Burkitt`s Lymphoma Presenting as Ileocolic Intussusception in a 8 year old boy

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ABSTRACT

Burkitt's lymphoma is an aggressive type of B-cell Non-Hodgkin lymphoma accounting for approximately 34% of lymphomas occurring in children. Non endemic Burkitt's lymphoma frequently involves abdomen and present with symptoms of bowel obstruction secondary to ileo-caecal intussusception caused by tumor growth, obstruction or bleeding and mimicking acute appendicitis. Histologically Burkitt's lymphoma is characterized by diffuse atypical lymphoid infiltrate composed of monomorphic lymphoid cells and "starry-sky" appearance. We are reporting the case of an 8 year old Nepali male who presented to our hospital with intussuseption secondary to mass which was histologically and innumohistochemically consistent with Burkitt's lymphoma.

KEY WORDS

Burkitt, Childhood, Gastrointestinal, Lymphoma

INTRODUCTION

Primary tumors of the gastrointestinal (GI) tract in children are rare comprising of less than 5% of all pediatric neoplasms.[1] Non-Hodgkin's lymphoma (NHL) remains the most common malignancy of the GI tract in children, out of which Burkitt's lymphoma is the most common tumor of the small intestine.[2] Three variants of Burkitt's lymphoma have been described endemic, sporadic and immunodeficiency associated. Endemic form is more common primarily is seen in Africa and typically presents in jaw, orbit or paraspinal region and is associated with Epstein Barr virus (EBV). The sporadic or non-endemic form is rare, seen worldwide, frequently as primary intestinal lymphoma and not associated with EBV.[3] Primary intestinal lymphoma presents as acute abdomen and abdominal mass involving the terminal ileum, caecum or mesentery; atypical presentation mimicking acute appendicitis may be seen. Some patients present with symptoms of bowel obstruction secondary to ileo-caecal intussusception caused by tumor growth, obstruction or bleeding and mimicking acute appendicitis.[4] In up to 17.5% of patients presenting with primary abdominal Burkitt lymphoma, intussusception is the presenting feature.[5] We present a eight years male child who presented with ileocolic intussusception and was diagnosed to have Burkitt's lymphoma.

CASE REPORT

A 8-year-old male child presented to the surgery outpatient department with complain of acute onset of perumbilical abdominal pain, low-grade fever, and nonbilious, nonbloody vomiting for two day. There was a history of intermittent periumbilical, non radiating, dull aching pain for 14 days. There was no history of any bladder or bowel disturbances. On examination, the child was febrile with pulse rate 90/min and blood pressure 110/80 mmHg. Abdomen was distended and tender with guarding and rigidity on examination. Baseline laboratory investigations showed that the child was anemic with hemoglobin 10.1%

gm. All other routine laboratory investigations were within normal limits. Ultrasonography revealed telescoping of small bowel loops in the distal segment, thickened caecal wall with maintatined vacularity and multiple enlarged mesenteric lynph nodes. With a clinicoradiological diagnosis of ileocecal intussusception patient was immediately taken up for exploratory laparotomy. Operative findings were telescoping of terminal ileum approximately 6 cm into the colon, intraluminal mass approximately 3 x 3 cm at ileocolic junction and few enlarged lymphnodes with rest of the mesentry being normal. The resection of the affected area of ileocecal junction along with the mass was done with end- to-end ileocolic anastomosis. We received a resected bowel consisting of 12 cm segment of ileum and cecum with appendix. External surface was congested and on cut section a gray-white solid tumor was identified measuring 4 cm × 3 cm × 1.5 cm protruding in the lumen. The overlying mucosa was ulcerated, and the tumor was involving full thickness of the bowel.

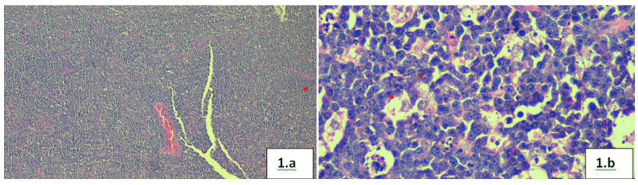

Figure 1a. H and E Section shows diffuse infiltration of monotonous lymphoid cells, 10X. 1b. H and E Section showing medium sized cells with scant cytoplasm, squared- off cytoplasmic borders, round nuclei with fine chromatin, and multiple nucleoli. Interspersed in between these tumor cells were histiocytes giving a "starry-sky" appearance, 40X.

