Intestinal Perforation in the Context of Thoracoamniotic Shunting and Congenital Diaphragmatic Hernia

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Abstract

A fetus was diagnosed by prenatal ultrasound with bilateral intrauterine pleural effusions that were subsequently drained in utero by insertion of bilateral thoracoamniotic shunts. Serial prenatal ultrasound scans were consistent with a left-sided diaphragmatic hernia. On the first day of life, the infant underwent an exploratory laparotomy for intestinal obstruction, with radiographic findings of pneumatosis intestinalis. Intraoperative findings were suggestive of prenatal bowel and diaphragm perforation, which might have occurred as a complication of thoracoamniotic shunting.

Keywords

- intestinal perforation
- thoracoamniotic shunt
- diaphragmatic hernia

Introduction

Fetal pleural effusions are uncommon and are detected prenatally with an incidence rate of approximately 1 in 10,000 to 15,000 pregnancies. They are a nonspecific sign and are frequently associated with congenital malformations, chromosomal abnormalities, chylothorax, anemia, heart defects, cardiac arrhythmias, and viral infections.[1] Congenital diaphragmatic hernia (CDH) has an incidence of 1 per 2,000 to 3,500 deliveries. Left-sided CDH commonly contain stomach, bowel, and spleen and is associated with a generalized pulmonary hypoplasia.[2] The clinical presentation depends on the extent of pulmonary hypoplasia and may vary from cyanosis and respiratory distress immediately after birth to late presentation of respiratory distress in the first few days of life when the herniated bowel loop fills with air and compresses the ipsilateral lung parenchyma.[2] Typical signs include abnormally sited cardiac impulse, absent breath sounds, and scaphoid abdomen. The diagnosis is commonly made on prenatal ultrasound and confirmed on postnatal chest radiography. Medical management consists of initial resuscitation with a view to avoid face mask ventilation, preoperative stabilization, and ventilatory care aiming in reducing the risk of volutrauma and persistent pulmonary hypertension of the newborn, while the surgical treatment consists of reduction of the herniated contents and closure of the diaphragmatic defect.

Case Report

The mother was a 38-year-old, gravida III, para II with an uncomplicated previous obstetric history. Early anomaly ultrasound scan in a quaternary level fetal medicine unit demonstrated hydrothorax with bilateral pleural effusions. At 18 weeks of gestation, bilateral thoracoamniotic shunts were inserted. A repeat ultrasound scan at 33 weeks of gestation showed increased amniotic fluid with a deepest

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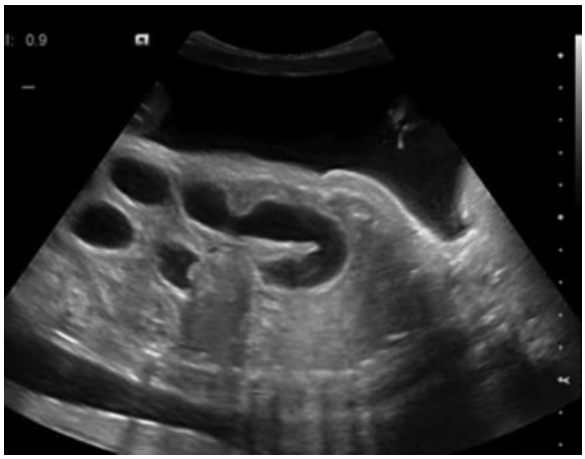

Fig. 1 Fetal ultrasound scan performed at 34 weeks. Note loops of the small bowel visualized in the chest cavity.

vertical pool of 9 cm; suspicion of oesophageal atresia was raised. Subsequent ultrasound scan performed at 34 weeks demonstrated mediastinal shift with the heart axis deviated to the right and loops of the small bowel and the stomach were visualized in the chest cavity. The findings were consistent with left-sided CDH (►**Fig. 1**). The fetal liver was below the diaphragm. The increased amniotic fluid did not warrant amniodrainage. Planned delivery was booked at 38 weeks; however, delivery was performed at 36 weeks of gestation by emergency caesarean section because of spontaneous onset of labor.

A male infant was born in good condition, cried at birth, and was intubated at 4 minutes of age. The right-sided thoracoamniotic shunt had spontaneously fallen out in utero, whereas the left-sided device was still in situ and immediately clamped to avoid development of a pneumothorax. A wide bore nasogastric tube was inserted, and the infant was electively intubated and transferred to the neonatal intensive care unit. Blood gas analysis revealed a cord arterial pH of 7.44 with a base excess of – 1.0 mmol/L and cord venous pH of 7.38 with a base excess of – 0.8 mmol/L. Umbilical arterial and venous catheters were inserted and intravenous antibiotics were initiated. The infant was ventilated with low pressure and oxygen requirements.

The initial chest and abdominal X-ray that was performed to confirm umbilical catheters positions showed a general opacification of the left hemithorax, interspersed with areas of air, and a solitary dilated loop of bowel below the stomach that contained a wide bore nasogastric tube, with no evidence of perforation. A further abdominal radiograph performed at 14 hours of age revealed dilated central loops of bowel with bubbles of gas suggestive of intramural gas and suspicious for pneumatosis intestinalis (►**Fig. 2**). At this point, intravenous metronidazole was added to the antibiotic regimen. The left-sided prenatally inserted chest drain was removed at 15 hours of age and the wound closed and dressed as per surgical advice. Echocardiography was performed at 17 hours of age and it revealed a structurally normal neonatal heart with no evidence of persistent pulmonary hypertension.

