Case Report

Burkitt's lymphoma masquerading as intestinal obstruction: An uncommon entity with variable clinical presentation

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ABSTRACT

Small bowel lymphoma is a comparatively rare disease. Usually neither typical nor explicit symptoms are determined during its course and the first manifestation can be noticed when a complication occurs. Burkitt's lymphoma is a high-grade, aggressive and rapidly growing B-cell neoplasm, which has low long-term survival rates. Sporadic Burkitt's lymphoma accounts for 1-2% of lymphomas in adults and for 40% of lymphomas in children in the worldwide population. The abdomen is the most frequent site of onset in nonendemic (sporadic) Burkitt's lymphoma. Symptoms are often misleading and make diagnosis difficult. We present a case of a 5-year-old male child who presented with symptoms of acute intestinal obstruction. Emergency therapeutic and diagnostic laparotomy was performed and biopsy from thickened ileal wall was taken. Histopathologically, it was diagnosed as non-Hodgkin's lymphoma possibly Burkitt's lymphoma and confirmed on immunohistochemistry.

Key words: Burkitt's lymphoma, obstruction, small intestine, sporadic

INTRODUCTION

Small intestine tumors constitute 2-6% of all neoplasms in gastrointestinal tract. The most cases of benign tumors are leiomyoma, adenoma, lipoma, hemangioma, neurogenic tumors, polyps, etc., The malignant tumors are presented by their four forms: Adenocarcinoma, sarcoma, lymphoma, and carcinoid.[1] Small intestinal lymphoma is a comparatively rare disease, consists of <1-2% of all malignant tumors of gastrointestinal tract.[2] Burkitt's lymphoma presents an undifferentiated, highly malignant tumor of B-lymphocyte. Since its discovery in 1958, three forms of the disease have been recognized; an endemic variety confined to the African continent and nonendemic, sporadic American forms, and third form in immunodeficient patients. Sporadic Burkitt's lymphoma accounts for 1-2% of lymphoma in adult and up to 40% lymphoma in children in United States and Western Europe, with abdomen as most frequent site.[3,4]

Access this article online	
Quick Response Code:	Website: www.ccij-online.org
	DOI: 10.4103/2278-0513.138090

CASE REPORT

A 5-year-old male child presented to Pediatric Surgical Department of our institute with the chief complaints of pain, abdominal distension, and abdominal fullness since 1 month. The abdominal distension rapidly increased for the last week along with onset of fever and vomiting. There was no past history of similar complaints.

The patient's height was 100 cm and his weight was 18 kg. On examination, patient had fever, high pulse rate with normal blood pressure and respiratory rate. There was no icterus, cyanosis, or peripheral lymphadenopathy. The systemic examination of cardiovascular system, respiratory system, and central nervous system was within normal limits. Perabdominal examination revealed abdominal pain and distension. No definite lump was palpable.

Ultrasonography of abdomen revealed mildly dilated small gut loops with collections in peritoneal cavity. No lymphadenopathy seen. Other abdominal organs were normal. Radiologically, he was diagnosed as a case

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of subacute intestinal obstruction with peritonitis and contrast-enhanced computed tomography (CECT) abdomen was advised. CECT abdomen revealed diffuse peritoneal thickening in the mid and lower abdomen with thickening of small bowel loops and thickened stomach wall with hypodense texture [Figure 1]. The possibilities of lymphoma and peritoneal mesothelioma were kept; and biopsy was advised with clinical correlation.

The child was diagnosed as acute abdomen secondary to a malignant pathology in intestine and an emergency laparotomy was done under general anesthesia. At laparotomy, the small intestinal wall was diffusely thickened, which was extending up to the stomach, making the patient difficult to operate. Biopsy from the thickened ileal wall performed and abdomen was closed in view of impossibility of resection and spread out disease. The biopsy was sent to the Department of Pathology of our institute for histopathological examination.

The postoperative period was uneventful.

Pathological finding

Gross examination

we received a wall like piece measuring 7 × 5 cm with wall thickness of 0.5-0.8 cm. External surface showed areas of congestion. Internal surface showed growth involving the whole piece reaching up to the resected margins.

