Case Report

An uncommon presentation of Burkitt's lymphoma as a primary gastrointestinal tumor causing intussusception: A case report with review of literature

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ABSTRACT

Burkitts's lymphoma is the commonest subtype of B-cell non-Hodgkins lymphoma in childhood. Though gastrointestinal tract is the commonest extra nodal site, intussusception secondary to Burkitt's lymphoma is an uncommon presentation. We report a case of an 8-year-old female child who presented with acute abdomen. A tumor arising from ileocaecal valve was the lead point causing intussusception that was reduced, and a limited right hemi colectomy was performed. A final diagnosis of Burkitt's lymphoma was made based on immunohistochemistry.

Key words: Burkitt's lymphoma, intussusception, primary gastrointestinal tumor

INTRODUCTION

Primary tumors of the gastrointestinal (GI) tract are rare in children and represent <5% of all pediatric neoplasms.[1] The rarity of the disease and variable clinical presentation prevent early detection when the possibility of cure exists. Non-Hodgkin's lymphoma (NHL) remains the most common malignancy of the GI tract in children.[2] 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 variety of NHL, which has low long-term survival rates but responds favorably if diagnosed accurately and treated early.

Access this article online	
Quick Response Code:	Website: www.ccij-online.org
	DOI: 10.4103/2278-0513.157812

CASE REPORT

An 8-year-old female child presented with complaints of intermittent abdominal pain in the right lower quadrant for 1-week and 10–15 episodes of vomitings for 2 days. On examination, there was tenderness in right iliac fossa with guarding and rigidity. Baseline investigations revealed 38% neutrophils, 51% lymphocytes and serum lactate dehydrogenase of 623 U/L. Ultrasonography was suggestive of ileocolic intussusception and the child was posted for laparotomy. On exploration, ileocolic intussusception was found secondary to a firm mass at the ileocaecal junction and a limited right hemicolectomy was done with end to end ileocolic anastomosis.

A fleshy polypoid lesion involving the full thickness of bowel and measuring approximately 2.5 cm × 2 cm at ileocaecal junction was found [Figure 1] which on microscopy showed diffuse infiltration of mucosa, sub mucosa and part of muscularis propria with monomorphic lymphoid cells [Figure 2]. No germinal centers were identified here while adjacent ileum and appendix showed reactive lymphoid tissue with prominent germinal centers. A preliminary pathological diagnosis

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of NHL was made which was confirmed as Burkitt's lymphoma as the tumor cells were diffusely positive for CD20 [Figure 3], CD10 [Figure 4] and negative for Bcl12 on immunohistochemistry. Positron emission tomography of the whole body showed metabolically active foci in the thymus and bilateral level II and level V lymph nodes. The patient was subsequently subjected to adjuvant chemotherapy with two cycles of cyclophosphamide, vincristine, prednisolone and doxorubicin regime.

DISCUSSION

Burkitt's lymphoma is the most frequent subtype of NHL in childhood and accounts for approximately 34% of these cases. It is the most rapidly growing tumor in children, with a doubling time of approximately 24 h, so prompt recognition and initiation of therapy are essential. It occurs more often in boys than girls with male-to-female ratios ranging from 1.3:1 to 8.8:1.[3]

Although Burkitt's lymphoma commonly involves the head and neck in children, the GI tract, genitourinary tract, gonads, mesentery, peritoneum, and retroperitoneum are also potential sites of involvement. Intrinsic involvement within the GI tract has been reported to occur in 22.5% of cases, abdominal or pelvic masses in 45% of cases, and hepatic lesions in 17% of cases, with the predominance of these cases occurring in children.[4]

Burkitt's lymphoma primarily affects the ileocaecal region in children younger than 16 years old. The terminal ileum is the most commonly reported location in children, likely because of the high concentration of lymph tissue in that region of the bowel.[4] In that, the tumor does not elicit a desmoplastic response, bowel obstruction is most often caused by intussusception.[5]

Patients with GI Burkitt's lymphoma may present with abdominal pain or distension, GI 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.

