

## **Case Report**

# **A Rare Case of Cecocolic Intussusception Secondary To Burkitt's Lymphoma**

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**Abstract:** Intussusception is the invagination of one portion of the intestine into another. It rarely occurs after the first 5 years of life. Etiologies of childhood intussusception differ depending on age at presentation. Burkitt's lymphoma is a rare pathological lead point in the older child with intussusception. Here we report a rare and interesting case of Cecocolic Intussusception Secondary to Burkitt's Lymphoma. The child was managed successfully with surgery followed by adjuvant chemotherapy.

**Keywords:** Intussusception, Burkitt's Lymphoma.

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## **INTRODUCTION**

Intussusception is the invagination of one portion of the intestine into another. It rarely occurs after the first 5 years of life and is classically associated with intense intermittent abdominal pain, vomiting, bloody mucoid diarrhea, and a palpable abdominal mass. Etiologies of childhood intussusception differ depending on age at presentation: in younger infants (less than 18 months of age) the cause appears to be an inflamed enlarged intestinal lymph tissue patch acting as the lead point, whereas in older children the lead point may be, amongst others, a hemangioma or other mural neoplasm or a Meckel's diverticulum [1-3]. Most intussusceptions in children (90%) are ileocolic and idiopathic in nature. Burkitt's lymphoma is a rare pathological lead point in the older child with intussusception. Here we report a rare and interesting case of Cecocolic Intussusception Secondary to Burkitt's Lymphoma.

## **CASE REPORT**

A 11 year old boy presented to the casualty with sudden onset of severe lower abdominal pain since 1 day, history of low grade fever since 1 day and vomiting since 1 day. On examination the child was mildly dehydrated and febrile, Pulse Rate 98 /min and Blood Pressure 110/70mmHg. On Per Abdominal examination there was Tenderness at the McBurney Point with rebound tenderness and localized guarding. There was no palpable abdominal mass. Bowel sounds

were hyper peristaltic. Other Systems were within normal limits. A Clinical diagnosis of Acute Appendicitis was made and the child was started on analgesics and antibiotics. Routine laboratory investigations revealed Elevated Total counts - 12500 cells/cu mm Differential count - N = 62, L = 29, E = 06, M = 02, B = 01. All other laboratory investigations were normal. An Ultrasound Abdomen was performed in which there was right lumbar and iliac probe tenderness. A bowel mass measuring 10 x 4 cms was noted in the right iliac fossa 'Pseudo Kidney Sign' and 'target sign' were noted. Appendix was not visualised. An erect X ray abdomen was taken which showed multiple air fluid levels. A diagnosis of acute intestinal obstruction secondary to intussusception was made.

The child was posted for Emergency Laparotomy and proceeds. A lower abdominal right paramedian incision was taken and abdomen opened in layers. Intraoperatively cecocolic intussusception was identified. The intussusception was reduced by the classical method. After reducing the intussusception a Cecal Pedunculated Polyp measuring 4 x 3 x 2 cms was found as the lead point. A cecotomy was performed and the polyp was excised with adequate surgical margins. A retrocecal inflamed Appendix measuring 6 cms with mesenteric lymphadenopathy was also noted. Appendectomy was performed. Both the specimens were sent for histopathological examination. The

postoperative period was uneventful and child completely recovered.

The histopathological examination of the cecal polyp revealed a Non Hodgkins Lymphoma – Lymphoblastic pattern Burkitt’s type. All surgical margins were free of tumour tissue. The paraffin blocks of the cecal polyp were then subjected to immunohistochemistry. They showed positivity for CD 20, CD 10 and Bcl 6. Ki 67 proliferative index of more

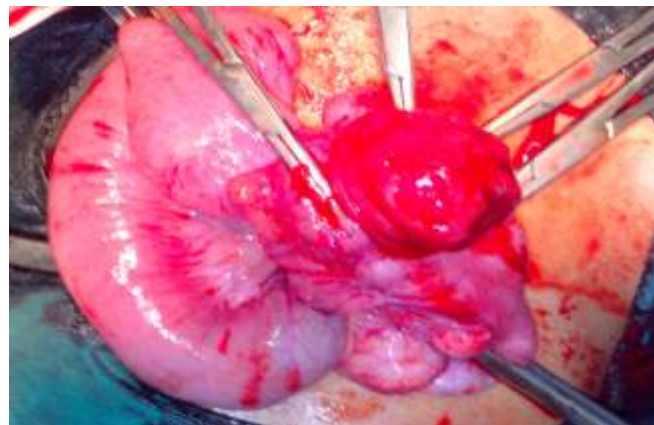
than 95% indicated the high proliferative nature of the tumour. They showed immunonegativity for Bcl2/CD3.

On follow up, a CT thorax and CT Abdomen were done which were negative for Lymphomas.

