Abstract
Intestinal malrotation is an anomaly of the midgut, resulting from an embryonic defect during the phases of herniation, rotation, and fixation. The objective is to report a case of complex diagnostics and approach. The diagnosis was made surgically in a patient presenting with hemodynamic instability, abdominal distension, signs of intestinal obstruction, and pneumoperitoneum on abdominal X-ray, with suspected grade III necrotizing enterocolitis. During surgery, a volvulus resulting from poor intestinal rotation was found at a distance of 12 cm from the ileocecal valve. Hemodynamic instability and abdominal distension recurred, and another exploratory laparotomy was required to correct new intestinal perforations. Therefore, early diagnosis with surgical correction before a volvulus appears is essential. Abdominal Doppler ultrasonography has been promising for early diagnosis.

Keywords: Intestinal obstruction, Acute abdomen, Gastrointestinal tract.

CASE REPORT
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INTRODUCTION

Intestinal malrotation (IMR) is an anomaly of the mid-gut, resulting from an embryonic defect during the phases of herniation, rotation, and fixation. It may present as non-rotation, incomplete rotation, reverse rotation, or mesocolic hernia.

The condition affects 0.2%–1% of the population and is symptomatic in 1/2500–6000 of cases. In 30%–60% of cases, it is associated with other malformations and diseases such as intestinal atresia, Meckel’s diverticulum, intussusception, Hirschsprung’s disease, mesenteric cyst, anomalies of the extrahepatic biliary ducts, congenital heart disease, congenital diaphragmatic hernia, and abdominal wall defects (omphalocele and gastroschisis).

Embryological development of the intestine is complex. Initially, there is intestinal herniation outside the abdominal cavity, with one counterclockwise rotation (270°) in relation to the axis of the superior mesenteric artery. At approximately 12 weeks of gestation, the midgut returns to the abdominal cavity and the duodenum–jejunal junction (DJJ) attaches to the posterior wall of the abdomen, with its left side on the vertebral column and the Treitz ligament, while the cecum is affixed to the lower right quadrant. IMR results from failure in extracelomic intestinal rotation, often with DJJ located in the upper right quadrant and the cecum in the upper abdomen. This anomalous fixation results from adhesive bands in the gallbladder, duodenum, and right abdominal wall. This produces a narrow mesenteric base, which is predisposed to intestinal volvulus. Less often, the intestine makes one clockwise rotation (90°), bringing the duodenum forward and the colon backward, forming a tunnel which can partially obstruct the mesenteric vessels.

IMR classically manifests in newborns as vomiting bile. Volvulus occurs in the mesenteric base, causing the superior mesenteric vessels to twist. Continuous ischemia causes symptoms such as hematochezia, irritability, pain, and abdominal distension. It can progress to intestinal necrosis, which manifests as abdominal erythema, signs of peritonitis, septic shock, and death. Older children may present chronic volvulus, with abdominal pain (colic), abdominal distension with intermittent vomiting, diarrhea, gastrointestinal bleeding, and malnutrition.

Prenatal diagnosis of isolated IMR is difficult and is usually accomplished by ultrasonographic observation of complications of the volvulus (intestinal distension, meconial peritonitis, and ascites). After birth, the key symptom is vomiting bile. In general, the first test that is performed is a simple X-ray of the abdomen, which shows signs of intestinal obstruction, which, in severe cases, is accompanied by intestinal pneumatosis, which suggests necrotizing enterocolitis. Currently, the gold standard for IMR diagnosis is gastrointestinal contrast radiography to assess the position of DJJ. In doubtful cases, serial contrast X-rays of the small intestine and contrast enema may be ordered to visualize the cecum, whose position near DJJ suggests IMR. Abdominal ultrasonography has a lower sensitivity and specificity than the tests previously mentioned. In cases of neonates with a history of vomiting bile and peritonitis, laparotomy can be performed after initial measurements are taken for diagnosis and treatment, even without a radiological diagnosis.

At diagnosis, patients generally need urgent surgical treatment due to the high risk of the disease. However, the recommended treatment in asymptomatic patients remains controversial. Surgical correction was described by Ladd in 1936. The procedure is based on counterclockwise correction of the rotation of the volvulus, dividing the Ladd bands, lengthening the mesenteric bundle, and positioning the small intestine in the lower right quadrant and the large intestine in the upper left quadrant. Laparoscopic correction was described by Zee and Bax in 1995 and is similar to the open surgery. Laparoscopy has been used in stable patients, whereas the open technique is preferred when there are volvulus and related complications. Cecal appendectomy, which is recommended by many authors for IMR treatment, is controversial because many others contraindicate it.

CASE REPORT

We present the case of a 22-year-old mother, gravida 2 para 1. The firstborn was a healthy 5-year-old girl at the time of the second pregnancy. Prenatal consultations did not show complications, and obstetric ultrasonography results performed 5 days before delivery were normal. The grandfather reported that the patient went to the emergency room on June 24, 2016, after her water broke at 26 weeks, and was diagnosed with premature amniotic rupture. Tocolysis was initiated and a bed for the newborn was requested in the neonatal intensive care unit (NICU). Lack of fetal heartbeat was noted and intrapartum fetal death was diagnosed. Labor was then induced and the fetus was expelled at 11:00 am (6/26/2016). According to the mother, the newborn had a gestational age of 26 weeks and weighed 915 g; the newborn had no heartbeat or fetal movements. Death was confirmed; the newborn was placed in a box, where he remained for 2 hours. However, the nursing team noticed respiratory movements and alerted the medical team, which began resuscitation and intensive care. After resuscitation, the newborn was transferred to the pediatric emergency room of a tertiary university hospital, where he was admitted at 7:00 pm (6/26/2016).

