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A RARE CASE SERIES OF MIDGUT MALROTATION MASQERADING AS INTESTINAL OBSTRUCTION IN ADULTS

General Surgery

KEYWORDS: malrotation of midgut, intestinal obstruction

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Importance:

The presentation of intestinal malrotation in adults is rare, and occurs in approximately 0.2%[1]. Because presentation is nonspecific and the index of suspicion for malrotation progressively decreases in the older population, the clinical diagnosis is usually not considered in the initial evaluation.

We report a series of 2 cases in adult patients presented over a period of 3years with an acute presentation of midgut malrotation which highlights the dilemmas of preoperative diagnosis.

CASE REPORT 1:

A 35-year old male patient was admitted to the Emergency department with history of abdominal pain since 2 days, acute in onset and colicky in nature. Associated with bilious vomiting and constipation since 2 days. Previous history of tuberculosis present 2 years back and completed anti-tubercular therapy (ATT). There was no previous surgical history.

On general examination, patient conscious and coherent, moderately built and moderately nourished. Vital signs PR: 106 bpm, BP: 100/60 mm of Hg, TEMP: 99° F, RR: 18 cpm, Spo2: 96 % at room air.

Systemic examination: Per abdomen: The abdomen was distended with tenderness in the umbilical, epigastric, left hypochondrium. Hernial orifices were intact. Tinkling bowel sounds present.

P/R Examination: Rectum was collapsed. No faecal staining.

Investigations:

Routine admission blood tests include complete blood picture, serum electrolytes, Renal function tests (RFT's) , liver function tests(LFT's), clotting profile and an arterial blood gas (ABG) were done. However, a full blood count (FBC) demonstrated a haemoglobin level of 14.2 g/dl with a slightly raised total white cell count (WBC) of 13,400 and a neutrophilia of 82%.

Chest X-ray: Normal lung fields, no hilar lymphadenopathy.
X-ray Erect Abdomen: Multiple air fluid levels were seen.
Ultrasound abdomen and pelvis: Dilated small bowel loops, no ascites.

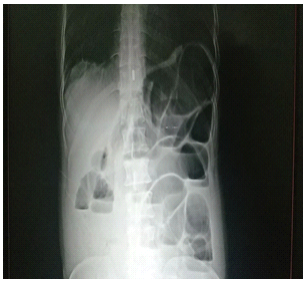

Fig 1: Photograph showing multiple air fluid levels on X-ray erect abdomen.

Preoperative diagnosis:
Acute intestinal obstruction (?? Adhesions due to tuberculosis)

Operative findings:
Caecum along with appendix was identified on the left side, Ladd's bands were identified from the right anterior abdominal wall to the duodenum and caecum, small bowel loops were dilated, and transverse colon was collapsed.

Procedure done:
Ladd's band was released and appendicectomy was done. Intestine is replaced into the abdominal cavity, with the small bowel lying entirely on the right side with the colon on the left.

POST OPERATIVE DIAGNOSIS: MALROTATION OF MIDGUT

Post operative recovery: Uneventful, Patient was observed for 1 day in ICU. Oral feeds allowed on day 3. Discharged on day 6.

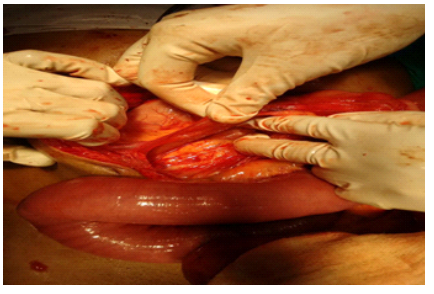

Fig 2: Ladd's bands were identified from the right anterior abdominal wall to the duodenum and caecum.

Fig 3: Photograph showing Caecum along with appendix was identified on the left side.

CASE REPORT 2:

A 22-year old male patient was admitted to the Emergency department with history of abdominal distention and colicky abdominal pain since 3 days, associated with bilious vomitings, since 4 days, history of dyspepsia present. Patient passing flatus but not

passing stools since 2 days. The patient had experienced similar symptoms in the past and had no previous surgical history.

On general examination, the patient Vital signs: PR:98 bpm, BP: 100/70 mm of Hg, TEMP: 99° F, RR: 16 cpm, Spo2: 98% at room air.

