CASE REPORT

Sporadic Burkitt's lymphoma masquerading as an intussuscepted Meckel's diverticulum in a 9-year-old child

Jessica Yan-Seen Ng,¹ Richard James Thompson,¹ Alfred Lam,² Sonu Nigam²

¹Surgery, Gold Coast University Hospital, Southport, Queensland, Australia

²Pathology, Gold Coast University Hospital, Southport, Queensland, Australia

Correspondence to
Dr Jessica Yan-Seen Ng,
jessica.ng@health.qld.gov.au

Accepted 26 June 2018

SUMMARY

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 (BL) 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

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 Hirsch-Schönlein purpura). Sixty per cent of presentations generally occur in children younger than 1 year and 80%–90% younger than 2 years. Sporadic Burkitt's lymphoma (SBL) 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 SBL as a differential diagnosis in children of an atypical age group presenting with intussusception.

We report the unusual case of a 9-year-old boy of Fiji-Indian descent who presented with abdominal pain and was found to have SBL 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

A 9-year-old boy 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. On 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

Biochemical analysis of bloods for electrolytes, renal and liver function on admission to emergency yielded a urate of 0.25 mmol/L and lactate dehydrogenase of 342 U/L, and full blood count showed a platelet of 449 U/L and an erythrocyte sedimentation rate 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).

Figure 1 Thickened and oedematous intussusciptum (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.

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To cite: Ng JY-S, Thompson RJ, Lam A, et al. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2018-224333

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

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 preoperative 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 appendectomy were performed (figure 2).

Histopathological diagnosis of 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, Epstein-Barr virus (EBV) and 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

Figure 3 Burkitt's lymphoma $\times 4$.

Figure 4 Burkitt's lymphoma $\times 40$.

bowel wall. The infiltrate comprised monomorphic medium to large lymphoid cells showing frequent mitosis and monomorphic medium to large lymphoid cells. A starry star appearance was noted (figures 3 and 4).

TREATMENT

A diagnosis of stage IV disease was made for which the patient received five cycles of chemotherapy (vincristine, prednisolone, cyclophosphamide, rituximab, methotrexate, doxorubicin).

OUTCOME AND FOLLOW-UP

Patient is now disease free at 5 months of follow-up.

DISCUSSION

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)). Sixty per cent 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's lymphoma and comprises 30% of non-endemic paediatric lymphomas. It exists in three 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% in 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 sporadic BL among 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 a risk factor for sporadic childhood BL in the USA.²

Risk factors that 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, central nervous system 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 than CT when diagnosing BL.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 Brichon *et al*; if the lymphoma is not visualised 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 and when the tumour is localised, 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.

Contributors All authors have contributed to this paper and share collective responsibility and accountability for results. JY-SN: primary author and constructor of most of the body of report. RJT: provided the case and pictures for the report. Also helped in the editing process for submission. AL and SN: contributed to histopathology.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

Learning points

- ▶ In conclusion, our case was unusual in that the patient did not present with the typical clinical features of Burkitt's lymphoma (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 to 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.

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