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Case Report

DELAY IN THE DIAGNOSIS OF SYMPTOMATIC INTESTINAL MALROTATION IN A NEW BORN PRESENTED WITH MIDGUT VOLVULUS AT 6MONTHS OF AGE – A RARE CASE REPORT

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ABSTRACT

Intestinal malrotation is a relatively uncommon condition with diverse outcomes. Familiarity with variations in the presentation of malrotation is imperative as early diagnosis and prompt subsequent surgical intervention are essential to optimizing outcome. The most frequent clinical sign in the neonate is bile-stained emesis. We report a case of unsuspected malrotation that were diagnosed in neonates with a history of nonbilious emesis who were assessed for presumed gastroesophageal reflux or aspiration. Gastroesophageal reflux is a common condition among newborns, and can be a subtle presentation of malrotation. Clinicians should consider malrotation as a possible cause of reflux, particularly in infants with unusually pathologic or persistent symptoms necessitating ongoing treatment for reflux. This is very important as delay in the diagnosis of intestinal malrotation may result in midgut volvulus which questions the viability of major portion of intestine.

INTRODUCTION

Malrotation of the intestines occurs in 0.5% of the population and can lead to devastating consequences, including volvulus, ischemic bowel, short bowel syndrome, or even death.[1,2] The condition results from abnormal rotation of the intestines during embryological development and is most often diagnosed in the first year of life.[1] While it is commonly agreed that the Ladd procedure is the treatment of choice for symptomatic malrotation, there are still differences in opinion about whether or not to treat incidental findings of malrotation with prophylactic surgical intervention.

During normal embryologic development, the gut tract expands so rapidly that the embryonic cavity cannot accommodate its size. As a result, the intestines extrude out into the amniotic space outside of the fetal abdomen. As the bowel returns to the abdomen by the 8th to 10th week of gestation, the loops of intestine normally rotate 90° counterclockwise around the superior mesenteric artery. They continue to rotate an extra 180° counterclockwise, resulting in a complete rotation of 270°. This motion returns the bowel into the posterior abdomen with the duodenojejunal junction (location of the ligament of Treitz) placed to the left of the midline and the cecum placed in the right iliac fossa.

Intestinal malrotation occurs when the normal embryological process of bowel rotation happens incorrectly or incompletely. Symptomatic malrotation occurs in as many as 1 in 200 live births, but the asymptomatic incidence is unknown because many of these cases are never diagnosed.[1] With incomplete intestinal rotation, the duodenojejunal portion of the small intestines is positioned vertically and the cecocolic portion of the intestine is rotated 90° instead of the full 180°. The cecum is positioned in the mid to upper left portion of the abdomen instead of the right lower quadrant, and peritoneal bands, also known as Ladd bands, fix the cecum to the lateral wall of the abdomen.[2] Occasionally, these bands can cause intermittent intestinal obstruction if they compress the duodenum.

CASE REPORT

A 6 month old female of 3kg body weight, was referred to Tripura Medical College and BRAM hospital as a case of intestinal malrotation with midgut volvulus. It was a term baby delivered via spontaneous vaginal delivery with a birth weight of 2.7 kg. Her birth was uncomplicated, and she was discharged home after a 48-hour stay in the hospital. After discharge, she was breastfed exclusively, had consistent wet diapers and stools, and continued to grow and gain weight. However, at 2 weeks of age, she presented to his primary care physician (PCP) with increasingly frequent spit ups and decreased stools for 1 week. Per the mother, the spit ups had started on the second day of life, but they had initially improved significantly with head elevation after feeds. There was a history of repeated hospital admissions and being treated conservatively for increasing amount and frequency of spit-up and was discharged. Over the last few weeks, the patient

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developed abdominal distension with vigorous bilious vomiting for which the mother consulted PCP who ordered an upper gastrointestinal (GI) contrast study with follow through as shown in figure 2 and subsequently admitted the patient to the hospital for severe gastroesophageal reflux, failure to thrive, and intestinal malrotation.

