



Case Report

## Sporadic Burkitt Lymphoma of the Small Bowel Presented As Acute Abdomen

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### ABSTRACT

Burkitt lymphoma is high grade, aggressive, rapidly growing B- cell neoplasm. Sporadic Burkitt lymphoma accounts for 1 -2% of lymphoma in adult and upto 40% of lymphoma in children in the world wide population. The abdomen is the most frequent site of onset in non-endemic (sporadic) Burkitt lymphoma.

We are presenting a case of a 6 year old female presented with acute abdomen as pain, distention of abdomen, nausea and vomiting with problem of defecation. Imaging studies suggestive of intussusception of small bowel as the cause of acute abdomen. Emergency therapeutic and diagnostic laparotomy and biopsy of appendicular mass was performed. Histopathologically it proved as diffused small non cleaved [Burkitt lymphoma] and confirmed on immunohistochemistry.

Small intestinal intussusception as acute abdomen caused by Burkitt lymphoma is a rare clinical entity.

**Key words:** Burkitt lymphoma, Sporadic , Acute abdomen.

### INTRODUCTION

Small intestinal tumor consist of 2 - 6% of all neoplasm in gastrointestinal tract.

<sup>[1]</sup> Small intestinal lymphoma is a comparatively rare disease, consist of less than 1 – 2 % of all malignant tumor of gastrointestinal tract. <sup>[1]</sup>

Burkitt's lymphoma presents an undifferentiated, highly malignant tumor of the 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. <sup>[2]</sup> Sporadic Burkitt's lymphoma accounts for 1 -2% of lymphoma in adult and up to 40% of lymphoma in children in the United States and Western Europe. <sup>[3]</sup>

Burkitt's lymphoma is an aggressive, highly malignant and rapidly growing B cell neoplasm. It frequently presents in the abdomen in nonendemic, sporadic Burkitt lymphoma regions. <sup>[3]</sup>

Herein, we are presenting a case of a 6 year old female presented as a rare clinical entity of acute abdomen due to intussusception caused by Burkitt's

lymphoma. The clinical diagnosis is difficult as Burkitt's lymphoma is very nonspecific in its presentation.

## **CASE REPORT**

A 6 year old female child presented to surgical department of our hospital with chief complaints of pain in abdomen, fullness of abdomen since 2 months. Patients gave history of fever and problem with defecation since 3 -4 days. There was past history of similar complaints with nausea and vomiting 1 month back. On clinical examination patient had fever, high pulse rate, pallor with normal blood pressure and respiratory rate. There was no icterus, cyanosis, or peripheral lymphadenopathy. Systemic -RS, CVS and CNS examination were within normal limits. Per abdominal examination revealed distended abdomen with lump in right iliac fossa m. 5x5 cms and other lump in the lumbar region m. 3x2 cms, both were firm in consistency. Per rectal examination was not significant. The emergency laboratory test were as Hb – 7.5 gm/dl, BT,CT,PT and INR were within normal limits. Urine showed no abnormality. HIV and HBsAg were nonreactive. Ultrasonography of abdomen revealed concentric lump of bowel in umbilical region suggestive of intussusception with minimal free fluid in abdomen. Mild hepatomegaly was also seen. Clinically diagnosed as acute abdomen due to intussusception with intestinal obstruction, a emergency laparotomy was done under general anesthesia. At laparotomy, large entangled whitish mass with adhesion noted at ileocecal junction which was difficult to operate. The origin was not found in view of entangled mass but a protruded mass was noted [?appendix] and it was resected. Multiple grey white adherent mesenteric lymph nodes were also seen. Abdomen closed in layers in view of impossibility of resection and spread out

disease. The resected protruded mass was sent for histopathological examination. Post-operative period was uneventful. The patient was referred to higher center for chemotherapy.

### ***Pathological findings:***

#### ***Gross***

We received single whitish mass m.9x4x2cms .Externally showed elongated appearance with congested blood vessels [Fig. 1]. On cut section, grayish white homogenous appearance with smooth consistency was noted. At one end of the mass showed lumen (? Appendicular) m.1.2 cms in diameter with adjacent homogenous whitish appearance with friable consistency. [Fig. 2]

#### ***Microscopy***

Multiple sections showed small intestinal mucosa with tumor underneath it. The epithelium was free from tumor. Tumor was seen reaching upto the payers' patches[Fig.3]. Tumor composed of diffuse infiltrate of monotonous small to medium sized lymphoid cells with round to oval nuclei and 2 -3 small nucleoli with scant basophilic cytoplasm.[Fig. 4] Few macrophages with characteristic 'starry sky' appearance was noted [Fig.5]. Few mitotic figures with vascular and lymphatic emboli were seen. Neoplastic lymphoid cells in one bit showed infiltration throughout the muscular layer. Light microscopic diagnosis was given as Diffuse, small non cleaved lymphoma with differential diagnosis of Burkitt lymphoma and MALT lymphoma.

