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Case Report

An Atypical Presentation of Ileocolic Intussusception Secondary to Burkitt's Lymphoma

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ABSTRACT

Burkitt's lymphoma is an uncommon cause of intussusception. Intussusception rarely occurs after the first 6 years of life. Etiology of childhood intussusception differs depending on age at presentation. Here we present an interesting case of a 7-year-old male child who presented with pain in abdomen and constipation since 15 days with distension of abdomen since 2 days. X-ray abdomen revealed multiple air fluid levels suggestive of intestinal obstruction. On Ultrasonography and CT scan abdomen, diagnosis of ileo-colic intussusceptions was made. Patient was posted for laparotomy and resection and anastomosis of the involved segment was done. Histopathological examination revealed features suggestive of Burkitt's lymphoma. Post operatively patient was given chemotherapy. Patient was followed up post operatively and post chemotherapy for six months and has no complaints at present.

Keywords: Burkitt's lymphoma, Intussusception, Intestinal obstruction, Ileocolic.

INTRODUCTION

Intussusception is the most common cause of intestinal obstruction in infants between 6 and 36 months of age. Approximately 60 percent of children are younger than one year old, and 80 to 90 percent are younger than two years. Intussusception is less common before three months and after six years of age. Intussusception caused by Burkitt lymphoma as a cause of acute abdomen is rare, with often misleading symptoms that make the diagnosis more difficult. [1] Many paediatric patients with Burkitt lymphoma present with intussusception as a first clinical sign, a presentation that potentially leads to the disease detection at an earlier stage. [2] The gastrointestinal tract is the most common extra-nodal site of, [3] Burkitt's lymphoma is a diffuse, undifferentiated, malignant monoclonal B-cell lymphoma with two major clinical presentations. American Burkitt's differs from the African type described by Burkitt in 1958 in its increased propensity for widespread involvement, especially within the abdominal cavity. [4] Pathologically, Burkitt's lymphoma and enteropathy-associated T-cell lymphoma, with MALT type are the most common lymphoma of the small intestine and account for 42.5% of the lymphomas. [5]

We hereby present a case of ileocolic intussusception in a 7 year old male child patient due to intestinal Burkitt's lymphoma.

PRESENTATION OF CASE

A 7 year old male child presented to surgery department of our hospital with

chief complaints of pain in abdomen, constipation since 15 days and distension of abdomen since 2 days. Patient was apparently alright 15 days back when he started complaining of abdominal pain, insidious in onset gradually and progressive with no relieving factors. Patient complained of aggravated pain since 2 days with complaints of not passing stool. Patient had no complaints of nausea, vomiting, malena or hematochezia.

On physical examination, vitals were normal. Patient's abdomen was distended with hyperperistaltic bowel sounds and mild tenderness all over the abdomen. Mass could be appreciated on palpation in right lumbar region. Per rectal examination was within normal limits. The patient was hypoproteinemic but not hypoalbuminemic.

Ultrasonographic examination of abdomen and pelvis showed evidence of heterogenous mass of approximate size 7.1x 4.6 cm in right hypochondrium giving bowel within bowel appearance and target sign with evidence of vascularity on doppler imaging suggestive of ileocolic type of intussusception with free fluid in abdomen and pelvis.

CT scan abdomen revealed heterogenously enhancing hypodense mass of approximate size 12 x 10 x 8.5 cm in right abdomen involving terminal ileum, ileo-caecal junction, caecum, ascending colon, hepatic flexure and proximal part of transverse colon forming a loop within loop appearance suggestive of ileo-colic intussusception with right pleural effusion. (Figure 1) Patient underwent exploratory laparotomy for the same. Ileocolic intussusception was reduced. Thickened omental lymph nodes were present of size approximately 3 x2 cm. In terminal part of ileum 3 x 3 cm size lymph node was present in mesentery with thickened mesentery. Biopsy was taken from omental lymph node. Right hemicolectomy with end to end anastomosis done. Right hemicolectomy and biopsy specimen was sent for histopathological examination which revealed Non-Hodgkin's lymphoma, diffuse high grade (Burkitt like) (Figure 2 and Figure 3)