Microscopy revealed small intestinal mucosa infiltrated by a diffuse atypical lymphoid infiltrate composed of monomorphic lymphoid cells. These cells were medium sized with scant cytoplasm, squared- off cytoplasmic borders, round nuclei with fine chromatin, and multiple nucleoli. Abundant mitotic and apoptotic cells were noted. Interspersed in between these tumor cells were histiocytes giving a "starry-sky" appearance (Fig. 1a and 1b). Tumour necrosis were also seen. A preliminary diagnosis of high-grade NHL possibly Burkitts lymphoma was made. Immunohistochemistry panel showed positivity for CD20, Bcl-6, CD 10. C-MYC was positive in more than 90% cells and Ki-67 labeling index was 98-100% (Fig. 2). Immunohistochemistry panel showed negativity for of CD3, tdt and Bcl2 (Fig. 2). A diagnosis of High grade B cell Non Hodgkin Lymphoma, Burkitt Lymphoma was confirmed.

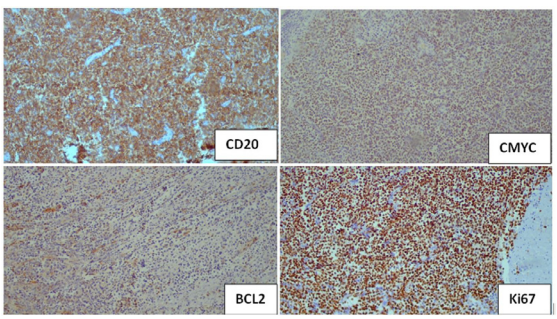

Figure 2. Immunohistochemistry panel showing positivity for CD20. C-MYC was positive in more than 90% cells and Bcl2 is negative. Ki-67 labeling index was 98-100%.

DISCUSSION

Lymphomas constitute the third-common cause of pediatric malignancies.[1] Burkitt's lymphoma (BL) is an aggressive type of B-cell non-Hodgkin lymphoma that was first defined by Dr. Dennis Burkitt in 1958 as a sarcoma of the jaw in Ugandan children. BL is genetically characterized by a chromosomal translocation that results in deregulation of the c- MYC oncogene.[3]

Burkitt lymphoma accounts for 34% of Non-Hodgkin lymphomas that occur in childhood and has a male predominance.[6] Our patient was a male child as well. Most patients with BL present with primary involvement of the abdomen or the head and neck region.[7] Endemic form of BL is mostly localized in the mandibular and maxillary bones in African descent children while non- african children present with the abdominal form of BL involving the bowel.[8] In our case also the child presented with abdominal form of BL. When primary disease involves abdomen patient present with abdominal mass with symptoms of gastrointestinal hemorrhage, abdominal pain, nausea, and intestinal obstruction caused by direct compression or involvement of the bowel lumen. In up to 18% of patients presenting with primary abdominal Burkitt lymphoma, intussusception is the presenting feature.[9,10] Some have symptoms mimicking acute appendicitis.[5] Similar clinical feature of acute abdomen secondary to intussusception, was present in our case.

Imaging modalities used for diagnosis of gastrointestinal malignancy, including BL are GI Sonography, CT scan, Nuclear medicine imaging studies, MR enterography. Utilization of these various imaging modalities helps narrow the differential diagnosis in pediatric patients presenting with abdominal masses. However, other forms of Non-Hodgkin lymphoma, including lymphoblastic lymphoma, blastic mantle cell lymphoma, and diffuse large B-cell lymphoma can exhibit similar imaging characteristics and thus mimic BL.[5]

Histopathological examination with immunohistochemical staining is usually diagnostic for BL. Histology is characterized by diffuse infiltrate of medium sized cells with round to oval nuclear outlines, two to five small but distinct nucleoli and small amount of intensely basophilic cytoplasm. Numerous mitotic figures and apoptotic cells are present. The prominent "starry-sky" appearance is caused by benign phagocytic histiocytes engulfing the nuclear debris resulting from apoptosis. Occasional the cytoplasmic borders of individual cells "square off" against each other. Neoplastic cells of BL express pan B-cell antigens CD19, CD20, CD22 and CD79a, germinal center markers CD10 and BCL6. MYC protein expression is present in most cells. CD5, CD23 and BCL2 are negative. The growth fraction can be assessed by Ki67 or MIB1 which is nearly 100% of tumour cells. In most cases BL demonstrate translocation t(8;14) (q24;q32) in which MYC is tranlocated from chromoseome 8 to chromosome 14.[3]

The St. Jude staging system is routinely used for pediatric patients. Treatment is stratified based on patient age and stage. In pediatric patients with complete surgical resection of disease, it is recommended that patients chemotherapy of moderate-intensity (i.e. cyclophosphamide, vincristine, prednisolone, doxorubicin). Cycles of chemotherapy depends upon the stage of the disease. The overall cure rate for sporadic Burkitt lymphoma approaches 90% in pediatric and young adult populations.[11]

Burkitt lymphoma is a highly malignant, rapidly growing and aggressive neoplasm which can present as a primary tumors of the gastrointestinal with non-specific clinical presentation like diffuse abdominal pain, weight loss, or present with large intra-abdominal mass, intestinal perforation or recurrent intussusception. Surgical excision followed by histomorphological and immunohistochemical assessment remains the gold standard for diagnosis.

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