Immunohistochemistry was done for further confirmation. Lymphoma cells exhibits immunopositivity for CD10 and CD 20 with 100% proliferative index for Ki 67 [Fig. 6]. The lymphoid cells were immunonegative for Cyclin D1, BCL 2, TdT and CD3[Fig.7] ,CD 21and SOX 11. Final histopathological impression given was as Burkitt lymphoma of the small intestine.



Figure 1 – External surface of excised elongated protruded whitish mass with congested blood vessels.



Figure 2 – Cut section showed grey white homogeneous mass with lumen of entrapped appendix.

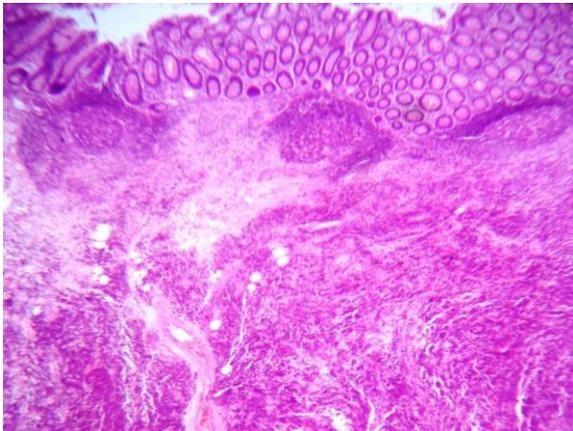


Figure 3- Light microscopy showed small intestinal mucosa and underneath showed tumor. (H &E stain, x100).

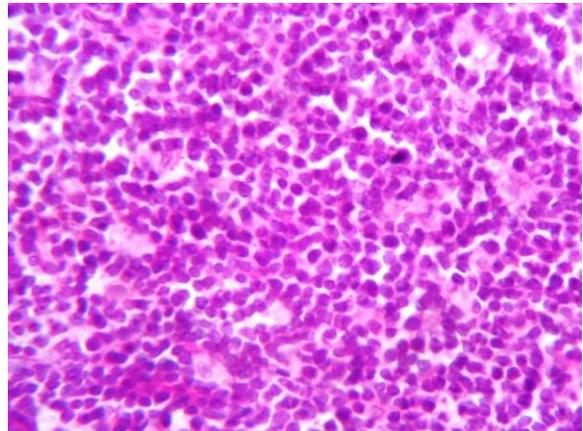


Figure 4 - The tumor composed of diffused infiltrate of monotonous small to medium sized lymphoid cells with round to oval nuclei and with small nucleoli. (H &E stain, x400).

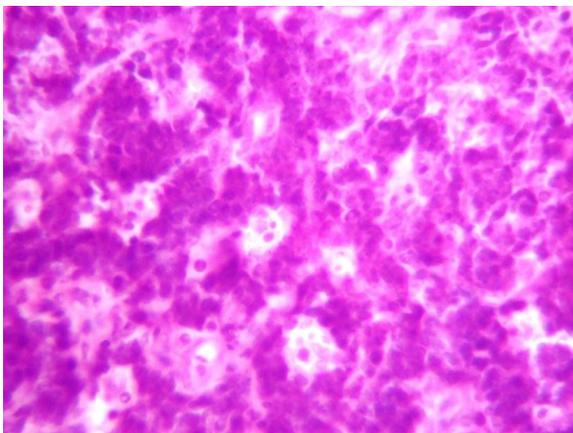


Figure 5 – Individual monomorphic lymphoid cells and few macrophages with characteristic 'starry sky' appearance (H &E stain, x400)

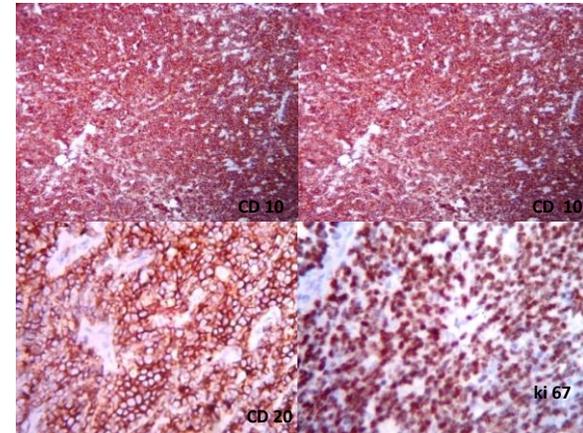


Figure 6- The tumor cells show diffuse immunoreactivity for CD 10, CD20 and Ki-67.

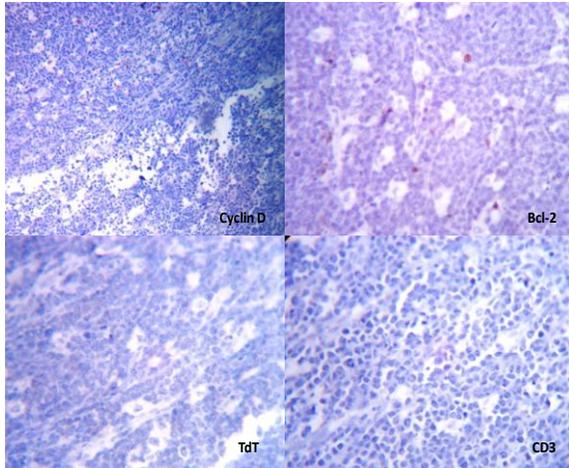


Figure 7- The tumor cells show immuno-nonreactivity for Cyclin-D, Bcl-2, TdT and CD 3.

