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A CASE REPORT MALROTATION OF GUT IN ADOLESCENTS

General Surgery

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ABSTRACT

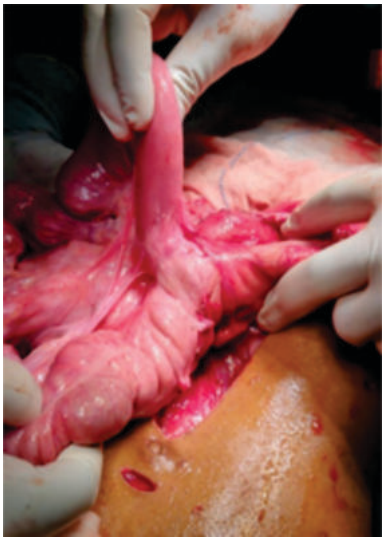
Introduction: Malrotation of the midgut is generally regarded as paediatric pathology with the majority of patients presenting in childhood. The diagnosis is rare in adults, which sometimes leads to delay in diagnosis and treatment. A high index of suspicion is therefore required when dealing with patients of any age group with abdominal symptoms.We present a case of a 14-year old who presented with an acute abdomen with preoperative computed tomography scan and operative findings confirming midgut rotation , The duodenum, small bowel, caecum and appendix were abnormally located, with the presence of classical Ladd's bands. There was no evidence of intestinal volvulus. The patient underwent exploratory laparotomy with an uneventful postoperative recovery. A review of the literature is presented to highlight the rarity of intestinal malrotation and the controversies surrounding its management in the adult population.

KEYWORDS

Midgut malrotation, acute abdomen, Ladd's bands, computed tomography scan

In congenital intestinal malrotation an impaired embryological development of the gut causes incomplete rotation and fixation of the intestines to the abdominal wall[1]. The fulfillment of the third embryonic rotationincludes the traversing of the duodenum to the left side of the abdomen, forming the ligaments of Treitz, and the migration of the ileo-caecal junction to the lower right abdominal quadrant. The fixation of the full-length bowel is complete during the twelfth week[2]. Congenital malformations such as diaphragmatic hernia, omphalocele or gastroschisis are associated with a similar but secondary incomplete rotation and fixation of the intestines [3] The inadequate fixation of the bowel alongside remaining embryonic fibrous adhesions, the Ladd's bands [4], may give rise to a variety of intestinal malfunction.

In the worst case scenario, malrotation may develop into a midgut volvulus with torsion causing high risk of ischemia and necrosis of the parts of the intestine supplied by the superior esenteric artery. This life-threatening condition is well known among pediatric surgeons and is always considered when physicians treat critically ill infants with abdominal symptoms and unknown diagnoses.



Malrotation has primarily been diagnosed in early childhood, with estimated onset of symptoms during the first year of life in 90 % of the cases [4,5,6]

There are recent reports of manifestation later in life, both as emergency conditions or more chronic gastrointestinal symptoms [4,5,6].

The exact incidence of intestinal malrotation is thus still difficult to determine. It was earlier described to be approximately 0.2 % (Stewart et al. 1976; Donnellan and Kimura 1996; Clark and Oldham 2002)[7,8,9], but an incidence up to 1 % has been reported (Adams and Stanton 2014). Improved radiological facilities, including multi-detector CT-scans, provide new possibilities to identify anatomical aberrations

Midgut malrotation is a congenital anomaly in the embryological development of the foetal intestinal rotation. It has been estimated that it affects approximately 1 in 500 live births [3]. that more than 90% of patients will present by the time of their first birthday [10].

