Sporadic Burkitt’s Lymphoma Masquerading As An Intussuscepted Meckel’s Diverticulum In A 9-Year-Old Child

**SUMMARY**  
*Up to 150 words summarising the case presentation and outcome (this will be freely available online)*

We report the case of a 9-year-old boy who presented with abdominal pain and was found to have an intussusception with a Sporadic Burkitt’s Lymphoma (SBL) lead point. Our case was unusual in that the patient did not present with the typical clinical features of BL, nor was he in a high-risk demographic for this uncommon disease.

**BACKGROUND**  
*Why you think this case is important – why did you write it up?*

In children, 75% cases of intussusception are idiopathic and 25% are due to underlying disease (most commonly due to Meckel’s diverticulum, polyps then either duplication cyst or Henoch-Schonlein Purpura). 60% of presentations generally occur in children younger than 1 year and 80-90% younger than 2 years. Sporadic Burkitt’s Lymphoma is a rare cause of intussusception, especially in older children (2.5 cases per million person years)[1], [2]. Herein, we stress the importance of considering Sporadic Burkitt’s Lymphoma as a differential diagnosis in children of an atypical age group presenting with intussusception.

We report the unusual case of a 9-year-old male of Fiji-Indian descent who presented with abdominal pain and was found to have Sporadic Burkitt’s Lymphoma as the lead point for an intussusception after surgery. He did not present with typical clinical features, nor was he in a high-risk demographic.

We discuss the diagnosis, management and treatment of our patient to bring attention to the uncommon cause of a common paediatric surgical presentation. We propose intraoperative lymph node biopsies to be performed and suggest intraoperative specimens be sent for histopathology as fresh tissue (as opposed for formalin fixed) to allow for flow cytometry to be performed in an urgent setting. Moreover, our case highlights the importance of communication, rapid referral and efficient communication in producing a favourable prognosis.

**CASE PRESENTATION**  
*Presenting features, medical/social/family history*

A 9-year-old male of Indian descent, originally from Fiji, was referred to the emergency department from his general practitioner with a 2-week history of worsening right-sided lower abdominal pain. Pain had been present for 6 months and was mild and colicky in nature. The patient had become anorexic. His symptoms were associated with 2 days of nausea and vomiting and fresh red blood mixed into soft bowel motions. He was well, had no sick contacts, had not recently travelled and had no recent dietary changes. Blood tests and imaging (abdominal ultrasound, abdominal x-ray) performed by his general practitioner were all unremarkable.

On examination, the patient appeared pale and exhibited episodes of colic. He was afebrile and all vital signs were within normal ranges. Upon abdominal examination, he was very distended- his abdomen was tender to touch and a mass was palpable in the right lower quadrant, localised guarding and percussion tenderness were present. Bowel sounds were scant and there was no palpable lymphadenopathy. The remainder of his examination was unremarkable.

**INVESTIGATIONS**  
*If relevant*

Biochemical analysis of bloods for electrolytes, renal and liver function on admission to emergency yielded a urate of 0.25mmol/L, LD of 342 U/L, full blood count showed a platelet of 449U/L and an ESR of 50, tests were otherwise normal. An abdominal ultrasound was performed which showed an ileo-ileocolic intussusception. The lead point was favoured a Meckel’s remnant due to the presence of increased vascularity in the wall. (Figure 1)

The decision to proceed to an exploratory laparotomy through McBurney’s incision was made due to acutely increasing pain and the unavailability of pneumatic or contrast reduction at the time. Intraoperative
findings included an ileo-ileocolic intussusception in the terminal ileum, which was in keeping with a clinically anticipated Meckel’s Diverticulum on the basis of pre-operative imaging. Bowel was viable and there was no pus or active bleeding identified. Attempts were made to reduce the intussusception, however these were not successful. Consequently a wedge resection with end-to-end anastomosis and an appendicectomy were performed. (Figure 2)

Histopathologic diagnosis of Burkitt’s Lymphoma (BL) was provided 3 days later and an urgent transfer was organised to the regional paediatric oncology unit for staging and treatment. Viral serology for HIV, EBV, hepatitis B and C were non-reactive.

In the appendix, the submucosa showed reactive lymphoid follicles. Sections from intussuscepted small bowel showed a diffuse expansile infiltrate which was present throughout the bowel wall. The infiltrate comprised of monomorphic medium to large lymphoid cells showing frequent mitosis and monomorphic medium to large lymphoid cells. A starry star appearance was noted. (Figure 3, Figure 4)

**DIFFERENTIAL DIAGNOSIS *If relevant***

**TREATMENT *If relevant***

A diagnosis of Stage IV disease was made for which the patient received 5 cycles of chemotherapy (Vincristine, Prednisolone, Cyclophosphamide, Rituximab, Methotrexate, Doxorubicin).