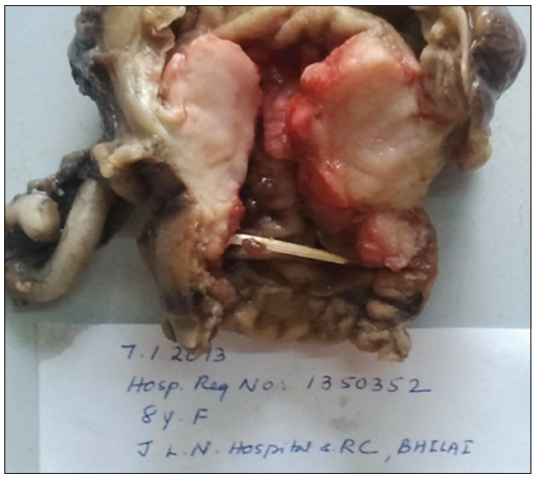

Figure 1: Resected specimen

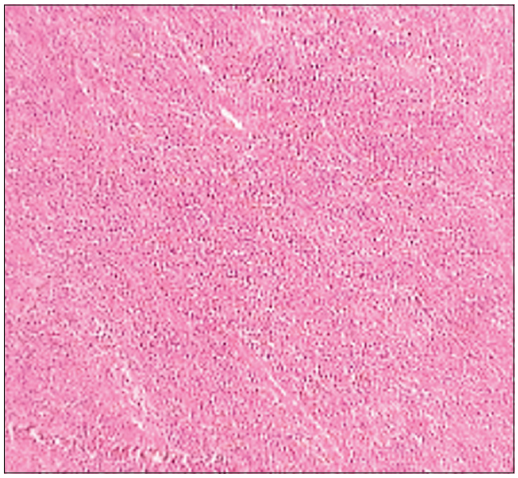

Figure 2: Histopathological examination of specimen in H and E stain

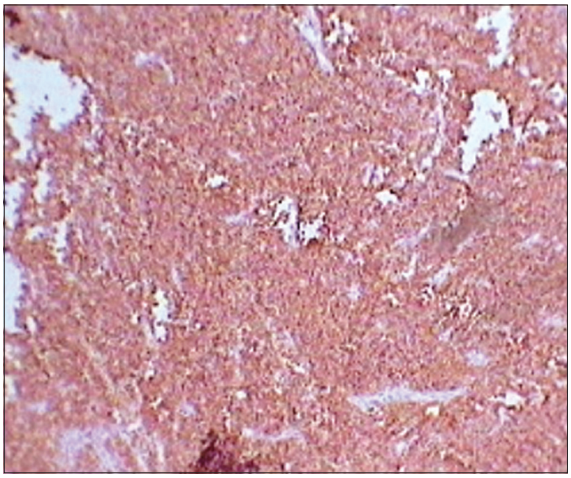

Figure 3: CD 20 positive

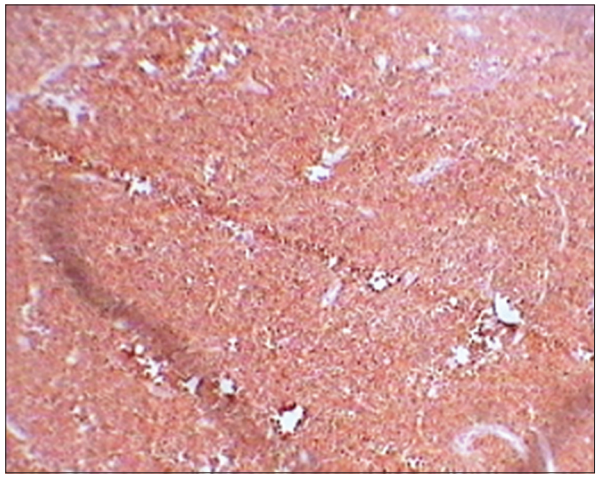

Figure 4: CD 10 positive

These acute abdominal symptoms often lead to emergency laparotomies before a diagnosis of Burkitt's lymphoma can be made.[6]

The disease 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 the 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.[7]

The characteristic morphology (diffuse proliferation of uniformed, small noncleaved cells with cytoplasmic vacuoles and starry sky appearance) accompanied by translocation of MYC and Ig gene required for the diagnosis of Burkitt's lymphoma.[8]

Imaging findings on computed tomography include a focal mass and diffuse thickening of the bowel wall. Cavitary lesions, communication with the bowel and perforation with abscess formation can occur but are uncommon.[9] Ultrasound findings include the doughnut sign of intussusception; the pseudo kidney sign, with or without intussusception, due to diffuse bowel wall thickening with a central echogenic region caused by intraluminal air;[10] or the target sign when occult tumor of the cecum invaginates into ascending colon. Bowel wall thickening may be due to lymphedema or tumor infiltration.

Surgery is required as the treatment to confirm the diagnosis and to relieve the common presenting symptoms of intestinal obstruction, abdominal mass, intussusception, or acute abdomen. Complete resection is associated with improved survival.

The primary treatment of Burkitt's lymphoma is chemotherapy. Several studies have suggested that sporadic variety may be best managed by combined modality of chemotherapy and surgical extirpation.[11] Before the era of combination chemotherapy, Burkitt's 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. More recently the combination of high-dose chemotherapy, whole body irradiation, and bone marrow transplantation has been advocated.[12]

CONCLUSION

Intestinal obstruction in pediatric age group, though a very common entity, intussusception secondary to a GI lymphoma is quiet an uncommon condition. The purpose of our report is not only to share our experience with the medical fraternity, but also to emphasize the clinical condition as one of the uncommon differential diagnoses of intestinal obstruction in pediatric age group.

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Cite this article as: Bokka SH, Mohammed F, Sharma S, Mohanty MK. An uncommon presentation of Burkitt's lymphoma as a primary gastrointestinal tumor causing intussusception: A case report with review of literature. Clin Cancer Investig J 2015;4:567-9.

Source of Support: Nil, **Conflict of Interest:** None declared.