

Adult midgut malrotation with left renal and seminal vesicle agenesis – A case report

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Abbreviation:

1. OPD – Outpatient department
2. CT – Computed tomography
3. NCCT – Non-Contrast Computed Tomography
4. NPO – Nil Per Oral
5. SMV – Superior Mesenteric vein
6. SMA – Superior Mesenteric artery
7. USG - Ultrasonogram

Abstract:

Malrotation is a disease occurring more often in children. It can also present in adults with a nonspecific array of symptoms. Current statistics of malrotation are about 1 in 500 live births, symptomatic presentation of malrotation occurs in 1 in 6000 live births. This case report is about a midgut malrotation in an adult male.

Malrotation in adults is often subacute with nonspecific presentation and hence it is often missed. A CT scan makes a diagnosis of the abdomen. Symptomatic patients can undergo Ladd's procedure by laparoscopic or open technique. Asymptomatic patients beyond the age of 20, if operated on may be ineffective or possibly harmful also. Intestinal malrotation in an adult should be made as a differential diagnosis when a patient presents with abdominal pain, especially in small bowel obstruction in a virgin abdomen. It is important to consider the patient's age before prophylactic surgery on malrotation which was identified incidentally.

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I. Introduction:

Intestinal malrotation is the failure of the midgut to rotate 270° counterclockwise during development. Consequently, the duodenum and caecum are joined together through Ladd's bands, precipitating midgut volvulus, intestinal obstruction and ischemia. Malrotation occurs in around 1 in 500 live births^[1], while symptomatic malrotation occurs in 1 in 6000 births instead^[2]. Although malrotation is a disease that is more prevalent in the paediatric population and presentation in adults is rare, few cases have popped up in recent years. In a study of 170 patients with malrotation, 31% were infants, 21% were children, and 48% were adults^[3]. Our case report is about a 31-year-old male who had congenital malrotation that went unnoticed, despite recurrent abdominal pain, until his emergent presentation to our academic institution.

Case Presentation:

A 31-year-old male, non-smoke and non-alcoholic came to the general surgery Outpatient Department (OPD) of Sree Balaji Medical College & Hospital with chief complaints of lower abdomen pain for four months which was a pricking type of pain, non-radiating. He had a history of nausea and vomiting for the past four months. His bowel habits were normal—no history of fever, or chills. The patient's past medical history revealed that he presented with 1 episode of acute abdomen 4 months back for which he had gone to a private clinician and has

been managed conservatively. He also had a history of vague abdominal pain that was episodic and occurred every few months over the past few years and was responsive to analgesics.

On examination, the patient was conscious and oriented to time, place and person. His vitals were stable. Per abdomen was soft, mild tenderness was elicited in the epigastric and right lumbar region. Per rectal examination revealed a normal stool-filled cavity, stool was semisolid in consistency and anal sphincter tone was normal. Other system examinations were found to be normal. The patient has been advised to do a Non-contrast CT (NCCT) of the Abdomen which gave an impression of “Left renal and left seminal vesicle agenesis, evidence of malrotation of midgut with twisting of mesentery/ bowel loops around the vessels (Whirlpool sign) which is shown in Fig:1.1 and malpositioning of the caecum and ascending colon noted in Fig 1.2 and 1.3, Suggestive of Midgut volvulus”.

