

Case Report

A Rare Case of Congenital Anterior Diaphragmatic Hernia

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ABSTRACT

We report a case of a newborn with the rare anterior diaphragmatic hernia with herniation of the liver into the pericardial sac that posed a diagnostic dilemma in our Neonatal Intensive Care Unit (NICU). An

anterior diaphragmatic hernia should be suspected in any newborn with an anterior mediastinal mass or a pericardial effusion. A MRI of the chest helped to confirm the diagnosis. The relevant literature is reviewed.

KEY WORDS: congenital, diaphragmatic, hernia, neonatal

INTRODUCTION

Congenital diaphragmatic hernia (CDH) occurs approximately once in every 2000 births^[1]. Posterolateral defects are the most common accounting for 75-80% of hernias^[2]. Anterior hernia of the foramen of Morgagni is the most uncommon type of CDH accounting for only 1-6% according to previous studies^[3]. A defect in the central tendon, the rarest of the CDH entities causes the recognizable triad of bilateral pulmonary compression with or without hypoplasia, massive pericardial effusion without cardiac compromise and an intra-pericardial herniation of the liver^[4]. One such case who delivered in our hospital and posed a diagnostic problem is reported here.

CASE REPORT

A preterm baby boy was born normally to a 28 year old P O + 3 + 4 + 3 mother at 35 weeks gestation. Antenatal ultrasound examination at 17 and 20 weeks gestation had detected bilateral pleural effusion. Subsequent serial ultrasound examinations at 22, 24, 26 and 28 weeks showed bilateral pleural effusion with pulmonary hypoplasia and there was suspicion of an eventration of the right hemidiaphragm or a central diaphragmatic hernia with liver herniation into the right side of the chest. Intrauterine pleural aspiration was done and 35 ml of fluid was removed which was reported to be serous inflammatory fluid. Amniocentesis confirmed 46XY karyotype. At 33 weeks gestation, pericardial effusion was detected in addition to

the above findings. Parents were counseled about the poor prognosis. A baby boy weighing 2.650 kg was delivered normally at 35 weeks gestation. He required immediate intubation for irregular gasp like respiration. Initial chest X-ray showed bilateral pleural effusion (Fig. 1). As ventilation was difficult, bilateral thoracentesis was done. Fifty milliliter of xanthochromic fluid was removed from the left side and less than 10 ml from the right side. He developed bilateral pneumothoraces which were drained. At 10 hours of age he was shifted to high frequency oscillatory mode of ventilation.

An ultrasound examination of the thorax on day two and nine showed re-accumulation of the pleural fluid on both sides. Computed tomography (CT) scan of the chest on day 8 was reported as loculated air collection in the anterior chest cavity with thin septae compressing the mediastinal structures and both lungs posteriorly and shifting the heart to the left. Echocardiography on day 16 was reported as near systemic pulmonary hypertension, poorly contracting dilated right ventricle and a moderate pericardial effusion. Cardiologist advised conservative management as he was hemodynamically stable. TC 99 lung perfusion scan on day 20 revealed poor uptake in the right lower lobe with poor perfusion and a normal left lung, suggesting a focal pathology in the right lower lobe.

High resolution CT scan of the chest done on day 23 revealed a fluid filled cyst in the anterior mediastinum and bilateral dependant atelectasis

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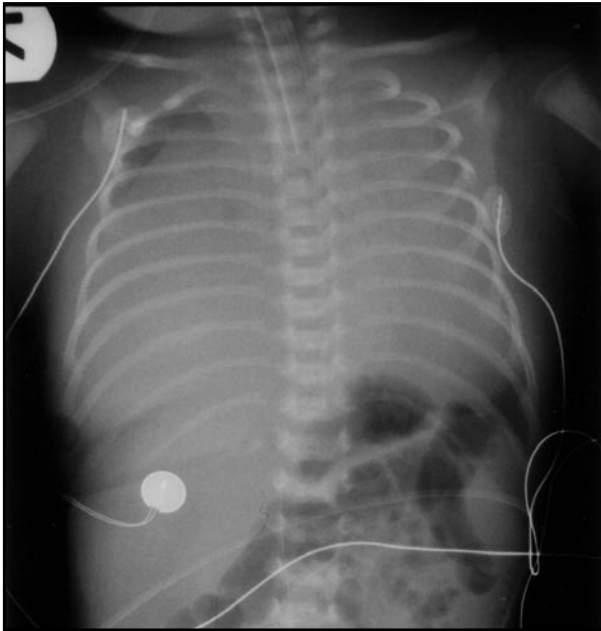


