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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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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.

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

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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 $\times$ 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% [8]. The prognosis of

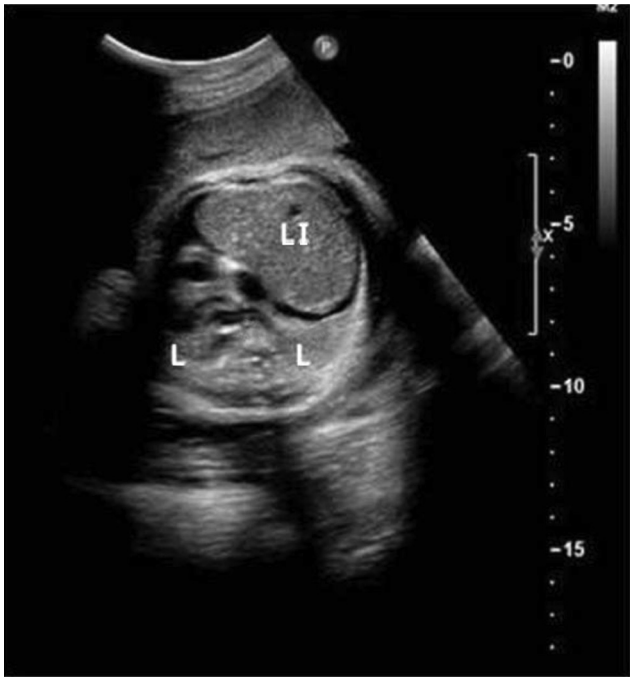

Fig. 1 Transverse view of the fetal thorax at 30 weeks of gestation shows deviation of the heart by the herniated liver (LI). Pericardial effusion is also present in this section without evidence of lung (L) compression.

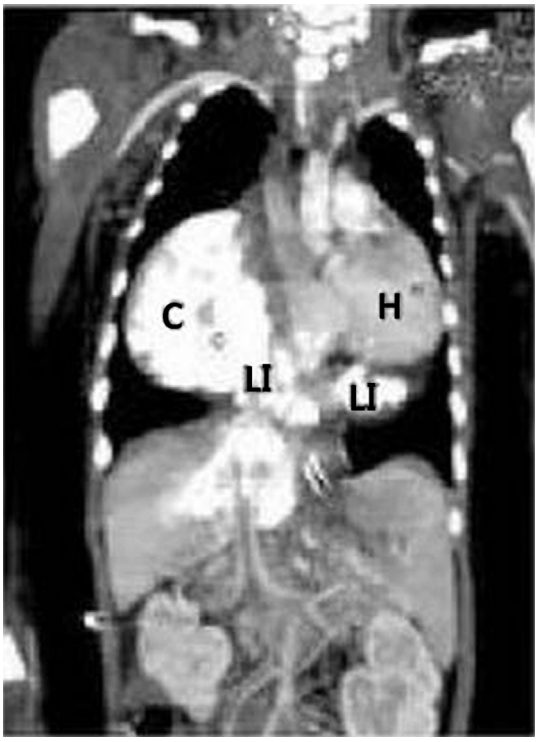

Fig. 2 Day 2 after birth, computed tomography reveals that the herniated content includes the left lobe and the anterior segment of the right lobe of the liver and parts of the hepatic flexure of the colon. C = colon; H = heart; LI = liver.