Ileosigmoid knotting in a child: Index case in Botswana and review of the literature

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Ileosigmoid knotting (ISK) is a rare form of complex intestinal obstruction where a loop of ileum wraps around the base of the sigmoid colon, or vice versa. We report the case of an ill-looking 8-year-old boy who presented with abdominal pain and vomiting. Abdominal examination revealed distension and features of diffuse peritonitis. Complicated appendicitis was at the top of the list of differential diagnoses. The patient was resuscitated and prepared for surgery. At laparotomy, haemorrhagic fluid with gangrenous ileum wrapping around gangrenous sigmoid colon was found. *En bloc* resection of the gangrenous ileum and sigmoid colon with primary ileo-ileal and descending colorectal anastomoses were done. The patient was discharged on the 10th postoperative day. Although extremely rare in this patient population, surgeons must be able to recognise and manage ISK in children.

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Ileosigmoid knotting (ISK) is an uncommon cause of bowel obstruction where a loop of ileum and a loop of sigmoid colon form a knot.[1-9] ISK was first reported by Parker in 1845.[1-5] ISK contributes to 0.5 - 1.7% of all cases of intestinal obstruction.[1] Most cases are reported in adults, typically in the third and fourth decades of life, and are most commonly seen in Africa and Asia.[1-9] ISK is unusual in childhood.[2,3,5-9] ISK is five times more common in males than in females.[1-3,5] The aetiology of ISK is multifactorial. Contributing factors include a long ileal mesentery, a long and narrow-based mesosigmoid, a heavy meal or high fluid intake after a long period of fasting, and a floating caecum.[2-5,7-9] Shepherd[10] is credited with the description of the mechanism for ISK: the loop of sigmoid containing faeces and gas presses on the loop of ileum, which is pushed down and wraps around the base of sigmoid loop. The apex of the loop then passes underneath the initial wrap, completing the knot. As the loops distend, they increase the tightness.[3,7]

To the best of our knowledge, only 14 paediatric cases of ISK have been reported in the literature.

Patient information

An 8-year-old boy presented with a 1-day history of abdominal pain and vomiting. On examination, he appeared seriously ill. The clinical findings were in keeping with diffuse peritonitis. He was resuscitated and prepared for surgery. His history and findings seemed most in keeping with complicated appendicitis (with perforation and peritonitis). After the patient was resuscitated with intravenous fluids and given antibiotics (cefotaxime, vancomycin and metronidazole), he was taken for laparotomy. Although ultrasound, radiograph and computed tomography (CT) scan services were available in the hospital, these were not performed, as the clinical findings were in keeping with an acute surgical abdomen requiring urgent laparotomy.

Upon entering the abdomen, haemorrhagic fluid was found. The patient was found to have ileosigmoid knotting, where the gangrenous ileum was tightly wrapped in a clockwise direction around the base of the equally gangrenous sigmoid colon (Fig. 1). *En bloc* resection was done (Fig. 2). Intestinal continuity was established by ileo-ileal and descending colon-rectal end-to-end anastomoses. The resected ileum was 30 cm in length, and the distal margin was 10 cm from the ileocaecal valve. The mesenteries of both ileum and sigmoid were normal. On the third postoperative day, the nasogastric tube drainage was minimal, and it was removed. On the fourth postoperative day, the patient was tolerating a liquid diet, had passed stool and was fully ambulant. He was discharged back to the referring public hospital on the tenth postoperative day, and subsequently discharged home. He remains asymptomatic 4 months after surgery.

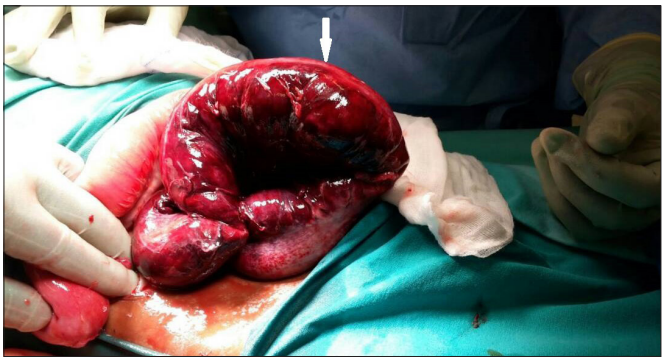

Fig. 1. Ischaemic and macerated sigmoid colon (white arrow).

