Ileal Atresia with Meconium Peritonitis: Fetal MRI Evaluation

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ABSTRACT

We report a case of ileal atresia with meconium peritonitis evaluated by fetal MRI. Prenatal ultrasounds in the third trimester initially demonstrated a cystic abdominal mass that resolved with development of dilated bowel loops. Fetal MRI at 32 weeks gestation identified a perihepatic collection with several dilated small bowel loops and normal sized meconium filled rectosigmoid consistent with distal bowel perforation and loculated meconium peritonitis. Following delivery, the infant presented with bowel obstruction. Contrast enema revealed a normal sized rectosigmoid with small ascending and transverse colon. A distal ileal atresia type IIIa was documented at surgery.

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CASE REPORT

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A 28-year-old Hispanic G3P2 female was referred at 32 4/7 weeks of gestation for further evaluation of cystic abdominal mass seen on fetal sonographic examination. By report, at 30 weeks an abdominal mass had been identified. This mass resolved on follow up with dilated bowel loops developing. Follow up ultrasound and fetal MRI were requested at our prenatal center for further assessment. Ultrasound revealed a poorly defined echogenic collection adjacent to the right lobe of the liver (Figure 1). No shadowing calcifications were identified. Several small bowel loops were dilated, there was no free fluid, and amniotic fluid was normal. Fetal MRI demonstrated a loculated high signal perihepatic collection measuring 4.7 x 3.6 x 1 cm most consistent with a meconium pseudocyst (Figure 2). Several dilated fluid filled loops of bowel high signal on T2w were identified. The rectosigmoid was distended with T2-weighted low signal, T1-weighted high signal material consistent with meconium. A diagnosis of distal jejunal or proximal ileal atresia with meconium peritonitis was suggested. The family consulted with surgery, and recommendations were made for close follow up and transfer to our pediatric hospital after delivery.

Premature labor resulted in a repeat Caesarean section at 34 6/7 weeks of gestation. Apgars were 8 and 9 at 1 and 5 minutes, respectively and the male infant weighed 2511 grams. The infant was noted to have abdominal distension and developed bilious emesis soon after. The patient was transferred to our children's hospital for further evaluation. The scout radiograph for a contrast enema performed on the second day of life (Figure 3) showed several loops of dilated small bowel. Contrast demonstrated a large meconium plug in the rectosigmoid region with the remaining colon small in caliber consistant with a microcolon containing several smaller proximal meconium plugs. No contrast refluxed into the small bowel. Abdominal exploration revealed a mesenteric defect measuring 4 cm in length through which several loops of small bowel had herniated. The distal segment of small intestine was dilated and bulbous with a focal point of perforation at the atretic site. Distal ileal atresia type IIIa was diagnosed and repaired successfully.

DISCUSSION

Vascular compromise of the developing bowel in utero may lead to atresia or stenosis. The initiating event leading to atresia may be identified in roughly a quarter of cases and may include hypotension, vascular accident, volvulus, malrotation, intussusception, meconium ileus and internal hernia (2). Atresia occurs most commonly within the jejunum or ileum with an approximate incidence of 2.5 in 10,000 live births (1). Ileal atresia frequently presents in the newborn with severe abdominal distension and bilious vomiting. Intestinal obstruction is now more frequently diagnosed prenatally with dilated bowel loops and polyhydramnios in up to 25% of cases (2). The dilated bowel loops are present proximal to the atretic site and microcolon is occasionally noted distal to the site of obstruction depending on how distal the obstruction is. When the obstruction is in the proximal jejunum, succus entericus continues to be produced distal to the obstruction entering the colon which is normal in size. If the obstruction is in the distal ileum, no succus entericus enters the colon resulting in a microcolon Meconium peritonitis - a chemical peritonitis secondary to intestinal perforation in utero - is a complication of ileal atresia, occurring in 5% of cases with a mortality of about 5% (2,3). Meconium peritonitis may be diagnosed prenatally by the presence of peritoneal fluid collections, which can loculate and calcify (4). Prenatal assessment is useful for planning delivery and neonatal management.

Our case demonstrates the typical features of meconium peritonitis where meconium from perforated bowel begins to loculate into a cystic collection (4). MRI was useful in confirming the presence of a perihepatic collection that became more difficult to visualize sonographically over time. The constellation of a perihepatic collection and dilated bowel loops suggested the diagnosis of a jejunal or ileal atresia with perforation. Fetal MRI was also useful in the assessment of bowel pathology as supported by other investigations (5). The dilated fluid filled loops of bowel suggested a small bowel atresia while the presence of a prominent rectum filled with meconium (high signal on T1-weighted and low signal on T2-weighted sequences) excluded the diagnosis of meconium ileus and total aganglionosis (Hirschsprung's of the entire colon) which should present with severe microcolon of the colon, sigmoid and rectum (6). Hirschsprung's disease is typically not diagnosed in utero. Postnataly the diagnosis can be suggested when the sigmoid diameter is greater than the rectal diameter (rectosigmoid ratio less than one) . Perhaps in the future this diagnosis may be suggested by assessing the rectosigmoid ratio by fetal MRI (7). Fetal MRI's role in improving the specificity of prenatal ultrasound in the detection of meconium peritonitis and other bowel abnormalities in utero such as microcolon is now being recognized in the literature (8).

