CASE REPORT

Adult ileocolic intussusception caused by Burkitt lymphoma

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SUMMARY

Ileocolic intussusception due to Burkitt lymphoma is extremely rare in adults. A man aged 17 years presented with a history of recurrent abdominal pain over the past 3 weeks. The abdomen was distended with diffuse tenderness, and bowel sounds were present. Abdominal ultrasound and CT scans showed evidence of small bowel obstruction with marked wall thickening in the ileocecal region and 'target' signs suggestive for intussusception. At laparoscopy, a mass involving the caecum and the terminal ileum was found, along with multiple locoregional nodes, which was highly suggestive of malignancy. A typical en bloc right colectomy with intracorporeal ileocolic anastomosis was performed. Histopathological examination showed a high-grade B-cell Burkitt lymphoma that was confirmed by immunohistochemistry. The patient was subsequently treated with adjuvant combination chemotherapy and is alive and disease-free at the 3-year follow-up.

BACKGROUND

Less than 5% of bowel obstruction in adults is caused by intussusception, that is, telescoping of one segment of bowel into the immediately distal segment. It is unlikely that a general surgeon will encounter more than one or two bowel intussusceptions throughout his career.[1 2] Ileocolic and ileocaecal intussusceptions represent between 10% and 15% of the cases. In the majority of patients, intussusceptions are secondary to benign and malignant (primary and metastatic) tumours, Meckel diverticula, foreign bodies and adhesions acting as a lead point.[3]

Sporadic Burkitt lymphoma accounts for 20–30% of non-Hodgkin's lymphomas, but is extremely rare in the adult western population. It is a high-grade B-cell tumour, genetically characterised by a chromosomal translocation that causes dysregulation of the c-MYC oncogene. Surgical resection of intestinal Burkitt lymphoma is the initial therapy of choice, and patients with localised disease may have a >90% 5-year survival rate.[4] The present case highlights the importance of preoperative imaging and prompt laparoscopic management in a young adult patient who presented with a history of recurrent abdominal pain and subacute bowel obstruction.

CASE PRESENTATION

A Caucasian man aged 17 years, with no significant medical history, was admitted to the emergency department for recurrent right side and periumbilical abdominal pain accompanied by nausea over the past 3 weeks. He was afebrile and haemodynamically stable. On physical examination, there was diffuse abdominal tenderness without evidence of a palpable mass or peritonism. Peristalsis was present. Biochemical and haematological investigations were normal except for the C reactive protein (CRP) (1.3 mg/dL) and a relative lymphocytopenia (20%). The patient was discharged with prescription of symptomatic therapy. After 1 week, he was admitted again with worsening abdominal symptoms and nausea.

INVESTIGATIONS

Physical examination showed marked abdominal distension and diffuse tenderness with mild rebound pain. The ear temperature was 36.5°C, blood pressure 130/70 mm Hg, heart rate 87 bpm and oxygen saturation 98% on room air. The rectal digital exploration and guaiac stool test result were negative. Laboratory findings showed increased CRP (19 mg/dL) and lymphocytopenia (15%). The transabdominal ultrasonography showed the presence of free pelvic fluid, markedly distended bowel loops and 'doughnut' sign suggestive for intussusception (figure 1). The abdominal CT scan confirmed the presence of bowel obstruction with segmental wall thickening and increased mucosal contrast enhancement with a typical 'target' sign. Multiple regional lymph nodes were also visible (figure 2).

DIFFERENTIAL DIAGNOSIS

Appendicitis, Meckel' diverticulitis, Crohn's disease, benign and malignant (primary or

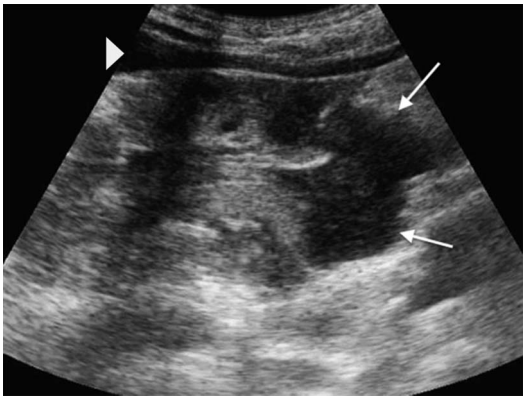

Figure 1 Abdominal ultrasonography (longitudinal view) showing free pericolic fluid (arrowhead) and bowel wall thickening with a hypoechoic 'doughnut' image suggestive of ileocolic intussusception (arrows).

