Reminder of important clinical lesson

Not all infantile respiratory distress in winter is bronchiolitis: congenital lobar emphysema

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The authors report the case of a 4-week-old male infant presented during the winter period with respiratory distress. He had a 3 day history of cough and coryza, and a 2 day history of breathlessness and reduced feeding. He had evidence of tachypnoea, subcostal recession and hypoxia on examination. An initial diagnosis of bronchiolitis was made. The authors explore how the correct diagnosis of congenital lobar emphysema (CLE) was reached, highlighting key clinical signs and investigations. He had evidence of a hyperinflated right middle lobe, with collapse of right upper and lower lobes and left upper lobe with associated mediastinal shift on chest x-ray (CXR) and CT scan. He was referred to the regional Paediatric Cardiothoracic Centre where right middle lobectomy was performed with complete resolution of his respiratory distress and re-expansion of the compressed lobes on CXR. Current literature concerning CLE is reviewed.

BACKGROUND

Bronchiolitis is one of the most common reasons for admission to a paediatric unit in the winter months. It presents with cough, coryza, poor feeding and low-grade temperature with widespread crackles, wheeze and respiratory distress on examination. However, not all infants presenting with respiratory distress in winter have bronchiolitis, and this diagnosis needs to be considered carefully especially in the neonatal period. This case is reported to emphasise the importance of reviewing a diagnosis, particularly when the clinical course is not typical, and to encourage an analytical approach and challenge to the statement 'common things are common' in this situation. It also serves as a reminder that not all congenital lung lesions are detected antenatally, or are apparent clinically at birth, and can present in the neonatal period or later in infancy, childhood or even adulthood. Congenital lung lesions need to be considered in the differential diagnosis of unexplained respiratory symptoms in a child.

CASE PRESENTATION

A male infant presented at 4 weeks of age, during winter, with a 3 day history of cough and coryza and a 2 day history of breathlessness and reduced feeds. There was no history of fever. Both parents were reported to be recovering from an upper respiratory tract infection. He was born at term by normal vaginal delivery with a birth weight of 3.3 kg. There had been no concerns on antenatal scan. He required no resuscitation at birth, but had had a brief cyanotic colour change at 2 h of age which did not require any active treatment and was said to be due to 'mucous'. He had been otherwise well until presentation. He was the first child to teenage parents. There was no family history of note and no smokers or pets at home.

On examination, the infant was apyrexial and with significant respiratory distress. He had a respiratory rate

of 65/min, oxygen saturations of 88% in air and marked subcostal recession. There was decreased air entry bibasally. He was tachycardic with a pulse of 170/min, had normal heart sounds with no murmurs, a normal precordium to inspection and normal peripheral pulses. His capillary refill was less than 2 s. His abdomen was soft with a 2 cm liver edge palpable. He was alert and handled well.

A chest x-ray (CXR) was performed and demonstrated a hyperinflated lobe on the right with collapse of the adjacent right lobes and collapse/consolidation of the left upper lobe (LUL) and mediastinal shift (figure 1). A venous gas showed evidence of respiratory acidosis with a pH 7.26, pCO₂ 9.41 and base excess +2.6. He had a normal full blood count (FBC) and C-reactive protein (CRP) <1. Supportive nasal HumiCare was commenced in light of the work of breathing and respiratory acidosis on blood gas. This system delivers high concentrations of humidified oxygen at a flow rate high enough to generate a continuous positive end expiratory pressure. Intravenous amoxicillin was commenced in view of possible consolidation. The working diagnosis at presentation was bronchiolitis with a possible secondary bacterial lower respiratory tract infection (LRTI).

The infant was reviewed on the consultant ward round the following morning. His maternal grandmother was present and she reported her concern that he had 'always breathed heavily'. The nursing staff were concerned that his respiratory distress had not improved despite supportive measures including HumiCare and intravenous therapy to replace enteral feeding. Key features were identified which were not felt to be in keeping with the diagnosis of bronchiolitis with secondary bacterial LRTI:

1. Respiratory examination revealed predominantly unilateral chest signs with reduced air entry throughout the right side. Air entry was also diminished at the left base.

Figure 1 Chest x-ray on presentation.

- There was no evidence of crepitations or wheeze on auscultation, as would be expected in bronchiolitis.
2. CXR demonstrated unilateral lobar hyperinflation with compression of adjacent lobes and mediastinal shift. The typical CXR in bronchiolitis would be expected to show bilateral hyperinflation with patchy atelectasis, although lobar collapse/consolidation is also associated.
 3. Nasopharyngeal aspirate (NPA) was negative for all respiratory viruses.
 4. The infant remained afebrile with a normal FBC and CRP.
 5. There was a history of chronic respiratory symptoms as reported by the maternal grandmother.

The diagnosis of congenital lobar emphysema (CLE) was considered in view of the clinical signs and the presence of a grossly overinflated lobe on the right side on CXR. A CT scan of the chest was performed and demonstrated a hyperinflated right middle lobe (RML) with collapsed right upper and lower lobe, collapse of the LUL and mediastinal shift (figure 2). A diagnosis of CLE of the RML was made. The infant was discussed with a Tertiary Paediatric Cardiothoracic Centre and transferred for definitive surgical management.

INVESTIGATIONS

CRP <1 mg/l.