While prenatal ultrasound is extremely sensitive in detecting cystic masses, the differential remains broad including ovarian cyst, duplication cyst, mesenteric cyst, meconium pseudocyst, hepatic cysts or renal cysts (8). Zangheri et al. explain the limitation of prenatal ultrasound detection of calcifications which "are an ultrasonographic entity, leaving the obstetrician with scant data to use in counseling the parents regarding its prognosis" (9). For this reason, fetal MRI can be a useful adjunct in the further delineation of the location of the cyst and can aid the assessment of other abnormalities.

TEACHING POINT

Meconium pseudocyst can be a secondary complication of ileal atresia in utero due to leakage of meconium into the peritoneal cavity. Fetal MRI is a useful adjunct in the assessment of abdominal cysts and bowel pathology, and in planning delivery and postnatal intervention including the need for postnatal imaging and surgical exploration.

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FIGURES

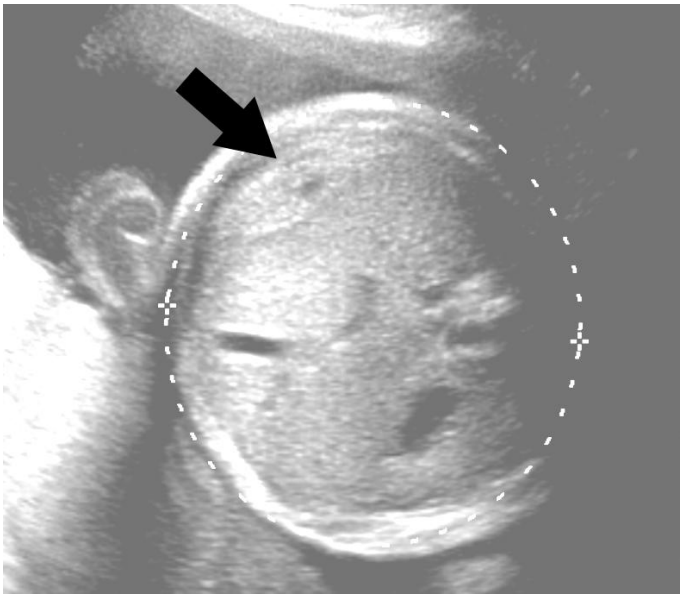
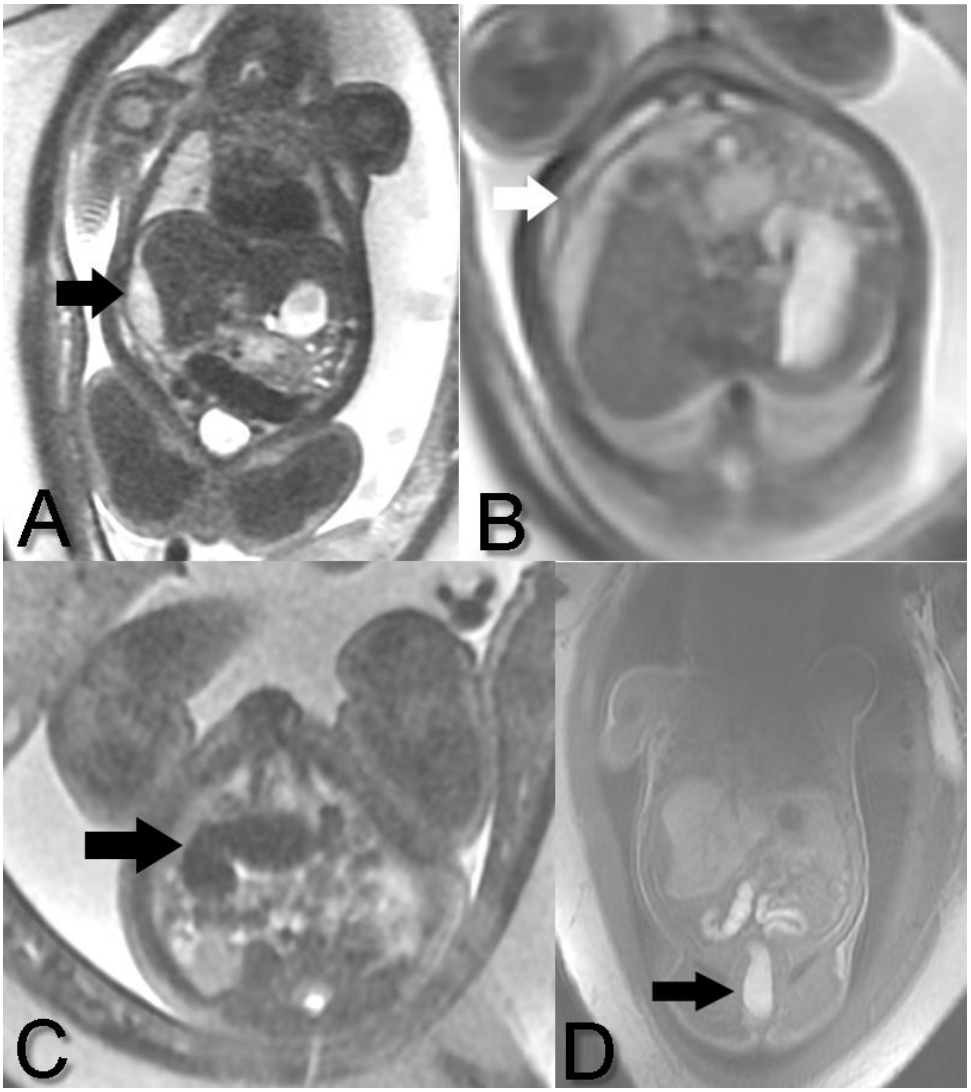


