

## Case Report

# A rare presentation of midgut malrotation as an acute intestinal obstruction and perforation in an adult

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### ABSTRACT

Midgut malrotation is a congenital anomaly seen usually in childhood. Its presentation as an acute intestinal obstruction is extremely rare in adults usually identified intra operatively. A high index of suspicion is always required when dealing with any case of acute intestinal obstruction. We report a case of young adult who presented with symptoms of acute intestinal obstruction and was diagnosed intra-operatively as cecal volvulus with impending perforation caused by midgut malrotation. Malrotation of the intestinal tract is seen due to aberrant embryology. The presentation of intestinal malrotation in adults is very rare. Contrast enhanced Computed tomography (CT) can show the abnormal anatomy clearly. Anomalies like midgut malrotation can present as an operative dilemma and awareness regarding these conditions can help surgeons deal with these conditions.

**Keywords:** Midgut malrotation, Acute intestinal obstruction, Cecal volvulus, Perforation

### INTRODUCTION

Midgut malrotation is a congenital anomaly referring to either lack of or incomplete rotation of the foetal intestines around the axis of the superior mesenteric artery during foetal development. Most patients usually present with bilious vomiting in the first month of life because of duodenal obstruction or a volvulus. It is rare for this condition to present in adulthood. It has been estimated that it affects approximately 1 in 500 live births.<sup>1</sup>

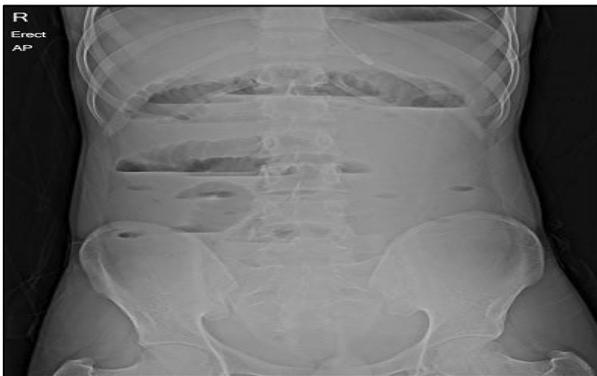
More than 90% of patients will present by the time of their first birthday. It has been reported that the incidence of malrotation in adults is approximately between 0.00001% and 0.19%.<sup>2,3</sup> In adults, the diagnosis in the pre-operative period is usually not thought of owing to a very low index of suspicion.

### CASE REPORT

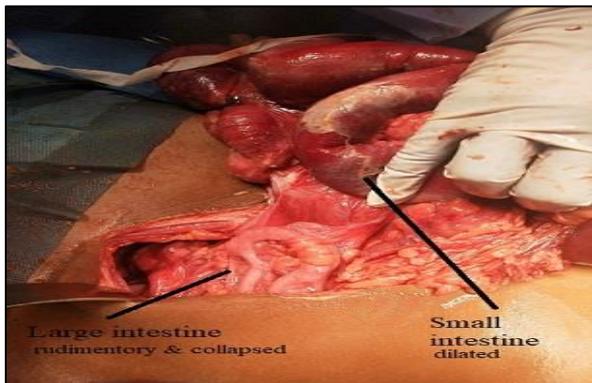
A 22-year-old male patient was admitted in the emergency ward with complaints of generalized pain abdomen since 4 days acute onset gradually progressive, colicky in nature. The patient also vomited thrice on the day of presentation with vomitus non-bile stained. He was also complaining of severe anorexia and inability to pass stools and flatus for 2 days. There was history of similar complaints since childhood days of pain abdomen and vomiting which relieved on itself without any medication and he denied any history of tuberculosis, peptic ulcer disease or any other chronic medical illness. He had no history of any abdominal surgery.