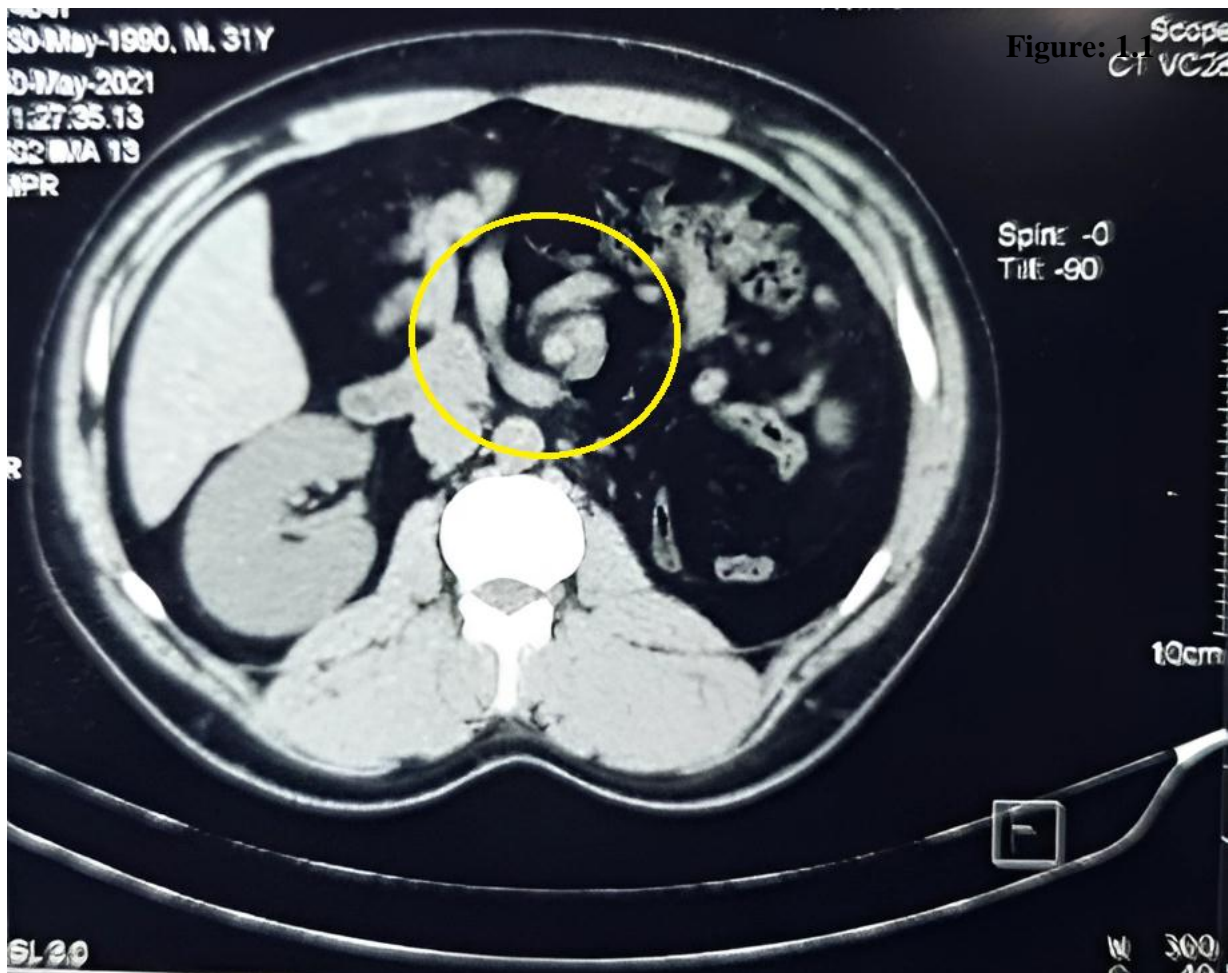
