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Case Report

# Burkitt Lymphoma with Aberrant BCL2 expression Masquerading as Ileocolic Intussusception in a Pediatric Patient

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## Abstract

Objective- The aim of this case report of a patient presenting with diffuse abdominal pain, who was operated for ileocolic intussusception and then diagnosed as Burkitt's lymphoma.

Case Report-A 10-year-old female presented in our emergency department with complaints of abdominal pain. Ultrasonography of the abdomen showed evidence of ring like bowel within bowel appearance, seen in the right hypochondrium suggestive of intussusception. Exploratory laprotomy was done, revealing short segment ileo-colic intussusception with large multiple mesenteric lymph nodes at root of mesentery, reaching a maximum size of 3 x 3 cm. Reduction of ileocolic intussusception was done and mesenteric lymphnode biopsy taken. The histopathologic examination of the excised mesenteric lymphnode was suggestive of a high-grade lymphoma, Burkitt lymphoma. The immunohistochemistry studies showed tumor cells were strongly positive for PAX5, BCL2, BCL6, CD10, CMYC and negative for CD3, Tdt, CD34. The Ki-67 was >90% which was suggestive of high grade mature B cell Non- Hodgkin lymphoma- Burkitt lymphoma. Burkitt lymphoma is an aggressive, highly malignant and rapidly growing B-cell neoplasm. Patients often originally present with nonspe¬cific symptoms, such as abdominal pain, gastrointestinal bleeding, anemia, or signs of acute abdomen. The lym¬phoma can be localized in mesenteric lymph nodes or extraperitoneal space. The most common extranodal location of Burkitt lymphoma is near the ileum or the cecum. High index of suspicision is necessary in children who are presenting with acute abdomen/ intussusception.

Conclusion-Burkitt lymphoma is the most common pediatric gastrointestinal malignancy and pediatric surgeon should consider it when a child comes with unspecified clinical presentation with diffuse abdominal pain.

**Keywords:** burkitt lymphoma; intussusception

## Introduction

Burkitt lymphoma is an aggressive, highly malignant and rapidly growing B-cell neoplasm. Burkitt lymphomas represent 8-10% of all tumours in children under 15 years of age [1]. It was first described by Dennis Burkitt in 1958. Three clinical variants have been described – endemic, sporadic and immunodeficiency associated. The endemic form is more commonly seen in African region, and develops due to a chromosomal translocation between chromosome 8 and 14 [2]. It presents as tumour of jaw (maxilla & mandible) or other facial bones and may be associated with Epstein Barr virus (EBV) as well as frequent concomitant malaria infection. The sporadic form is less frequent but is seen worldwide mostly in United States. This results from a chromosome 8 translocation involving the c-myc oncogene and commonly presents as an abdominal mass involving the terminal ileum, caecum or mesentery, with ascites and atypical presentation mimicking acute appendicitis. The third form of Burkitt lymphoma is seen in immunocompromised patients, and typically presents with diffuse lymphadenopathy, bone marrow and central nervous system involvement. Intussusception caused by Burkitt lymphoma, with often misleading symptoms that make the diagnosis more difficult (3). It is the most common cause of intussuception in children above 4 years. Burkitt lymphoma has a high proliferation rate which gives the histological appearance of a "starry sky", the blue lymphoid cells being the sky and numerous tingible body macrophages, due to the high proliferation, being the stars. (4). Burkitt lymphomas have an excellent prognosis, inspite of the stage of the disease due to the better response to the chemotherapeutic regimen. The aim of this

case study is to highlight the rare presentation of Burkitts lymphoma with intususscception in paediatric age group.

## Case Presentation

A 10 year old female child presented in our emergency department with complaints of abdominal pain and backache for last 2 days. There was a history of similar episodes of abdominal pain for the last 2 months. On admission the child was pale and irritable, with a scaphoid abdomen, diffuse pain and tenderness in right upper quadrant and guarding. Bowel sounds were present. Laboratory investigations showed anaemia with a hemoglobin level of 6.8 g/dl and total leukocyte count of 13,900/cu mm with 75% neutrophils and 20%, lymphocytes,; erythrocyte sedimentation rate of 18 mm/hr and platelet count of 274,000/µl. Urine microscopy was normal. Her X- ray abdomen erect was unremarkable. Ultrasonography of the abdomen showed evidence of ring like bowel within bowel appearance seen in the right hypochondrium. Exploratory laparotomy was done under general anesthesia which revealed short segment ileo-colic intussusception with large multiple mesenteric lymph nodes at root of mesentery. The enlarged mesenteric lymph nodes had a maximum size of 3 x 3cm. Reduction of ileo-colic intussusception was done and mesenteric lymphnode biopsy taken. Two units of blood were transfused.

