

Case Report

A rare acute presentation of intestinal malrotation in the elderly

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ABSTRACT

Intestinal malrotation, a congenital anomaly of the midgut, typically presents in pediatrics age group with the most feared complication of midgut volvulus and its catastrophic sequelae. Midgut volvulus secondary to intestinal malrotation is a rare presentation in adults more so in the elderly. Its rarity portends a diagnostic dilemma for both the surgeon and radiologist. We report a 65-year-old man admitted following a referral with a 12-hour history of acute abdomen with an initial diagnosis of acute pancreatitis but later had laparotomy with intraoperative findings of a midgut volvulus and gangrenous ileal segment secondary to intestinal malrotation. This case emphasizes the importance of early diagnosis to prevent the disastrous complication of this disease. The rarity of this condition portends a formidable diagnostic challenge in adults hence the awareness of its possibility and a high index of suspicion is crucial in diagnosis.

Keywords: Intestinal malrotation, Midgut volvulus, Elderly

INTRODUCTION

Intestinal malrotation is a defect that occurs during gastrulation (between 5 to 10 weeks of fetal life) resulting in a failure of the bowel to rotate 270° counter clockwise around the axis of the superior mesenteric artery (SMA) as it elongates and herniates into the base of the umbilical cord. This failure may result in non-rotation, incomplete rotation, reversed rotation or fixation problems (anomalous fixation of the mesentery) of the midgut. Intestinal malrotation is estimated to occur in about 0.2% of adult population or less even though the cases documented in the elderly in literature presumes that some cases may go undiagnosed all through life.¹⁻³ Midgut volvulus secondary to malrotation usually occurs in the first week of life and during infancy with about 90% of cases presenting within the first year of life.^{2,4} For this reason, this condition is typically considered to occur in the pediatrics age group and hardly ever considered as a possible differential diagnosis in adults let alone the elderly. This may eventually lead to avoidable morbidity and possibly mortality in this group of patients due to the catastrophic events that follows delayed management of an

acute presentation. Most cases are diagnosed incidentally following radiological investigations for other pathologies while others are diagnosed intraoperatively.² Preoperative diagnosis of midgut volvulus secondary to malrotation in the elderly is laden with enormous challenges since management of this condition is usually not part of the armamentarium of the general surgeon but rather that of the pediatric surgeon.

We report the case of this elderly man with midgut volvulus secondary to intestinal malrotation to highlight the challenges of diagnosis and management of the potentially fatal sequelae that may evolve due to this diagnostic dilemma.

CASE REPORT

A 65-year-old clergyman presented at the Surgical Emergency Department (ED) following a referral with sudden onset constant periumbilical pain which subsequently became generalized. The pain had started 12 hours prior to presentation. He had associated multiple episodes of non-bilious vomiting of recently ingested meal

and anorexia but there was no abdominal distension, jaundice or change in bowel habits. He had no fever, no similar episode of symptoms in the past or previous abdominal surgery. He had no symptoms suggestive of abdominal ventral wall hernias. There was no prior weight loss. Systemic review was unremarkable. He neither smoked cigarette nor took alcoholic beverages. He had no co-morbidity and he was not on any routine medications.



Figure 1: Abdominal X-ray supine view.



Figure 2: Abdominal X-ray erect view.

Examination findings revealed a conscious elderly man who was in obvious distress from pain. He was not pale, afebrile, anicteric and was well hydrated on intravenous fluids. Other general examinations findings were unremarkable. Vital signs were within normal limits. He had no abdominal distension but had marked epigastric and periumbilical tenderness. There was no palpable organomegaly or intraabdominal masses. Ascites was not clinically demonstrable. Bowel sounds were hypoactive with normal digital rectal examination. Further systemic examination was unremarkable.

He had an initial diagnosis of acute pancreatitis and was being managed accordingly on nil per oris, intravenous fluids and parenteral antibiotics and analgesics. His admitting full blood count (FBC) was normal (with a Hb 15 g/dl, white cell count (WBC) of $8 \times 10^9/l$, and platelets $190 \times 10^9/l$). Electrolytes, urea, creatinine and liver function tests were within normal range. Serum amylase and lipase

results were still pending as at the time initial review. C-reactive protein (CRP) studies are not routine tests at our centre and so were not requested for. Plain abdominal x-ray showed the ileum and ascending colon with faecal loading, two air-fluid levels and scanty air in the rectum as shown in Figures 1 and 2. Abdominal ultrasound demonstrated features of moderately dilated bowel loops with moderate ascites. Diagnosis remained elusive, as such an abdominal computed tomography (CT) scan was ordered but this was not done due to financial constraints.



Figure 3: Midgut volvulus 720° counter-clockwise twist.



Figure 4: Gangrenous bowel segment.

