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Case Study

Compound Volvulus: An unexpected surgical finding

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Key Learning Points

- CT findings suggestive of an ileosigmoid knot – mesenteric whirl sign
- The importance of multi- modality work up for a patient when there is a clinical diagnostic dilemma, which includes different imaging modalities and surgical exploration
- First encounter of an ileosigmoid knot that led to a literature search and education of myself of this rare condition
- I was a registrar at the time of managing this patient, this diagnostic dilemma assisted me in refining my clinical decision making and surgical skills as well as surgical decision making intra operatively.

Introduction

Ileosigmoid knot (ISK) also known as compound volvulus or double volvulus is a rare entity. The description of this condition is that the ileum wraps around the base of the sigmoid, forming a knot leading to bowel obstruction and ischaemia of both the ileum and sigmoid. The ileum or the sigmoid can be the active component and can wrap in either a clockwise or counter-clockwise direction[1,2,3]. However it needs to be differentiated from sigmoid volvulus, which is amenable to endoscopic reduction whereas compound volvulus typically progresses to necrotic ileum and sigmoid, which cannot be endoscopically reduced. The first case of ISK was described by Parker while it was first reported in Africa by Burkitt in 1952. Since then more than 300 cases have been reported[1,2].

Based on studies done in Uganda, where it is more commonly seen in males with an average age of 40 years. It was noted to be more common in non- whites, along the volvulus belt, namely, Africa, Asia and the Middle East[1,4]. Due to its common occurrence in Turkey (80 cases), they developed a new classification system which serves as a treatment algorithm and prognosticator[3]. To date there has only been one reported case of ISK in South Africa in a HIV positive patient that was complicated by mucormycosis which resulted in a mortality[5]. Below we describe a case in an academic hospital in South Africa in a HIV positive patient with a favourable outcome.

Case Study

A 27 year old male presented with an acute one day history of abdominal pain radiating to the back post an alcoholic binge. No history of diarrhoea, vomiting or constipation was noted. Of note he was newly diagnosed with RVD (retroviral disease) during this admission with a CD4 count of 29 cells/uL [332 – 1642] and a viral load of 20800 copies/ml. He was not on any ARVs (anti-retrovirals) drugs. He had no prior surgical history. Clinically his vitals were a blood pressure 143/42mmHg, pulse rate 52 bpm and apyrexial. On abdominal examination he had no abdominal distension, was soft, diffusely tender but not peritonitic. On rectal examination, normal coloured stools were noted. All other systems were normal. Biochemically on a room air arterial blood gas his metabolic status was assessed as a respiratory alkalosis, normal values in square brackets (pH 7.520 [7.350 – 7.450], CO2 26.3 [35 – 49], HCO3 21.3 , Be 0.1) with a mildly raised lactate (2.4 [05 – 1.2]). Formal haematological and biochemical analysis showed a White blood count 7.68 [3.92 – 10.4], Haemoglobin 14.6 [13.4 – 17.5], platelets 335 [171 – 388], Urea 3 [2.1 – 7.1], creatinine 59 [64 – 104], amylase 52 [30 – 120], lipase 16 [13 – 60] and CRP 4 [<10].

The patient was resuscitated according to primary surgical principles with intravenous fluids, transurethral catheterisation and nasogastric drainage (nil drainage noted).

Radiological images showed a chest Xray with no free air under the diaphragm. On an erect abdominal xray, few air fluid levels indicative of small bowel obstruction, but also a paucity of bowel gas which could imply fluid in the abdomen and no air in the rectum (erect film), see figure 1. No features of large bowel obstruction or the typical coffee bean of a sigmoid volvulus were noted.

A working diagnosis of alcoholic pancreatitis was made, however the clinical history and biochemistry were conflicting. Therefore a CT abdomen was done, where a mesenteric whirl sign was noted and the suspicion of necrotic bowel was likely, see figure 2.

With significant free fluid and the CT images suggestive of necrotic bowel, an exploratory laparotomy was planned.

