Ileosigmoid Knot

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Abstract Ileosigmoid knotting, also known as compound volvulus or double Volvulus, is a rare cause of acute intestinal obstruction. In this condition the ileum wraps around the base of the sigmoid colon and forms a knot. The condition is serious, generally progressing rapidly to gangrene of both ileum and sigmoid colon. Ileosigmoid knotting is an unusual entity in the West, but is comparatively common in certain African, Asian and Middle Eastern nations. Awareness of the condition is essential for prompt diagnosis and optimal management. This article will focus on the etiopathogenesis, presentation, diagnostic modalities, surgical interventions and outcome with review of articles and case reports published till date.

Keywords Ileosigmoid knot · Compound volvulus · Intestinal obstruction

Introduction

Ileosigmoid knotting (ISK) is a rare cause of intestinal obstruction that rapidly progresses to gangrene of the ileum as well as the sigmoid colon. Preoperative diagnosis is difficult because of its infrequency and atypical radiographic findings. Even the surgery textbooks, such as *Bailey & Love's Short Practice of Surgery* and *Shackelford's Surgery of the Alimentary Tract*, give only a brief description of ISK. It is essential to differentiate it from sigmoid volvulus because endoscopic reduction may be possible in sigmoid

volvulus, but not in ISK. In recent years, computed tomography (CT) has emerged as a preoperative diagnostic tool. Generalized peritonitis and sepsis are the main causes of poor outcome. After hemodynamic stabilization, immediate surgical intervention is the only viable option.

History

ISK is very rare in many parts of the world. Parker [1] is credited with having described the first case of ISK. Ekehorn (1903) and Faltin (1909) in their studies classified the ileosigmoid knot according to bowel involved and arrangement of loops.

Kalleo (1932) reviewed a total of 84 cases in the world literature and added 77 additional cases from Finland, making a total of 161 cases. At that time no cases were reported from Africa and America. The first patient from the Asian subcontinent was reported by Paul in 1940, an account of which was given by Shepherd [2], and from Africa by Burkitt in 1952. The first case from India was reported by Dunkertey (1953). Since then, over 300 cases have been reported in English literature [6, 11, 14, 16, 17].

Etiopathogenesis

Three factors are responsible for the ileosigmoid knot [3, 7, 16]:

- I. A long small bowel mesentery and freely mobile small bowel.
- II. A long sigmoid colon on a narrow pedicle.
- III. Ingestion of a high bulk diet in the presence of an empty small bowel.

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Fig. 1 Mechanism of ISK formation

When a semiliquid bulky meal progresses into the proximal jejunum, it increases the mobility of the intestine and the heavier segments of the proximal jejunum fall into the left lower quadrant. The empty loops of ileum and distal jejunum twist in a clockwise rotation around the base of a narrow sigmoid colon.

Fig. 2 **a** Supine X-ray. Dilated sigmoid loop (*small arrow*), medially deviated and distended descending colon (*large arrow*) and dilated loops of small intestine (*arrow head*). **b** Line diagram showing anatomical relations in ileosigmoid knot

Further peristalsis forms an ileosigmoid knot with two closed loop obstructions (Fig. 1), one in the small bowel and other in the sigmoid colon. Evidence for the mechanism is suggested by studies carried out on Baganda people in Uganda who eat once a day and on Muslims who eat a single daily meal during the Ramzan fast [2, 6]. ISK is predominately seen in males (80.2%) with a mean age of 40 years.

Besides the above anatomic prerequisites, the literature reveals the evidence of other secondary causative factors including late pregnancy, transmesenteric herniation, Meckel diverticulitis with a band, ileocecal intussusception, and floating cecum. While ISK is predominately reported in certain African, Asian, and Middle-East nations, it is a rare occurrence in the white population [5, 10, 11, 13].

Classification

ISK has been categorized into the following three types:

- Type I, the ileum (active component) wraps itself around the sigmoid colon (passive component) in a clockwise or anticlockwise direction (type A when clockwise and type B when anticlockwise).
- Type II, the sigmoid colon (active component) wraps itself around a loop of ileum (passive component) in a clockwise or anticlockwise direction.
- Type III, the ileocecal segment (active component) wraps itself around the sigmoid colon (passive component).

The most common type of ISK reported is type I (53.9–57.5%), followed by type II (18.9–20.6%), type III (1.5%), and others undetermined. The direction of torsion is clockwise in 60.9–63.2% of cases [2, 6, 11].