Fig.1: Initial chest X-ray showing bilateral pleural effusion

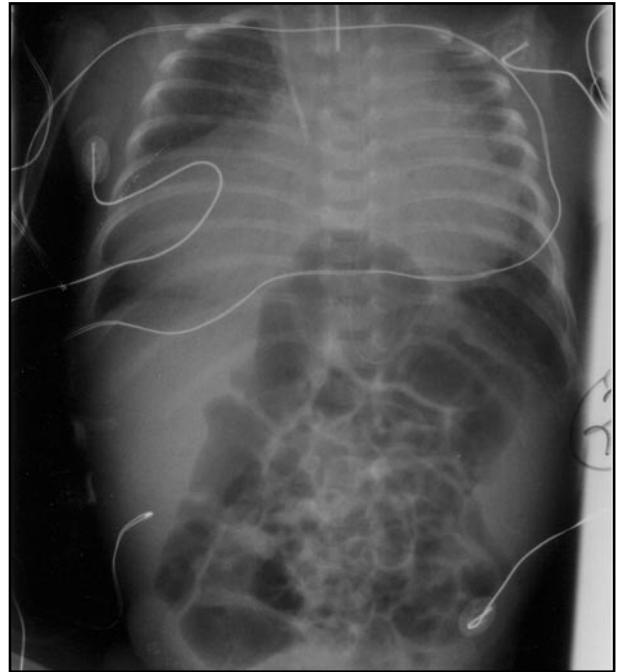


Fig.2: Postoperative chest X-ray showing the right para-cardiac mass and the bowel loops in the lower mid-thorax

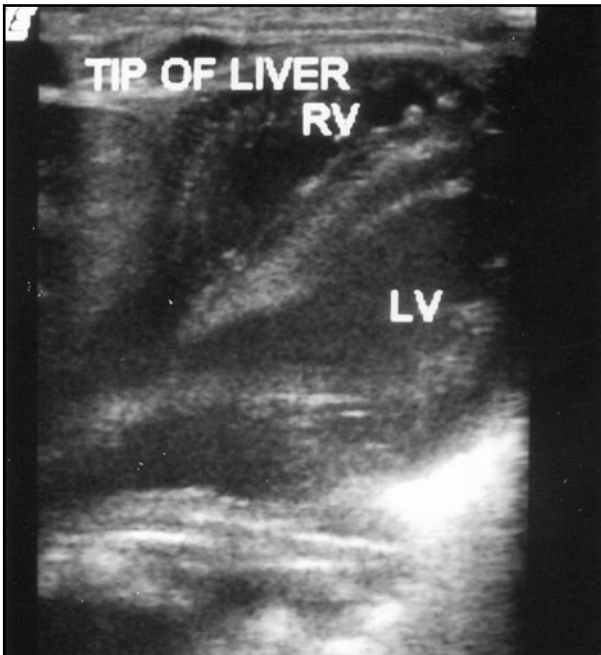


Fig.3: Post operative chest ultrasound showing the liver herniation

with pneumonic consolidation of the right lower lobe. Thoracic surgical consultation was sought. An exploratory left thoracotomy was done on day 36. At operation a large pericardial effusion was detected. 200 ml of green coloured pericardial fluid was drained. Left hemi-diaphragm and thymus were reported normal. A pericardial window was made. Post-operative chest skiagrams showed clearing of the cardiophrenic angles but a rounded opacity in the right lower zone contiguous with the right cardiac border was more clearly visible (Fig. 2). In some of the post-op CXR films bowel loops could be seen extending up to the lower mid-thorax. He

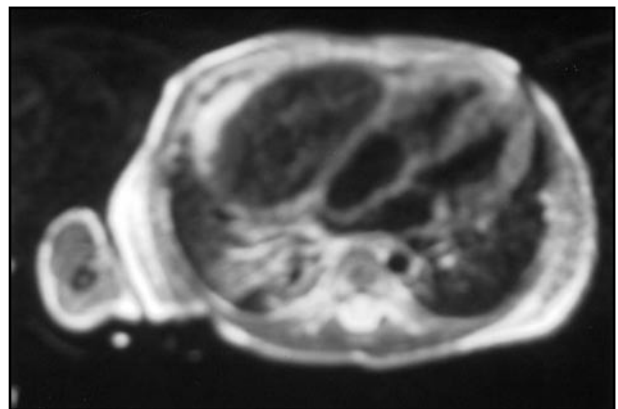


Fig.4: MRI chest showing the liver adjacent to the heart

continued to be ventilator dependant.