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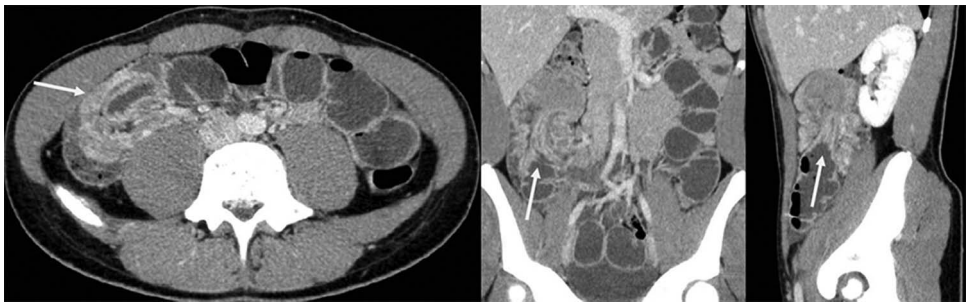

Figure 2 Abdominal CT scan showing distended bowel loops and thickening of the colonic wall with increased mucosal contrast enhancement ('target' sign) suggestive of ileocolic intussusception.

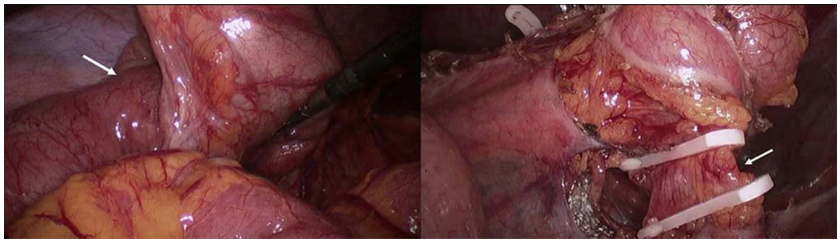

Figure 3 Intraoperative laparoscopic findings: left, ileocolic intussusception (arrow); right, the ileocolic vascular pedicle is clipped at its origin (arrow) to allow en bloc lymphadenectomy. Enlarged nodes are visible above the clips.

secondary) tumours could be considered in the differential diagnosis. Given the history of intermittent abdominal pain and distension, and the results of imaging, the decision was made to proceed with surgical exploration.

TREATMENT

Informed consent was obtained and the patient was transferred to the operating theatre. Under general anaesthesia, a three-port laparoscopic exploration was performed that revealed the presence of ileocolic intussusception with a caecal mass and multiple enlarged pericolic and mesenteric lymph nodes. The proximal small bowel loop was distended, oedematous and with initial ischaemic changes (figure 3). Intraoperative frozen section was not available. Considering the site of intussusception, the high risk of perforation and the high probability of underlying malignancy, en bloc laparoscopic right colectomy with intracorporeal, side-to-side and isoperistaltic semimechanical ileocolic anastomosis was performed. The specimen (figure 4) was extracted through a 5 cm mini-laparotomy in the right lower quadrant. The total operative time was 220 min. Histological examination was suggestive for Burkitt lymphoma (CD20+, CD10+, Bcl6+, CD3−, Bcl2−, CD34−), with a high mitotic index (Ki-67 in 100% of cells) without myeloperoxidase expression. Intracapsular metastases were found in 2 out of the 26 removed lymph nodes (figure 5).

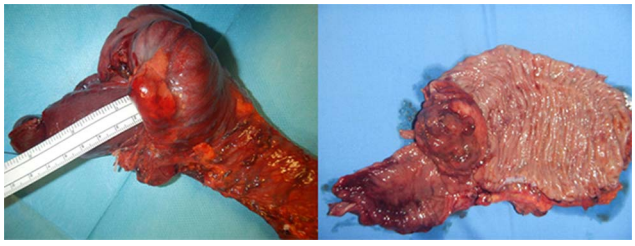

Figure 4 Surgical specimen: left, the terminal ileum appears invaginated for a length of 3 cm into the caecum; right, Burkitt lymphoma originating from the ileocaecal valve.

OUTCOME AND FOLLOW-UP

The postoperative course was uneventful. The patient was discharged home on postoperative day 6. Subsequent staging with PET–CT scan, bone marrow biopsy and rachicentesis showed no evidence of metastatic disease. Six weeks after operation, adjuvant combination therapy was initiated using cisplatin, cytarabine, vincristine, VP-16, adriamycin and intratechal methotrexate; a total of six cycles of therapy were administered. The patient is alive with excellent performance status and is disease-free at 3 years after surgery.