Fig. 3 **a** CT shows dilated loop of ileum (*white arrow head*) with mesenteric whirl sign (*arrow*) and medially deviated descending colon and cecum (*black arrow head*). **b** Gas filled ileal loop (*arrow head*) and whirl sign (*small arrow*) and distended sigmoid colon (*large arrow*)

Clinical Presentation

Usually, ISK presents as acute abdominal events. It can rapidly progress to gangrene of the ileum as well as of the sigmoid colon that may result in fatal complications such as, generalized peritonitis, sepsis, dehydration, and electrolyte imbalance. The predominant symptoms and signs of presentation include abdominal pain and tenderness (100%), abdominal distension (94%–100%), nausea and vomiting (87–100%), rebound tenderness (69%), and shock (0–60%), where it was specified [2, 6].

Diagnosis

Despite the critical condition, preoperative diagnosis is not easy. In recent years preoperative diagnoses are being made more often, but it was a rarity in the past in most cases. The diagnostic difficulty is partly caused by the unfamiliarity of this rare entity and the confusing and self-contradictory features of the disease. Although clinical features such as vomiting suggest small bowel obstruction, the radiographic findings are that of colonic obstruction, which is uncommon in small bowel obstruction [4, 6, 8]. Radiographically, ISK is often mistaken for simple sigmoid volvulus. However, unlike sigmoid volvulus, attempts to deflate the distended colon using a sigmoidoscope or a flatus tube, often fails and may be dangerous in ISK. This is because the ileum tightly envelops the base of the sigmoid colon, defying any such attempt. Combining the three features—clinical picture of small bowel obstruction, radiographic evidence of predominantly large bowel obstruction, and inability to insert a sigmoidoscope—could possibly form a useful diagnostic triad and raise the suspicion of ISK [8] (Fig. 2).

The radiographic findings of ISK, which include a double loop of dilated sigmoid shadow and multiple air

fluid levels in the small intestine, are sporadically described and are difficult to identify as such because of unfamiliarity [11, 12]. Nonetheless, it is important that they should at least raise the suspicion of ISK (Fig. 2).

In the recent years, contrast-enhanced computed tomography is emerging as a sensitive tool for the diagnosis of ISK. The findings in a CT scan suggestive of ISK include ‘the whirl sign’ created by the twisted intestine and sigmoid mesocolon in ileosigmoid knot, medial deviation of the cecum, and descending colon. In addition, others have noted radial distribution of the intestine and mesenteric vasculature and consider it to be helpful diagnostic information [9, 10, 12]. However, CT examination may not be possible in all patients because of nonavailability or some patients could be in a very poor

Fig. 4 Ileosigmoid knot with gangrenous terminal ileum (*arrow*) and sigmoid colon (*arrow head*)

Fig. 5 **a** Straight X-ray Abdomen showing Typical Sigmoid volvulus. **b** Line diagram showing anatomical relations in Sigmoid Volvulus

condition. Whenever possible, a CT examination could be useful in making a diagnosis in these otherwise difficult patients (Figs. 3, 4 and 5).

Laboratory reports usually show drop in hemoglobin, leucocytosis, raised BUN, and electrolyte imbalance suggestive of peritonitis.

