CASE REPORT

Persistent respiratory distress in a neonate: a diagnostic dilemma

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SUMMARY

We present a 17-day-old term, female baby who was referred to our centre for persistent respiratory distress. She was managed for pneumonia and pneumothorax at the primary care centre. On detailed clinical examination at admission, a possibility of congenital lobar emphysema (CLE) was considered. A CT chest was performed, and diagnosis of CLE was confirmed. The infant was managed with lobectomy. The respiratory distress settled within a few hours after the surgery, and the baby was discharged in stable condition.

BACKGROUND

Respiratory distress is common in neonates, and most centres are able to manage respiratory distress. However, the problem arises when respiratory distress does not resolve, and one needs to find the cause of persistent respiratory distress and tailor the management accordingly. One of the rare causes of persistent respiratory distress is congenital lobar emphysema (CLE). CLE is a lung malformation and requires emergency surgical treatment, if symptomatic. However, some of the patients can be managed conservatively. It should be diagnosed as early as possible and must be differentiated from

pneumothorax as the treatments for the two conditions are different.

CASE PRESENTATION

A female baby of 2.8 kg was born to a primi mother by caesarean section for non-progression of labour at a private hospital. Antenatal ultrasounds were reported to be normal, and liquor was clear as per documents submitted to us by the parents. The infant had a difficult transition at birth and required resuscitation, details of which were not known to parents and were not documented on the discharge sheet. As per the verbal report from the parents, the baby was managed with brief mechanical ventilation and was treated with an empiric antibiotic for possible sepsis/pneumonia. Needle aspiration was attempted for suspected pneumothorax with no improvement in the symptoms. Later, a repeat thoracentesis followed by tube thoracostomy was performed for suspected unresolved pneumothorax causing persistent respiratory distress. The baby was transferred to tertiary care centre on day of life for the unresolved respiratory distress of unknown aetiology. On examination, at the time of admission, the baby had tachypnoea with a respiratory rate of 74 breaths/min, and was having subcostal/intercostal retractions. On auscultation, there were decreased breath sounds on the left side of the chest. On cardiovascular examination, the heart sounds were better audible on the right side. A chest X-ray was ordered which revealed overinflation of the left upper lobe and herniation into the right side of the chest with mediastinum shift to the right, raising the possibility of left upper lobe lobar emphysema (figure 1). CLE can be differentiated from pneumothorax by extension of pulmonary vessels to the periphery of the hyperinflated lobe, and there is no visualisation of a pleural line unlike in a pneumothorax. CT scan confirmed the diagnosis with features of hyperinflation of the left upper lobe with vascular attenuation. Furthermore, it was associated with shifting of mediastinum with herniation of left upper lobe (figure 2). The baby was started on nasal flow cannula and was requiring a FiO_2 of 0.3 to maintain a normal saturation of 90%–95%. On investigation, sepsis screen was negative, and blood culture was sterile. Blood gas analysis revealed mixed acidosis with pH of 7.17, pO_2 55, bicarbonate of 18, a base deficit of 4 and pCO_2 of 55. Packed red blood cell transfusion was planned in view of anaemia (packed cell volume

Figure 1 X-ray chest suggestive of congenital lobar emphysema with mediastinal shift to the opposite side.

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Figure 2 CT chest showing congenital lobar emphysema of the left lung.

24) and oxygen requirement. The baby was operated on day 3 of admission (day 19 of life). Left side lobectomy with the placement of intercostal drain (ICD) was done. Postoperative course was uneventful, except for brief ventilation support of 12 hours. The ICD was removed on the fourth postoperative day. Feeds were started immediately after extubation and increased to full feeds in next 24 hours. The baby was discharged on breast feeds in stable condition on day 28 of life. Cross-lateral and decubitus radiographs may further aid in distinguishing the two.

INVESTIGATIONS

- ▶ Chest X-ray.
- ▶ CT thorax chest.
- ▶ Echocardiography which was normal.

DIFFERENTIAL DIAGNOSIS

1. Congenital cystic airway malformation
2. Sequestration of lung.
3. Pneumothorax.
4. Bronchogenic cyst.
5. Pneumonia.
6. Mucous plug.
7. Childhood interstitial lung disease.

TREATMENT

1. Lobectomy.
2. ICD.
3. Antibiotics.

OUTCOME AND FOLLOW-UP

On follow-up, the baby is thriving well and gaining weight. There are no signs of respiratory distress at 6 months of age.

DISCUSSION

CLE is a rare congenital malformation of the lung with a prevalence of 1 in 20 000 to 1 in 30 000. There is trapping of air in the lung during the expiratory phase of respiration due to deficient cartilage (video 1).¹ The most common site of involvement in CLE is the left upper lobe followed by right middle lobe and right upper lobe. CLE usually presents as repeated episodes of respiratory distress during the first 6 months of life. On chest

Video 1 Intraoperative finding, suggestive of congenital lobar emphysema.

examination, there is asymmetry of breath sounds and retractions. For diagnosis, X-ray chest, CT thorax and MRI can be done. CT findings suggestive of CLE include²:

1. Herniation of lobe into contralateral side.
2. Mediastinal shift to opposite side.
3. Collapse of part of the ipsilateral lung.

In 10% of CLE cases, there may be other associated malformations like congenital heart disease.^{3 4}

There are published reports of CLE being diagnosed beyond the neonatal period also.⁵ Similar to our index case, there is a published report of CLE presenting as recurrent pneumothorax in an adolescent.⁶ Treatment of CLE in babies with respiratory distress comprises surgical resection of the affected lobe. However, a conservative approach is reasonable in children who have no or negligible symptoms. A few newborn children who are asymptomatic during childbirth may likewise develop respiratory manifestations later in life which may require surgical resection.^{7 8}

Our case report highlights the fact that both CLE and pneumothorax are close differentials of hyperinflation of the lung, especially of the left upper lobe. The changes on X-ray such as extension of pulmonary vessels and lack of a pleural line should be recognised early. This would in turn prevent unnecessary thoracocentesis and lead to a more timely diagnosis. Furthermore, it

Learning points

- ▶ Lung malformation, although rare, is an important cause of persistent respiratory distress in infants.
- ▶ Congenital lobar emphysema (CLE) may mimic pneumothorax. Definitive diagnosis is important to avoid unnecessary interventions like needle aspiration.
- ▶ When in doubt, CT is required to confirm the diagnosis of CLE (the radiological features that distinguish pneumothorax from CLE have already been discussed in case presentation).

also stresses on the fact that although lung malformations are rare, they should be considered in a child with persistent respiratory distress.

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