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An unusual hernia: congenital pericardial effusion associated with liver herniation into the pericardial sac

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Abstract To our knowledge there have been only two previous cases of diaphragmatic hernia into the pericardium diagnosed antenatally. We describe our pre- and post-natal radiological findings in such a case, although the final diagnosis eluded us until after delivery.

Case report

A 31-year-old primigravida with a history of infertility and normal findings on ultrasound examination carried out at another hospital at 31 weeks gestation was scanned for fetal growth at 37 weeks. Both hemithoraces were found to be transonic apart from a small echogenic area postero-inferiorly, thought to represent lung tissue. A mass was noted immediately adjacent to the right heart border, and its echogenicity was similar to that of the liver (Fig. 1 a). A provisional diagnosis of a right diaphragmatic hernia of the liver with massive bilateral pleural effusions was made.

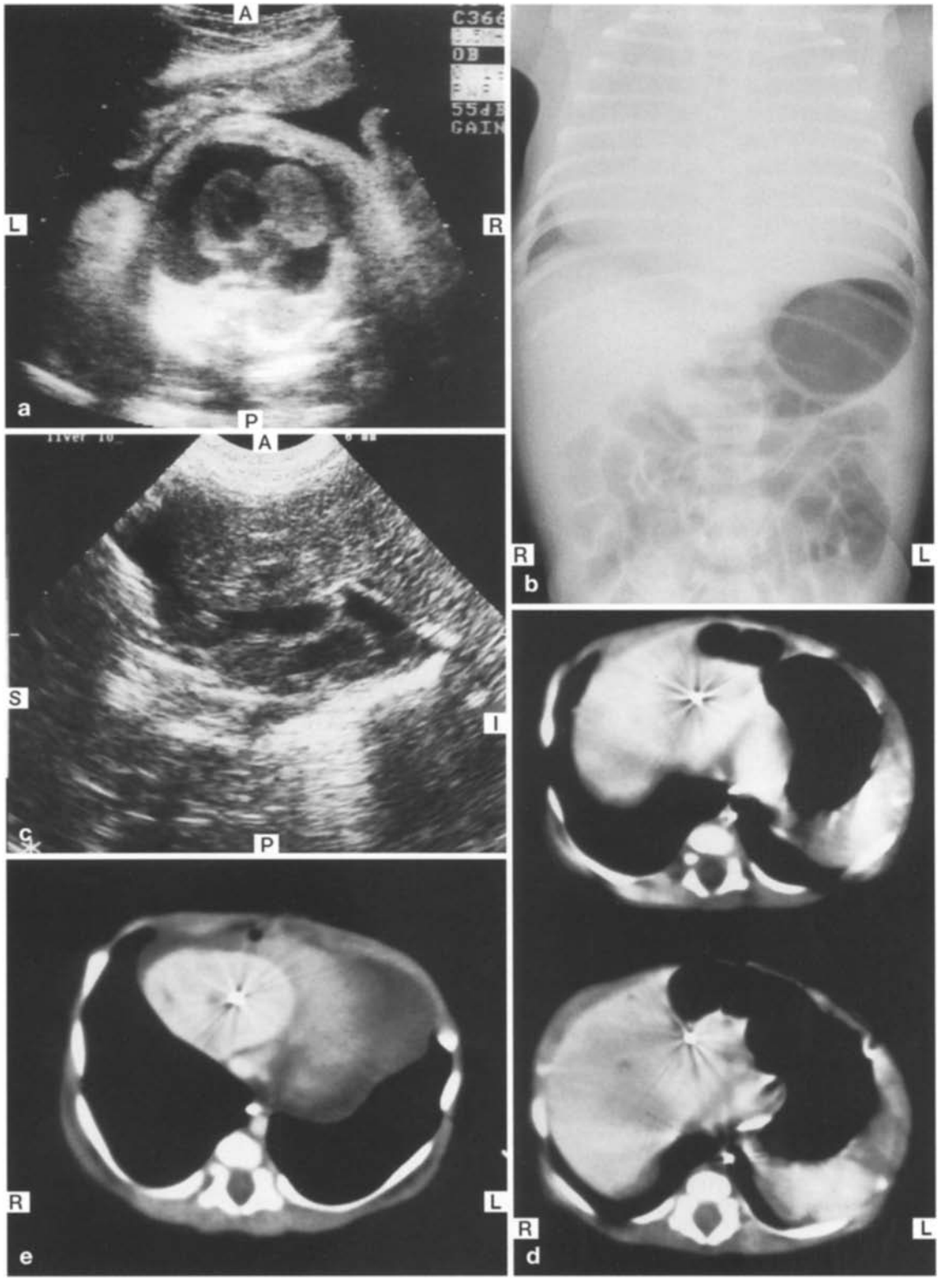
Following discussion with the paediatricians, an elective caesarian section was performed at 38 weeks to avoid intrapartum complications and provide optimal post-natal support. A paediatric team was available to carry out immediate drainage of the effusions. Surprisingly, the Apgar scores at 1, 5 and 10 min were 5, 7 and 9, respectively. The baby only required intermittent positive pressure ventilation by bag and mask for 3 min and proved to be stable subsequently. There were no features of respiratory distress or cardiac tamponade. A chest film shortly after delivery showed massive cardiac enlargement, the appearance being suggestive of a pericardial effusion (Fig. 1 b). The effusion was confirmed on cardiac ultrasound and a mass within the pericardium was identified anteriorly on the right in continuity with the liver and with liver echogenicity (Fig. 1 c). While the possibility of a pericardial tumour was considered, appearances were thought more likely to represent herniated liver. Sixty milliliters of bloodstained serous fluid was obtained on pericardiocentesis. The baby was transferred to the Royal Hospital for Sick Children and a contrast medium enhanced CT of the thorax and upper abdomen confirmed herniation of part of the liver into the pericardial sac. Contrast medium was identified within an umbilical vein catheter, both subdiaphragmatically and within the intrapericardial segment of the liver (Fig. 1 d, e).

