Case Report

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Laparoscopic management in an adult patient with intestinal obstruction secondary to congenital midgut malrotation: a case report and review of the literature

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ABSTRACT

Intestinal malrotation is the failure of the fetal intestine to complete a 270-degree counter clockwise rotation around the superior mesenteric artery, resulting in positional and fixation abnormalities of the small and large intestine. Adult presentation is uncommon with an estimated incidence of 0.2 to 0.5%. There is no consensus on treatment in patients with asymptomatic intestinal malrotation in adults. The aim of this document was to report and review a case of intestinal malrotation conditioning intestinal obstruction in an adult patient with successful laparoscopic management. We conclude that laparoscopic resolution is feasible, reproducible and may ensure a faster recovery in a selected group of patients.

Keywords: Malrotation congenital, Intestinal obstruction, Band of Ladd, Case report, Laparoscopy

INTRODUCTION

Intestinal malrotation represents one of the most common anomalies of the gastrointestinal tract encountered by radiologists and surgeons. It is defined as the inability of the fetal intestine to complete a 270-degree counterclockwise rotation around the superior mesenteric artery (SMA) at 10 weeks of gestation. This results in anomalies of position and fixation of the small and large intestine. Consequently the duodenum and cecum unite through Ladd's bands, precipitating midgut volvulus, intestinal obstruction and ischemia. Adult presentation is uncommon with an estimated incidence of 0.2 to 0.5%. The aim of this report was to present the case of a 44 year-old adult with intestinal obstruction due to malrotation with successful laparoscopic management in an emergency setting with literature review.

CASE REPORT

A 44 year-old male patient with a history of open appendectomy 3 years prior to hospitalization, presented to the emergency department with 6 days history of insidious, progressive epigastrium pain radiating to the lower back. It was associated with nausea, vomiting and obstipation.

On physical exam, he was afebrile and hemodynamically stable. His abdominal exam revealed a distended abdomen with significant voluntary guarding. His abdomen was diffusely tender with the point of maximal tenderness in the right and left upper quadrants.

Blood investigations on admission showed an elevated white blood cell count of 15.3×103/µl with 79% neutrophils, hemoglobin 19.80 g/dl, hematocrit 55.80%, abnormal renal function with urea 204 mg/dl, creatinine

of 6.47 mg/dl, electrolyte imbalance including sodium 120 mEq/l, potassium 2.7 mEq/l, chlorine 54 mEq/l, phosphorus 10.92 mEq/l, magnesium 2.9 mEq/l and also abnormal liver function tests and pancreatic enzymes with total bilirubin 7.52 mg/dl, direct bilirubin 0.92 mg/dl, indirect bilirubin 6.60 mg/dl, albumin 5.36 g/dl, ALT 123 IU/l, AST 66 IU/l, GGT 97 IU/l, FA 74 IU/l, DHL 318 IU/l, amylase 342 U/l and lipase 841 U/l.





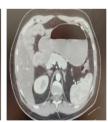


Figure 1: A computerized tomography (CT) scan of the abdomen was performed revealing duodenum that does not cross the midline in its third part, duodenojejunal junction in the right hemiabdomen; cecum located in the pelvis and lateralized to the left, with left ascending colon, findings suggestive of intestinal malrotation.

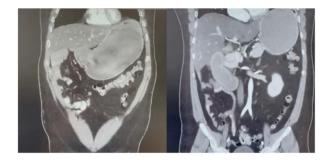


Figure 2: New CT scan was performed showing dilatation of the gastric chamber and duodenum apparently up to the third part with a transition zone without being able to visualize the etiology, with contrast medium advancing to the sigmoid colon.



Figure 3: The cecum, ascending and descending colon were found towards the left paracolic gutter of the patient.

He was admitted with the diagnosis of acute pancreatitis by the internal medicine service for medical management. During his hospital admission he continued with oral intolerance, as well as significant abdominal distension, generalized abdominal pain of moderate intensity, absence of bowel movements, constipation and inability to pass gas. A computerized tomography (CT) scan of the abdomen was performed revealing duodenum that does not cross the midline in its third part, duodeno-jejunal junction in the right hemiabdomen; cecum located in the pelvis and lateralized to the left, with left ascending colon, findings suggestive of intestinal malrotation (Figure 1).

A panendoscopy was performed which reported 100% stenosis of the lumen in the third part of the duodenum, secondary to extrinsic compression.

Conservative treatment was started with hydration, analgesia and placement of a nasogastric tube, however after 3 days the patient continued with intolerance to the oral route, as well as lack of bowel movements and obstipation and a new CT scan was performed showing dilatation of the gastric chamber and duodenum apparently up to the third part with a transition zone without being able to visualize the etiology, with contrast medium advancing to the sigmoid colon (Figure 2).

Considering the unfavorable clinical evolution, it was decided to perform a diagnostic laparoscopy, identifying as trans-operative findings: Ladd's bands at the level of the third part of the duodenum which conditioned fixation of the third part of the duodenum towards the retroperitoneum, where the transition zone was found. The cecum, ascending and descending colon were found towards the left paracolic gutter of the patient (Figure 3). Due to these findings it was decided to perform a laparoscopic Ladd's procedure, which consisted in the release of the described bands and rectification and fixation of the small bowel with the "GEA" extracorporeal knot technique.

Postoperative evolution was adequate, the patient successfully recovered tolerating the oral administration 24 hours after surgery and presented bowel movements and evacuations. He was discharged on the third postoperative day.