Systemic examination: The abdomen was distended with tenderness in the umbilical, epigastric. Hernial orifices were intact. Tinkling bowel sounds present.

Investigations:
Complete blood picture demonstrated a haemoglobin level of 15 g/dl with a slightly raised total white cell count (WBC) of 12,400 and a neutrophilia of 72%, hematocrit slightly elevated, serum sodium of 126 mEq/mol, serum potassium 3mEq/mol. Blood urea, serum creatinine, liver function tests (LFTs), clotting profile were within normal limits. However X-ray Erect Abdomen: Multiple airfluid levels noted. Ultrasound abdomen: Dilated small bowel loops , no free fluid.

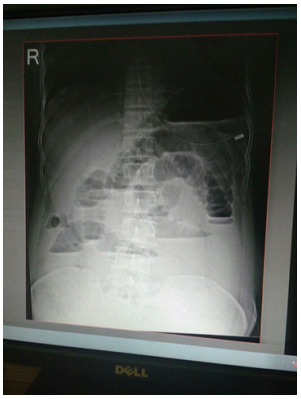

Figure 4: X-ray erect abdomen at admission

PRE OPERATIVE DIAGNOSIS: SUBACUTE INTESTINAL OBSTRUCTION
Management:
The patient was managed conservatively for 48 hours on:

- IV fluid resuscitation (with isotonic saline solution).
- Nasogastric tube decompression.
- Broad spectrum antibiotics (given prophylactically).
- Urine output monitoring by placement of foley's catheter.
- Serial electrolyte level measurements, as well as hematocrit and WBC counts (to assess the adequacy of fluid replacement).
- Inspite of conservative management, clinical detoriation of the patient and increasing small bowel distention in abdominal radiographs prompted in operative intervention.

Figure 5: X-ray erect abdomen before surgery

Operative findings:
Ladd's bands were identified from the right anterior abdominal wall to the caecum along with appendix was on the left side , small bowel loops were dilated , transverse colon was collapsed

Fig 6: Photograph showing Ladd's band attached to the caecum.

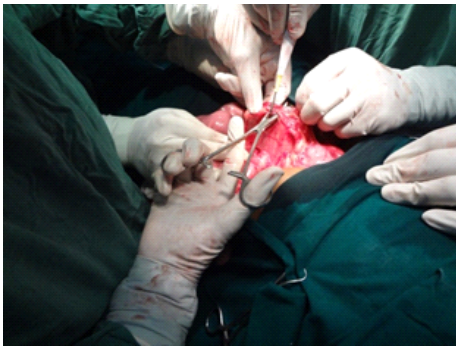

Fig 7: Photograph showing Ladd's bands being released.

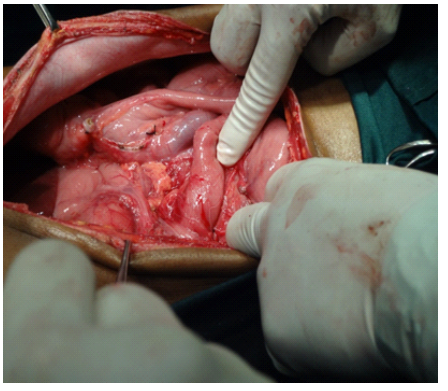

Fig 8: Photograph showing the small bowel placed on the right side while the colon is placed on the left.

POST OPERATIVE DIAGNOSIS: MALROTATION OF MIDGUT
Procedure done: Ladd's bands are divided, widening the base of the mesentery, and placing the duodenum and proximal jejunum along the right gutter and the caecum in the left abdomen. Appendectomy performed. Intestine is replaced into the abdominal cavity, with the small bowel lying entirely on the right side while the colon is on the left.

POST OPERATIVE PERIOD: Uneventful.

DISCUSSION:
In adulthood, intestinal malrotation is estimated to occur in 0.2% to 0.5%.[1,2] A literature review by von Flue et al. cites 40 cases from 1923 to 1992.[4] Intestinal malrotation can be broadly defined as any deviation from the normal 270 degree counterclockwise rotation of the midgut.[21]

The first description of the embryologic process of intestinal rotation and fixation was not published until 1898. In 1923, **Dott** was the first to describe the relationship between embryologic intestinal rotation and surgical treatment.[3] In 1936, **William E. Ladd** wrote the classic article on treatment of malrotation. His surgical approach, now

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known as the Ladd procedure, remains the cornerstone of practice today.[5]

Embryology[21]:
The primitive gut is formed during the fourth week of fetal human gestation.