**OUTCOME AND FOLLOW-UP***

Patient is now disease free at 5 months follow up.

**DISCUSSION Include a very brief review of similar published cases***

In children, 75% cases of intussusception are idiopathic and 25% are due to underlying disease (most commonly due to Meckel’s diverticulum, polyps then either duplication cyst or Henoch-Schönlein purpura (immunoglobulin A vasculitis)). 60% of presentations generally occur in children younger than 1 year and 80-90% younger than 2 years.

BL is a highly aggressive B cell Non Hodgkin Lymphoma and comprises of 30% of non-endemic paediatric lymphoma. It exists in 3 distinct clinical forms- endemic, sporadic and immunodeficiency associated. The sporadic form presents with abdominal symptoms and most often results in massive disease. It is characterised by early onset (27% in children 3-5, 25% children 6-8 and 6% in children 0-2) and show a male predominance of 79%. The mean age of diagnosis is 7.8 and is more common in Caucasians (81%). Literature reports a higher rate of SBL amongst Whites and Asians/Pacific Islanders than among Blacks. By ethnicity, BL incidence was higher among non-Hispanic Whites than Hispanic Whites. Coloured and black children presented with more advanced disease and at a younger age (5-6 years) than white children (8-9 years) and the survival ratios between the groups differ markedly [3]. Mbulaiteye et al suggests that white race may be risk factors for sporadic childhood BL in the United States [2].

Risk factors which may predispose one to BL include early age of exposure to common childhood infections like EBV (sporadic variant shows only 20% association with EBV) and socially disadvantaged populations [2].

Clinical features normally seen with BL in the paediatric population include massive disease and ascites, jaw or facial bone involvement (25%), localised lymphadenopathy, CNS involvement (15-30% of cases) and symptoms related to bowel obstruction or gastrointestinal bleeding as seen in our case [4].

Our patient did not present with typical symptoms and thus, difficulties were experienced when making a diagnosis. Brichon et al suggest that abdominal sonography is the most efficient examination for the diagnosis of intestinal intussusception and sometimes of the primitive lesion [1]. This is supported by Brekalo et al who indicate that echography is more advantageous that computerized tomography when diagnosing Burkitt's lymphoma [5]. However, radiographic signs on abdominal sonography did not reveal the aetiology of lead point of the patient’s intussusception.
The management of our patient is consistent with the recommendation of Chirug et al; if the lymphoma is not visualized with ultrasonography, an emergency laparotomy is necessary for the diagnosis of the lymphoma and the intestinal resection in case of necessity [1]. The prognosis of paediatric patients with BL presenting with intussusception is favourable as such patients often have completely resectable disease [6], when the tumour is localized, total resection results in a good outcome [7]. In this case, a desirable outcome for the patient was ultimately achieved as a result of the combination of medical and surgical treatment.

LEARNING POINTS/TAKE HOME MESSAGES 3 to 5 bullet points – this is a required field

- In conclusion, our case was unusual in that the patient did not present with the typical clinical features of BL, nor was he in a high-risk demographic for this uncommon disease.
- We stress the importance of considering BL as a differential diagnosis in children of an atypical age group presenting with intussusception, especially in ethnicities where disease is more common.
- There may be a role for intraoperative lymph node biopsies and in sending intraoperative specimen as fresh tissue (as opposed for formalin fixed) to allow for flow cytometry to be performed in an urgent setting in patients where there is a high index of suspicion of lymphoma.
- The importance of communication and rapid referral and efficient communication is essential in producing a favourable prognosis.

REFERENCES Vancouver style (Was the patient involved in a clinical trial? Please reference related articles)


FIGURE/VIDEO CAPTIONS figures should NOT be embedded in this document

Figure. 1 Thickened and oedematous intussuscipiens (large bowel) with preserved vascularity. Intussusceptum (small bowel/mesentery) demonstrated preserved vascularity. Lead point (large cystic lesion with peripheral vascularity and a triple wall signature) within the intussusception near the hepatic flexure. No free intra-abdominal fluid, evidence of appendicitis or enlarged lymph nodes.

Figure. 2 Intraoperative view of intussuscepted segment of bowel. Small patches of bruising. No obvious mesenteric lymph nodes visible.

Figure. 3 Burkitt’s Lymphoma x 4

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