Ileosigmoid knotting in a child

The first case report in a French girl

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SUMMARY

Ileosigmoid knotting occurs when the ileum wraps around the base of an elongated sigmoid colon. It is rare in developed countries, especially in children. The authors report the first case of ileosigmoid knotting in a Caucasian French girl.

Introduction

Ileosigmoid knotting (ISK) is an extremely rare cause of bowel obstruction and ischemic necrosis; it consists of a loop of the ileum encircling the loop of the sigmoid colon [1-3]. This unusual entity is fairly rare in Europe and most cases have been reported in certain African, Asian and Middle Eastern countries [4, 5]. Typically, ISK occurs in adults and only a few cases have been described in children.

We describe the first case of ISK in a 9-year-old Caucasian French girl.

Case report

A 9-year-old French girl was admitted to our unit at Robert Debré Hospital for profound shock. She had been in severe abdominal pain and vomiting in the early morning. On admission, she was pale, with hypovolemia. On physical examination her temperature was 37 °C, the abdomen was extremely distended with diffuse tenderness. X-ray examination showed intestinal loops and a markedly dilated loop in the upper right quadrant (figure 1a). Tomodensitometry showed marked ascites and thickening of the intestinal walls (figure 1b). After volume repletion, she was taken to the operating room for urgent laparotomy without a precise diagnosis. The operation was performed 12 hours after the onset of symptoms. At laparotomy, a large quantity of blood-stained fluid was noted and the ISK was observed. A loop of strangulated ileum was found tightly encircling the congested and dilated loop of the sigmoid colon forming a knot (figure 1c).

The 22 cm sigmoid colon was obviously necrotic and was resected. Two meters of the distal ileum was also ischemic with, 50 cm frankly necrotic. No perforation was visualized. The 50 cm of distal ileum was resected and the ileocecal valve was preserved. Ileostomy and colostomy were performed on each side of the resected areas. A second intervention was not necessary and 3 weeks after the initial operation, the stoma was closed after baryiom opacification to rule out any intestinal stonies. The total duration of parenteral nutrition was 3 weeks and the patient was discharged. She is in good condition after 15 months of follow-up.

Discussion

Since the first report by Parker et al. in 1845 [6], cases of ISK have been described in several countries, especially in developing countries but, to the best of our knowledge, never in France. The young girl in this case had no history of recent immigration, but the hypothesis of a possible origin in Africa. ISK occurs predominantly in previously healthy adults mainly in the fourth or fifth decade [4, 5] and only a few cases of ISK have been reported in children [7-10]. ISK usually occurs in males (5:1 male preponderance) either in adults or children [5]. The present case is the first reported case of ISK in a French female child since all other reported children cases have occurred in males. Chirdan et al. described 4 cases of ISK; the author described 4 ISK and 2 sigmoid volvulus with only one girl, and no additional details.

In that study, the median age at presentation was 4.5 years old (range 2 weeks-15 years). In two other studies [9], patients were 6 year-old boys. The etiology of ISK is complex. It involves anatomical factors as in our case which was considered to have an anatomic basis. These include a long small intestinal mesentery with a mobile small intestine [1, 5] and a long sigmoid colon on a narrow pedicle [4]. Other factors involved in ISK are late pregnancy, transmesenteric herniation, Meckel’s diverticulitis with a band, ileocecal intussusception and floating caecum and ingesting high-bulk foods and liquids after prolonged fasting [1, 4, 5]. The diagnosis, before surgery is often difficult to establish. Indeed, the symptoms of ISK are not specific and the accurate preoperative diagnosis is made in less than 20% of cases [5, 10]. Even during surgery, ISK can be misdiagnosed because the macroscopic features of this unusual condition are not familiar to surgeons. It is thus important to consider the diagnosis of ISK in children with a strangulated ileum. Characteristic radiological features including a double closed loop obstruction, with...
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the small intestinal loops in the upper left quadrant and the sigmoid loop in the right [1, 5] are often misdiagnosed pre-operatively. Abdominal tomodensitometry also exhibits characteristic findings which may reveal a sigmoid volvulus including characteristic whirlpool signs created by the twisted intestine and mesocolon created both by the twisted intestine and mesentery and medial deviation of the descending colon with a beak appearance on its medial border [11, 12]. CT can also reveal signs of bowel ischemia caused by strangulation, such as pneumatosis. However, an ileosigmoid knot is not easily detected because the ileal twist is higher in the abdomen than the location of a sigmoid volvulus. The whirl is visible on more contiguous slices than that of the sigmoid volvulus. Compared with an abdominal X-ray, CT can detect medial deviation of the distal descending colon with a pointed appearance of its medial border, which is a distinct feature of the ileosigmoid knot. These features were noted retrospectively in our case. Alver performed a new classification for ISK. Type I: the ileum (active component) wraps itself around the sigmoid colon (passive component) in a clockwise or counterclockwise direction, Type II: the sigmoid colon (active component) wraps itself around a loop of ileum (passive component) in a clockwise or counterclockwise direction, Type III: the ileocecal segment (active component) wraps itself around the sigmoid colon (passive component). We think in our case report corresponds to ISK type I.

The mortality rate is high, 25 to 47% [1, 4, 5, 13], mainly due to toxic shock from bowel gangrene. Prompt laparotomy and resection of the non viable or doubtful intestines in a patient with antibiotic therapy and adequate fluid resuscitation are key factors to diminish the mortality rate. In our case, both the clearly gangrenous ileum and sigmoid were resected; some authors recommend performing sigmoid resection even if it appears to be macroscopically viable [3, 4] whereas in some reported cases, the sigmoid is not resected [10]. Even though this pathology is rare in adults and even rarer in children, it should be known and always rapidly treated.

REFERENCES