Adult midgut malrotation is very rare and its incidence has been reported to be between 0001% and 0.19% [10,11]Most adult diagnoses of midgut malrotation are made in asymptomatic patients; either on imaging investigations for unrelated conditions or at operations for other pathology.are a small proportion of affected adults who may present with acute or chronic

We report a case of an adult patient with an acute presentation of midgut malrotation which highlights the dilemmas of preoperative diagnosis, as supported by a review of the literature

Symptomatic malrotation was treated by corrective surgery according to the technique originally described by Ladd. If a volvulus was present, the intestines were de-rotated in a counter clockwise manner and all Ladd's bands were carefully removed and dissected. If needed, the mesentery was broadened and the adhesions surrounding the mesenteric vessels dissected in order to avoid future recurrence of volvulus. When the dissection was done, the small bowel was placed to the right and the colon to the left side of the abdominal cavity in a "non-rotational" position. Two different surgeons registered data from medical charts on these surgical details independently

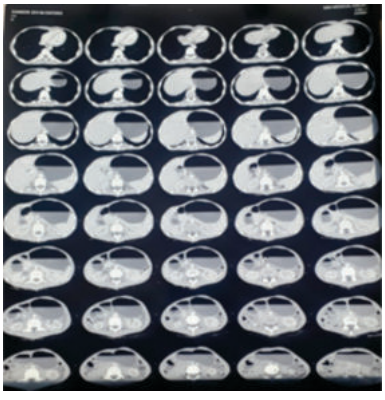
CASE REPORT

A 14 year old was admitted to the Surgery department with a three day history of acute onset, cramp like abdominal pain. There was associated nausea but no vomiting.Absent Bowels sounds with no flatus had been passed for 24 hours.

On examination, the patient was afebrile and haemodynamically stable. The abdomen was moderately distended with significant tenderness in the epigastrium and left hypochondrial regions. There was no evidence of peritonitis. A chest Radiograph did not reveal air under the diaphragm. Abdominal radiograph showed non-dilated gas filled loops of bowel in the central and upper abdominal regions.

The patient was resuscitated with intravenous fluids, analgesia . Under ultrasonography largely distended stomach was found and he was managed conservatively and further investigation was done to diagnose it as a case of malrotation with Ladd band causing gastric out let syndrome .

Patient had very poor nutritional status with S.albumin 1.8 ng /dl. Patient was nutritionally build up and taken for surgery after s.albumin 3.0 Finding at operation included dilated small bowel in the upper abdomen, partial torsion and the caecum was the left side of the abdomen tethered by torted omentum, and loops of small bowel occupying the right paracolic gutter and the right iliac fossa.adhensolysis was done The anatomical malrotation was left uncorrected.Ladd band exisced



However, a significant number of cases remain quiescent during childhood. Incidental diagnosis may then occur in adulthood; when imaging investigations are carried out for other symptoms or, during surgery for unrelated pathology. It has been reported that the incidence of malrotation in adults is approximately 0.2%. However, it is probable that this rate will rise with future developments in diagnostic imaging. but evidence from post mortem studies suggest that gut malrotation may affecth up to 1 in 6000 [10,11].

Midgut malrotation is broadly considered a deviationhp from the normal 270 degree counterclockwise rotation of the gut during embryonic development. During week 4 of foetal development, the embryonic gut, consistinurther in 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.the midgut undergoes a 270 degree counterclockwise 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 ofTreitz. duodeno-jejunal flexure(DJF) and jejunum to reduce first and lie to the left. The 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 [3,11-12].

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 [3,11,12]. Ladd's bands compress the duodenum and can potentially cause duodenal obstruction.