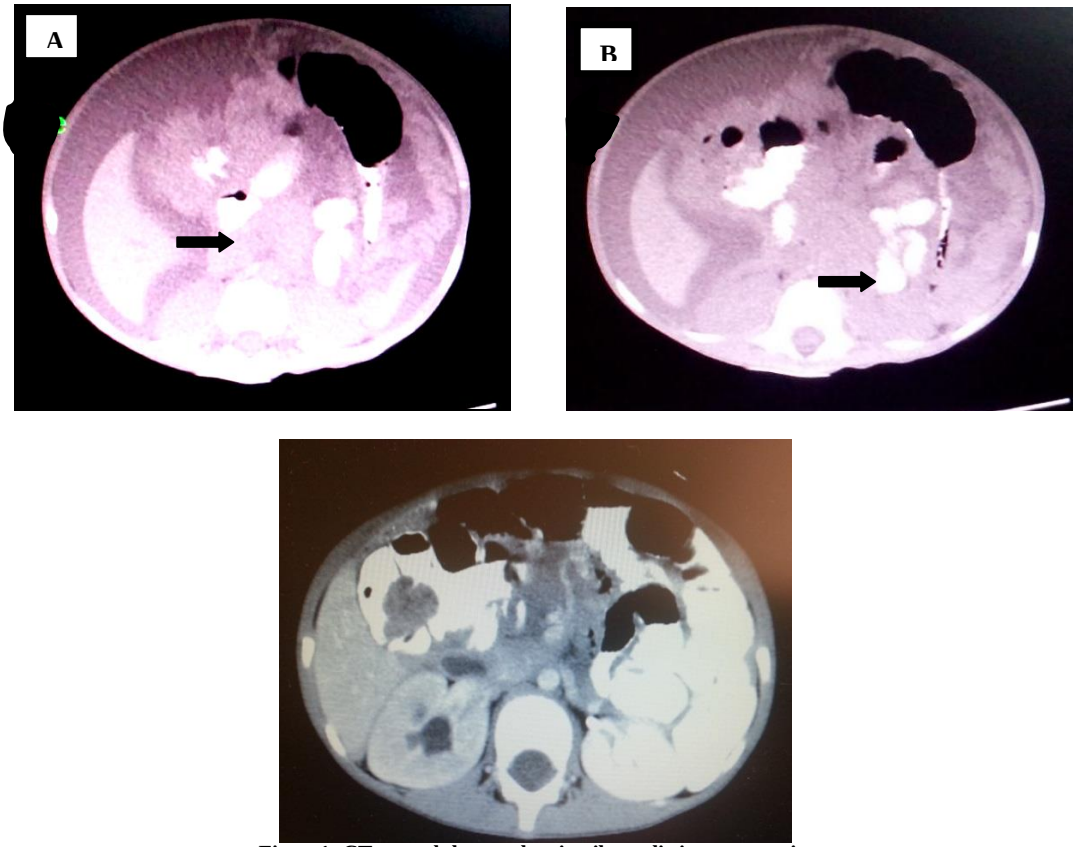

Figure1: CT scan abdomen showing ileo-colic intussusceptions

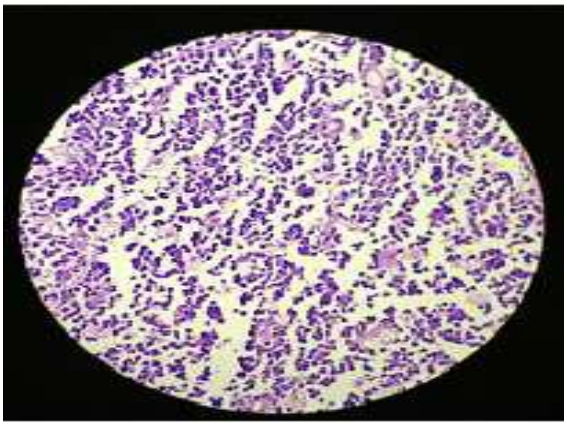

Figure 2: Histopathological appearance of Burkitt's lymphoma

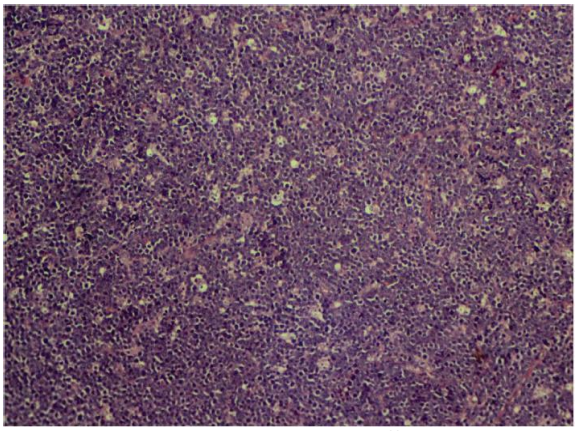

Figure 3: Lymphoma cells arranged in diffuse sheets admixed with numerous macrophages giving a "starry sky" pattern (H&E, x100)

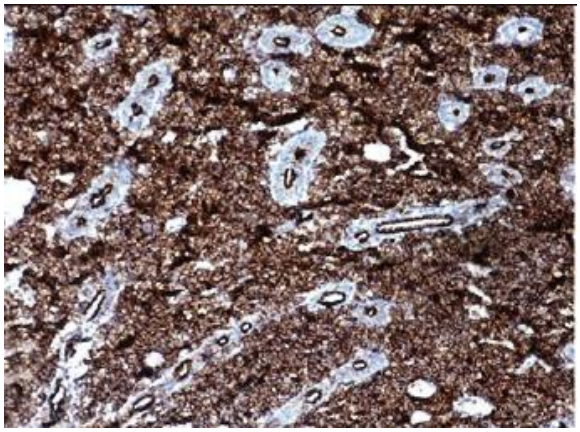

Figure 4: Photomicrograph showing positivity for CD10 (immunohistochemistry [IHC], ×100

Immunohistochemistry was done for confirming the diagnosis. Immunophenotype of malignant cells revealed: CD20 (+), CD10 (+), TdT (-), Bcl2 (-), and CD5 (-) with about 100% proliferative indexes for Ki-67 (Figure 4). The final histopathological diagnosis was Burkitt's lymphoma

Postoperatively patientwas administered chemotherapy-CHOP regimen [cyclophosphamide, doxorubicin hydrochloride, vincristine and prednisone]. Post-operatively patient was followed for six months, had no complaints and was doing well.