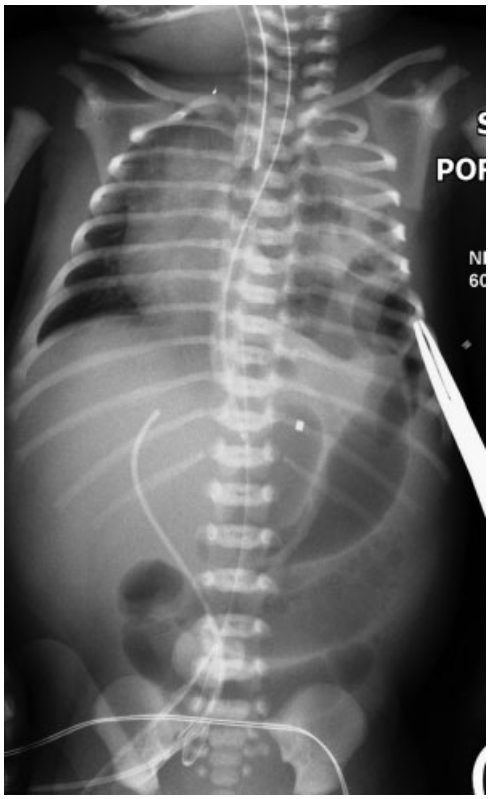

Fig. 2 Chest and abdomen radiograph at 14 hours of age, showing dilated bowel loops with overlying gas bubbles. The square radio-opacity over the 11th rib represents the steel tip at the end of the in utero–inserted shunt.

In view of a developing metabolic acidosis with a base excess of – 8.9 mmol/L, abnormal bowel gas pattern, and possible intramural gas on the abdominal radiograph, the infant was taken to theater at 21 hours of age for exploratory laparotomy, reduction of bowel from the thoracic cavity, and closure of left congenital diaphragmatic defect. During operation, the entire small bowel was found to be located in the chest, and there was a large radial defect in the left hemidiaphragm. The findings were not typical of a CDH that is usually characterized by a defect in the foramen of Bochdalek (a transverse postero-lateral defect). There was no hernial sac, and the lung tissue did not look hypoplastic following reduction of the intestinal contents back into the abdominal compartment. The bowel was matted and coated in prenatal intestinal contents in a manner similar to that seen in meconium peritonitis (but in the chest), the adhesions were sufficient to be causing a high obstruction of the bowel, accounting for the postnatal intestinal obstruction. The surgical procedure consisted of separation of all adhesions, pulling down of the intestine in the abdominal cavity and closure of the diaphragmatic defect. Postoperatively, sedation and paralysis were gradually discontinued, ventilatory support was weaned as tolerated, and the infant was subsequently extubated on the 4th day of life. He was discharged home at day 30 and was followed up in clinic. By 6 months of age, he was clinically well with no recurrent respiratory

symptoms. His follow-up chest X-ray exhibited extensive bilateral consolidation with possible cavitation.

Discussion

Prenatal interventions aiming at draining pleural effusions into the amniotic cavity can be undertaken in utero to allow for normal lung growth and development and to prevent caval compression and development of hydrops fetalis.[3] Fetal thoracoamniotic shunting involves the insertion of a drainage tube though the fetal chest wall into the fetal pleural cavity allowing continued drainage of fluid from the pleural effusion into the amniotic cavity. It is performed under local anesthesia and ultrasound guidance. Effective drainage leads to lung expansion and resolution of hydrops and polyhydramnios.[4] Recognized intrauterine complications include unilateral arm edema,[5] intrathoracic displacement of the shunt, preterm birth and fetal loss.[4]

Neonatal intestinal perforation might happen spontaneously or secondary to preexisting gastrointestinal pathology such as duodenal atresia, pyloric atresia, malrotation, tracheoesophageal fistula or diaphragmatic abnormalities such as diaphragmatic eventration.[6] Intestinal perforation complicating CDH is a rare condition. When it occurs, it usually represents late-presenting cases that happen during childhood or early adult life.[7] Manning et al[8] have reported a series consisting of five cases of CDH presenting with gastrointestinal complications, one of whom presented in the neonatal period with intestinal perforation. A case of a fetus with gastric perforation associated with CDH has also been reported.[6] It has been speculated that the mechanical activation of the gastrointestinal tract and swallowing of air that occur after birth might lead to deterioration of blood supply to the herniated bowel by enlargement of the bowel volume, thus damaging the bowel wall by means of ischemia and mechanical pressure.[6] Findings of meconium peritonitis, characterized by calcifications, fetal ascites, and pseudocyst are generally attributed to prenatal intestinal perforation.

The presence of bilateral pleural effusions at anomaly scan might have been an initial sign of diaphragmatic hernia, which can be visualized by ultrasound before other signs arise. Attempting to drain them possibly resulted in intestinal perforation within the chest cavity.

Alternatively, the operative findings raise the suspicion that the prenatal intervention might have resulted in a traumatic diaphragmatic perforation on the left-hand side. This was then possibly complicated by bowel migrating up through the defect into the left hemithorax resulting in intestinal perforation, and possible meconium peritonitis.

On the contrary, gastric or intestinal perforation due to incarceration is not unusual in CDH. Several cases have been reported in the literature.[6,9–11]

Clearly, it is impossible to distinguish whether these findings should be attributed to a mechanical complication of diaphragmatic hernia or to an intrauterine intervention, as no published data have been found in the literature linking intrauterine thoracoamniotic shunting to intestinal perforation or diaphragmatic penetration.

The authors think that it is important to highlight the potential differential diagnosis that might result in the context of prenatal thoracoamniotic shunting and CDH.

Conflict of Interest

None

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