Figure 1: Ultrasound. 28 year old woman at 32 4/7 weeks gestational age underwent prenatal sonographic examination. Sonographic imaging (using 3.5 MHz transducer) demonstrates an ill-defined perihepatic region of inhomogeneous increased echogenicity (arrow) in the fetal abdomen.

Figure 2 (bottom): MRI. 28 year old woman at 32 4/7 weeks GA underwent fetal MRI using a 1.5T scanner. T2 weighted imaging (TR: 5000, TE 160) A perihepatic collection of fluid, is seen on coronal (A) and axial (B) images consistent with a loculated meconium pseudocyst (arrows). Mildly dilated fluid filled small bowel loops were present as well. (C) Axial image of the pelvis demonstrates a low signal meconium-filled dilated rectosigmoid colon. (D) Coronal T1 weighted sequence (TR:100, TE: 4.2) confirms the presence of a distended meconium filled rectum excluding the diagnosis of microcolon from total colonic aganglionosis.



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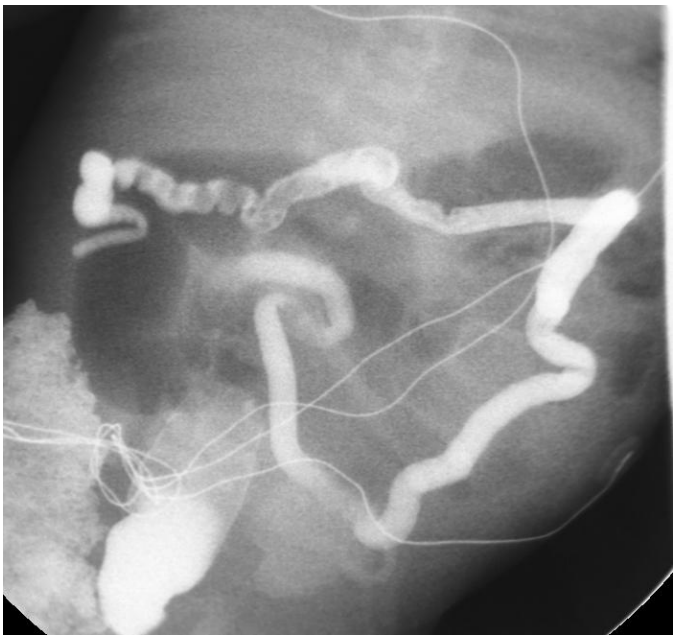

Figure 3: Fluoroscopy. 2 day old infant with microlon. Postnatal contrast enema using 100 cc of Visipaque 270 and 20 cc of normal saline demonstrates a normal sized rectum but microcolon of the sigmoid and remaining colon. Meconium plugs fill the transverse colon. Dilated air filled loops of small bowel are noted as well, consistent with small bowel obstruction. Findings suggest a distal small bowel atresia as the normal sized rectum makes meconium ileus unlikely.

ABBREVIATIONS

GA - gestational age
MRI - magnetic resonance imaging

KEYWORDS

Ileal atresia, meconium peritonitis, fetal abdominal masses, fetal MRI

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