On the initial physical examination in the hospital, the patient was awake and appeared emaciated and dehydrated. His abdomen was soft, nontender, distended with visible loops and had active bowel sounds and no organomegaly as shown in figure 1. The rest of his physical examination was also normal. Overall, he appeared stable, nontoxic, and in no acute distress. On doppler ultrasonography whole abdomen alteration in the relationship of superior mesenteric artery (SMA) and vein (SMV) with whirlpool sign positive suggestive of intestinal malrotation with midgut volvulus. A nasogastric tube (NGT) was placed, and the patient was kept NPO (nil per orally) with intravenous fluid and broad spectrum antibiotics. Patient was prepared for emergency laparotomy on the next day. On laparotomy after evisceration midgut volvulus was de-rotated, duodenum was kocherised, nasogastric tube was advanced upto jejunum to exclude any duodenal web, ladds bands were released, widening of mesenteric base was done and lastly appendectomy was performed as shown in figure 3 and figure 4. The small bowel was placed in the right side and large bowel was placed in the left side. Abdomen closed in layers. After 2 days of surgery patient passed stool with bowel movements restored and decreased abdominal distension. The NGT was removed on post operative day 4. The infant began tolerating oral feedings without subsequent emesis. On postsurgical day 7, the infant was discharged home, where he continued to tolerate feedings and gain weight.

Post operative day 14, the child was asymptomatic with body weight of 5kg, tolerating full oral feeds with occasional spit ups as shown in figure 5.

Figure 1 Showing distended abdomen with visible bowel loops

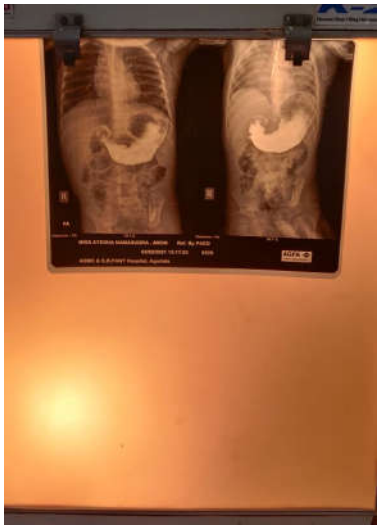

Figure 2 Upper GI contrast study and follow through

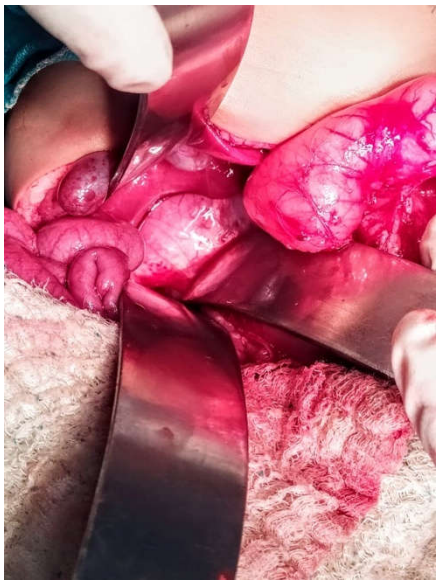

Figure 3 intra-operative picture showing distended stomach and duodenum

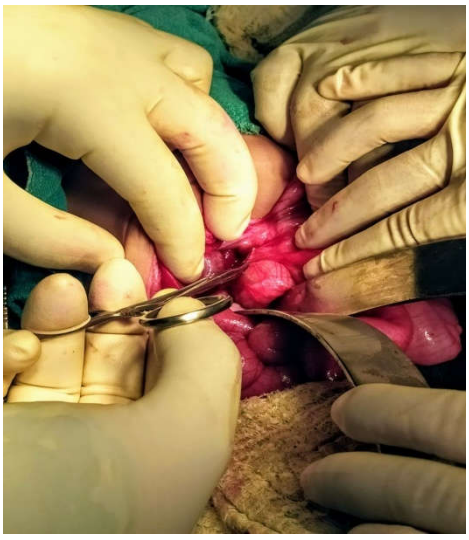

Figure 4 Showing kocherisation of duodenum and division of ladds bands

Figure 5 Showing post-operative day 14 for follow –up

DISCUSSION

Malrotation can cause a wide range of clinical symptoms. Infants with malrotation that has progressed to volvulus

present with bilious vomiting and symptoms of acute bowel obstruction, including obstipation and abdominal distension.[2] They may also present with hypovolemia with or without septic shock.[2] These cases most commonly present in the first month of life and 90% present within the first year. Malrotation may also manifest as recurrent episodes of bilious or nonbilious vomiting, failure to thrive, solid food intolerance, malabsorption, diarrhea, bloating, and abdominal pain.[3] Some infants may present with nonspecific findings, which mimic colic, gastroesophageal reflux, pancreatitis, or biliary obstruction.[3-5] As the children get older and the symptoms become less specific, diagnosis becomes more challenging and less timely. The upper GI series, which has a sensitivity of approximately 96% in infants, is the imaging study of choice when diagnosing malrotation patients.

