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Case report

Isolated fetal ascites as a presentation of intestinal atresia

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Abstract

Isolated fetal ascites is a rare condition and its natural history needs further research. We describe a case of progressive fetal isolated ascites diagnosed at 22 weeks of gestation associated with hyperechogenic bowel in a primiparous woman. Genetic abnormalities, infection and metabolic causes were excluded. An elective C-section was performed at 29 weeks of gestation after no clinical improvement was obtained with paracentesis. After birth, X-ray and ultrasonography showed high-volume ascites and enlarged liver. On the 5th day of life, no meconium had passed and an abdominal X-ray was repeated: pneumoperitoneum was diagnosed.

Laparotomy identified intestinal atresia type IIIB associated with meconial peritonitis and a derivative jejunostomy was performed. There were complications caused by late-onset sepsis and necrotizing enterocolitis. Reconstruction of jejunostomy was successfully performed on day 114. At 15 months of age, the infant is in good health and has normal psychomotor development.

Keywords

Intestinal atresia, ascites, echogenic bowel, peritonitis, pneumoperitoneum, preterm infant.

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Introduction

Isolated fetal ascites is a rare condition and is defined as fluid accumulation in the abdominal cavity without fluid accumulation in other body cavities or involvement of subcutaneous tissue. It is a separate entity from hydrops fetalis and although the causes and outcome for hydrops fetalis have been well described and documented, the natural history of isolated ascites is not well understood [1, 2].

Case report

Isolated fetal ascites associated with hyperechogenic bowel was diagnosed at 22 weeks of gestation in a 28-year-old primiparous woman with asymptomatic microcephaly. She used cannabis in the first weeks of pregnancy before she knew she was pregnant. Other family history was irrelevant.

Infection screening was negative; karyotype and array were normal; Niemann-Pick disease and peroxisomal and lysosomal storage diseases were excluded. Genitourinary and cardiac abnormalities were also excluded by ultrasonography.

At 28 weeks, paracentesis was performed due to worsening of ascites, with drainage of 215 ml of non-chylous yellow fluid. The quality of the ascitic fluid sample sent for analysis was considered inadequate. As there was no clinical improvement, an elective C-section was performed at 29 weeks of gestation. Apgar score was 7/8/9, weight was 1,510 g (percentile 92), height 37.5 cm (percentile 57) and head circumference 27 cm (percentile 92).

X-ray and ultrasonography after birth showed high-volume ascites and enlarged liver. Paracentesis was performed, confirming a transudate without other chemical disturbances.

Non-invasive ventilation was started at birth; however, on the 2nd day of life, invasive ventilation was required for increased oxygen requirement and abdominal distension. Because no bowel movement was registered until the 5th day of life, an abdominal X-ray was repeated and pneumoperitoneum was diagnosed (**Fig. 1**).

Exploratory laparotomy revealed type IIIB jejunoileal atresia associated with meconial peritonitis; derivative jejunostomy was performed (**Fig. 2**). On day 45, due to worsening of abdominal distension, a new surgical intervention was required. As a result, fecal peritonitis with transmural necrosis and pneumatosis was diagnosed. The use of an antibiotic for an extended period was necessary for late-onset sepsis and necrotizing enterocolitis.

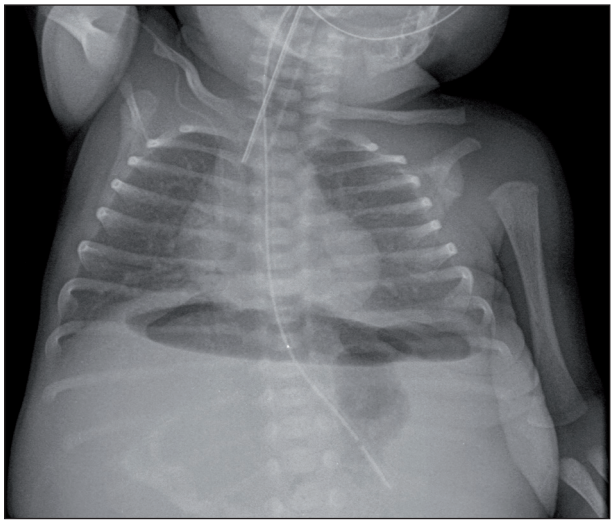

Figure 1. Abdominal X-ray on the 5th day of life revealing pneumoperitoneum.