Table 1 Clinical presentations and symptomatology as noted by various authors

Author (yrs)	No. of cases	Age mean	Sex [M:F]	Duration of symptoms in hours [mean/range]	Symptoms						Pre-diagnosis
					P	D	V	C	M	S	
Shepherd et al. (1967)	92	42 (11–75)	78:14:00	18 (6–76)	92	90	?	?	?	?	?
Kakar et al. (1981)	11	38	10:01	?	11	3	11	?	?	4	2
Puthu et al. (1991)	7	40	4:03	36 (8–72)	7	7	7	–	–	2	2
Miller et al. (1992)	1	41	0:01	2	1	0	1	.	.	0	0
Alver et al. (1993)	68	49 (18–79)	57:11:00	33 (24–144)	68	64	59	64	.	38	0
Akgun et al. (1997)	16	45 (20–90)	11:05	100 (1–144)	16	15	14	.	.	9	?
Kedir et al. (1998)	9				9	9	9	9	.	5	.
Raveenthiran et al. (2001)	7	43 (30–60)	6:01	53 (6–240)	7	6	5	7	.	4	5
Tamura et al. (2004)	1	78	1:00	?	1	1	.	.	.	.	1
Hashimoto et al. (2004)	2	32 (4–60)	2:00	26	1	2	.	.	1	.	1
Atamanalp et al. (2004)	63	47 (7–75)	47:16:00	46.6 (12–120)	63	60	53	63	9	38	0
Hirano et al. (2005)	1	75	1:00	28	1	1	1	.	.	.	1
Berrebi et al. (2006)	1	9	0:01	12	1	1	1	.	.	1	0
Atamanalp et al. (2006)	9	10.6 (7–16)	7:02	45.3 (24–72)	9	9	9	9	3	3	0
Aguka et al. (2006)	1	25	M	96	1	1	1	.	.	.	0
Kotisso et al. (2006)	15	45	M>F	83	15	15	15	15	.	.	
Bawa et al. [15]	1	26	1:00	>240	1	1	1	1	.	.	0
Alwi et al. (2009)	2										
Machado et al. (2009)	1	60	M		1	1	1				
Burrah et al. (2010)	1	60	M	48	1	–	–	1	–	–	?

P pain abdomen, D distention, V vomiting, C constipation, M melena, S shock

Table 2 Predisposing factors and pre-operative investigation done for diagnosis as by various authors

Author	No.	Predisposing factors						Preoperative investigations	
		LP	PS/Ad	MD	FC	IH	NS	X-ray	CT
Shepherd et al. (1967)	92	.	.	.	.	.	92	NS	.
Kakar et al. (1981)	11	.	.	.	.	.	.	8	.
Puthu et al. (1991)	7	.	.	1	.	.	.	7	.
Miller et al. (1992)	1	.	1	.	.	.	.	1	.
Alver et al. (1993)	68	4	12	1	1	4	.	56	.
Akgun et al. (1997)	16	.	.	.	.	.	16	16	.
Kedir et al. (1998)	9	.	.	.	.	.	.	.	.
Raveenthiran et al. (2001)	7	.	.	.	.	.	7	7	.
Tamura et al. (2004)	1	.	1	.	.	.	.	1	1
Hashimoto et al. (2004)	2	.	.	.	.	.	2	2	2
Atamanalp et al. (2004)	63	2	10	.	1	.	.	41	0
Hirano et al. (2005)	1	.	.	.	.	.	1	1	1
Berrebi et al. (2006)	1	.	.	.	.	.	1	1	1
Atamanalp et al. (2006)	9	.	.	.	.	.	9	9	0
Aguka et al. (2006)	1	.	.	.	.	.	.	1	0
Kotisso et al. (2006)	15	.	.	.	.	.	.	15	0
Bawa et al. [15]	1	.	.	.	.	.	1	1	0
Alwi et al. (2009)	2	.	.	.	.	.	.	2	0
Machado et al. (2009)	1	.	.	1	.	.	.	1	1
Burrah et al. (2010)	1	–							

LP late pregnancy, PS/Ad previous abdominal surgery, MD Meckel's Diverticulum, FC floating caecum, IH internal herniation, NS nothing significant

Management

The initial management involves aggressive resuscitation with fluid and electrolytes and the correction of acid-base imbalance if any. After hemodynamic stabilization, operative intervention should be undertaken as early as possible. Appropriate antibiotic therapy is commenced early and continued for 5–7 days after the operation. The usual antibiotic combination includes cephalosporins, aminoglycosides, and metronidazole [5, 7, 8].

The anatomical and pathological changes dictate the operative procedure. In 73.5–79.4% of the cases, gangrenous bowel was encountered, whereas in 20.6–26.5% both small and large bowels were assessed to be viable in surgery. In 52.9–60.3% cases, both the small intestine and sigmoid colon were gangrenous. Paradoxically, the incidence of bowel gangrene was 90.9% in those who presented within 24 h of their symptoms. Among those who presented after 24 h after their initial symptoms, bowel gangrene was seen in 57% [2, 6] (Tables 1 and 2).