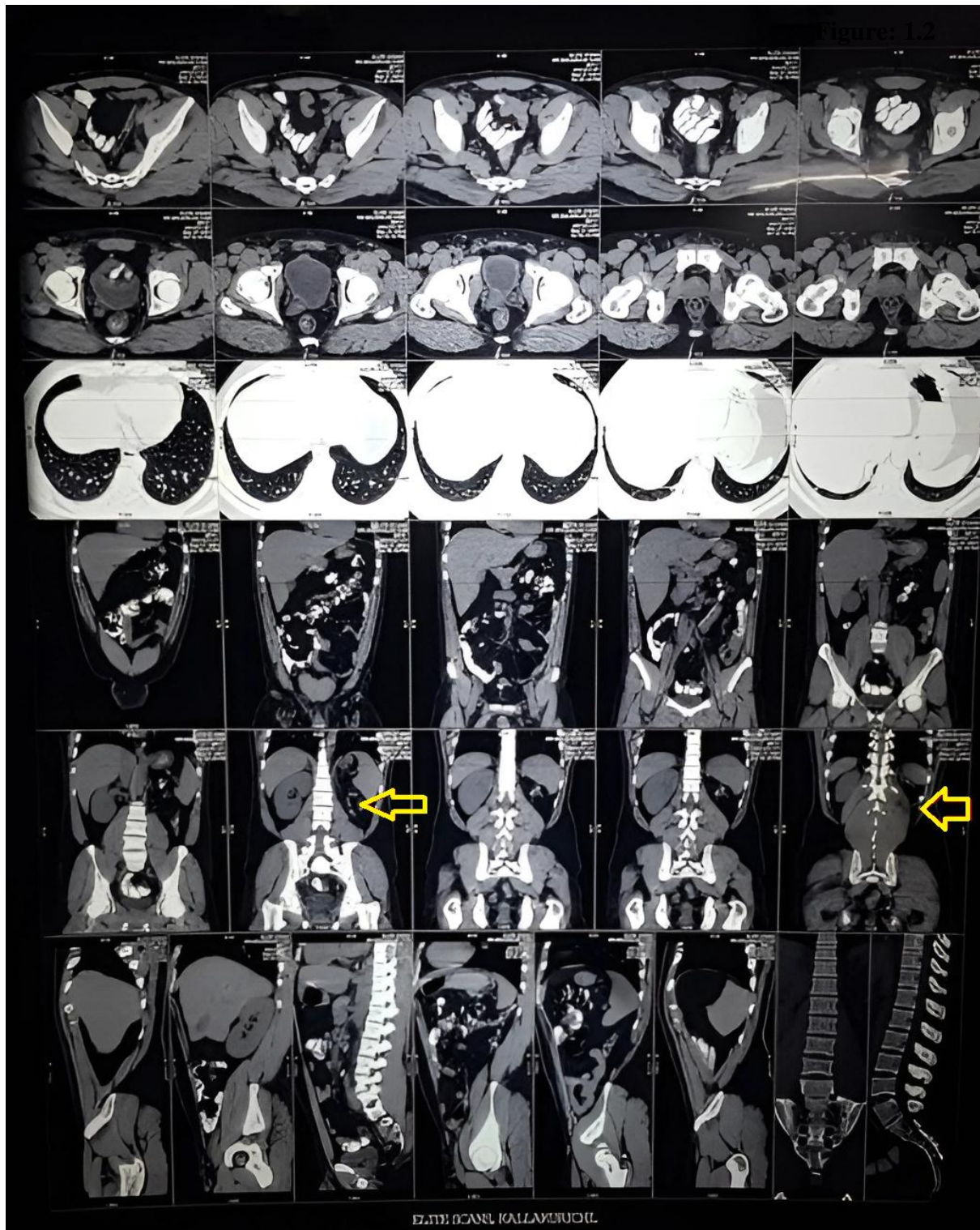