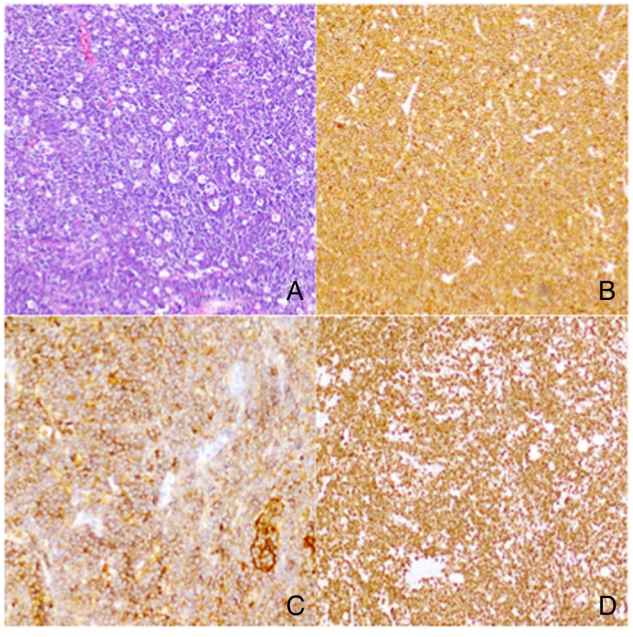

Figure 5 (A) H&E stain. Neoplastic cells are uniform, medium sized, with squared-off basophilic cytoplasm. Nuclei are round to oval, slightly indented, with coarse chromatin. Typical 'starry sky' appearance, due to blue neoplastic cells and histiocytes. (B, C) Intense and widespread immunohistochemical staining with anti-CD10 (C) and anti-CD20 (B). (D) One hundred per cent of cells stain positive for Ki-67, indicating the high proliferative activity of Burkitt lymphoma.

DISCUSSION

Intussusception is rare and accounts for only 1–5% of bowel obstruction in adults; almost 90% of adult intussusceptions are secondary to a pathological condition that acts as a lead point, with a 65% risk of an associated malignancy.[5] Although the gastrointestinal tract is the most frequent site of extranodal lymphoma, sporadic Burkitt lymphoma is a very rare cause of ileocolic intussusception in adults presenting with acute abdomen.[6 7] Ultrasonography and CT scan of the abdomen were diagnostic of intussusception in our young adult patient who presented with a 3-week history of recurrent abdominal pain. While ultrasonography is operator-dependent, CT scan is very reliable to identify the causative lead point.[8] At laparoscopy, the diagnosis of ileocolic intussusception with signs of initial bowel ischaemia of the terminal ileum and also the presence of multiple enlarged mesenteric and pericolic lymph nodes was confirmed, and therefore an en bloc right colectomy with oncological criteria was performed.

Unlike the paediatric population, attempts at reduction in the intussuscepted bowel are generally not indicated in adults for a number of reasons: (1) the ileocaecal valve acts as a constricting ring possibly leading to bowel ischaemia and strangulation; (2) malignancy is often associated with ileocaecal intussusceptions; (3) manipulation of the bowel may result in perforation and consequent contamination and tumour dissemination. Among the most compelling arguments in favour of primary en bloc resection in these patients is that they often present at an early stage and with completely resectable disease.[9]

There has been a debate in the past regarding the role of surgical reduction. In 1956, Roper[10] recommended manual reduction of all enteric intussusceptions in adult patients prior to definitive treatment. Recently, the role of manual reduction has been revisited. Chiang and Lin[11] reviewed 72 patients older than 16 years of age and found a 24% prevalence of ileocaecal or ileo-caecal-colic intussusceptions. Most of these patients underwent reduction followed by resection. One patient, who underwent reduction only because no leading point was found at initial operation, required right colectomy 2 years later for adenocarcinoma. Honjo *et al*[12] reviewed 44 adult patients who presented with bowel intussusception and advocated reduction followed by limited resection. This approach was successful in 4 of 15 patients with ileocaecal intussusception. Overall, 14 right colectomies were performed, but it is not clear whether a laparoscopic approach was used.

Primary resection should be the initial therapy of choice in ileocolic intussusceptions presenting with a history of prior episodes of intestinal obstruction and evidence of a mass at surgical exploration.[13 14] It has been clearly shown that resection can safely be performed even in emergent situations without prior bowel preparation.[15] Compromised blood supply is an additional reason to proceed directly with resection without any attempt at reduction. In addition, reduction is a traumatic manoeuvre. There are multiple and well-recognized advantages for using laparoscopy in the acute care surgery setting: less postoperative pain and discomfort, shorter hospital stay, and earlier return to daily activities. In addition, the quality of resection and the patients quality of life after laparoscopic bowel surgery have proven satisfactory.[16] Attempts at bowel reduction in ileocolic intusssusceptions in adult patients should not have any role[17 18] because it is not reasonable trying to spare a longer resection and then perform an anastomosis on traumatized, edematous and ischemic bowel edges.

Learning points

- Ultrasound and CT scans are often diagnostic of ileocolic intussusception.
- Ileocolic intussusception due to Burkitt lymphoma is extremely rare in adults.
- Reduction in ileocolic intussusception should never be attempted.
- Laparoscopy may confirm the diagnosis and allows performing a safe and complete primary oncological resection.
- Adjuvant combination therapy is recommended even in patients with localised disease to increase the chance of long-term survival.

Contributors DB and EA collected the patient's data and wrote the first manuscript draft. LB supervised and approved the final version of the manuscript.

Competing interests None declared.

Patient consent Obtained.

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