On admission to the emergency room, the newborn was in poor overall condition, with cold and cyanotic extremities, oxygen saturation of 85% in ambient air, rhonchi and rales on auscultation, heart rate of 130 bpm, and heart murmurs, and was administered oxygen by a hood. Orotracheal intubation (OTI) was performed, followed by aspiration and surfactant administration (200 mg/kg); capillary glycemia was checked (low) and 2 mL of 10% glucose was prescribed. Antibiotic therapy...
was initiated with ampicillin, sulbactam, and gentamycin for early neonatal sepsis, and dobutamine was initiated due to distributive shock. The child was transferred to the NICU.

Serum test results on admission were AST = 488.5 U/L, ALT = 89.2 U/L, total bilirubin = 2.768 mg/dL, indirect = 2.201 mg/dL and direct = 0.567 mg/dL, CK = 443 U/L, CKMB = 426 U/L, hemoglobin = 12.00 g/dL, leukocyte count = 28250 cells/mm³ (2% myelocytes, 6% metamyelocytes, 17% rods, 66% segmented, 1% eosinophils, 0% basophils, 23% lymphocytes, and 10% monocytes), non-reactive serology (toxoplasmosis, rubella, syphilis, cytomegalovirus, HIV, and HTLV), negative bacterial blood culture, and slight bilateral reticulogranular infiltrate and normal abdomen on radiography of the thorax and abdomen Figure 1. Subsequently, echocardiography conducted on June 28, 2016, showed 1.8-mm PCA; transfontanellar ultrasonography on June 30, 2016, showed ventricular dilation and absence of bleeding.

On July 04, 2016, the patient developed hemodynamic instability and abdominal distension. Simple abdominal X-ray showed intestinal obstruction, pneumoperitoneum, and signs of grade III necrotizing enterocolitis Figure 2. Laparotomy was then indicated.

The procedure revealed meconium peritonitis with intestinal perforation associated with the midgut volvulus from IMR at a distance of 12 cm from the ileocecal valve and signs of ischemia, but no necrosis. Perforation suturing was formed along with cleaning of the abdominal cavity, excision of Ladd’s adhesions between the duodenum and transverse colon and the angle of the liver, appendectomy, repositioning of the bowel loops, and closing of the planes and the supraumbilical transverse incision on the right Figures 3 to 6.

After recovery, the patient again developed hemodynamic instability and abdominal distension. Another simple X-ray of the abdomen showed pneumoperitoneum Figure 7; therefore, another exploratory laparotomy was performed (7/13/2016). This time, two perforations were revealed, one 20 cm from the duodenum near the site of the lysis of adhesions from the first surgery and another
at the site of the previous suture, which was blocked by intestinal loops. There was also duodenal semi-occlusion by bilious residue. The upper perforation was sutured, and 5 cm of the loop at the site of the second perforation was sectioned with termino-terminal anastomosis. There were no signs of peritonitis.

The infant developed distributive shock and coagulation disorder in the immediate postoperative period. He was diagnosed with cystic hygroma in the parietal region on transfontanellar ultrasonography (7/21/2016). An electrocardiogram (7/26/2016) showed right chamber overload. A dilated eye exam (7/29/2016) showed avascular peripheral retina. The patient developed pulmonary pneumonia associated with bronchodyplasia (8/01/2016). He also required blood transfusions due to anemia (8/04/2016). Elective extubation was attempted (8/11/2016), and the patient was kept on non-invasive ventilation (NIV), but he developed apnea, hypothermia, and bradycardia, and OTI was again required. With a diagnosis of late sepsis and positive blood cultures for *Serratia marcescens*, the patient was treated for pneumonia (8/16/2016). Elective extubation was performed (8/24/2016) with the patient maintained on NIV, but reintubation was necessary on 8/25/2016 due to respiratory distress and chest X-ray showing total atelectasis of the right lung.

The patient had hemodynamic instability requiring vasoactive drugs and treatment for sepsis with positive blood cultures for *Staphylococcus haemolyticus*. He was treated for bacterial conjunctivitis (9/04/2016, 9/19/2016, and 10/15/2016). A dilated eye exam (9/09/2016) showed stage 2 retinopathy of prematurity, Zone III, which was treated with laser photocoagulation (9/09/2016 and 9/23/2016). Elective extubation was performed again (9/12/2016), with worsening respiratory pattern (9/21/2016) and reintubation. The patient also underwent bilateral herniorrhaphy with orchiopexy (9/22/2016). He was definitively extubated (10/15/2016) and maintained a good respiratory pattern on NIV, despite respiratory stridor and chest X-ray showing atelectasis on the right lung. On 10/19/2016, he was transferred from the ICU to the ward in good general condition, breathing spontaneously in ambient air, weighing 2310 g and using a nasogastric catheter, with good progression to oral diet.

Echocardiography was conducted again (10/03/2016), with the following results: POF 2.5 mm, PCA 2.5 mm, and normal ejection fraction; electroencephalography (11/08/2016) showed epileptiform activity in the right middle temporal region and diffuse disorganized pattern. The patient progressed uneventfully in the ward, the nasogastric catheter was removed (11/10/2016), and the infant was discharged (11/11/2016). At the time of this report, he is doing well and followed up at the outpatient clinic after discharge from the ICU.
IMR requires surgical correction before volvulus occurs, but diagnosis rarely occurs during this period. Abdominal Doppler ultrasonography permits locating the superior mesenteric vessels and checking the position of the vein in relation to the artery, which has shown promising results for early diagnosis.
REFERENCES