Intraoperative findings was an ileo-sigmoid knot (a necrotic sigmoid volvulus with viable descending

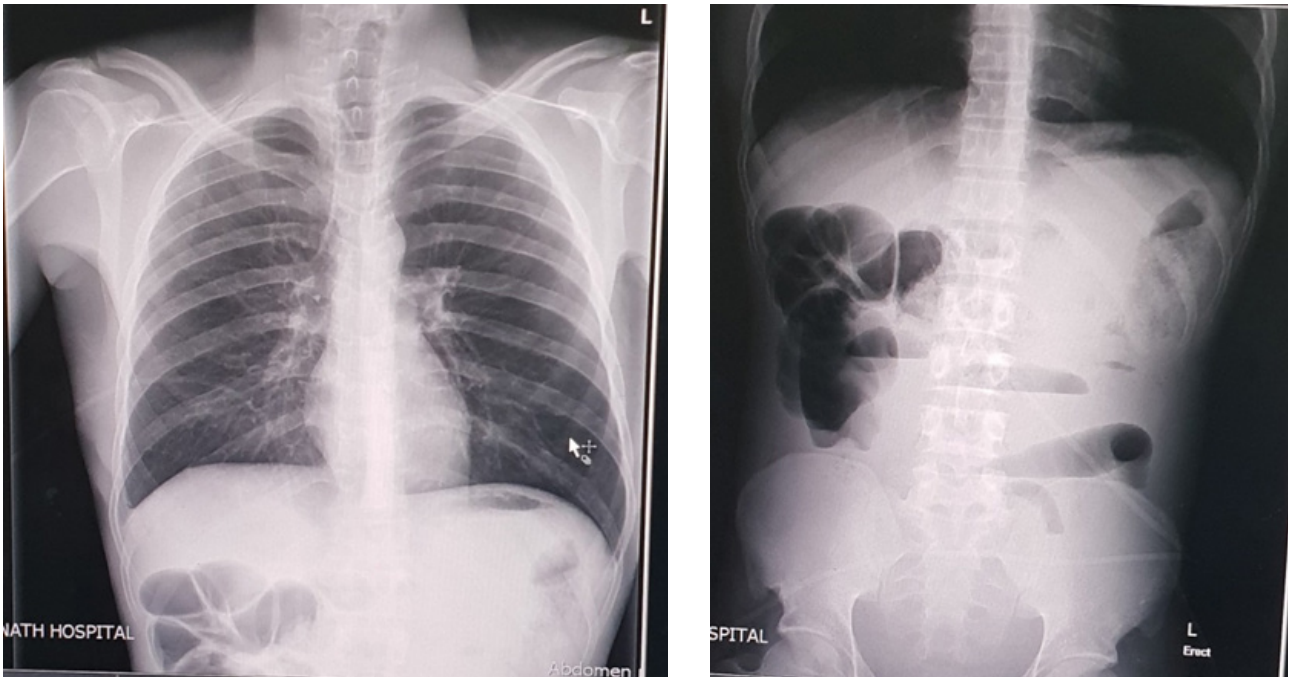

Figure 1: Erect chest (left) and abdominal Xray (right).

colon wrapped around necrotic loops of ileum) and serosanginous peritoneal fluid approximately 800ml. The sigmoid and ileum were necrotic. See figure 3. Resections of necrotic bowel was done with an ileocolic anastomosis (side to side stapled) and a Hartmann's procedure with an end colostomy due to the calibre change. Patient did well intraoperative not requiring inotropic support, the patient was extubated post operatively and sent to the ward. He had an uneventful stay with ERAS management principles. He was educated by the stoma sisters and discharged 6 days post operatively.

At a year follow – up the patient had initiated ARVs and is doing well. He is still awaiting reversal of his colostomy, due to the workload experienced at our hospital with regards to malignancies; patients with benign disease tend to have long waiting times for surgical procedures.

Discussion

ISK presents clinically with abdominal pain, tenderness and distension, nausea and vomiting. Progresses rapidly to gangrenous bowel and therefore can present with peritonitis or septic shock[1]. In reviewing some of the clinical presentation, it is as the literature states non-specific as in the patient in the case study above[2,4,6,7]. As with the variable clinical picture, the plain film radiological images can also be variable ranging from features of a sigmoid volvulus, large bowel obstruction or small bowel obstruction on abdominal xrays[7].

Typically a preoperative diagnosis is a rarity due to the diagnostic dilemma. It has been suggested that a triad of the conflicting diagnosis could raise suspicion of ISK. The triad includes, clinical picture of small bowel obstruction, large bowel obstruction radiologically and the inability to reduce a suspected sigmoid volvulus endoscopically[1]. The most important differential to consider is sigmoid volvulus as it impersonates it radiologically but a compound volvulus is not amenable to endoscopic reduction[2].

If the patient clinically allows for further diagnostics a CT can be done, features suggestive of a ISK is the "whirl sign", due to the twisted mesentery and the medially deviated caecum and descending colon as was the case in the patient described above[1,6,7].