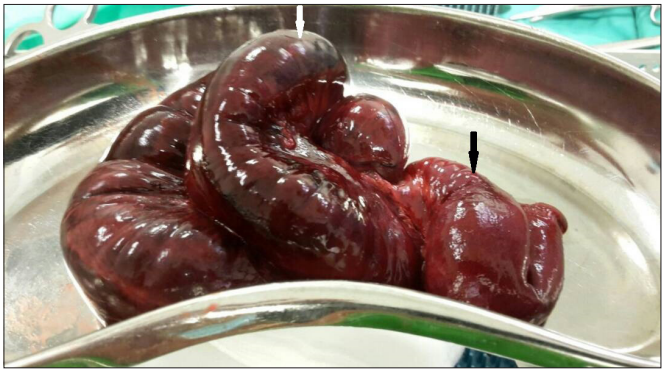

Fig. 2. En bloc *resected sigmoid colon (white arrow) and gangrenous ileum (black arrow).*

Discussion

Alver *et al.*[11] classify ISK into three types: type I, where the active ileum wraps around the base of the sigmoid colon; type II, where the active sigmoid colon wraps around the ileum; and type III, where the active ileocaecal segment wraps around the sigmoid colon.[2] Each type is subclassified into two subtypes based on whether the active component rotates in a clockwise or counter-clockwise direction.[2,4,9] Both ileum and sigmoid colon are gangrenous in 43% of cases, ileum alone in 33%, sigmoid alone in 4%, and neither is gangrenous in 20% of cases.[4,9] The most commonly reported presentations in the literature are abdominal pain (100% of cases), nausea and vomiting (87 - 100%), abdominal distension (94 - 100%), rebound tenderness (69%) and shock (0 - 60%).[1,4,9] Leukocytosis, elevated blood urea nitrogen, a drop in haemoglobin and electrolyte imbalance have been variably reported in laboratory investigations.[3,4] According to the reviewed literature, most cases of ISK are diagnosed intraoperatively. This is the result of a combination of factors: the rarity of the condition, the nonspecific presentation and the atypical radiographic findings.[1-4,7-9] Sometimes, a double closed loop bowel obstruction with a sigmoid loop in the right upper quadrant and a small bowel loop on the left may be seen on plain abdominal X-ray.[2-4,7,9] CT may variably show a whirl sign (twisted intestine and mesentery), along with medial deviation of the caecum with a beak appearance of the descending colon, or a dilated loop of sigmoid colon with convergence of the superior mesenteric vessels toward the knot.[1-4,7,9]

In our literature review, we identified only 14 reported cases of ISK in children.[2,5,7,9] Paediatric patient ages ranged from 6 to 16 years, with a mean age of 9.6 years, and a male-to-female ratio of 11:2. As in the adult literature, one or both limbs of the intestine were gangrenous in the majority of paediatric cases. Based on our review of the paediatric literature, the operative procedure depended on the findings and the stability of the patient.

Untwisting of the ISK is not advised. Not only is it difficult, but it carries a risk of rupture of the gangrenous bowel and ensuing endotoxaemia.[6,7,9] Depending on the patient's haemodynamic status, the intraoperative findings and the surgeon's judgment, resection of the gangrenous segments may either be followed by primary anastomosis or creation of stomas.[1,3,4,6,7,9]

Although the diagnosis of ISK was rarely established preoperatively in the articles reviewed, key elements in the success rate of management were felt to include judicious fluid and electrolyte resuscitation, correction of acid-base imbalances, adequate antibiotic coverage, prompt laparotomy, appropriate intraoperative decision-making and aggressive perioperative care.[2-5,7,9] Mortality rates are described to be between 19% and 47%, the most common cause being septic shock.[2-5,7,9] In patients aged >60 years, surgical intervention beyond 24 hours of presentation, and extent of gangrene, are associated with a higher mortality.[3,7] Long-term morbidity includes adhesive obstruction, chronic diarrhoea and malnutrition.[6]

As in most case reports, our patient was male.[1,3,5-7,9] Also in keeping with most other reports, both ileum and sigmoid colon were gangrenous.[2,4-6,9]

As stated, most cases of ISK are diagnosed intraoperatively.[1-4,7-9] In our case, complicated appendicitis was at the top of the list of differential diagnoses. Our sense was that the patient required urgent laparotomy, hence no imaging studies were ordered.

Paediatric patients with ISK usually present as an emergency, and are in a physiologically compromised state.[1-9] The diagnosis is rarely established preoperatively. Depending on the patient's physiologic status, the intraoperative findings and the experience of the surgeon, resection of gangrenous segments and primary anastomosis and/or creation of stoma(s) are reasonable options.[1,3,4,6,9] Re-look laparotomy is recommended in those patients where the viability of the residual bowel is in question.[5,7]

Conclusion

Although ISK is very rare in children, its recognition and appropriate surgical management are crucial to achieve the best possible outcomes: both general and paediatric surgeons should be familiar with the intraoperative findings and the appropriate surgical options.

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