Fig 1.1 shows malposition of superior mesenteric vein and superior mesenteric artery – Whirlpool sign.



(Fig 1.2 CT image showing absence of left kidney – Renal agenesis)

The patient was advised to do, x-ray erect abdomen and all other routine investigations. Thereafter, he was advised with a plan of Ladd's procedure. After getting proper consent from the patient, he was admitted and planned for Ladd's procedure electively.

Under General anaesthesia, a midline laparotomy incision was made and the abdomen opened in layers. Ladd's bands were identified and released as shown in Fig:1.4 and 1.5. A bowel walk was done and appears normal with caecum in the midline and the entire small bowel on the right side. Repositioning of intestinal loops was done along with caecopexy shown in Fig:1.6 with appendicectomy.

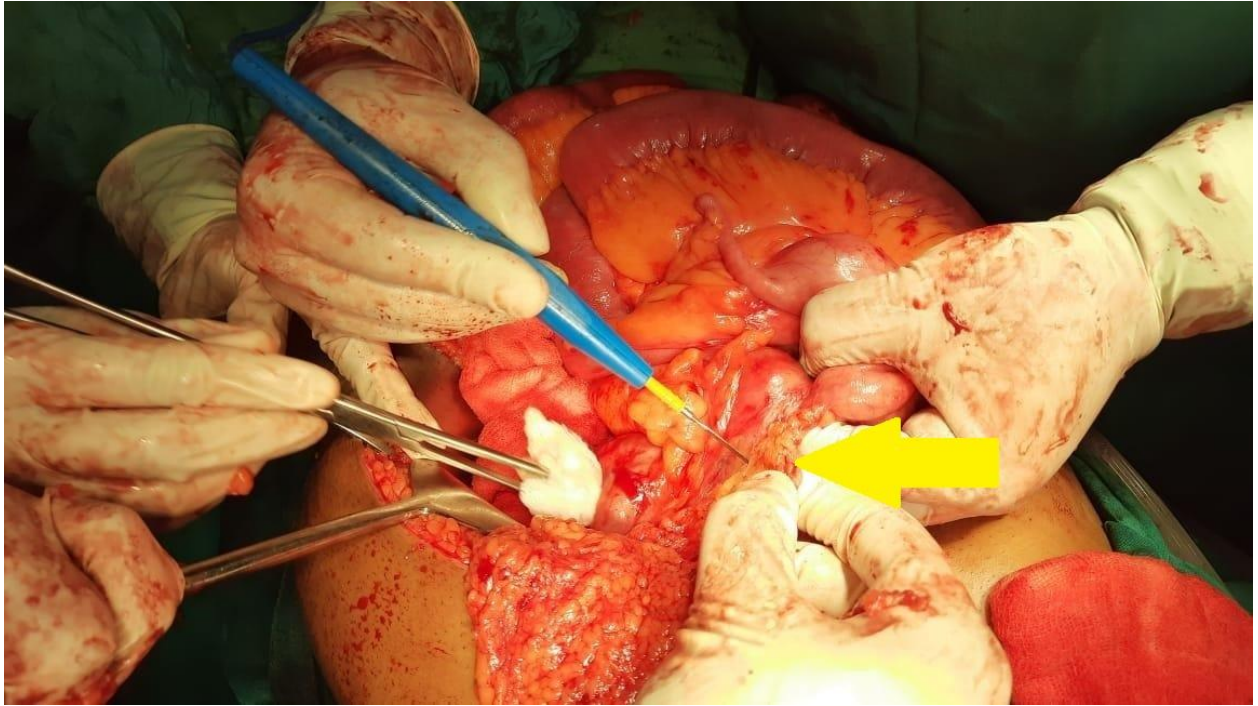


Fig 1.3 shows presence of Ladd's band and Ladd's band release.