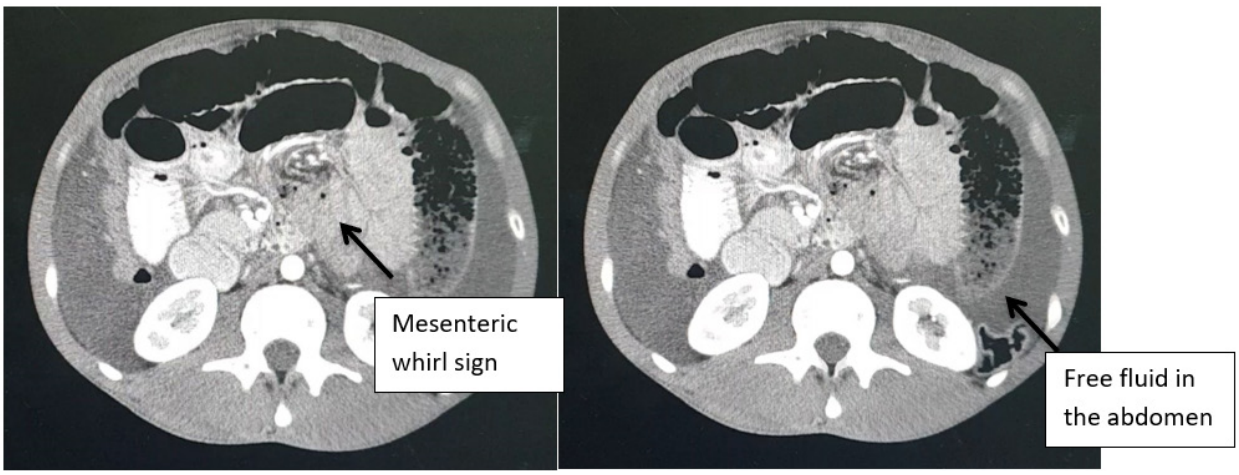

Figure 2: CT abdomen showing mesenteric whirl sign on axial images.

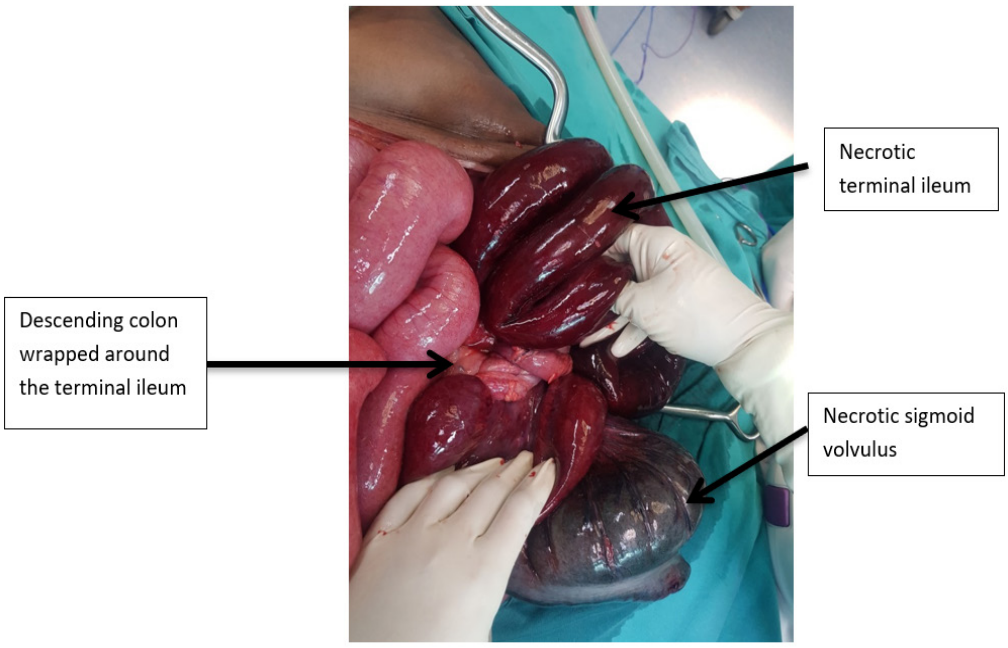

Figure 3: Intraoperative findings – necrotic sigmoid volvulus and terminal ileum with descending colon wrapped around terminal ileum.