Further review after 24 hours however showed progressively worsening abdominal pain, generalized tenderness and vital signs instability including tachycardia and fever. Peritonitis from complicated acute pancreatitis was entertained to keep-in-view ruptured viscus. He was then resuscitated and had an emergency exploratory laparotomy 28 hours after presentation at the emergency department. Intraoperative findings showed features of intestinal malrotation: the Duodenojejunal (DJ) flexure on the right of midline, the dorsal mesenteric attachment was narrow, and peritoneal folds crossed from colon and

caecum to duodenum, midgut volvulus with 720 degree twist in the clockwise direction, gangrenous segment extending from the proximal half of the ileum to the terminal ileum as shown in Figure 3 and 4. He had the gangrenous segment resected and an ileocolic anastomosis was done but no attempt was made at correcting the intestinal malrotation.

He subsequently developed sepsis, progressed to septic shock with multiorgan dysfunction syndrome (acute kidney injury and acute respiratory distress syndrome) necessitating hemodynamic monitoring, vasopressor support, mechanical ventilation and hemodialysis. He had a re-look laparotomy on suspicion of anastomotic leak with intraabdominal septic collection on 10th day postop however, findings were negative. His clinical state subsequently deteriorated to Glasgow coma scale (GCS) of 3T with coagulopathy. He improved with aggressive intensive care. He had 10 units of whole blood and 3 units of fresh frozen plasma transfused during admission. He was discharged home on 38th post-operative day. He was well at 9 months of follow up.

DISCUSSION

Midgut volvulus secondary to intestinal malrotation is a rare condition in the elderly with very few cases reported in literature.^{3,5-14} The first case in the elderly was probably reported in 1992 by Izes et al at the Lahey Clinic Medical Centre, USA.¹⁵ Its rarity therefore portends a challenge in diagnosis and this mostly dependent on a high index of suspicion. The case we presented here is unusual in two ways in that; firstly, the patient did not report any previous episode of recurrent abdominal pain and secondly, he did not have bilious vomiting which made the diagnosis even more puzzling.

From embryogenesis, midgut development starts with differentiation of the primitive intestinal tract into foregut, midgut and hindgut. Intestinal loop can be divided into two limbs; DJ and the caecocolic (CC), which rotate separately but in parallel with the SMA serving as the fulcrum and the omphalomesenteric duct at the apex. Midgut rotation commences following herniation of the midgut due to disproportionate growth and elongation of the midgut into the extraembryonic coelom at the fourth gestational week. The rotation involves the pre-arterial and post-arterial limbs rotating separately in a 270° anticlockwise direction around the SMA (with the first 90° rotation occurring outside the abdomen, the second 90° during the return of the bowel and the last 90° occurring in the abdomen). The DJ limb passes posterior to the SMA to lie on the left while the CC limb lies on the right, with subsequent fixation of the ascending and descending colon. Disruption of any of these steps lead to the spectrum of presentation encountered clinically.

Intestinal rotation was first clearly described and divided into stages by Frazer et al in their 1915 paper while Dott later translated this into clinical presentations based on

alteration of these embryological processes.^{16,17} The commonest rotational anomalies that occurs include non-rotation, incomplete rotation and reverse rotation. In non-rotation, the 270° rotation does not occur, as such, the DJ limb lies to the right while the CC limb lies on the left of the midline. A midgut volvulus then ensues as a result of narrow mesenteric base there formed and duodenal obstruction due to abnormally positioned caecal limbs. In incomplete rotation, normal rotation is arrested at about 180° thereby leading to the caecum lying in the right upper abdomen with presence of obstructing peritoneal bands (Ladd's bands). In reverse rotation anomaly, a 90° clockwise rotation occurs causing the transverse colon to lie to the right of the SMA passing through a tunnel posterior to the small bowel mesentery posterior to the duodenum which can predispose to volvulus with transverse colon obstruction.¹⁸

Midgut malrotation in elderly presents in several ways ranging from acute to chronic presentations with chronic being commoner. The pathophysiology of the chronic symptoms may relate to the effect of compression of Ladd's bands running from the caecum and/or ascending colon to the right abdominal wall. Symptoms are usually atypical and non-specific which makes diagnosis on clinical grounds more elusive due to it being rarely considered in adults. They include recurrent abdominal pain, vomiting, diarrhoea, early satiety, bloating and dyspepsia occurring over several months or years and are either managed by the physicians for irritable bowel syndrome or labelled as having psychiatric disease. However, the patients with acute presentation usually present with features of acute bowel obstruction with or without previous history of symptoms suggestive of chronicity of symptoms.

Kotobi et al noted that majority of patient in their series had chronic presentations with misdiagnosis for a long time and inadequate surgeries when they did, resulting in recurrence of symptoms.⁸ Izes et al and Haak et al also reported that patients had chronic presentations although the patient reported by Haak et al had features of acute intestinal obstruction during presentation prior to diagnosis.^{5,15} Fernandez-Moure et al however in a case report described a nonagenarian with acute presentation and no history suggestive of chronicity similar to the patient we are reporting.³ Similar presentation was also reported by Singh et al and Emanuwa et al.^{2,4}