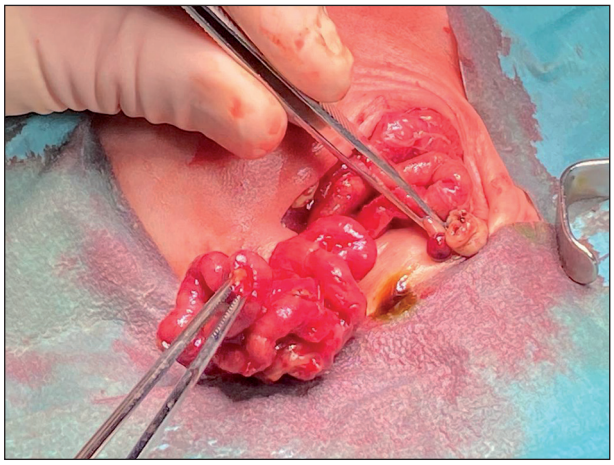

Figure 2. Picture of the exploratory laparotomy revealing type IIIB jejunoileal atresia.

Reconstruction of jejunostomy was successfully performed on day 114 of life.

Cystic fibrosis and metabolic diseases were excluded. The infant presented comorbidities of moderate bronchopulmonary dysplasia, anemia needing multiple blood transfusions and cholestasis.

She is now 15 months old and has adequate growth and psychomotor development.

Discussion

The prevalence of isolated fetal ascites is not clear. The use of a systematic protocol for diagnostic work-up should be followed, as an etiology or associated disorder could be determined in 84% to 92% of cases [1, 3]. The etiology includes intrauterine infections, chromosomal and metabolic anomalies, structural anomalies involving the gastrointestinal, genitourinary or cardiac system

and abnormal lymphatic drainage or congenital lymphatic dysplasia [1-3]. Catania et al. identified a high prevalence of fetal structural anomalies; among them, gastrointestinal anomalies accounted for 33% of cases, followed by urinary anomalies in 21.5% of cases and cardiac anomalies in 13.7% of cases [3]. Meconium peritonitis secondary to bowel obstruction and perforation is the most common gastrointestinal problem causing isolated fetal ascites and intestinal atresia should be included in the differential diagnosis [1-4]. In rare cases it can be transitory, resolving pre- or postnatally [2, 3, 5, 6].

The detection of isolated fetal ascites before the 24th week of gestation is associated with an increased risk of intrauterine demise and adverse postnatal outcome. When the diagnosis is made after this gestational age it is associated with an increased risk of gastrointestinal diseases, especially meconium peritonitis [1, 2]. In this case, the diagnosis was reported prior to 24 weeks of gestation and was presented as meconium peritonitis.

While previous studies suggest that prenatal paracentesis may reduce the abdominal circumference and facilitate neonatal resuscitation, more recent studies have found it to be questionable [1, 7, 8]. In this case report there was no improvement with paracentesis.

The prognosis for isolated fetal ascites depends on the underlying pathology but generally has been reported to be better than for hydrops fetalis. Zelop and Benacerraf described survival rates as high as 94% and El Bishry 80% [1, 2, 9].

The incidence of intestinal atresia ranges from 1.3 to 3.5 per 10,000 live births, of which 20% are associated with a chromosomal anomaly [10, 11]. Jejunal or ileal atresia occurs in approximately 0.7 per 10,000 births, each representing 20% of small intestine atresias [10-12]. Colonic atresia should also be included in the differential diagnosis of isolated fetal ascites [4].

Type III jejunoileal atresia is the most common and typically is an acquired lesion thought to result from vascular disruption leading to ischemic necrosis of a segment of the fetal intestine [9]. It may present prenatally with polyhydramnios, ascites, dilated bowel loops and increased bowel echogenicity [10, 13, 14].

Affected infants typically develop abdominal distension and vomiting in the first 2 days after birth and most of them fail to pass meconium [10, 12]. Osmulikevici et al. also described a case of isolated fetal ascites in a newborn with apple peel jejunal atresia and we found no other case in the literature [15].

Fewer than 5% of infants with jejunoileal atresia have associated chromosomal anomalies [10, 15, 16]. It can be associated with cystic fibrosis, gastroschisis, cardiac malformations, immunodeficiency, midgut volvulus during gestation, use of vasoconstrictive medications during pregnancy and inherited thrombophilia. It also can occur in families [10, 17]. In the present case, it should be noted that the mother used cannabis in the first weeks of pregnancy before she knew she was pregnant and previous studies describe the causal association between cannabinoid consumption and intestinal atresia [18, 19].

The treatment is surgical, and prognosis depends on the initial vascular compromise and remaining bowel length and the occurrence of postoperative complications. Most mortality occurs in infants with medical conditions such as prematurity or respiratory distress syndrome, associated anomalies, or complications such as short gut syndrome [10, 15, 16].

Declaration of interest

The Authors declare that there is no conflict of interest.

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