Physiological Herniation:
Development of primary intestinal loop is caused by rapid elongation of cephalic limb. Stage I occurs during 5-10 week's gestation i.e, period of physiologic herniation.

Due to rapid growth and expansion of liver, the abdominal cavity temporarily becomes too small to contain all intestinal loops and they enter into the extra embryonic cavity in the umbilical cord during sixth week of development.

Rotation of gut:
Coincident with growth in length the primary intestinal loop rotates around an axis formed by SMA. When viewed from front this rotation is counter clockwise and it is approximately 270^0, when it is complete. Even during rotation elongation of small intestinal loop continues and forms number of coiled loops. The large intestine lengthens considerably but does not participate in coiling phenomenon. The rotation occurs during herniation (90^0) as well as return of intestinal loops into the abdominal cavity (180^0).

Retraction of herniated loops:
During 10^{th} week, i.e stage II, the herniated loops begin to return to abdominal cavity. Proximal part of jejunum, 1^{st} part to re-enter abdominal cavity, comes to lie on the left side. The later returning loops gradually settle more and more to right. The caecal bud which appears at about 6^{th} week as a small conical dilatation of caudal limb of primary intestinal loop is the last part to re-enter abdominal cavity.

Temporarily it lies in the right upper quadrant directly below the right lobe of liver. From here it descends into the right iliac fossa placing the ascending colon and hepatic flexure on right side of abdominal cavity. During this process, the distal end of caecal bud forms the appendix.

Mesenteries of intestinal loops
Stage III lasts from 11 weeks' gestation until term. It involves the descent of the cecum to the right lower quadrant and fixation of the mesenteries. The right and left colons are anchored to the posterior abdominal wall by mesenteric attachments.

When the caudal limb of loop moves to right side of abdominal cavity, the dorsal mesentry, twists around the origin of the SMA. Later when the ascending and descending mesenteries obtain their definitive position, their mesenteries press against the peritoneum of posterior abdominal wall.

After fusion of these layers, the ascending and descending colon are permanently anchores in retroperitoneal position the appendix, lower end of caecum, sigmoid, however, retain their free mesenteries. Transverse mesocolon however, fuses with posterior wall of greater omentum, but maintains its mobility.

Intestinal torsion anomalies can be classified according to the stage of occurrence. Stage 1 is mainly the omphalocele; stage 2 includes malrotation, non-rotation, reverse rotation and hyper-rotation of the gut ; stage 3 anomalies include mobile cecum, mobile duodenum and free mesentery of the small bowel . Potential hernial pouches form when the mesentery of the right and left colon and the duodenum do not become fixed to the retroperitoneum.[5]

Clinical presentation of intestinal malrotation can be unspecific. Dietz *et al.*, in 2002, described the Cleveland Clinic series of 10 adults with this form of intestinal anomaly.[6] They described two different groups of patients, according to their symptoms. The first group of patients presented chronic symptoms and the second group had symptoms related to the acute onset of obstruction, mainly due to adhesion of the Ladd's bands or volvulus.[6] Moldrem et al reported that 48.5% of their thirty-three patients presented with an acute abdomen.[7] Fu et al reported that 6 of 12 patients in their series presented with chronic intermittent abdominal symptoms.[8] Diagnostic delays are common in this group of patients because of the nonspecific nature of the presentations.