Histopathological examination showed structure of small intestine revealing diffuse infiltration with a neoplastic lymphoid infiltrate. The infiltrate composed of intermediate to large sized cells, with round to slightly irregular nuclei. The nuclei displayed a clumped chromatin pattern and have one or several peripherally localized nucleoli [Figure 2]. The cells had a scant amount of basophilic cytoplasm. Many mitotic figures and apoptotic cells along with few macrophages were also evident. Based on this microscopic finding, diagnosis of lymphoma was made with possibility of Burkitt's lymphoma.

Immunohistochemistry was done for confirming the diagnosis. Immunophenotype of malignant cells: CD20(+), CD10(+), TdT(−), Bcl2(−), and CD5(−) with about 100% proliferative index for Ki-67 [Figure 3]. The final histopathological diagnosis was Burkitt's lymphoma of the small intestine.

DISCUSSION

Primary non-Hodgkin's lymphoma (NHL) of gastrointestinal tract is the most common extra-nodal lymphoma in pediatric age group. Yet, the overall incidence is very low. The rarity of the disease as well as variable clinical presentation prevents early detection when the possibility of cure exists.[5]

First described by Dennis Burkitt in 1958, Burkitt's lymphoma is a highly aggressive NHL often presenting in extra-nodal sites or as an acute leukemia.[6] Burkitt's lymphoma usually is encountered within young people and

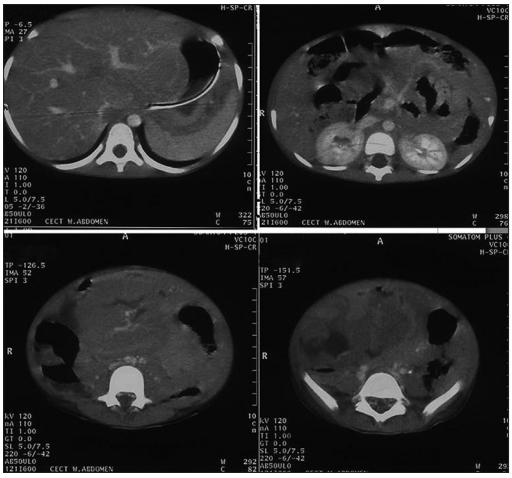

Figure 1: Contrast-enhanced computed tomography (portal venous phase) shows thickened stomach wall and thickening of the small bowel loops

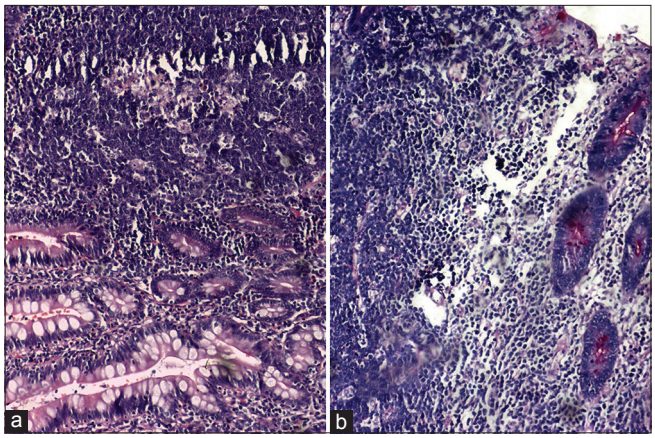

Figure 2: Photomicrograph showing diffuse infiltration of intermediate to large sized cells with round to irregular nuclei with clumped chromatin and one to several peripherally localized nucleoli (a) H and E, ×100 (b) PAS, ×100

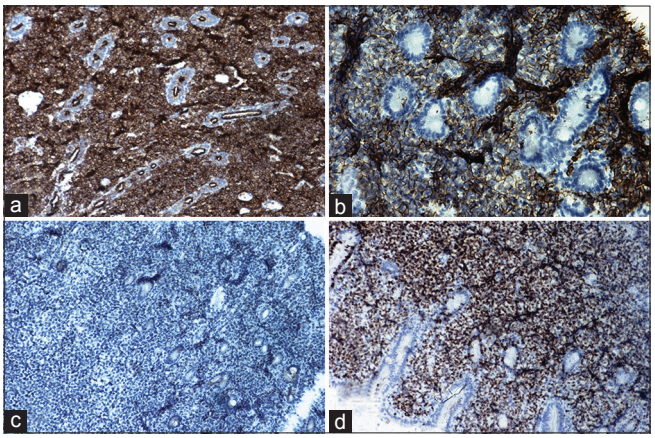

Figure 3: Photomicrograph showing (a) positivity for CD10 (immunohistochemistry [IHC], ×100), (b) positivity for CD20 (IHC, ×200), (c) negativity for TdT (IHC, ×100), and (d) 100% proliferation index for Ki-67 (IHC, ×200)

