Prenatal and neonatal sonographic imaging of a central diaphragmatic hernia

Abstract A case of a central diaphragmatic hernia diagnosed prenatally is reported. The prenatal sonographic findings included central herniation of most of the liver into the chest and hydrops. The hernia was successfully repaired. However, the infant died secondary to respiratory distress syndrome.

Introduction

This report describes the prenatal and neonatal ultrasound findings in an infant with a congenital central diaphragmatic hernia, the rarest type of hernia. We found no previous description of a central diaphragmatic defect in a fetus.

Case report

A 24-year-old G5P1 woman presented at 22 weeks of gestation for fetal sonography. Marked bilateral hydronephrosis and bilateral pleural effusions were found. The liver was herniated into the chest in a central fashion; the dome of the liver in the right hemithorax was just inferior to the thoracic apex (Fig. 1 a). The left lobe of the liver was also herniated into the left chest, although not as severely as the right lobe (Fig. 1 b). The diaphragm was not identifiable. The stomach was identified in the left upper quadrant of the abdomen.

The heart was deviated to the left. Fetal echocardiography revealed a partial atrioventricular canal defect. There was mild polyhydramnios. Amniocentesis revealed a 46 XY karyotype. The mother refused termination of pregnancy. A male infant was delivered via cesarean section at 33 weeks of gestation due to poor variability on the fetal heart tracing. He was intubated and treated for respiratory distress syndrome.

X-ray of the abdomen and chest after birth demonstrated a large central mass silhouetting the heart and filling the entire thorax (Fig. 1 e). An ultrasound examination performed on the day of birth revealed that the entire liver was herniated into the chest. The heart was displaced posteriorly. The pericardium was not identified. Intra-abdominal ascites communicated with fluid seen around the heart and bilateral pleural effusions (Fig. 1 d). Only the posterolateral portions of both hemidiaphragms were identified, seen as echogenic, discontinuous, curvilinear structures (Fig. 1 e). The spleen and stomach were seen in their normal locations in the left upper quadrant of the abdomen, beneath the visualized portion of the left hemidiaphragm. Also noted was mild bilateral hydronephrosis.

At 7 days of age, the infant underwent operative repair via a bilateral subcostal incision. Operative findings included nonrotation of the intestine, herniation of the liver into the chest, and a large amount of peritoneal fluid. The pericardial-peritoneal membrane, which was the hernia sac, was excised, leaving a small posterior rim of diaphragm on both sides. There was no diaphragm anteriorly between the mid-axillary lines. The heart was in the mid mediastinum, and the pleural cavities and phrenic nerves were compressed laterally. The defect was repaired with a 1-mm Gore-Tex patch, after wide opening of both pleural cavities. The infant died at 4 months of age from pulmonary failure. An autopsy was not performed.

Discussion

Congenital diaphragmatic hernia (CDH) occurs approximately once in every 2400 births [1]. The diaphragm has four developmental components: (a) the septum transversum, the earliest component to form; (b) the dorsal esophageal mesentery; (c) the pleuroperitoneal membrane; and (d) the body wall [2].
Fig. 1a  Sagittal ultrasound image of the right fetal chest; dome of the liver (arrow) was adjacent to the thoracic apex, with a small crescent of pleural fluid in the intervening space (arrowhead).
b  Sagittal ultrasound image of the left fetal chest; left lobe of the liver (arrow) was herniated into the thoracic cavity, although not as severely as the right lobe. Pleural effusion was present. c  X-ray of the chest and abdomen reveals a large central intrathoracic mass (arrows). d  Transverse ultrasound image of the neonatal chest demonstrated bilateral pleural fluid which extended around the heart (curved arrow) and communicated with ascitic fluid. The liver (arrow) and gallbladder fundus (arrowhead) were identified at this level. e  Transverse neonatal ultrasound of the right upper quadrant of the abdomen; a small rim of diaphragm was seen posterolateral to the liver, with an abrupt termination (arrows).