DISCUSSION

Burkitt Lymphoma (BL) is a high grade Non-Hodgkins Lymphoma (NHL) that is characterized histopathologically by a mass of diffuse small non-cleaved B cell lymphocytes. [6,7] BL is of greatest importance in sub-Saharan Africa where it is the most common childhood (2-16 years, mean 7 years) cancer, accounting for up to 36% of childhood cancers and 70% of childhood lymphomas [8] but, overall constitutes 5% of lymphomas for both adult and childhood populations.

NHLs are generally clonal malignancies of the multiple cellular components of the normal lymph node, spleen and thymus. Burkitt lymphoma arise as a clonal transformation occurring at specific stage of normal B cells differentiation during antigenic stimulation in accordance with Murphy's law which holds that for every cell type and stage of differentiation there will occur neoplastic counterparts. [9]

The WHO recognizes the Classical (Endemic) Burkitt and two other variants.

African or Endemic Burkitt lymphoma: The actual age range of occurrence of endemic Burkitts is 2-16 years but the most common age of occurrence is 4-7 years with a male: female ratio of 2:1. The lesion usually involves bones of jaw and other facial bones, as well as kidneys, gastrointestinal tract, ovaries and other extra-nodal sites. [10]

-African or sporadic: It accounts for 1-2% of lymphoma in adults and up to 40% of lymphoma in the children in the US and Western Europe. Lymph node involvement is more common among adults than amongst children. [11] Rarely, patient may also have malignant pleural effusion and

ascites. [12] The disease has many forms of clinical presentations which is nonspecific and the patient may have symptoms such as colicky abdominal pain, nausea, vomiting, distension. It is diagnosed in children usually as an abdominal mass. The tumour causes either indirect symptoms, due to pressure phenomena or direct involvement of the bowel lumen leading to either intestinal obstruction or intussusceptions.

Intussusception is one of the most common emergencies in children that require the attention of a surgeon. Especially in older children, in 10% of the cases, there is a pathological lesion (Meckel's diverticulum, polyp, and lymphoma) at the lead point of the intussusceptum. [13] Intussusception is the invagination of a bowel loop with its mesenteric fold (intussusceptum) into the lumen of a contiguous portion of bowel (intussuscipiens) as a result of peristalsis. [14] Intussusception is primarily a disease of infants and children with 90% showing no specific cause. [15] The classic clinical triad described for intussusception is made up of abdominal colic, "red currant jelly" stools and a palpable abdominal mass; in children older than 2-3 years the presentation of intussusception is subtler and the classic triad of symptoms may not be present. [16] Ultrasonography is the most efficient examination for the diagnosis of intestinal intussusceptions with a reported accuracy of up to 100%, and combined with clinical observation, is usually sufficient to diagnose the intussusceptions. [17] There are several proposed tools to aid in the diagnosis of intussusceptions. However, CT scan remains the most useful radiologic method for diagnosing intestinal intussusceptions according to several studies. [14,18]

Surgery, chemotherapy, radiotherapy and radioimmunotherapy are the different modalities for the management of GI lymphoma and can be applied in different combinations. [18] The successful outcome depends on early postoperative diagnosis and aggressive chemotherapy with supportive care since the clinical picture is nonspecific.

Our case presentation is the first case of Burkitt's lymphoma with abdominal involvement in a child, causing ileocolic intussusception diagnosed here in rural area like Sewagram.

CONCLUSION

In our case we report a child who presented with ileo-colic intussusception secondary to diffuse high grade Burkitt's lymphoma, was managed by resection of the involved segment followed by adjuvant chemotherapy. Burkitt's lymphoma of the ileum is a rare clinical entity in surgical practice. Our case report highlights the importance of identifying pathological lead points in older children and more importantly the fact that Burkitt's lymphoma should be considered a possibility in the Indian population. Children tend to do well with limited surgical resection and chemotherapy.

Adequate surgical resection, combined with modern regimens of intensive chemotherapy, has reduced the complication rate and overall morbidity of this disease. However, the diagnosis should be made promptly, and intussusceptions should be considered at all ages in children presenting with recurrent colicky abdominal pain. The presence of pathological lead point of an intussusception should be sought in patients over 2 years of age.

Conflict of Interests

The authors declare that there is no conflict of interests regarding the publication of this paper.

Author's Contribution

Dr. Mangesh Hivre is the main author, drafted the manuscript and collected data related to the subject. Dr. Bhupendra Mehra and Dr. Dilip Gupta gave advice on tropical perspective, Dr. Akshay Pednekar, Dr. Tushar Nagtode and Dr. Anurag Yadav helped with literature search. All authors contributed equally to this work.

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