CASE REPORT

Prenatal Diagnosis and Management for Congenital Intrapericardial Diaphragmatic Hernia with Massive Cardiac Effusion: A Case Report and Literature Review

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Received 12 June 2014; accepted 10 July 2014
Available online 28 November 2014

KEY WORDS
cardiac effusion, intrapericardial diaphragmatic hernia, prenatal diagnosis

Congenital intrapericardial diaphragmatic hernia with massive pericardial effusion is a rare type of Morgagni hernia. Since 1980, there have been only 16 reported cases. We report on the imaging features of such a case that was diagnosed in utero. The prognosis of congenital intrapericardial diaphragmatic hernia is better than the other types of congenital diaphragmatic hernia, but lung hypoplasia due to compression by the pericardial effusion is not uncommon. Early intervention and treatment should be given to improve the perinatal outcome once the prenatal diagnosis has been made. We have summarized current diagnostic methods and management for this rare phenotype, after reviewing previous case reports and articles relating to the intervention for congenital diaphragmatic hernia.

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Introduction

Congenital intrapericardial diaphragmatic hernia with massive pericardial effusion is a very rare phenotype of congenital diaphragmatic hernia (CDH). Since 1980, there have been only 16 cases reported [1–7]. One common feature of all cases is an anterior diaphragmatic defect, or Morgagni hernia, which accounts for only 2% of CDH. Lung
hypoplasia due to compression from the massive pericardial effusion is often found, and this necessitates early intervention to improve the perinatal outcome. After reviewing previous case reports and related articles, we have summarized the current diagnostic methods and management for this rare phenotype.

Case report

A 26-year-old primigravida woman was referred to our hospital for fetal body fluid accumulation and suspected anomaly at the 22nd week of her pregnancy. The level II ultrasound examination showed a fetus with no detectable cardiac anomaly except for bilateral pleural effusion and possible diaphragmatic hernia. Maternal blood test showed no sign of infection, and cordocentesis showed normal karyotype (46, XY). The patient and her family opted to proceed with the pregnancy after consulting with the genetic and perinatal team. A fetal ultrasonography at the 27th gestational week highly suspected right-sided diaphragmatic hernia; the result of the rest of the prenatal examination was normal. At the 30th gestational week, ultrasonography showed a massive pericardial effusion with the liver herniating into the pericardial space pushing the heart laterally and the lung posteriorly (Fig. 1). The measured lung area/head circumference ratio (LHR) of the fetus was 1.2. The heart showed no sign of cardiac tamponade or malformation, and the lung was in normal development with no compression. After counseling was given to the patient and her family, it was decided that no intervention would be given during the prenatal course of the fetus without performing pericardiocentesis. Instead, a corrective surgery in the early neonatal period was offered to avoid the preterm delivery of a low-birth-weight infant.

At the 37th week, the patient developed premature rupture of the membrane and came to the hospital for delivery. The fetal monitor showed normal findings; however, the labor course was prolonged with no progression, and so a Cesarean section was performed. A 2950-g male baby was delivered with Apgar score 1 at 1 minute and Apgar score 5 at 5 minutes. An endotracheal tube was immediately inserted, and the newborn was transferred to the neonatal intensive care unit. A chest film of the newborn on Day 2 after birth showed increased opacity at the anterior mediastinum and herniated mass on the right side. The abdominal computed tomography showed moderate pericardial effusion, herniated anterior segment of the right lobe of the liver, the left lobe of the liver, and parts of the hepatic flexure of the colon into the right chest (Fig. 2). Surgery was arranged, and a huge retrosternal defect of the diaphragm (7 cm x 5 cm) was found (Fig. 3). The surgical procedures included repositioning of the liver and closure of the defect. Two days after the operation, the baby was successfully weaned from the ventilator. He was discharged 7 days later with stable vital signs. No other complications have been reported to us during follow-up; he recently celebrated his 6th birthday.

Discussion

CDH occurs in about one of every 2400 live-births, with a mortality rate ranging from 40% to 90%. The prognosis of
CDH is determined by the severity of the pulmonary hypoplasia and associated cardiac anomaly. The two important causes of morbidity and mortality are persistent pulmonary hypertension and persistent fetal circulation. Normally, the lung area increases four times more than the head circumference between the 12th gestational week and 32nd gestational week [9]. The effect of gestational age on lung size measurement can be taken into account by expressing the measured LHR of the index case (observed) over the appropriate normal mean (expected) for gestation of the same side lung. Unfortunately, LHR is not an early predictor; studies have shown that the best validated prognostic indicator of CDH is the observed/expected LHR, which is determined between the 32nd gestational week and 33rd gestational week [8].

Among the various theories about the formation of CDH, Allan and Greer [10] proposed a plausible explanation for Morgagni hernia, which states that failure of closure of the pleuropерitoneal folds is the culprit. The medially extended folds fail to fuse with the primary esophageal mesentery, the dorsal portion of the liver, and septum transversum, allowing the possibility of liver herniation through the peritoneopericardial canal into the pericardial space and resulting in pericardial effusion. Explanations for the pericardial effusion include transudation from congested viscera, lymph accumulation due to thoracic duct compression, and mechanical irritation of the pericardial lining [1].