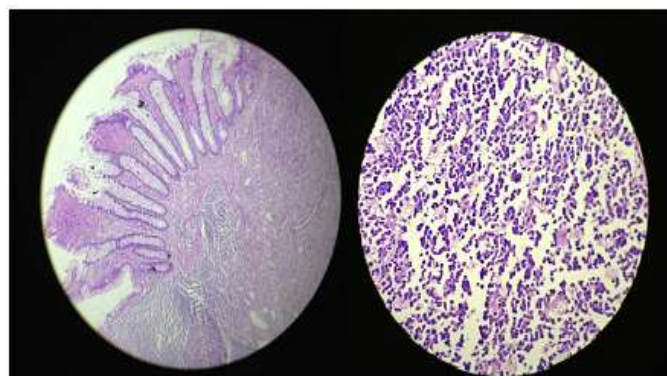
After consultation with a medical oncologist, the child was started on CHOP Regimen with Methotrexate. The Child has currently completed 4 cycles of the 21 day cycle chemotherapy and is doing well.



**Fig.1:** Intraoperatively cecocolic intussusception was identified



**Fig. 2:** After reducing the intussusception a Cecal Pedunculated Polyp measuring 4 x 3 x 2 cms was found as the lead point.



**Fig.3:** Microscopic picture of Burkitt lymphoma invading the Submucosa and Muscularis Propria with areas of normal colonic tissue

## DISCUSSION

Burkitt's lymphoma is a highly aggressive non-Hodgkin B- cell lymphoma mostly presenting as extra nodal disease. Children and immune compromised patients are most often affected by the disease [1]. Burkitt's lymphoma is classified into three clinical forms: a) Endemic form which is common in Africa and presents mostly as rapidly growing tumour involving facial bones. b) Non-endemic or the sporadic form which typically presents as abdominal mass or ascites. c) The third form occurs in immune compromised patients and it is commonly seen in patients with HIV/AIDS. It most commonly presents as diffuse lymphadenopathy [4].

The sporadic form commonly presents with abdominal swelling as a large mesenteric, retroperitoneal or pelvic mass, tenderness, pain or fullness. Some patients present with symptoms of bowel obstruction secondary to ileocaecal intussusception caused by tumour growth, obstruction or bleeding, mimicking acute appendicitis [5, 6]. Malignant lymphoma comprise approximately 1 to 4% of all the gastrointestinal malignant neoplasms [7].

Intussusception is a common cause of abdominal pain in children. Although most cases are idiopathic, about 10% of cases have a pathologic lead point. Burkitt's lymphoma is not a common etiology. Intussusception caused by Burkitt lymphoma, as a cause of acute abdomen, is rare, with symptoms which often mislead and make diagnosis more difficult [8]. The rapidity of volumetric doubling of this neoplasm frequently justifies an acute abdomen presentation that may mimic other less-rare diseases [5-8].

Burkitt's lymphoma occurring in adults, involving the terminal ileum with ileal or ileocolic intussusception's has been reported as acute abdomen requiring an emergency surgery, reported as case or as small numbers in series, treated with small bowel resection or right hemicolectomy [6, 8-13]. However due to rapid proliferative rate and good response to chemotherapeutic regimens a limited resection is now advocated and surgical role is confined to relieve obstruction.

For patients with limited diseases, including localized extra abdominal or completely resected abdominal disease (NCI Stages A and AR) a long term survival rate > 90% can be achieved with combination chemotherapy [14-21]. Treatment options include: COMP - Cyclophosphamide + Vincristine (Oncovin) + Methotrexate + Prednisone. CHOP-plus methotrexate. NHL-BFM90-Prednisone + dexamethasone + vincristine + doxorubicin + cyclophosphamide + ifosfamide + etoposide + cytarabine + methotrexate. French LMB-89- high dose cyclophosphamide + high dose methotrexate /

leucovorin + cytarabine + vincristine + prednisone + doxorubicin CCG-5961- Reduction in intensification of the French LMB-89 regimen.

Prognosis in children correlates with the bulk of disease at the time of diagnosis [5]. Survival rate has been improved significantly with the appropriate management of the metabolic consequences of rapid cell turnover and with combination chemotherapy and CNS prophylaxis [23]. Patients with limited (A,AR) disease have an excellent prognosis with a survival rate greater than 90% [24]. In adults, particularly those with advanced stage of this disease, do more poorly than children with the disease [5].

## CONCLUSION

In our case report a child who presented with a cecocolic intussusception secondary to a pathological lead point a solitary cecal polyp which was later identified as Sporadic Burkitt's Lymphoma was successfully managed by cecotomy with resection of polyp followed by adjuvant chemotherapy. Our case report highlights the importance of identifying pathological lead points in older children and more importantly the fact that Sporadic Burkitt's Lymphoma should be considered a possibility in the Indian population. Children tend to do well with limited surgical resection and chemotherapy.

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