Treatment of symptomatic malrotation entails surgical intervention. The Ladd procedure, the current standard of care for malrotation, involves lengthening the mesentery and positioning the small and large intestines in the right and left sides of the abdomen, respectively. This surgical intervention aims to reduce the risk of volvulus in the future as well as alleviate other symptoms caused by malrotation. On the other hand, physicians still disagree on the best treatment for asymptomatic malrotation, which is often found incidentally on imaging done for other conditions.

Malrotation, and subsequent progression to volvulus, presents most commonly in the first month of life, and 90% of volvulus cases occur in the first year of life.[6] As children get older, their risk of volvulus significantly decreases. Less than a third of malrotation cases in older children lead to the complication of volvulus, and this fraction continues to decrease as individuals continue into adulthood.[7] Therefore, some argue that the risk of volvulus past a certain age does not outweigh the risks associated with surgery. The Ladd procedure, the current treatment for malrotation, is associated with numerous complications in 8% to 14% of patients including intestinal obstruction.[8,9] Despite the procedure, recurrent volvulus can occur in up to 8% of patients.[10] The risks associated with surgery are greater in individuals with other comorbidities, such as heterotaxy syndrome.[2,8,11] This leads many to maintain that the risks of surgical intervention do not outweigh the benefits, especially in those with other significant comorbidities, such as major cardiac disease.[8]

Nevertheless, most surgeons agree that the benefits of a Ladd procedure to correct intestinal malrotation offsets the risks.[12] A Ladd procedure not only prevents the life-threatening complication of volvulus but also relieves other GI symptoms associated with malrotation. The lifetime risk of volvulus secondary to malrotation has been estimated to be up to 20%.[5,13] While the risk of malrotation progressing to volvulus decreases with age, current literature reveals multiple reports of adolescents and adults presenting with volvulus.[7,10,13-15] Since predicting volvulus is so challenging and it can become life-threatening in a short amount of time, a Ladd procedure is the only way to prevent volvulus in individuals with malrotation.

Furthermore, even if malrotation never progresses to volvulus, it can cause chronic GI symptoms, including chronic abdominal pain, malabsorption, diarrhea, solid food intolerance, common bile duct obstruction, and abdominal distension.[3] Fifty percent to 70% of individuals with malrotation eventually develop symptoms associated with their anomaly.[3,7,16] These chronic and unspecific symptoms are frequently misdiagnosed as irritable bowel syndrome, biliary disease, peptic ulcer disease, or psychiatric disorders.[17] These frequent misdiagnoses result in the patients suffering with these chronic complaints for months, if not years. This anatomical anomaly can also lead to delayed diagnosis of appendicitis due to the unusual location of the appendix, and this delay in diagnosis and treatment leads to increased incidences of complications, such as appendiceal rupture, in these individuals.[17] Therefore, in addition to preventing volvulus, the Ladd procedure can alleviate other chronic GI symptoms.

Last, the Ladd procedure leads to better outcomes, including reduced morbidity and mortality, when performed in nonemergent situations.[10] Elective Ladd procedure can be performed laparoscopically with shorter operative times (63 minutes vs 76 minutes) and potentially less postoperative adhesions and other subsequent complications.[18] Once malrotation has progressed to volvulus, an open procedure is preferred to a laparoscopic one. Furthermore, waiting for malrotation to progress to the emergent situation of volvulus may result in severe morbidity for these patients including short bowel syndrome and death.

CONCLUSION

Malrotation is a relatively common anomaly that can lead to life-threatening volvulus and bowel ischemia. We present a case of malrotation found incidentally on imaging and use this case as a nidus for discussion over recommended treatment for incidental malrotation. While some physicians advocate against prophylactic surgical intervention in patients presenting with incidental findings of malrotation, many others argue that the benefits of Ladd prophylaxis outweigh the risks. In order to reach a consensus on how best to approach and treat incidental findings of malrotation, further investigation and discussion of this controversial topic is necessary.

Declaration of Conflicting Interests

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