Review Article

Review of Management of Ileosigmoid Knotting

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ABSTRACT

Ileosigmoid knotting (ISK) is a rare type of intestinal obstruction caused by knotting of the mesentery of the ileum or sigmoid colon that rapidly progresses to gangrene with a high risk of mortality and high morbidity. The incidence of ISK is not well established, but it is higher in regions with high rates of sigmoid volvulus and in countries along the sigmoid belt. Clinical presentation is that of both small-bowel and large-bowel obstruction and includes vomiting and nausea, abdominal pain, tenderness, and distention, with constipation. A contrast-enhanced computer tomography (CT) scan is the preferred modality for imaging. Management involves hemodynamic stabilisation with correction of shock using aggressive fluid resuscitation, electrolyte balance and commencement of antibiotics. Principles of surgery include resection of the knot, resection of the gangrenous bowel and establishing intestinal continuity. The outcome is generally complicated by peritonitis and sepsis that lead to mortality.

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Introduction

Ileosigmoid knotting (ISK), which is also known as double volvulus or compound volvulus, is a rare cause of intestinal obstruction [1-3]. It results in a closed-loop obstruction caused by the knotting or intertwining of the mesentery of the ileum or sigmoid colon that rapidly progresses to the strangulation of one or both loops of the bowel [1-4]. ISK causes a double-loop obstruction that rapidly progresses to bowel gangrene and death if emergency surgery is not undertaken [5]. Parker is credited with describing the first case of ISK [6]. The incidence of ISK is ranked at 4% in developed countries and 50% in developing countries [7]. The incidence of ISK is not well known but is higher in areas with a high incidence of sigmoid volvulus [5]. ISK is most seen in males, and it presents with a peak incidence in the third to fourth decade of life [1].

The aetiology of ISK remains controversial and is commonly seen in the sigmoid belt, namely Africa, Asia, and the Middle East, but in East Africa, ISK is commonly seen among the young males of the Baganda tribe [1, 2]. ISK is common in areas of low socioeconomic status [2]. Preoperative diagnosis of ISK is difficult as the condition has an infrequent presentation and rare radiological features; however, a high index of suspicion should be used in areas with a high incidence of sigmoid volvulus where advanced imaging is available [4, 8]. ISK presents as an acute abdomen that requires emergency surgery and will result in death if not done [8]. This paper outlines the management of ISK in a resource-limited site without advanced imaging, at Ndola Teaching Hospital.

History of Sigmoid Knotting

The first description of ISK was by Riverius in the sixteenth century. A detailed account of this rare condition was given by Rokitansky in 1836 [9]. The first reported case describing a patient with ISK was by Parker in 1845 [6]. The first account of ISK in Africa was given by Burkit in 1952, who carried out studies among the Baganda people in Uganda (who eat once a day) and provided evidence for the description of the mechanism of ISK, and among Baganda Muslims (who eat a single meal per day during the Ramadan fast) [4].

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Clinical Presentation

ISK is common in males, at 82%, in their fourth decade. Though rare in females, it presents in late pregnancy [2, 4, 5]. ISK is common in African people because of geographical (sigmoid belt), racial and dietary factors [10, 11]. Predominant symptoms include:

i. abdominal pain (100%)
ii. abdominal distension (94%-100%)
iii. nausea and vomiting (87%-100%) and
iv. partial or absolute constipation [4].

Signs of ISK include:

i. abdominal tenderness (100%)
ii. rebound tenderness (69%) and
iii. shock (0-60%) [4].

Pathophysiology

The aetiology of ISK is controversial, but the following factors are thought to be responsible for ISK:

i. a long mesentry of the small bowel and a freely mobile small bowel,
ii. a long axis of the sigmoid colon with a narrow pedicle and mesentry base and
iii. ingestion of bulk food in the presence of an empty small bowel [3, 5].

Other factors include relaxation of the abdomen during sleep or postpartum period and Meckel’s diverticulum, which has been found to be 14-53% of ISK [1]. Other secondary causes include late pregnancy, trans-mesenteric herniation, ileocaecal intussusception, and a floating caecum [12].

Proximal jejunal mobility due to increased peristalsis is increased by semiliquid bulky ingested food that makes heavy segments of the jejunum fall on the left lower quadrant, while the empty loops of distal jejunum and ileum twist around the base of the redundant sigmoid colon [4]. Evidence for this mechanism is suggested by studies on the Baganda people of Uganda [10, 13]. ISK is essentially a closed-loop obstruction involving the ileum and sigmoid intestinal loops [1, 8]. The entanglement of the ileum and the sigmoid rapidly progresses to a vascular compromise resulting in gangrene of the ileum and sigmoid loops, and perforation [7].

Alver first classified ISK into four types based on the active and passive components of either the ileum or the sigmoid colon [13]. These four types were further subdivided based on the direction of torsion into A (a clockwise direction) and B (an anticlockwise direction). Alver’s classification includes:

i. Type I, where the ileum is an active component that wraps around the sigmoid,
ii. Type II, where the redundant sigmoid colon wraps around the ileum,
iii. Type III, where the ileocecal loop of bowel is wrapped around the sigmoid colon, and
iv. Type IV, where the active segment cannot be determined.

Al-Qahtani et al. report that the most common ISK is Type I with subsection A (clockwise direction of torsion) [5].

Investigations

The preferred imaging investigation in ISK is a contrast-enhanced CT scan, which reveals the classical “whirl sign” of sigmoid volvulus created by the twisting of the mesentery of the bowel, and the appearance of a “birds beak sign” formed by the efferent and afferent limbs of the sigmoid colon [14, 15]. The whirl sign is created by the twisting of the small bowel and sigmoid colon in many slices of the CT scan, compared to the twists found in the sigmoid volvulus [1]. The CT scan is not always applicable for most patients due to their poor conditions on presentation [5].