The pathogenesis of ISK is linked to 3 factors namely, a long small bowel mesentery thus enabling mobility, long sigmoid on a narrow pedicle and the ingestion of a high bulk diet with an empty small bowel[2]. Other causative anatomic characteristics include late pregnancy, trans-mesenteric herniation, Meckel's diverticulum with a band, ileocaecal intussusception and a floating caecum[1,2]. The most likely causative factor in our case based on operative findings is a long sigmoid on a narrow pedicle, which thus allowed the descending colon to wrap around the terminal ileum causing obstruction.

Many classifications have been proposed, based on operative findings as well as being a prognosticator for mortality. Mandal et al (2012), classification is based on the active and passive components wrapping around each other. The torsion is 360 in 52.9% and decreases to 19.1% with a torsion of 360 x2 and to 5.9% with a torsion of 360 x 3[1]. Our case can be described as a type 2, sigmoid colon the active component wraps around the ileum the passive component in a clockwise or anticlockwise direction, however most of the sigmoid colon was involved in its own volvulus. Type 2 has an occurrence of between 18.9 – 20.6%[1].

Another classification based on intraoperative findings correlated with mortality. Mortality is 6.8 – 8% for non-gangrenous bowel and 20- 100% for gangrenous bowel[5]. Our case thus had a favourable outcome despite the gangrenous bowel. The Turkish initially proposed a classification in 2009 based on the viability of the bowel and risk factors such as age and co morbid disease. This was then modified in 2017 to include age, ASA, bowel condition and the presence of gangrene. In this classification a surgical treatment algorithm and mortality and morbidity are also stated[3].

Based on the Atamanalp classification our patient would fall in the ISK III group with an A0 (age < 70 yrs), ASAII (presence of mild systemic disease), BI (presence of bowel ischaemia), GII (presence of double segment gangrene). However our patient fits neither into the ISKIIIA group, due to BI nor the ISK IIIB group due to the A0. However this classification system is contradictory. It places B 0 (good bowel condition) in the same subgroup ISK IIIA with G II which is the presence of double segment gangrene. Therefore, this is the shortfall of this classification system[3].

The common trend with ISK is the paradox in clinical presentation and intra operative findings, this theme is carried through to the management of these patients. Patients that present within 24 hours of the onset of symptoms have a greater incidence of gangrenous bowel, 90.9% and a higher mortality than those that present after 24 hours with a 57% incidence of gangrenous bowel and a lower mortality. One would believe that the longer the strangulation from the knot the greater the chance of encountering gangrenous bowel. The literature attempts an explanation attributing it to the geometric degree of rotation and not the duration of symptoms[2].

Due to the propensity for the rapid progression to gangrenous bowel, patients commonly present in shock and require aggressive resuscitation and intravenous antibiotics prior to surgical intervention.

Management is dependent on the operative findings of which a vast majority showed gangrenous bowel. The most common surgical procedure is an ileal resection and primary anastomosis, sigmoidectomy and Hartmanns procedure and colostomy as was the case in the above patient. Rarely is a total colectomy done[1,4]. Of note when we operated on our patient, the HIV status was not known and a primary anastomosis and Hartmann's procedure was performed. Subsequently to the patient's surgery, the HIV status was found with a CD4 count of 29, despite being immunocompromised, there were no complications with regards to his anastomosis. At the same facility a postoperative CD4 count was found to be predictive of anastomotic leak rates regardless of HIV sero-status, however this study was done in patients sustaining penetrating abdominal trauma[8]. At our facility we operate on many HIV positive patients and therefore knowing the status preoperatively may not have changed intra-operative decision making, one has to consider the morbidity of a high output ileostomy and an anastomotic leak as well as the access to healthcare institutions. The patient was stable

intra-operatively, not requiring inotropic support and therefore the decision for a primary ileocolic anastomosis and a Hartmann's stoma was made.

Of note, majority of the case reports indicate an operative finding of haemorrhagic peritoneal fluid associated with gangrenous bowel, as in the case described, the significance of this finding has not being investigated[2,4,5,7].

The perplexity of ISK stems from clinical presentation to radiological investigations and operative procedures and the prognosis of the patient. Despite the necrotic small bowel and sigmoid colon, our patient was not in septic shock pre-operatively, did not require intraoperative inotropic support or organ support post operatively in the background of being severely immunocompromised, we still had a favourable outcome.

Conclusion

In conclusion, when there is a diagnostic dilemma between clinical and radiological findings, ISK should have a high index of suspicion as seen in the above case. The HIV seropositivity did not adversely affect the above patient with an ileocolic anastomosis and was not a factor in the diagnostic dilemma that is characteristic of ISK.

Conflicts of interest

None.

Funding

None.

Consent

The patient has consented to the publication of this case study.

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