At thoracotomy on day 2 a large diaphragmatic hernia into the pericardium was found, containing the right lobe of the liver. A defect in the pericardium measuring 6×6 cm was identified in the central tendon and was subsequently closed. The post-operative follow-up period was uneventful and the infant is thriving normally several months later. Previous similar cases have also had a successful outcome following corrective surgery, perhaps helped by the absence of pulmonary hypoplasia, which is often seen with the more conventional diaphragmatic hernia [1–5].

Discussion

The ultrasound findings were puzzling; the presence of large effusions secondary to liver herniation being difficult to explain. Could the hernia be compressing the thoracic duct causing accumulation of lymph? Had the hernia been present at 31 weeks and been unrecognised? Were the effusions present at that time or had they developed subsequently? The possibility of herniation into the pericardium was not considered.

Congenital diaphragmatic hernia occurs in 1 in 200 to 1 in 5000 live births. Postero-lateral defects are most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◀**Fig. 1** **a** Transverse antenatal ultrasound scan at 37 weeks. Seen from above, a mass is demonstrated adjacent to the right cardiac border with apparent bilateral pleural effusions. *A*, anterior; *L*, left; *P*, posterior; *R*, right. **b** Immediate post-natal chest radiograph demonstrating apparently massive pericardial effusion. **c** Longitudinal scan showing mass of hepatic echogenicity continuous with the liver through the right hemidiaphragm, lying anterolateral to the heart. *I*, inferior; *S*, superior. **d** Umbilical vein catheter in the subdiaphragmatic liver. **e** Umbilical vein catheter within the intrapericardial segment of the liver with associated pericardial effusion

common accounting for 75–80 % of hernias, and left-sided hernias are eight times more common than right-sided. Anterior hernias of the foramen of Morgagni account for 1–68 % of all defects, and bilateral hernias account for around 1 % [1]. Herniation into the pericardial sac is the rarest form with only nine previously reported cases [2–5], and only two diagnosed antenatally [6]. These hernias are believed to represent developmental failure of the retrosternal portion of the septum transversum. This is first observed around the 8th week of fetal life, growing posteriorly to form part of the diaphragm. By the 10th week the fetal coelomic cavity is divided into the thorax and abdomen. Presenting signs of this condition have included respiratory distress and cyanosis soon after birth. Massive pericardial effusion is found but the absence of cardiac tamponade suggests the slow formation of the fluid with compensatory progressive distension of the fetal pericardium. It has been suggested that the development of pericardial fluid is the result of mechanical irritation of the pericardial mass. Venous obstruction in the liver leading to congestion and transudation could also be responsible.

Classically, intrapericardial tumours, although rare and usually benign (teratoma, haemangioma) are associated with congenital pericardial effusions. However, the possibility of a congenital diaphragmatic hernia should also be considered and included in the ultrasonic differential diagnosis, as a correct antenatal diagnosis enables optimal pre-, peri- and post-natal management.

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