Nine cases of intrapericardial diaphragmatic hernia had been reported between 1980 and 2002 [10]. Eight more cases, including ours, have been reported thereafter (Table 1) [1–7]. Summarizing these 17 cases, none had cardiac tamponade despite the massive pericardial effusion; this may be attributable to the slow accumulation process to allow continuous distension of the fetal pericardium [2]. Six cases showed signs of lung hypoplasia; two cases were electively terminated, two cases succumbed perinatally shortly after birth, and two cases survived. Comparing the two cases who have survived, Antinolо et al [3] performed two inutero pericardial fluid aspirations at the 21st gestational week and 23rd gestational week and successfully corrected the lung hypoplasia, whereas Kanamori et al [1] performed only one inutero fluid aspiration, and persistent pulmonary hypertension occurred and lasted for 4 days postpartum. Surgical repair of the diaphragmatic hernia was usually performed in the early neonatal period, but Ikeда et al [11] showed that it can be postponed later to 70 days postpartum if low birth weight is a cause for concern. Including our case, 10 have been diagnosed in utero.

Before making this diagnosis, other differentials such as diaphragmatic eventration, intrapericardial teratoma, trauma, or infection should be ruled out. Doppler ultrasound is a useful tool to investigate the content of the intrapericardial mass; if it is the liver, the hepatic vessels can be seen to continue from below the diaphragm up into the mass. Magnetic resonance imaging has a higher specificity than ultrasound in diagnosing CDH [8], and Ikeда et al [11] used it to quantify the pericardial effusion. Chest X-ray and computed tomography can be arranged perinatally; in our case, increased opacity over the anterior mediastinum and widening mediastinum on the postnatal chest film are the diagnostic signs. Alternatively, Kanamori et al [1] used an umbilical catheter to trace the liver into the thoracic cavity.

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**Table 1** Reported cases of congenital intrapericardial diaphragmatic hernia with cardiac effusion.

<table>
<thead>
<tr>
<th>Prenatal diagnosis</th>
<th>Lung hypoplasia</th>
<th>Cardiac finding</th>
<th>Outcome</th>
<th>Operation (d)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kanamori et al [1]</td>
<td>27 wk</td>
<td>+</td>
<td>PDA</td>
<td>Alive 4</td>
</tr>
<tr>
<td>Filly et al [2]</td>
<td>37 wk</td>
<td>–</td>
<td>VSD</td>
<td>Alive 8</td>
</tr>
<tr>
<td>Antinolо et al [3]</td>
<td>22 wk</td>
<td>+</td>
<td>–</td>
<td>Alive 0</td>
</tr>
<tr>
<td>Hara et al [4]</td>
<td>18 wk</td>
<td>+</td>
<td>Ventricle aneurysm</td>
<td>Aborted</td>
</tr>
<tr>
<td>Jain et al [6]</td>
<td>24 wk</td>
<td>+</td>
<td>–</td>
<td>Aborted</td>
</tr>
<tr>
<td>Akalin et al [7]</td>
<td>Postpartum 5 wk</td>
<td>+</td>
<td>–</td>
<td>Alive 30</td>
</tr>
<tr>
<td>Haino et al [5]</td>
<td>26 wk</td>
<td>–</td>
<td>–</td>
<td>Alive 21</td>
</tr>
<tr>
<td>Our study</td>
<td>30 wk</td>
<td>–</td>
<td>–</td>
<td>Alive 0</td>
</tr>
</tbody>
</table>

PDA = patent ductus arteriosus; VSD = ventricular septum defect.
As in common CDH, lung development is of the most important concern. Fetoscopic tracheal occlusion has been used to stimulate lung growth [8]; a balloon is percutaneously inserted into the trachea at the 26th–28th gestational weeks, and reversal of occlusion is planned at the 34th gestational week. The procedure carries a risk for preterm premature rupture of membrane. Kern et al [12] recently proposed ex utero intrapartum treatment to be used in intrathoracic volume expansion, extending its original use to guarantee sufficient oxygenation for a fetus at risk of airway obstruction; however, intraoperative maternal hemorrhage should be monitored because of the prolonged uterine relaxation. Recently, Antin˜olo et al [3] performed two fetal pericardiocentesis for pericardial fluid aspiration and successfully rescued the fetus from the fate of lung hypoplasia. All these methods shed light on the management for lung hypoplasia on future encounters.

In conclusion, lung development is the most important concern in these fetuses. Besides Doppler ultrasound, several instruments can be used to investigate the fetal condition and whether the lung is being compressed by the massive pericardial effusion. If lung hypoplasia is found, early intervention and treatment aiming to improve lung growth should be given to improve the fetal outcome.

References