DISCUSSION

Ladd's bands, are peritoneal tissue fibers that classically attach the cecum to the abdominal wall and create an obstruction of the duodenum. (Classical malrotation, the most common form, involves incomplete rotation of the duodenum (duodenojejunal malrotation) and colon (cecocolic malrotation), which causes the small bowel to be suspended in a narrow mesentery that is not attached to the retroperitoneum. Variations of rotation, such as isolated duodenojejunal or cecocolic malrotation, and reverse rotation, can occur, but are infrequent. (2,8,13,15)

The exact etiology of malrotation remains uncertain, but some studies have indicated a genetic component. An association between malrotation and mutations in the FOXF1 transcription factor and L-R asymmetry genes has been identified.^{8,15}

Intestinal malrotation has an incidence of 0.2% in live births, usually presenting in the first week of life. 190% of cases are diagnosed in the first year of life. 1,2,8 It is estimated that up to 1 of 200 live births have an asymptomatic rotational anomaly. 3 Adult presentation is uncommon with an estimated incidence of 0.2 to 0.5%. The male to female ratio is 2:1 and more than one third of cases are associated with a congenital anomaly. 3

The classic symptomatology in the newborn is intestinal obstruction, with vomiting of biliary characteristics. ^{2,4,13} As for the symptomatology in adults, it typically presents with progressive abdominal pain. ⁴ The chronic and nonspecific symptoms observed in adult malrotation, combined with its low prevalence, result in an incorrect, late diagnosis which increases morbidity and mortality. ¹ Other symptoms include vomiting, constipation, and oral intolerance. ^{1,9} Cases have been described in which intestinal malrotation is associated with acute pancreatitis, as in the present case. ^{1,10}

In pediatric patients the gold standard for diagnosis is a gastroduodenal series, which should demonstrate an abnormal location of the duodenal-jejunal junction. In the case of volvulus, an abrupt cut-off of contrast in the gastrointestinal tract may be seen.^{2,5} In adults an abdominal CT scan with oral and intravenous contrast usually gives the diagnosis. The classic finding associated with malrotation on ultrasound and CT is the whirlpool sign, created by collapsed bowel loops surrounding the mesenteric vasculature of the midgut.^{1,2}

Neville et al found that about one-fifth of patients had undergone a previous abdominal surgical procedure without a diagnosis of malrotation. ¹

Once symptomatic malrotation has been diagnosed, it must be surgically corrected; if there is a confirmed or suspected volvulus, emergency surgery is indicated to prevent complications.³ In the case of our patient, presenting clinical symptoms of intestinal occlusion, with no clinical or laboratory data of loop distress and with doubts about the etiology of duodenal compression, it was initially decided to provide conservative management with fasting, hydration and placement of a nasogastric tube; however, since he persisted with symptomatology, it was decided to perform surgical management.

The Ladd's procedure is considered the gold standard for the treatment of complicated intestinal malrotation and was described in 1936.^{2,3} It consists of reduction of the intestinal volvulus if present, division of the peritoneal bands causing the obstruction, repositioning of the small and large intestine to the right and left side of the abdomen respectively, and finally concludes with an appendectomy.³ Yousef El-Gohary et al performed a retrospective study in pediatric patients submitted to the Ladd procedure with a 5-year follow-up, in which they observed a low incidence in the recurrence of intestinal malrotation.¹¹

The laparoscopic approach was first reported by Zee et al in 1995. Since then, some case series have shown that the laparoscopic Ladd's procedure achieves a decrease in hospital stay and postoperative nasogastric tube use, optimizing patient recovery and with a conversion rate to laparotomy between 2% and 33%.

Oms in 2014 performed a retrospective study in which 83 patients aged 0 to 18 years were included, who underwent surgical treatment for intestinal malrotation. ¹⁴ Laparoscopic management was performed in 33 patients, and open management in 50 patients, the average surgical time was 63 minutes for the minimally invasive group, unlike the open management with a time of 76 minutes (p=0.588). The length of hospital stay was significantly longer after laparotomy (9 versus 16 days, p=0.002). The most common reoperation in open surgery was for bowel obstruction (5 versus 0%), not statistically significant.

Lane et al in 2015 performed a retrospective comparative study between adult patients undergoing a Ladd procedure with open versus laparoscopic approach. They included 22 patients in total, identifying decreased hospital stay in those who underwent a laparoscopic procedure $(5.0\pm2.5$ days versus 11.6 ± 8.1 days, p=0.0148).⁵

In 2013 Yuka et al performed a literature review on adolescent patients with intestinal malrotation undergoing Ladd's procedure.¹² It was performed in 27 patients, of which 12 were performed laparoscopically. The authors concluded that laparoscopic management was an adequate option for definitive treatment in patients with intestinal malrotation without volvulus.

In the case of our patient, it was decided to perform diagnostic laparoscopy, finding Ladd's bands at the level of the third part of the duodenum without associated volvulus, so dissection of the bands was performed, freeing the intestine to be subsequently rectified and fixed using extracorporeal stitches. The patient already had a history of previous appendectomy and, as in the cases described in the literature, a diagnosis of malrotation was not made in that surgical event.

CONCLUSION

Intestinal malrotation is considered an uncommon pathology, which usually occurs in pediatric patients, however the percentage in the adult population is not negligible and is usually associated with complications such as intestinal obstruction or midgut volvulus that

require prompt surgical resolution. Likewise, laparoscopy can offer a faster recovery and less morbidity in this group of patients, without increasing the rates of postoperative complications or long-term recurrence.

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