The literature shows various surgical options to manage this condition (Table 3). The various surgical procedures undertaken in the past and present include:

1. Ileal resection + Primary Anastomosis + Sigmoid Derotation (IR + PA + SD)
2. Ileal Resection + Primary Anastomosis + Mesosigmoidostomy (IR + PA + MS)

3. Ileal Resection + End Ileostomy + Sigmoid Resection + Primary Anastomosis (IR + EI + SR + PA)
4. Ileal Diversion + Sigmoid Resection + Primary Anastomosis (ID + SR + PA)
5. Ileal Resection + Primary Anastomosis + Sigmoid Resection + Primary Anastomosis (IR + PA + SR + PA)
6. Ileal Resection + Primary Anastomosis + Sigmoid Resection + Hartmann's Procedure/Colostomy (IR + PA + SR + HR/Col)

Of the above-mentioned procedures, the most commonly performed one was Ileal Resection + Primary Anastomosis + Sigmoid Resection + Hartmann's Procedure/Colostomy.

Surgical Principles to Follow

If both loops are viable, the knot may be undone by sigmoid enterotomy and traction of the sigmoid loop may be performed. This procedure may also be selected when the sigmoid colon alone is viable. When the ileum and the sigmoid colon are gangrenous, it can be difficult to untie the knot, and rupture of the gangrenous loop could lead to spillage of toxic bowel contents. Therefore, intestinal clamps should be applied before dissection or resection of the knot followed by resection of both the loops.

Table 3 Operative procedures as performed by different authors and their outcome

Author	No.	Procedures performed						Outcome	
		IR+PA+SD	IR+PA+MS	IR+EI+SR+PA	ID+SR+PA	IR+PA+SR+PA	IR+PA+SR+HR/Col	Good	Death
Shepherd et al. (1967)	92							48	44 (47.8%)
Kakar et al. (1981)	11	2	2				7	8	3 (27.2%)
Puthu et al. (1991)	7					4	2	5	2 (28.7%)
Miller et al. (1992)	1					1		1	
Alver et al. (1993)	68	10			18	26	33	47	21 (30.8%)
Akgun et al. (1997)	16	4	3	3	3	3		13	3 (18.7%)
Kedir et al. (1998)	9							5	4 (44.4%)
Raveenthiran et al. (2001)	7				4	3		7	
Tamura et al. (2004)	1					1		1	
Hashimoto et al. (2004)	2					2		2	
Atamanalp et al. (2004)	63	10	2	1		6	38	53	10 (15.9%)
Hirano et al. (2005)	1					1		1	
Berrebi et al. (2006)	1						1	1	
Atamanalp et al. (2006)	9	1				1	7	8	1 (11.1%)
Aguka et al. (2006)	1						1	1	
Kotisso et al. (2006)	15						14	12	3 (20%)
Bawa et al. [15]	1					1		1	
Alwi et al. (2009)	1								
Machado et al. (2009)	1					1		1	.

IR+PA+SD Ileal Resection + Primary Anastomosis+Sigmoid Derotation, *IR+PA+MS* Ileal Resection+Primary Anastomosis+ Mesosigmoidostomy, *IR+EI+SR+PA* Ileal Resection+End Ileostomy+ Sigmoid Resection+ Primary Anastomosis, *ID+SR+PA* Ileal Diversion+ Sigmoid Resection+Primary Anastomosis, *IR+PA+SR+PA* Ileal Resection+Primary Anastomosis+ Sigmoid Resection+Primary Anastomosis, *IR+PA+SR+HR/Col* Ileal Resection+Primary Anastomosis+ Sigmoid Resection+Hartmann's Procedure/Colostomy

Primary anastomosis of the small bowel is preferable, but if the terminal ileum is gangrenous and within 10 cm of the ileocecal valve, an end-to-end anastomosis should be avoided because of the risk of leak. The distal stump can be closed and end-to-side ileocecostomy can be performed as safer option.

Resection of the sigmoid colon is often advised in all instances even when it is viable. Recurrent volvulus or repeat knotting due to redundancy of the loop may cause gangrene after surgery. In the past, a Hartmann operation or a covering colostomy was advocated to avoid the risk of fecal leak from colonic anastomosis. However, recent data suggest that primary colonic anastomosis may be undertaken safely when the history is short and the remaining bowel is clean, well vascularized, and undistended. Intra-operative colonic irrigation, followed by resection and primary anastomosis, is another alternative. Avoiding a colostomy is always welcome, as it reduces the morbidity and cost of health care. In patients without sigmoid gangrene, mesosigmoidostomy is practiced by some to prevent recurrent ISK [3, 7, 11, 14].