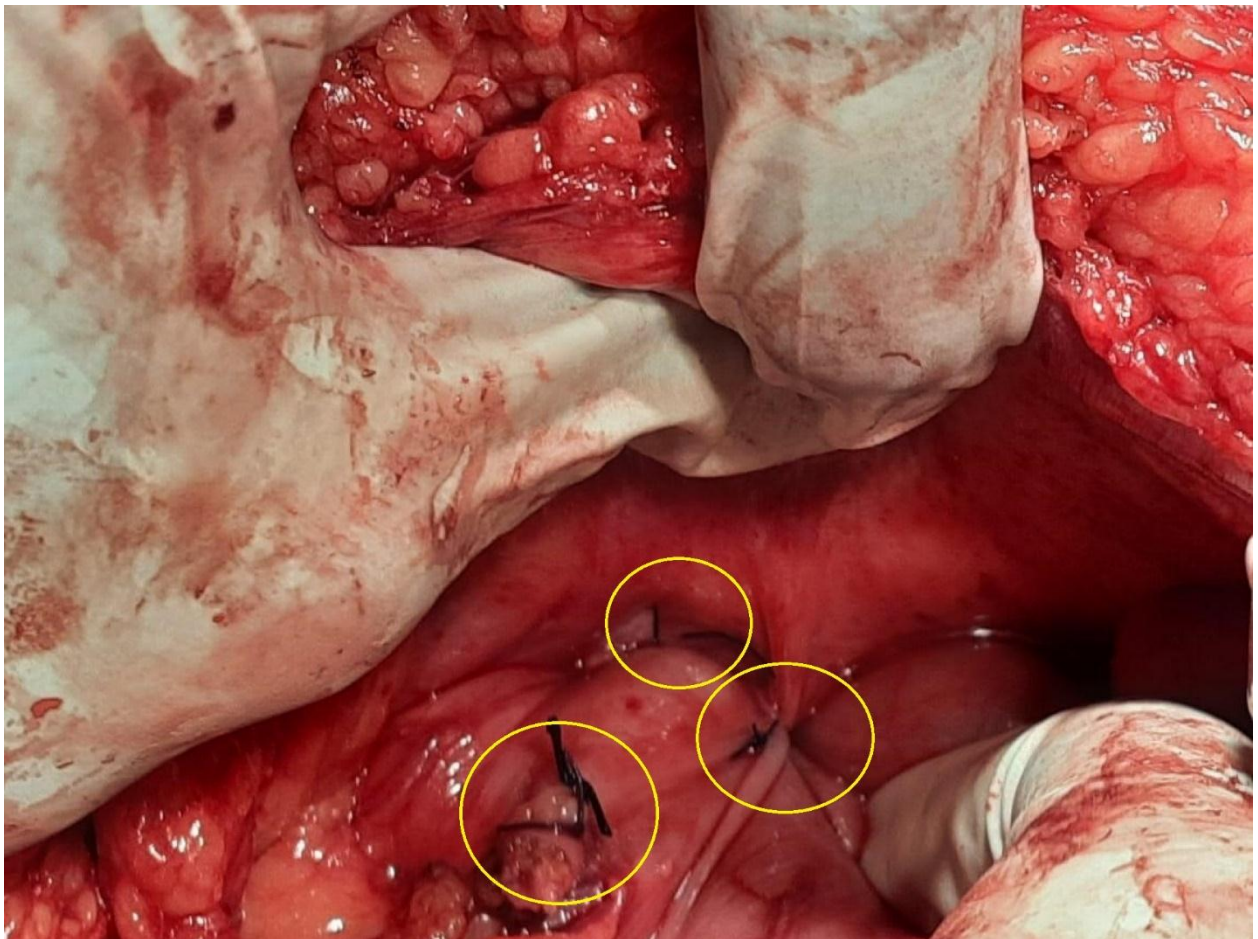


Fig 1.4 shows caecopexy after ladd's band release

Patient withstood the procedure well. Oral feeds were started after 48 hours of NPO and tolerated well. The postoperative period was uneventful. The patient was relieved of all preoperative symptoms after surgery.

II. Discussion:

Malrotation in adults is rare, it is most commonly seen in neonatal period [4]. Even though clinical symptoms are obscure, adult patients visit the hospital mostly with complaints such as vomiting and recurrent abdominal pain, probably due to chronic partial obstruction [5,6]. Some may present with malabsorption due to the inability to eat and protein loss associated with diarrhoea caused by chronic volvulus [7]. Imaging studies such as plain X-ray abdomen, Ultrasonography, and Computed tomography help in identifying the condition. Of all the investigations, contrast-enhanced radiograph is the most accurate method [8].

Typical radiological signs are corkscrew sign or whirlpool sign, which are caused by the dilatation of various duodenal segments at different levels and the relocation of the duodenojejunal junction due to jejunum folding [9].

In ultrasonography, the superior mesenteric vein (SMV) lies to the left or anterior to the superior mesenteric artery (SMA). Doppler USG may show the whirlpool sign with rotation of SMV around SMA which is typical for malrotation [10]. Besides, jejunal arteries lie to the right instead of to the left in computerized tomography scan as another diagnostic sign of malrotation [11].

Since malrotation commonly causes intestinal obstruction all patients deserve elective laparotomy [12]. Ladd's procedure has been the standard procedure of elective treatment of intestinal malrotation since 1936 [13]. Ladd's procedure consists of untwisting the midgut volvulus first followed by dividing the bands causing obstruction and then the segments of colon and small bowel are set to a neutral position. Appendectomy is added to the procedure to prevent future difficulty in diagnosis of appendicitis. If there are signs of large and small bowel ischemia resection and anastomosis of the ischemic segments will be needed.

In some cases, extensive resection of the small bowel will be needed, however short bowel syndrome and subsequent complications are unavoidable in these patients.

Laparoscopic Ladd's procedure for elective cases is effective, even superior to conventional procedure in some aspects [14]. However, the surgical option should always be patient-based. Presentation of adult midgut malrotation cases can be rare, even though a whirlpool sign in computed tomography of the abdomen may give suspicion of bowel twisting. An acute presentation is always presented with extensive bowel ischemia and necrosis and extension resection is needed.

In our case there was no signs of bowel ischemia and necrosis and hence untwisting the midgut was done and Ladd's bands were released along with appendectomy. In a case of adult midgut malrotation with volvulus presented as acute abdomen, timely recognition and management is the key to save life, if possible, but extensive small bowel resection may be unavoidable at times and may associate with fatal life-threatening consequences.

III. Conclusion:

This is a rare case of adult midgut malrotation with volvulus presented with abdominal pain. CT showing classical sign of midgut malrotation – Whirlpool sign with left renal agenesis and Seminal vesicle agenesis. It was effectively managed by Open Ladd's procedure with an uneventful post-operative period.

This case emphasizes consideration of malrotation as a differential diagnosis in adults presenting with abdominal pain. Patients might be asymptomatic or may have obscure symptoms, thus an elaborate physical examination and precise imaging studies will provide us accurate diagnosis and thus appropriate line of management to limit mortality and morbidity.

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