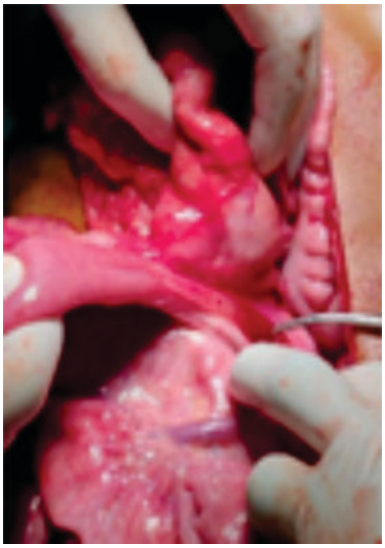
Midgut malrotation in adults presents in numerous ways and the symptoms are non-specific. The clinical diagnosis in adolescents and adults is difficult because it is rarely considered on clinical grounds. Many patients remain asymptomatic and the diagnosis is discovered incidentally during investigations or laparotomy for other unrelated problems in adult life. Wang and Welch [11] showed that 24 of 50 patients were clinically asymptomatic in their case series of adolescents and adults with malrotation.

literature: acute and chronic [10,12] Chronic presentation is more common in adults. This is characterised by intermittent crampy abdominal pain, bloating, nausea and vomiting over several months or years. The symptoms may be highly nonspecific Dietz et al [15] studied a series of 10 adults with bowel obstruction caused by intestinal malrotation. They reported that 5 adults presented with chronic features and that the duration of symptoms extended to 30 years. Fu et al [12] reported that 6 of 12 patients in their series presented with chronic intermittent abdominal symptoms. Diagnostic delays are common in this group of patients because of the nonspecific nature of the presentations. compression effect of Ladd's bands running from the caecum and ascending colon to the right abdominal wall [16] will generally show the duodenum and duodenojejunal flexure located to the right of the spine. The use of a contrast enema in conjunction with the UGI study has also been advocated as it can be used to demonstrate an abnormally located ileocaecum and right colon. Deviation from the normal positional relationship of SMV and SMA was originally described by Nichols and Li [14] as a useful indicator of the diagnosis of midgut malrotation. 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 . CT scan findings of gut malrotation and small bowel obstruction without volvulus, may show internal herniation secondary to Ladd's bands.

Choi et al[13] reviewed 177 patients over a 35-year period. They found that asymptomatic patients had a low risk of intestinal volvulus and therefore advised that routine investigations, screening and elective surgery were not necessary with close follow-up. However, it is increasingly argued that all suitable patients with intestinal malrotation should undergo surgical correction regardless of age as it is impossible to predict which patients will develop catastrophic complications .

The surgical management of intestinal malrotation was first described by William Ladd in 1936 [3] and this remains the mainstay of treatment. The classical Ladd's Procedure consists of 4 parts: division of Ladd's bands overlying the duodenum; widening of the narrowed root of the small bowel mesentery by mobilising the duodenum and division of the adhesions around the SMA to prevent further volvulus; counter clockwise de torsioning of the midgut volvulus if present and appendicectomy to prevent future diagnostic dilemma of an abnormally located appendix.

Dietz et al [15] also reported a complete symptomatic resolution in 8 of their 10 patients treated surgically. Variations of the technique used to manage intestinal malrotation have been introduced to prevent recurrent volvulus.



We offered a modified procedure to our patient by performing a division of Ladd's bands and an appendicectomy. There was no volvulus and we did not feel that the duodenum needed to be mobilised and straightened in this case. Our patient has been completely symptom free during 12 months of follow up.

A retrospective analysis of both open and laparoscopic Ladd's procedures by Stanfill et al performed at the Children's Hospital of Illinois, USA noted that short-term results were superior with the laparoscopic approach and can be achieved without any increase in the duration of the operation [15].

CONCLUSION

Intestinal malrotationa 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 intraabdominal symptoms. The presentation is usually nonspecific and this often leads to diagnostic and treatment delay with possible bowel ischaemia and necrosis. Evidence of which portends 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 an abdominal catastrophe and fatality. There are no reliable means of identifying which group of patients with intestinal malrotation will develop subsequent complications. In the light of this, many authors are now advocating early surgical intervention in the form of a standard and modified Ladd's procedure. There is evidence in the literature that the use of Ladd's procedure or ordinary division of Ladd's bands and adhesiolysis relieves symptoms and in fact, prevents recurrence in the majority of patients.

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