgkin's lymphoma of the intestine.

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