Cogenital Intrapericardial Herniation of Liver

NB MATHUR, ANURADHA BANSAL AND *SATISH KUMAR AGGARWAL

*From Department of Pediatrics and *Pediatric Surgery, Maulana Azad Medical College, New Delhi 110002, India.*

Correspondence to:
Dr NB Mathur,
Director Professor of Pediatrics,
Maulana Azad Medical College,
New Delhi 110002, India.
drnbmathur@vsnl.com
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Intrapericardial herniation of liver is a rare form of diaphragmatic hernia. We report a 30-hour old baby with right congenital anterior diaphragmatic hernia masquerading as congenital pneumonia with cardiomegaly. It is prudent to consider congenial anterior diaphragmatic hernia in any newborn with unexplained respiratory distress, cardiomegaly and pericardial effusion.

Key words: *Diaphragmatic hernia, Intrapericardial herniation, Neonate, Respiratory distress.*

Intrapericardial herniation of liver is the rarest form of diaphragmatic hernia [1,2] and only one such case has been reported from India [3]. We report a case of right congenital diaphragmatic hernia masquerading as congenital pneumonia with cardiomegaly and discuss the relevant diagnostic issues.

CASE REPORT

A 30-hour old female born to 28 year old primigravida mother at 34 weeks of gestation presented to us with respiratory distress since half hour of life. The baby was delivered through caesarean section for meconium stained amniotic fluid at a private nursing home. Antenatal period was uneventful with normal antenatal ultrasound. Birthweight was 1800 g and Apgar score was 7, 8, 9. In view of respiratory distress, the baby was managed as a case of congenital pneumonia with oxygen, intravenous fluids and antibiotics. The symptoms persisted beyond 24 hours of life and the baby was referred to our hospital. At admission she had a respiratory rate of 92 per minute with minimal intercostal and sub-costal retractions. Heart rate was 140 per minute and capillary refill time was normal. Oxygen saturation was 89% in room air and there was no evidence of congestive cardiac failure. Arterial blood gas revealed a pH of 7.42 with P_aO_2 of 61 mmHg, P_aCO_2 33 mm of Hg and bicarbonate 25 mEq/L.

On examination, the neonate had a cleft of soft palate and low set ears. Systemic examination was not contributory. She was active, alert and neonatal reflexes were present. She was started on oxygen, intravenous fluids and antibiotics were added in view of positive sepsis screen. Chest *X*-ray showed normal lung fields with massive cardiomegaly. Ultrasound of the chest revealed moderate pericardial effusion. A 2.4 × 1.8 cm fluid filled area was seen in right lower hemithorax continuing with pericardial cavity. There was a defect in the central tendon of diaphragm with herniation of superior part of right lobe of liver in lower hemithorax abutting the right atrium. Both the lungs were filled with air and solid organs were normal in morphology. CT scan of the chest confirmed these findings. Echocardiography also showed protrusion of liver into thoracic cavity and compressing the right atrium and right ventricle. There was a 3 mm atrial septal defect and mild bilateral peripheral pulmonary artery stenosis. Chromosomal analysis could not be done due to financial constraints.

The baby was operated on day 5 of life. A laparoscopic repair was planned. A defect (3cm diameter) in the central tendon of the diaphragm was visualized, through which a part of the right lobe of liver was herniating into the mediastinum. Rest of the viscera was normal. While dissecting the margins of the defect laparoscopically there was some bleeding from the diaphragm. For the sake of safety the procedure was converted into open operation. The bleeding was from a diaphragmatic vessel and could be controlled easily. The defect was defined and the liver delivered into the abdomen. A gush of clear fluid came from the pericardium, into which the liver was herniating. Pleura was not opened on either side. The defect was repaired with non absorbable sutures. The abdomen was closed.

The baby was shifted to NICU for elective ventilation and supportive treatment. The pericardial fluid analysis revealed 15 cells, all lymphocytes and 342 mg/dL protein suggestive of transudate. The culture was sterile. The baby remained stable for 24 hours but further course

could not be followed as she was taken against medical advice due to some family reasons.

DISCUSSION

Anterior diaphragmatic hernia of Morgagni is an uncommon entity believed to occur due to failure of development of retrosternal portion of septum transversum during 8th week of embryonic life [1]. The central tendon of diaphragm is a thin but strong aponeurosis situated near the centre of the vault formed by the muscle, but somewhat closer to the front than to the back of the thorax so that the posterior muscular fibres are longer. It is situated immediately below the pericardium, with which it is partially blended [4]. Hence, herniation through a defect in the central tendon of diaphragm results in intra pericardial herniation of abdominal contents.

Till date, only 13 cases of congenital anterior diaphragmatic hernia into the pericardium have been reported. Pericardial effusion was not clearly described in three of these. All of these cases showed pathological characteristics differing from those of congenital diaphragmatic hernia detected after early infancy. Congenital anterior diaphragmatic hernia is characterized by: (*i*) development during fetal period (*ii*) frequent complication by massive pericardial effusion and the absence of cardiac tamponade; (*iii)* less frequent respiratory problems, and (*iv*) hernial contents consisting of liver herniating into the pericardium [5].

Herniation of liver into the pericardial cavity usually leads to slowly developing pericardial effusion as described above [1-3,5-6]. Some cases may even present beyond neonatal period as persistent pneumonia [7,8]. Although the detailed pathophysiology of pericardial effusion remains unclear, the following mechanisms have been proposed; compression of the thoracic duct causing accumulation of lymph; venous obstruction in the liver leading to congestion and transudation, and, mechanical irritation [5]. In our case, the baby remained hemodynamically stable and did not require any ventilatory support preoperatively. This might be because of the absence of lung hypoplasia. Smaller hernial content as compared to the posterior hernia, and slow formation of pericardial fluid with compensatory progressive dilatation of fetal pericardium have been proposed to be the reasons for the absence of cardiac tamponade [5].

To conclude, it is prudent to consider congenial anterior diaphragmatic hernia as a possibility in any newborn with unexplained respiratory distress, cardiomegaly and pericardial effusion. Prompt diagnosis is crucial as this condition is amenable to surgical treatment. Minimal invasive surgery is gaining popularity for the treatment [9,10].

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