Capillary blood gases: On admission: pH 7.26, pCO₂ 9.41 kPa, base excess (BE) +2.6 mmol/l; 30 min after starting HumiCare: pH 7.33, pCO₂ 8.4 kPa, BE +7.1 mmol/l; 5 h after starting HumiCare: pH 7.33, pCO₂ 8.4 kPa, BE +7.1 mmol/l.

NPA: Negative for respiratory viruses.

CXR: Hyperinflated right lobe (considered to be either upper or middle) with collapse of the adjacent right lobes and collapse/consolidation of the LUL and mediastinal shift

CT chest: Grossly hyperinflated RML with collapsed right upper and lower lobe, collapse of the LUL and mediastinal shift.

Echocardiography: Normal structure and function.

DIFFERENTIAL DIAGNOSIS

The differential diagnoses to consider, given the asymmetrical clinical signs with reduced air entry on the right, mediastinal shift to the left and a hyperlucent right lobe with adjacent lobar collapse on CXR are:

- Pneumothorax: the CXR findings may be misinterpreted as pneumothorax, however, lung markings in this CXR extend to the periphery of lung fields.
- Localised pulmonary interstitial emphysema: the infant was a term baby who had not required neonatal intensive care so did not have the risk factors of prematurity, respiratory distress syndrome or mechanical ventilation.
- Congenital cystic adenomatous malformation: this most commonly affects the lower lobes and can present as an air or fluid-filled cyst or cysts.

TREATMENT

The infant underwent curative surgery in the form of a right middle lobectomy within 3 days of diagnosis. Intraoperative findings of a hyperinflated RML was confirmed. A decision was made not to perform bronchoscopy, as the diagnosis was clear from the chest CT and other clinical assessment modalities.

Figure 2 Thoracic CT scan.

OUTCOME AND FOLLOW-UP

The respiratory distress resolved and he had a normal CXR within 48 h of surgery. He was discharged home 72 h post operatively. He remained asymptomatic with a normal respiratory examination at 6 week follow-up.

DISCUSSION

CLE is a developmental abnormality of the lower respiratory tract defined as progressive lobar hyperinflation due to partial/complete obstruction of a bronchus.¹ It mainly affects the upper and middle lobes with the LUL being the most often affected: LUL 40–50%; RML 25–30%; right upper lobe 20%; right lower lobe 2–10%.^{1 2} CLE is three times more common in males than females, with an overall estimated prevalence of 1 in 20 000 to 1 in 30 000.^{1 3}

Fifty per cent of all cases have no definitive cause.⁴ Twenty five per cent of all cases are due to obstruction of the developing airway.¹ Intrinsic airway obstruction is most common, due to defects in the bronchial wall or bronchial atresia, resulting in collapse of the airway during expiration. This causes a ‘ball valve’ effect with air trapping on expiration and hyperinflation of the affected lobe. Extrinsic cause can include vascular anomalies or intrathoracic masses such as foregut cysts, teratomas or lymphadenopathy.

The hyperinflation of the lobe causes distortion of the terminal airspaces, rendering them non-functional tissue.¹ In contrast to adult-type emphysema it is important to note that it is the distortion and not destruction of the alveolar walls that causes CLE. This differentiation has led to a change in terminology to congenital lobar hyperinflation. The hyperinflation of the affected lobe also compresses surrounding lobes and thus decrease their functionality. Mediastinal shift can be present in severe cases of hyperinflation.

Fifty per cent of all postnatally confirmed cases of CLE will typically present within the first month; 25–30% at birth; 50% by 1 month of age; 90–100% by 6 months of age.^{1 2} There is a spectrum of symptoms ranging from children who are asymptomatic, with the diagnosis made as an incidental finding, to severe respiratory distress and cyanosis. It is also possible to diagnose CLE in the antenatal period during fetal screening and obstetric monitoring with ultrasonography, by detection of either an echogenic or cystic lung lesion.⁵

The diagnosis is a clinical diagnosis supported by radiological imaging. The CXR and CT scan in this case demonstrate the classical findings of a hyperinflated lobe compressing neighbouring lobes and herniating across the anterior mediastinum to cause mediastinal shift. Bronchoscopy can be helpful in diagnosing anatomical

abnormalities of the airways or in identifying the presence of external airways compression. An echocardiogram was performed as 14% of children with CLE have associated cardiovascular abnormalities.¹

CLE can be managed conservatively, with surgery reserved for the more severe cases. Studies have concluded that surgical management is appropriate for all infants <2 months and for those over 2 months with severe respiratory distress, mediastinal shift +/- lobar collapse on CXR or abnormal bronchoscopy findings.⁶⁻⁹ The appropriate surgical management is excision of the affected lobe.^{6,7} Conservative management involves serial clinical review with interval CXR's to monitor progression. Case studies have concluded that conservative management is appropriate for infants or older children with minimal respiratory distress.⁸⁻¹⁰ One study of 20 children in Toronto showed that 6 out of 11 symptomatic children who were managed conservatively showed clinical and/or spontaneous radiological improvement.¹⁰

Learning points

- It is important to constantly review a diagnosis, and although 'common things are common', always consider history, examination and investigation findings carefully and challenge this assumption if they do not 'add up'.
- Congenital lung lesions can present for the first time outside the perinatal period, in infancy, childhood or even in adulthood.
- Congenital lung abnormalities should be considered in the differential diagnosis of a neonate presenting with respiratory distress.
- History from grandparents can be invaluable, particularly when they are supporting young inexperienced parents with their first child.

Competing interests None.

Patient consent Obtained.

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