Postoperative recovery was uneventful. The histopathological examination of the excised mesenteric lymph node revealed atypical lymphoid infiltrate of sheets of monotonous population of small lymphoid cells with dispersed tingible body macrophages suggestive of a high grade Non-Hodgkin lymphoma possibly Burkitt lymphoma (Figure i). The immunohistochemistry studies showed tumor cells were positive for PAX5, BCL2, CD10, BCL6, C-MYC (Figure 2) and were negative for CD3, Tdt and CD34. This confirmed the diagnosis of a high grade mature B cell Non-Hodgkin lymphoma-Burkitt lymphoma. Burkitt lymphomas are usually BCL2 negative, or may show weak positivity. Our case showed a diffuse strong positivity for BCL2, BCL6 and MYC, (Fig.iv and v) which leads us to consider the newer entity of High grade B cell lymphomas (HGBL) with rearrangements of all three. These may be called as double hit or triple hit lymphomas. This diagnosis is made on FISH studies for the rearrangements. Our study is limited by the absence of FISH studies due to monetary restraints.

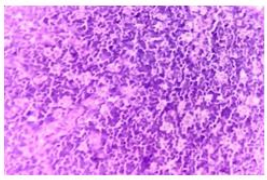
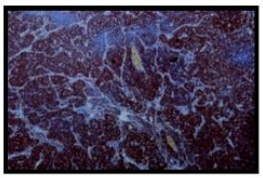


Figure (i)



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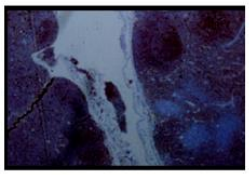

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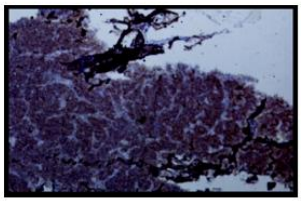

Figure (iv)

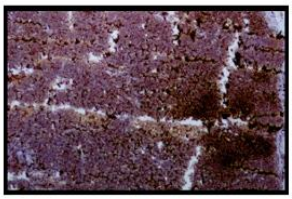

Figure (v)

**Figure (i):** Sheets of small atypical lymphoid cells, with interspersed tangible body macrophages; classic of Burkitt lymphoma. **Figure(ii)** shows PAX5 positivity, **Figure(iii)** CD10 positivity. **Figure (iv) c**-myc positivity. **Figure (v)** shows diffuse strong positivity for BCL2.

## Discussion

Intussusception is the most common pediatric surgical emergency seen in the early age group with a clinical triad of abdominal colic, red currant jelly stool and palpable mass. The incidence of primary gastrointestinal lymphoma is around 1-4 % of all gastrointestinal malignancies with Burkitt lymphoma accounting for 0.3-1.3% of all Non Hodgkins lymphomas (5). The peak age of presentation of gastrointestinal NHL in children is 5-15 years with males being 1.8-2.5 times more affected as compared to females (6). Distal ileum or ileocecal region is the most commonly involved in children. It is difficult to suspect a lymphoma in pediatric patients presenting with acute abdomen. Intussusception is the first clinical sign in nearly 18% of case with primary abdominal Burkitt lymphoma. In 80% cases abdominal pain is the most common symptom along with nausea, vomiting, constipation, diarrhea, malaise and weight loss (7). In our case the patient is a female with an age of ten years and the main presenting complaint was abdominal pain. In the study by Ein et al only 3 of 11 children were term survivors (8) while Puri et al. reported one death in the entire series of 292 children with Burkitt lymphoma who presented with intussusception (9). Ultrasonography is most preferred modality to detect intestinal intussusception with 100% accuracy (10), while computerized tomography, barium enema and colonoscopy is also helpful in diagnosis (11).

The 2016 revised WHO classification of lymphoid malignancies has included a new entity of a High grade B cell lymphoma (HGBL), characterized by *MYC* and *BCL2* and or *BCL6* rearrangements. HGBL with rearrangements in all three loci, *MYC*,*BCL2*, and *BCL6* as diagnosed by FISH are called Triple Hit Lymphomas (THL). The poor prognostic factors are elderly age group, CNS and marrow involvement and higher stage. Our patient did not have any of these. The importance of diagnosing the THL lies in its poor response to the R-CHOP regime (12).