SEQUELAE OF MIDGUT MALROTATION:

Acute midgut volvulus [6,7]	Chronic midgut volvulus [6,7]	Acute duodenal obstruction	Chronic duodenal obstruction	Internal herniation
-Sudden onset of bilious emesis -Diffuse abdominal pain out of proportion to physical examination	-Lymphatic and venous obstruction. -Recurrent abdominal pain and malabsorption syndrome	Vomiting's & abdominal distention, peritonitis	Vomiting's , gastroesophageal reflux, early satiety, abdominal discomfort	Vomiting's and constipation

INVESTIGATIONS:
The diagnosis of intestinal malrotation can be made by radiographic studies. Conventional radiography is neither sensitive nor specific for malrotation, although right-sided jejunal markings and the absence of a stool-filled colon in the right lower quadrant may be suggestive of this finding.[8,9,16]

Plain x-ray: Suggestive findings involve gasless abdomen, Double bubble, Pneumatosis Intestinalis with Distended bowel loops.[8,9]

UGI Contrast Series: {only in stable pts} is the 'Gold standard' investigation. Findings include abnormal position of the ligament of Treitz and duodenum remains to the right of the spine.[10,16]

USG: Reversal of the normal anatomic relationship between the SMA and superior mesenteric vein (SMV) is suggestive of malrotation. "Whirlpool sign" is consistent with a midgut volvulus.[8,9,11]

Contrast enema: Usually shows malposition of the right colon with the ileum entering the cecum from the right, but the cecum may assume a normal location. The contrast enema findings are also non-specific because cecal location can be variable without malrotation in up to 20% of patients.[8,9,10]

Angiography: It was used to demonstrate the characteristic corkscrew appearance of a whirling SMA and its branches; the 'barber pole sign' as well as extensive collaterals caused by proximal SMA occlusion.[15]

Computerized Tomography: Many cases of quiescent malrotation in adults are currently being detected on cross-sectional imaging (particularly CT) performed for various unrelated reasons. CT not only shows the intestinal malpositioning seen on barium studies but also depicts associated extra intestinal findings not evident on conventional examinations. Deviation from the normal relationship between the superior mesenteric artery (SMA) and superior mesenteric vein (SMV) is a useful indicator of malrotation.[13,14] 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.[14] However, abnormal orientation of the SMA-SMV relationship is not entirely diagnostic of malrotation.

UGI Contrast Series (only in stable pts): It is the 'Gold standard' investigation. Findings include abnormal position of the ligament of Treitz and duodenum remains to the right of the spine– that is, the duodeno-jejunal junction fails to cross the midline and lies below the

level of the duodenal bulb.[15,16]

MANAGEMENT:

Ladd procedure, initially described in 1936, is the classic surgical treatment for intestinal malrotation.[4,17,18] This procedure consists of 4 parts: counterclockwise detorsion of the midgut volvulus if present; division of Ladd's bands overlying the duodenum thereby relieving the cause of the intermittent obstruction; widening of the narrowed root of the small bowel mesentery by mobilizing the duodenum; and division of the adhesions around the SMA to prevent further volvulus. Appendectomy is performed due to possible difficulty in the diagnosis of future appendicitis, distant from the classic lower right quadrant position.[4]

There are recent reports of the use of the laparoscopic approach in the surgical treatment of intestinal malrotation. The technique appears to be safe and effective when performed by experienced laparoscopic surgeons, especially in the absence of volvulus.[17,18] Mazziotti et al. recently reported a series of malrotation patients managed successfully with laparoscopic intervention.[20] Moreover, Matzke et al. published their comparative results for open and laparoscopic Ladd procedures, concluding that the laparoscopic Ladd procedure is feasible, safe and as effective as the standard open Ladd procedure for the treatment of adults who have intestinal malrotation without midgut volvulus.[19] Similar conclusions are presented by Seymour et al. who also noted that long-term follow-up will be required to determine whether patients are at risk for either new symptoms or recurrent malrotation-related problems such as midgut volvulus.[22]

CONCLUSION:

The clinical diagnosis of malrotation after childhood is usually not considered; this oversight underscores the importance of recognizing this unsuspected condition on imaging. Regardless of patient age, surgical treatment of quiescent malrotation should be considered because surgery remains the only real safeguard against complications. In older patients who present with acute symptoms related to unsuspected malrotation, rapid imaging diagnosis and surgery may be life-saving.

Diagnosis requires a high index of suspicion. Specific findings that are diagnostic of malrotation can be detected through the use of both upper and lower gastrointestinal tract barium studies, CT scan and often emergency laparotomy. Complete resolution of acute obstruction or chronic abdominal pain is the result of a high index of suspicion for malrotation, appropriate diagnostic studies and aggressive definitive treatment.

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