A plain abdominal X-ray may show closed-loop obstruction, with the sigmoid colon (with omega sign) on the right and the small bowel on the left, a rare finding [1, 3]. Features of sigmoid volvulus with the omega sign can lead to attempts to perform endoscopic decompression by insertion of the rigid sigmoidoscope, which if performed can lead to perforation in the presence of bowel gangrene [4]. X-ray is highly inaccurate in the diagnosis of ISK preoperatively [4]. Clinical features of small-bowel obstruction and radiological features of large-bowel obstruction are contradictory and often lead to misdiagnosis [12]. Laboratory results show leukocytosis, low haemoglobin, raised blood urea nitrogen (BUN) and electrolyte imbalance suggestive of peritonitis [4].

Diagnosis

Preoperative diagnosis of ISK is difficult, commonly so when a CT scan is not used, due to the rarity of the condition and atypical radiological features. The diagnosis is therefore mostly arrived intraoperatively at laparotomy [12]. Commonly, the radiographic appearance of ISK is mistaken for sigmoid volvulus [4].

Raveenthiran described a diagnostic criterion that increases chances of a preoperative diagnosis, which includes the following:

i. clinical features of small bowel obstruction,
ii. radiological features of large bowel obstruction, and
iii. failure to insert a sigmoidoscope, to achieve endoscopic decompression [16].

The above three factors form a diagnostic criterion that increases suspicion of ISK. The lack of capacity for early diagnosis and intervention is associated with high mortality, reported to be between 20-100%, where bowel gangrene is present [10, 13, 17, 18].

Management and Treatment

The clinical presentation of ISK is that of an acute abdomen where emergency, accurate and curative management should not be delayed, since the condition rapidly progresses to gangrene of the bowel, demanding early diagnosis and operative treatment [17]. Management should begin with aggressive fluid resuscitation, electrolyte balance, correction of acid-base imbalance and a five-to-seven-day course of antibiotics. The usual combination of cephalosporin, aminoglycosides,
and metronidazole should be commenced immediately [19]. Care should be taken to manage the shock aggressively and promptly in the patient to allow early surgical intervention, since this has been shown to increase the survival of these patients [19].

Surgical Treatment

Immediate surgical intervention should be instituted as soon as hemodynamic stability and resolution of shock is achieved [19]. Surgery achieves the following:

i. resection or release of the knot,

ii. resection of gangrenous bowel loops,

iii. continuation of gastrointestinal continuity, and

iv. prevention of recurrence of ISK [18].

The detorsion of viable bowel can be done in selected patients in the absence of gangrene to allow definitive treatment. However, gangrenous bowel is resected and never untwisted to release the knot because of the risk of perforation or rupture leading to spillage of toxic intestinal contents into the abdomen [18]. Mandal et al. report the incidence of gangrene to be 90.9% for those reporting in the first 24 hours after the start of symptoms, while those reporting more than 24 hours after the start showed a 57% incidence of gangrene, indicating that early presentation has a higher incidence of bowel gangrene [4]. If both loops of bowel are viable, the knot may be undone via sigmoid enterostomy and traction of the sigmoid colon performed to allow primary anastomosis of both the small bowel and the sigmoid colon as definitive treatment [17].

Definitive surgical treatment includes the following:

i. ileal resection and primary anastomosis plus sigmoid resection and primary anastomosis—where both the small bowel and the sigmoid colon are viable, recent data suggests that primary sigmoid anastomosis can be safely undertaken if the bowel is clean, well-vascularized, and not distended,

ii. ileal resection and primary anastomosis plus sigmoid resection with Hartman’s colostomy, where gangrene is present, and

iii. ileal resection with loop ileostomy plus sigmoid resection with primary anastomosis, where gangrene is present [4].

Even though primary anastomosis of the small bowel is preferred, including in the presence of a small resected gangrenous segment, a surgeon should be mindful that if ileal gangrene is within 10cm of ileocaecal junction, then right hemicolectomy, not primary anastomosis, should be performed to avoid an anastomotic leak [4]. Mandal et al. report that the most common procedure performed is ileal resection with primary anastomosis plus sigmoid resection with Hartman’s colostomy. Resection of a viable sigmoid bowel should in all instances be to prevent a recurrence of ISK or sigmoid volvulus [17].

Outcome

ISK is a life-threatening condition caused by a rare intestinal obstruction that rapidly progresses to gangrene [17]. A poor outcome is usually the result of peritonitis and sepsis in ISK [5]. Avoiding colostomy reduces patient morbidity and the cost of health care [5, 19-21]. Reported mortality from ISK is reported to range from 0-48%, with a mean of 35.5% [4]. The incidence of mortality is related to advanced age, comorbidities, duration of symptoms, presence of gangrene and septic shock, leading to multiorgan failure [18]. Chalya et al. report that sepsis is the most common complication in ISK found in their study [7].

Prognosis

ISK has a poor prognosis due to the presence of gangrene and sepsis, resulting in mortality of 48% that can reach 100% [5, 13, 18].

Conclusion

ISK is a life-threatening cause of intestinal obstruction that rapidly progresses to gangrene of both the small bowel and the sigmoid colon, with a poor outcome and high mortality due to peritonitis and sepsis. Early fluid resuscitation to achieve hemodynamic stability, correction of electrolytes, early commencement of antibiotic therapy and early surgical intervention reduce the high mortality associated with ISK.

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Conflicts of Interest

None.

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