On physical examination, the patient was a well-nourished young adult male. The patient was having

tachycardia with 110 pulse rate and 110/60 mmHg of blood pressure (BP). The abdomen was distended with generalised tenderness and bowel sounds were absent on auscultation. Per rectal examination was empty with no growth. The patient's complete blood count demonstrated a haemoglobin level of 14.2 g/dl and white blood cell (WBC) of 3900 cells/cumm. All other biochemical parameters including blood sugar, serum electrolytes, liver function tests, kidney function tests, clotting profile were within normal limits, abdominal X-ray revealed multiple air fluid levels (figure 1) and ultrasound abdomen showing dilated bowel loops with distended caecum, diagnosis of acute intestinal obstruction was made and the patient was admitted and planned for urgent exploratory laparotomy.



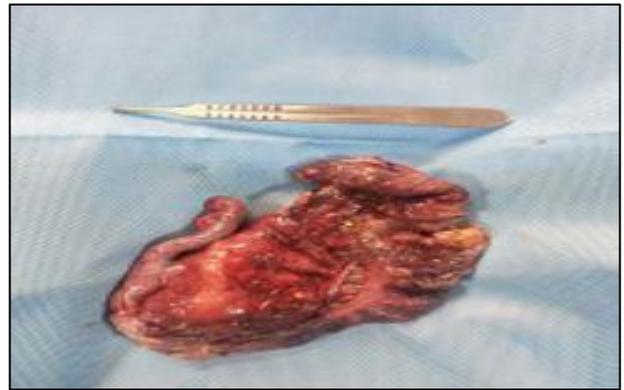
**Figure 1: X-ray erect abdomen.**



**Figure 2: Intra-operative picture.**



**Figure 3: Torsion site.**



**Figure 4: Resected specimen.**

The patient was resuscitated with intravenous fluids, analgesia and prepared for an emergency exploratory laparotomy. The abdomen was opened from a midline incision and a largely dilated loops of small intestine and caecum with appendix visualized. The whole small bowel loops were seen to be mainly concentrated on the right upper abdomen with duodenojejunal flexure present abnormally on the right side of midline and rudimentary large intestine in left side of midline (figure 2). On careful inspection a large cecal volvulus with torsion site was noted and the caecum was gangrenous and perforated (figure 3). Caecum was detorted bands were excised, mobilised and resected (figure 4). Ilio-transverse anastomosis with diversion ileostomy was done. Post-operative period was uneventful, and ileostomy was functional on post op day 1 and the patient was discharged on the eight post-operative day. The patient is on a regular follow up.

## DISCUSSION

The caecum forms the proximal portion of the colon. It is 6 cm in length and 7.5-8 cm wide. The caecum is covered by peritoneum, mostly there is no distinct mesentery and the mobility is limited. Rarely the caecum can be particularly mobile, which predisposes to cecal volvulus and may contribute to unusual clinical presentations of acute appendicitis.<sup>4,5</sup>

The cecal volvulus usually presents as terminal small intestine obstruction. Plain x-ray of the abdomen showed distal small bowel obstruction with a dilated caecum in the epigastrium or the left upper quadrant. Computed tomography (CT) scan helps identifying the site of obstruction. Surgery is aimed at decompressing the obstruction and fixing the caecum in the right lower quadrant. If strangulation is seen, right hemicolectomy is done.

Fukuya et al showed seven adult patients with midgut volvulus as a result of intestinal malrotation. Four patients presented with long histories of intermittent abdominal pain; three patients presented with acute onset of severe abdominal pain. Abdominal angiography in one of these

patients showed abnormal courses of mesenteric vessels to the volvulized segment of small bowel. These three patients with the acute onset underwent laparotomy, which showed ischemic segments of bowel.<sup>6</sup>

Malrotation of the intestinal tract is due to problems in embryology. Outcome of malrotation associated with midgut volvulus usually is catastrophic, an understanding of the anatomy, diagnostic criteria and appropriate therapy for this emergency condition is essential.<sup>7</sup>

Midgut malrotation is broadly considered a deviation from the normal 270 degree counter clockwise rotation of the gut during embryonic development. During week 4 of foetal development, the embryonic gut, consisting of a straight endodermal tube, develops vascular pedicles to be divided into the foregut, midgut and hindgut based on the anatomical blood supply.

