Right-sided congenital diaphragmatic hernia masqueraded as right lobar pneumonia in a term newborn infant

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DESCRIPTION
A full-term male infant born by normal vaginal delivery developed respiratory distress at 15 min of life. Antenatal history was unremarkable. The last fetal scan at 32 weeks gestation did not reveal any congenital anomaly. There was no maternal history of leaking liquor, fever, or group B streptococcus colonisation.

Physical examination showed tachypnoea, chest retractions and decreased air entry in the right lung. Oxygen saturation was 80%–82%, and blood gas showed respiratory acidosis. He was placed on continuous positive airway pressure of 7 cm water and FiO2 0.3. Vital signs were within normal limits. Blood count and C reactive protein levels were unremarkable. Chest X-ray (CXR) showed opacity in the lower zone of the right chest, suggesting lobar consolidation (figure 1). Antibiotics were initiated promptly. At 6 hours of life, with worsening respiratory distress and echocardiography showing evidence of pulmonary hypertension, he was intubated. Repeat CXR revealed multiple radiolucent bowel loops in the right hemithorax (figure 2), consistent with a diagnosis of right-sided congenital diaphragmatic hernia (R-CDH) as confirmed by ultrasound. Karyotyping was normal.

The infant remained haemodynamically stable while intubated with FiO2 0.3. Surgical repair was performed on day 2 via the transabdominal approach. Intraoperatively, viable loops of intestines were found in the chest and mobilised back into the abdomen. There was no significant liver herniation. The diaphragmatic defect was repaired by primary closure. Postoperatively, he was extubated on day 7, initiated on feeds on day 9, and discharged home on day 17 of life.

Congenital diaphragmatic hernia (CDH) occurs in 0.08–0.45 per 1000 births, attributed to the failure of fusion of pleuroperitoneal membranes during fetal development.1 Antenatal ultrasound screening identifies more than 60% of CDH by second trimester and prognosticates using serial lung-to-head ratio assessment and degree of liver herniation. Fetal MRI estimates fetal lung volumes and liver position.2

CDH occurs on the right in only 13% of cases.3 Infants with R-CDH often present with respiratory distress at birth due to lung hypoplasia and persistent pulmonary hypertension of newborn.4 Diagnosis of R-CDH can be challenging as clinical presentation and radiological findings may masquerade as lung pathology.3 6 In R-CDH, the liver may obturate the diaphragmatic defect and prevent extrusion of abdominal contents into the thorax, hence not visualised on plain radiographs and delaying diagnosis.

Initial CXRs of R-CDH may be misinterpreted as pneumonia, as demonstrated in our case. Pleural effusion may occur due to hepatic venous outflow obstruction causing congestion. Differential diagnoses include pneumatoceles, cystic adenomatoid malformation, congenital lobar emphysema and diaphragmatic evagination.7 When diagnosis is equivocal, alternative-imaging modalities such as
ultrasonography can be complementary. Ultrasound identifies parenchymatous organs and peristaltic movement of bowels in the hemithorax, and the absence of hyperechoic diaphragm and pulmonary A-lines in CDH. Doppler vascular flow aids in diagnosing thoracic liver herniation.

Intestinal malrotation is reported in 39% of infants during initial CDH repair and those with a missed association have higher risk of future abdominal complications.9

Overall survival in R-CDH is approximately 63%.10 The severity of pulmonary hypoplasia and pulmonary hypertension is a significant factor determining R-CDH outcome. The timing of surgical repair also contributes to subsequent morbidities.3

Fortunately, the initial stabilisation at birth, early diagnosis and surgical intervention gave our patient a favourable outcome.

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