Other forms of presentation reported in literature include left sided acute appendicitis (a dicey conundrum for the general surgeon in the elderly) and caecal volvulus as reported by Chuang et al and Hanna and Akoh respectively.^{10,19} In a review of literature of forty-nine patients found to have malrotation with colon cancer in Japan, Nakatani et al noted that about 69.4% of the patients were noted to have non-rotation anomalies and diagnosis was made by as incidental findings during radiological investigations.⁹ Patients were also noted to have midgut malrotation during resection of hepatopancreatobiliary

tumors in a case series following review of literature by Rittenhouse et al.¹¹

Diagnosis of malrotation of the gut in adults is usually by incidental findings during radiographs for other conditions or acute presentation even though this is still fraught with challenges by the radiologists in the elderly (since this condition is rarely considered in adults). Hanna and Akor reported in their case report that despite an abdominal CT that was done at presentation diagnosis was missed due to low index of suspicion in the elderly.¹⁹ Plain abdominal radiographs have little or no role in the diagnosis of these patients however abnormal gas-bowel distribution, right sided jejunal loops and absence of colon in the right lower quadrant may prompt further investigations. Unlike in the older children where the upper gastrointestinal series is considered the gold standard of diagnosis of malrotation, colour doppler ultrasound scan (USS) and abdominal computed tomography are preferred investigations in adults.^{1,2}

The “whirlpool sign” first described by Fisher on CT scan and by Pracros et al on USS demonstrated by clockwise encirclement of the SMA by coils of small bowel with mesentery and the superior mesenteric vein (SMV) is seen on doppler USS even though it may be associated with technical difficulties in adults.²⁰⁻²² Abdominal CT scan can demonstrate SMV in anomalous location posterior and left of the SMA even though patients with malrotation may have a normal SMA-SMV orientation, and an abnormality of SMA-SMV relationship may be due to a normal variant and not necessarily intestinal malrotation.^{7,23} Also, midgut and duodenum may be noted not to cross the spine. In the presence of associated volvulus, besides the above findings, the “whirl-pool sign” may also be demonstrated.

Other abnormalities such as small intestinal dilatation, circumferential bowel thickening, beak-like appearance, halo appearance or closed loops may be demonstrated. Other investigations include magnetic resonance imaging (which will show similar findings as CT scan however not a readily available investigation) and mesenteric angiography. The mesenteric angiography will demonstrate the abnormal relationship between the SMA and SMV and detect the patency of the vessels. It also demonstrates the characteristic corkscrew appearance of a whirling SMA and its branches- the “barber pole sign” first described by Buranasiri et al with extensive collaterals caused by proximal SMA occlusion.²⁴ This is no longer used for diagnosis due to its invasive nature and cost. CT scan remains the preferred investigation for diagnosis in adults.

Management was first described by William Ladd for paediatric patients in his 1932 landmark surgical paper and this hasn't changed much since then except for the laparoscopic approach that has been described by van Der zee and currently carried out routinely by surgeons skilled in this approach.^{25,26} The Ladd's procedure involve entry into abdominal cavity and evisceration of the bowel,

counter-clockwise detorsion of the bowel (in the presence of midgut volvulus), division of Ladd's caecal bands, broadening of the small intestine mesentery, incidental appendectomy and placement of small bowel along the right lateral gutter and colon along the left lateral gutter. He clearly noted that “timely and suitable surgical intervention offers the only chance for cure and restoration of the health of these patients”. Management of symptomatic adults with midgut malrotation clearly requires surgical intervention however the management of asymptomatic patients is more controversial.

Currently, there is no clinical means or investigation to determine if an asymptomatic elderly patient with malrotation will have an acute presentation such as midgut volvulus throughout life as such subjecting such patients to surgery following radiologic diagnosis has been an issue of debate.²⁷ Most surgeons however advocate surgical repair for these patients even though the risk of such should be weighed against potential benefit since the risk remains present throughout life.^{14,28-30}

Another issue of debate is whether an incomplete procedure (laparotomy, counter-clockwise rotation± resection and anastomosis) rather than carrying out the complete Ladd's procedure as fully described by Ladd in the very ill elderly patient with consideration of completion of the procedure later (as an elective procedure) or not at all.^{2,3,19} Also, one needs to keep in mind that the general surgeon may not necessarily be as familiar with the procedure when compared to the paediatric surgeon so a paediatric surgeon's assistance may be required. The decision to carry out an incomplete Ladd procedure in our patient seems appropriate when viewed retrospectively considering the morbidity encountered in the patient in the postoperative period. Whether a laparoscopic or open approach should be carried out will depend on the skill and experience of the surgeon and the mode of presentation with the open approach being advocated in associated volvulus except there is minimal chance of conversion to open by the surgeon.^{28,30} The short-term benefit of laparoscopic surgery is ever present however it can be technically challenging.

CONCLUSION

Intestinal malrotation with midgut volvulus; a rare condition in the elderly remains a difficult diagnostic challenge in the clinical setting since it is hardly ever considered a differential diagnosis of acute abdomen in this age group. Awareness of its possibility will aid in its possible early diagnosis which is key in prevention of the possible catastrophic sequelae associated with this condition. Even though our patient survived and has remained well all through follow up, he could have succumbed to the complications also, the morbidity encountered could have been prevented if he had an early diagnosis. Operative management of the condition may

require the assistance of a paediatric surgeon which the general surgeon should not hesitate to call upon if required.

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