## DISCUSSION

The gastrointestinal tract [GIT] is the most commonly involved site of extra nodal lymphomas.<sup>[4]</sup> The close association between chronic inflammation and specific GIT lymphomas not only provide interesting insight into the pathobiology of lymphomas but also poses unique diagnostic challenges.<sup>[4]</sup>

Burkitt lymphoma is highly aggressive non-Hodgkin lymphoma [NHL] often presenting as extra nodal site or as acute leukemia.<sup>[5]</sup> Burkitt's lymphoma was first described by Dennis Burkitt, British surgeon in the African malarial belt as round cell sarcoma in the jaws of African children.<sup>[5, 6]</sup>

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 --third variant in Immunodeficient patients. The endemic [African] and nonendemic [American] forms are immunologically similar, but difference exists in their anatomical and epidemiologic manifestations.<sup>[7]</sup>

In endemic areas, facial bones particularly jaws, maxilla and orbit is involved particularly in young children and associated with Epstein Barr Virus infection,

as well as frequent contaminant malarial infection.<sup>[7,8,9]</sup> In comparison, the sporadic form mainly present in lymphoid tissues of the gut, often presenting as masses in waldeyers ring or the terminal ileum, caecum or mesentery.<sup>[7, 9]</sup> Specifically, sporadic form most commonly present in the areas of ileocecal valve. This phenomenon is thought to be related to the influence of immunoproliferative lymph nodes and payer's patches in this area.<sup>[2, 5, 7]</sup>

The sporadic form of Burkitt's lymphoma commonly presents with abdominal swelling as a large mesenteric retroperitoneal pelvic mass, tenderness, pain or fullness.<sup>[5, 7]</sup> Some patient presents with symptoms of bowel obstruction secondary to ileocecal intussusception caused by tumour growth, obstruction or bleeding, mimicking acute appendicitis.<sup>[9, 10, 11]</sup>

Our patient had a clinical presentation of acute abdomen with abdominal pain, distention and obstruction, secondary to intussusception due to solitary Burkitt lymphoma in a 6 year old female child. In view of impossibility of resection and spread out disease, surgery was not carried out. Diagnostic biopsy of elongated protruded mass from the ileocecal region was carried out, which histopathologically and immunohistochemically confirmed as Burkitt's lymphoma. The patient was referred to higher center for chemotherapy in our case.

The primary treatment of Burkitt lymphoma is chemotherapy.<sup>[2,12]</sup> Several studies have suggested that sporadic variety may be best managed by combined modality of chemotherapy and surgical extirpation.<sup>[1, 2, 12]</sup> Before the era of combination chemotherapy, Burkitt lymphoma was an almost universally fatal disease. Rare cases of spontaneous remission have been described in African children but in most of the cases the outlook was grave.<sup>[13]</sup> More recently the combination of high dose

chemotherapy, whole body irradiation and bone marrow transplantation has been advocated. [13]

The WHO criteria for Burkitt's lymphoma includes demonstration of a translocation involving MYC oncogene on chromosome 8 with one of the immunoglobulin gene [2,14 or 22] in small to medium uniform B cells which express CD 10 and BCL 6 and lacks BCL 2 and TdT. The proliferation fraction by Ki 67 Immunostaining is 99% or higher. [4] In our case CD10 and CD20 was immunoreactivity with 100% Immunostaining by Ki67, whereas BCL2 and TdT were immunonegative .

The characteristic morphology [diffuse proliferation of uniformed, small non cleaved cells with cytoplasmic vacuoles and starry sky appearance] accompanied by translocation of MYC and Ig gene required for diagnosis of Burkitt's lymphoma. [4]

In our case, light microscopy and immunohistochemical studies have confirmed the Burkitt's lymphoma.

Regarding differential diagnosis of Burkitt lymphoma, main source of diagnostic difficulty are lymphoblastic lymphoma, diffuse large cell lymphoma. Clinical and histopathological diagnosis may differentiate them from Burkitt lymphoma and confirmed by immunohistochemistry. [13]

In majority of the children, the diagnosis of GIT lymphoma was made at laparotomy. [13] In our case, in view of impossibility of resection, the patient sent for combination chemotherapy to higher center after diagnosis of Burkitt lymphoma on light microscopy and immuno histochemistry.

## CONCLUSION

GIT lymphomas are common extra nodal lymphomas occurring in all age groups. Burkitt's lymphoma of small

intestine in a child, presented as acute abdomen, secondary to intestinal obstruction / intussusception is a rare clinical entity. Accurate diagnosis of the type of lymphoma by histopathology and immunohistochemistry is equally important for correct treatment and determining prognosis.

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