The midgut is supplied by the Superior mesenteric artery (SMA) and by the fifth week of embryonic life, it begins a process of rapid elongation and outgrows the capacity of the abdominal cavity. This leads to a temporary physiological herniation into the umbilical cord at about the sixth week of life with return to the abdominal cavity about 4 to 6 weeks later. During this period, the midgut undergoes a 270-degree counter-clockwise rotation around the SMA axis. This process leads to the formation of the duodenal C-loop, placing it behind the SMA in a retroperitoneal position and emerging at the ligament of Treitz.

The progressive reduction of the physiological midgut herniation commences at about week 10 of embryonic development. The duodeno-jejunal flexure (DJF) and jejunum to reduce first and lie to the left. The distal small bowel then follows and lies progressively to the right of the abdominal cavity. The descent of the caecum from its higher position in the right upper quadrant forms the latter part of this complex rotational development; it becomes positioned in the right lower abdomen. The ascending colon then assumes a retroperitoneal position, also on the right side. The base of the small bowel mesentery subsequently fuses with the posterior peritoneum in a diagonal fashion, from the ligament of Treitz at the DJF to the caecum, completing the whole process at about the eleventh week of foetal development.<sup>8-10</sup>

The failure of the normal physiological rotation of the midgut leads to various degrees of anomaly including the entire small bowel remaining on the right side of the abdomen, the caecum, appendix and colon on the left and an absent ligament of Treitz.

In addition, the small bowel mesentery may develop a narrow vertical attachment and the peritoneal fibrous bands fixing the duodenum and caecum to the abdominal wall may persist. These congenital bands extend from the right lateral abdominal wall, across the duodenum and attach to the undescended caecum and are known as Ladd's

bands.<sup>10,11</sup> Ladd's bands compress the duodenum and can potentially cause duodenal obstruction. The malrotation of the gut and abnormal location of the caecum produces a narrow superior mesenteric vascular (SMV) pedicle, as opposed to the normally broad-based small bowel mesentery. This narrow SMA take off and lack of posterior peritoneal fusion predispose to subsequent midgut volvulus and obstruction with potential vascular catastrophe.<sup>11,12</sup>

Midgut malrotation can be identified using plain abdominal radiograph, ultrasound scan (USS), CT scan, magnetic resonance imaging (MRI) scan and mesenteric arteriography. Conventional plain radiography is neither sensitive nor specific in the diagnosis of gut malrotation although right-sided jejunal markings and the absence of a stool-filled colon in the right lower quadrant may be suggestive, leading to further investigation. CT scan with or without upper Gastrointestinal tract (GIT) contrast study is now considered the investigation of choice; providing diagnostic accuracy of up to 80%.<sup>13,14,15</sup>

CT and MRI scans may show the SMV to be in an anomalous position; posterior and to the left of the SMA. In addition, they may show the abnormal anatomical arrangements of the midgut with the duodenum not crossing the spine. Deviation from the normal positional relationship of SMV and SMA was originally described by Nichols and Li as a useful indicator of the diagnosis of midgut malrotation.<sup>16</sup>

However, abnormal orientation of the SMA-SMV relationship is not entirely diagnostic of malrotation; it can also be seen in some patients without the pathology and a proportion of patients with malrotation may have a normal SMA-SMV relationship.

The CT appearance of midgut volvulus is diagnostic of malrotation. The shortened mesentery allows the small bowel and mesentery to twist and wrap around the narrowed SMA pedicle to create a distinctive 'whirlpool' appearance on CT scan. This pattern was first described by Fisher in a patient with midgut volvulus.<sup>17</sup>

## CONCLUSION

Intestinal malrotation is a rare condition but is considered an important cause of bowel obstruction in adults. The diagnosis of malrotation after childhood is difficult and usually not readily considered as the cause of intra-abdominal symptoms. The presentation is usually non-specific, and this often leads to diagnostic and treatment delay with possible bowel ischemia and necrosis. Identification of these shows a poor prognosis and death. Therefore, a high index of suspicion needs to be maintained and prompt surgical intervention must be considered in order to prevent a fatality.

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