is the most frequent type of the juvenile NHL. Most often it is located in ileum and ileocecal angle and consists only 5% of the small bowel lymphoma.[1] Burkitt's lymphoma is a B-cell lymphoma genetically characterized by a chromosomal translocation that results in deregulation of the c-myc oncogene.[6]

Three variants of Burkitt's lymphoma have been described as: Endemic (largely found in Africa), sporadic (nonendemic) subsequently described outside Africa, affecting mainly abdominal viscera and a third variant in immunodeficient patients. The endemic (African) and nonendemic (American) forms are immunologically similar, but difference exists in their anatomical and epidemiologic manifestations.[2]

In endemic areas, facial bones particularly jaws, maxilla, and orbit are involved particularly in young children and associated with Epstein-Barr virus infection, as well as frequent contaminant malarial infection. In comparison, the sporadic form tends to present in the lymphoid tissues of the gut, often presenting as masses in the Waldeyer's ring or the terminal ileum, or even with involvement of abdominal organs with the most common involvement of the distal ileum, cecum or mesentery.[7-9]

Patients with gastrointestinal Burkitt's lymphoma may present with abdominal pain or distension, gastrointestinal bleeding or intestinal obstruction resulting from direct compression of the lumen by an expanding mass or by an intussusception triggered by intraluminal projection of the tumor mass. These acute abdominal symptoms often lead to emergency laparotomies before a diagnosis of Burkitt's lymphoma can be made.[10]

Burkitt's lymphoma begins submucosally, presumably in lymphoid follicles or Peyer's patches, and grows submucosally as a soft, diffuse, infiltrating lesion that leaves the mucosa intact in early stages. Later, ulceration of mucosa may occur. Regardless of the extent of the lesion or its appearance, the involved bowel remains strikingly pliable, so that a lesion that looks as if ought to be obstructing the entire bowel may not particularly trouble the patient.[11]

Our patient had a clinical presentation of acute abdomen with abdominal pain and distension secondary to the diffuse thickening of the intestinal wall due to Burkitt's lymphoma in a 5-year-old male child. In view of impossibility of resection and spread out disease, surgery was not carried out. Diagnostic biopsy from thickened ileum was performed, which histopathologically and immunohistochemically confirmed as Burkitt's lymphoma of small intestine.

The characteristic morphological features (diffuse infiltration of all layers of intestine with a neoplastic lymphoid infiltrate consisting of intermediate to large sized cells, with round to slightly irregular nuclei displaying clumped chromatin pattern with one or several peripherally located nucleoli; and scant amount of basophilic cytoplasm along with many mitotic figures as well as many apoptotic cells giving starry sky appearance) along with immunohistochemical profile of the neoplastic cells (CD20+, CD10+, TdT−, Bcl2−, CD5−, and Ki-67 proliferation index of about 100%) helped us to confirm the Burkitt't lymphoma and to exclude the possibilities of lymphoblastic lymphoma and diffuse large cell lymphoma.[1,2]

CONCLUSION

Burkitt's lymphoma of the ileum is a rare clinical entity in surgical practice. The clinical picture is nonspecific. Preoperative diagnosis is difficult and may be suggested by ultrasound or computed tomography scan, and in some cases the presence of tumor mass may be an intraoperative surprise. Histopathological and immunohistochemical examination of paraffin embedded sections from the tumor is equally important for the confirmation of diagnosis; and for correct treatment and determining prognosis.

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Cite this article as: Gupta V, Sangwaiya A, Sharma J, Kaira V, Ratan K, Sen R. Burkitt's lymphoma masquerading as intestinal obstruction: An uncommon entity with variable clinical presentation. Clin Cancer Investig J 2014;3:441-3.
Source of Support: Nil, **Conflict of Interest:** None declared.