Outcome

The reported mortality from ISK varies from 0% to 48% (mean, 35.5%) (Table 3). The mortality figures are generally related to the duration of symptoms, the presence or absence of gangrene and the general status of the patient, including the presence of septicemic shock [2–7, 11]. However, some of the reports suggest that those patients who undergo surgery within 24 h after the onset of symptoms have a significantly higher incidence of mortality than those whose symptoms exceed 24 h [2, 6]. In spite of early judgment for prompt surgical intervention, the higher rates of both gangrene and mortality seem paradoxical and presumably reflect the fulminating clinical deterioration of patients due to early and extensive infarction of the bowel involved in a tight knot. However, other authors have found no correlation between the duration of symptoms and incidence of gangrene and consider that the incidence and onset of gangrene is to be related more to the geometric degree of rotation than the duration of symptoms [8, 16, 17]. The literature review reveals that there has been a

constant decline in the mortality with few reported cases of adverse outcome in the last 10 years (Table 3).

Summary

ISK is a rare yet life-threatening cause of acute intestinal obstruction. Unfamiliarity of the condition and diagnostic difficulties have contributed to the high morbidity and mortality in the past. Better understanding of the problem and increased probabilities of preoperative diagnosis with the help of contrast-enhanced computed tomography scanning have facilitated early diagnosis and intervention. Aggressive fluid resuscitation, preoperative antibiotics, prompt laparotomy, effective surgery including resection of gangrenous bowel with or without primary anastomosis, and better perioperative care of the shocked patient have optimized the survival of these patients.

References

1. Parker E (1845) Case of intestinal obstruction: sigmoid flexure strangulated by the ileum. *Edinb Med Surg J* 64:306–308
2. Shepherd JJ (1967) Ninety two cases of Ileosigmoid knotting in Uganda. *Br J Surg* 54:561–566
3. Kakar A, Bhatnagar BN (1981) Ileosigmoid knotting: a clinical study of 11 cases. *Aust NZJ Surg* 51:456–458
4. Puthu D, Rajan N, Shenoy GM, Pai US (1991) The ileosigmoid knot. *Dis Colon Rectum* 34:161–166
5. Miller BJ, Borrowdale RC (1992) Ileosigmoid knotting: a case report and review. *Aust NZJ Surg* 62:402–404
6. Alver O, Oren D, Tireli M, Kayabasi B, Akdemis D (1993) Ileosigmoid knotting in Turkey: review of 68 cases. *Dis Colon Rectum* 36:1139–1147
7. Akgun Y (1997) Management of Ileosigmoid knotting. *Br J Surg* 84:672–673
8. Raveenthiran V (2001) The Ileosigmoid knot: new observation and changing trends. *Dis Colon Rectum* 44:1196–1200
9. Tamura M, Shinagawa M, Funaki Y (2004) Ileosigmoid knot: computed tomography findings and the mechanism of its formation. *ANZ J Surg* 74:184–186
10. Hashimoto T, Yamaguchi J, Fujioka H, Okada H, Izawa K, Kanematsu T (2004) Two cases of Ileosigmoid knot: the youngest reported patient and CT findings. *Hepato-Gastroenterology* 51:771–773
11. Atamanalp SS, Oren D, Basoglu M, Yildirgan MI, Balik AA, Polat KY et al (2004) Ileosigmoid knotting: outcome in 63 patients. *Dis Colon Rectum* 47:906–910
12. Hirano Y, Hara T, Horichi Y, Nozawa H, Nakada K, Oyama AK et al (2005) Ileosigmoid knot: case report and CT findings. *Abdom Imaging* 30:674–676
13. Berrebi D, DeLagausie P, Azeinfish S, Chalard F, Peuchmaur M, Algrain Y (2006) Ileosigmoid knotting in a child: the first case report in a French girl. *Gastroenterol Clin Biol* 30:1414–1416
14. Atamanalp SS, Oren D, Yildirgan MI, Bosoglu M, Aydinli B, Ozturk G, Salman B (2008) Ileosigmoid knotting in children: a review of 9 cases. *World J Surg* 31:31–35
15. Bawa D, Ikenna EC, Ugwu BT (2008) Ileosigmoid knotting: a case for primary anastomosis. *Niger J Med* 17:115–117
16. Machado NO (2009) Ileosigmoid knot: a case report and literature review of 280 cases. *Ann Saudi Med* 29(5):402–406
17. Burrah R, Memon A, Pathan H, Ravikanth R, Kilpadi A (2010) Ileosigmoid knot—case report. *Indian J Surg* 72:140–142