The ideal treatment of gastrointestinal lymphoma is individualized based on the type of disease, its location and molecular profiling, so that a multidisciplinary approach with surgery and chemotherapy can optimize the chance of cure. Laparotomy is the gold standard in both diagnosis and treatment. Burkitt lymphoma is very sensitive to chemotherapy due to its high proliferation rate (11). Therapeutic agents include the following drugs: cyclophosphamide, methotrexate, cytarabine, iphosphamide, etoposide, vincristine, vindesine, adriamycin, doxorubicin and dexamethasone (13-14). Burkitt lymphoma patients who present with intussusception require shorter-duration and less intense chemotherapy than patients diagnosed in other ways. In our patient we did reduction of intussusception followed by chemotherapy.

## Conclusion

Burkitt lymphoma is the most common pediatric gastrointestinal malignancy and a pediatric surgeon should consider it when a child comes with non specific clinical presentation with diffuse abdominal pain. Making the diagnosis of Non Hodgkin lymphoma is clinically challenging and high index of suspicision is necessary in children who are presenting with acute

abdomen/intussusceptions regardless of age. Laparotomy is the gold standard in both diagnosis and treatment. A multidisciplinary approach is required for effective management which now includes molecular studies and cytogenetics for targeted therapies which will lead to better prognosis.

**Consent:** Informed consent was taken from the patient for the publication of this case report

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**Conflict of Interest:** the authors declare that they have no conflict of interest.

## Reference


1. Angotti R, Ferrara F, BurgioA et al. (2009). Intussusception in a child with sporadic Burkitt lymphoma. JSAS. 1, 1, doi: 10.4081/350.
2. Brodzisz A, Woźniak MM, Dudkiewicz E, Grabowski D, Stefaniak J, Wieczorek AP, Kowalczyk J. (2013). Ultrasound presentation of abdominal non-Hodgkin lymphomas in pediatric patients. J Ultrason. 13(55):373-378.
3. Hoxha FT, Hashani SI, Krasniqi AS, Kurshumliu FI, Komoni DS, Hasimja SM et al. (2009). Intussusceptions as acute abdomen caused by Burkitt lymphoma: a case report. Cases Journal. 2:9322.
4. Chung EM, Biko DM, Arzamendi AM, Meldrum JT, Stocker JT. (2015). Solid tumors of the peritoneum, omentum, and mesentery in children: radiologic-pathologic correlation: from the radiologic pathology archives. Radiographics. Mar-Apr;35(2):521-546.
5. Bălănescu NR, Topor L, Malureanu D, Stoica I. (2013). Ileocolic intussusception due to Burkitt lymphoma: a case report. J Med Life6(1): 61-64.
6. Gupta A, Kumar S, Hai A, Agarwal S. (2008). Primary non hodgkin lymphoma of large bowel presenting as ileocolocolic intussusception in a child : A rare clinical presentation. The Internet Journal of Pediatrics and Neonatology. 10:1.
7. Shakya VC, Agrawal CS, Koirala R, et al. (2009). Intussusception due to non Hodgkin's lymphoma; different experiences in two children: Two case reports. Cases J 2:6304.
8. Ein SH, Stephens CA, Shandling B, Filler RM. (1986). Intussusception due to lymphoma. Journal of Pediatric Surgery. 21(9):786-788.
9. Puri P, Guiney EJ. (1986). Small bowel tumors causing intussusception in childhood. Journal of Pediatric Surgery. 21(5):461.
10. Puri P, Höllwarth M, (compilers). (2009). In: Pediatric Surgery Diagnosis and Management. New York: Springer; pp. 485-490.
11. Aydede H, Sakarya A. (2002). Ileocolic intussusception due to ileal lymphoma (Case report). Turk J Gastroenterol. 13:236–238.
12. Chen BJ, Fend F, Campo E, Quintanilla-Martinez L. (2019). Aggressive B-cell lymphomas from morphologyto molecular pathogenesis.Ann Lymphoma. 3:1.
13. Chieng JHC, Garrett J, Ding SL, Sullivan M. (2009). Clinical presentation and endoscopic features of primary gastric Burkitt lymphoma in childhood, presenting as a protein-losing enteropathy: a case report. Journal Of Medical Case Reports. (3):7256.
14. Attarbaschi A, Mann G, Dworzak M, Trebo M, Mühlegger N, Reiter A, Horcher E, Gardner H. (2002). The role of surgery in the treatment of pediatric B-cell non-Hodgkin's lymphoma. Journal of Pediatric Surgery. 37(10):1470-1475.






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