A repeat ultrasound examination of the chest on day 45 showed evidence of the liver herniation into the anterior mediastinum through a defect in the anterior diaphragm measuring 4.2 cms (Fig. 3). A MRI chest done on the same day confirmed the liver herniation into the anterior mediastinum (Fig. 4) and he was referred abroad for surgery.

On day 100, a patch repair of the central diaphragmatic hernia and a fundoplication was done. He was successfully extubated and sent back to Kuwait.

DISCUSSION

The foramen of Morgagni (space of Larrey) extends from the sternum medially to the 8th rib laterally and Morgagni hernia occurs because of the failure of complete fusion of the sternal and costal parts of the hemi-diaphragm^[1,5]. The most

commonly herniated viscera are the liver, spleen and the omentum. Large and small bowels are less commonly seen. Among Morgagni hernias, a particularly rare form is a central defect involving both the diaphragm and the pericardium and in these cases the viscera herniate into the pericardial sac^[1]. These hernias are believed to represent the developmental failure of the retrosternal position of the septum transversum^[2]. Presentation may be with respiratory distress and cyanosis soon after birth or with a massive pericardial effusion without cardiac tamponade^[2]. This suggests the slow formation of the pericardial fluid with compensatory progressive distension of the fetal pericardium. The accumulation of fluid may be due to mechanical irritation of the pericardium^[2] and a compromised hepatic venous outflow involving the herniated liver (Budd-Chiari like effect)^[4].

The postero-anterior and lateral CXR are diagnostic of Morgagni hernia, if the hernial sac contains air filled bowel loops above the diaphragm. This can be confirmed by an upper GI series or a barium enema^[6]. But, the diagnosis could be difficult if the viscera above the diaphragm are solid with liver or omentum, confusing it with a low anterior mediastinal mass^[6]. A well defined shadow in the right cardiophrenic angle may be seen on a PA CXR film as was seen in this case after the pericardial fluid was drained. In such cases other diagnostic imaging such as ultrasonography, radionuclide liver / spleen scan, CT or MRI may be needed to confirm the diagnosis^[5,6]. Liver herniation above the diaphragm can be sonographically distinguished from a pericardial tumor by the appearance of the hepatic vessels coursing into the mass which has the same echogenicity as the liver^[1,7].

Echo and CT scan are useful investigations. Continuation of the mass with the liver and the same texture of both shadows help to make the diagnosis of the intra-pericardial herniation of the liver^[8].

Though there was suspicion of an anterior diaphragmatic hernia in the earlier antenatal ultrasound examinations, the combination of bilateral pleural effusion and pericardial effusion at the later antenatal ultrasound examinations and the postnatal CXR films made the precise diagnosis of the case difficult. CT scan of the chest done twice and TC 99 ventilation perfusion lung scan did not help in the diagnosis. Once the pericardial fluid was drained at open thoracotomy, the chest X-ray films and ultrasound examination of chest led to the diagnosis of the anterior diaphragmatic hernia which was confirmed by the MRI.

Fetal MRI has been found to be clearly superior to ultrasound in demonstrating the liver herniation into the chest in the antenatal period^[9]. Emergency

pericardiocentesis and surgery are not always required immediately after birth, even when the condition is suspected by prenatal diagnosis^[10].

Morgagni hernias detected in the neonatal period are often associated with congenital anomalies^[11]. These include congenital heart disease (80%), malrotation (26%), omphalocele (15%) and Down syndrome (14%)^[12]. This case did not have any other major anomaly.

CONCLUSION

The possibility of a congenital anterior diaphragmatic hernia with liver herniation should always be considered in the antenatal and post-natal diagnosis of a pericardial effusion or an anterior mediastinal mass. Fetal MRI and post-natal MRI can help in the early diagnosis and optimal management of such cases.

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