CASE REPORT

Congenital lobar emphysema: diagnostic and therapeutic challenges

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SUMMARY

Congenital lobar emphysema (CLE), a rare condition that usually presents in the neonatal period, can be a diagnostic and therapeutic challenge for the treating clinician. If unrecognised, it is a significant risk at the time of anaesthetic induction. We describe a case of CLE in a 3-month-old boy who was initially treated for suspected aspiration pneumonia at the referring hospital. We highlight the importance of careful consideration of common childhood respiratory illness as well as pneumothorax in the differential diagnosis, and the significance of appropriate preoperative anaesthetic management. We also emphasise the importance of acknowledging a mother's concerns when taking a paediatric history.

BACKGROUND

Congenital lobar emphysema (CLE) is a rare disease with a varied presentation. The main considerations in managing these children are: the need to make an accurate diagnosis in order to exclude more commonly encountered childhood respiratory diseases; to distinguish CLE from pneumothorax; and to avoid positive pressure at induction of anaesthesia. Children with CLE face a significant anaesthetic risk and require expertise in treatment.

CASE PRESENTATION

A 3-month-old boy was admitted to the referring hospital after a choking episode of postbottle-feed at home.

During the choking episode, he went floppy and blue for ~15 s. Otherwise, he had been well, and was afebrile. On presentation to the referring hospital, he was tachypnoeic with associated increased work while breathing. He was treated for suspected aspiration pneumonia with intravenous antibiotics and nasal prong oxygen for 1 week. Although he improved clinically, there was ongoing concern regarding his tachypnoea and persistent abnormal chest radiograph findings. Chest X-ray on presentation to our institution is presented in [figure 1](#). Of note is the fact that the boy's mother had reported on a number of occasions to her local medical officer her son's 'fast breathing' around 5–6 weeks of age.

The boy was born at term, via normal vaginal delivery, weighing 3900 g (85th centile). No resuscitation was required at birth, and admission to the neonatal unit was not required. No abnormalities were detected antenatally. There was no significant family history.

The boy was transferred to our facility late in the evening and admitted via the emergency

department. On arrival, heart rate was 152, respiratory rate was 65–70, and oxygen saturations 99% in room air. He was afebrile. Weight was 5100 g (3rd centile) which represented a significant fall from his birth percentile. Venous blood gas composition was: pH 7.36, pCO₂ 55 mm Hg, pO₂ 45 mm Hg, HCO₃ 30.3 mmol/L, base excess 3.7 mmol/L. He was started on high flow humidified oxygen in the emergency department for increased work of breathing. A CT chest scan was scheduled for the following day. He remained clinically stable in the paediatric ward overnight, and examination the next day revealed a thin, bright, alert, smiling infant. Respiratory rate was 50–55 on 4 L/min of humidified oxygen. Oxygen saturations were 99%. There was an increased anteroposterior diameter of the chest wall, reduced air entry in the right mid-to-lower zones, and widespread expiratory wheezes. There were mild-to-moderate subcostal/intercostal recessions and tracheal tug. Although the heart sounds were normal, the apex beat was displaced laterally. Heart rate was 140.

He underwent inhalational anaesthetic induction with sevoflurane and lignocaine 10 mg airway topicalisation maintaining airway pressures at <10 cm H₂O. During the CT, ventilatory support was maintained on the Dräger Primus anaesthetic machine circuit with a pressure support setting of 12 cm H₂O above a positive end-expiratory pressure of 4 cm H₂O, and a brief breath hold during

Figure 1 Right middle zone hyperlucency, right upper and lower zone collapse, and associated mediastinal shift to the left.

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Figure 2 Significant hyperexpansion and hyperlucency of the right middle lobe with contralateral shift of mediastinal structures. The right upper and lower lobes are compressed by the overinflated right middle lobe resulting in collapse.

inspiration with airway pressures below 20 cm H₂O. CT findings are displayed in [figure 2](#). The patient was extubated after the CT and taken to the postanaesthetic care unit, with the plan of elective surgery on the next day as the CT scan took place late in the evening. However, the patient's work of breathing increased in the postanaesthetic care unit; after surgical review of the images and the patient, a decision was made to proceed to emergency lobectomy. The patient was then taken to the operating theatre and emergency thoracotomy took place with the patient again undergoing inhalational induction with sevoflurane supplemented with ketamine in 0.2 mg/kg boluses up to 0.6 mg/kg, fentanyl 0.5 µg/kg, and local anaesthetic infiltration. Delivery of the emphysematous lobe is displayed in [figure 3](#). Pressure support ventilation was continued until delivery of the lobe, at which time positive pressure ventilation was instituted. The patient remained intubated in the Paediatric Intensive Care Unit until the following morning.

The resected lung lobe was received fresh in the pathology laboratory in a hyperinflated state. The lobe was perfused with formalin for optimal fixation, and large amounts of froth were observed to flow from the bronchi. Unlike most lung specimens, no further expansion of lung occurred during formalin perfusion, compatible with initial hyperinflation. Histopathological examination showed overdistention of normally formed alveoli, in the pattern of lobar hyperinflation ([figure 4](#)). Unlike emphysema in adults, there was no tissue destruction of alveolar walls. There were no features of aspiration pneumonia, infection, proximal bronchial stenosis/obstruction or proximal cartilage abnormality. A definitive cause for the emphysematous lobe was not found.

Figure 3 Delivery of the right middle lobe through the thoracotomy site.

OUTCOME AND FOLLOW-UP

The boy was discharged from hospital 2 days postoperatively; 2-month follow-up in the outpatient clinic revealed a thriving boy with no respiratory symptoms.

DISCUSSION

CLE is a rare anomaly of the lower respiratory tract that is characterised by hyperinflation of one or more pulmonary lobes.¹ Unilateral involvement of the left upper and right middle lobes is usual, although bilateral cases have been reported.^{1–5} For unknown reasons males are affected more commonly than females.

Lower airway obstruction, either intrinsic or extrinsic, leads to the creation of a 'ball-valve' mechanism in which a greater volume of air enters the affected lobe during inspiration than leaves it during expiration, producing air trapping. Bronchial cartilage dysplasia or deficiency leading to bronchomalacia is the commonest cause; however, as in our patient, a definitive cause is not found in up to 50% of cases.^{1 3} Extraluminal compression from bronchogenic cysts, vascular anomalies, and tumours also occur. Endobronchial causes of lobar emphysema are also well recognised, including mucus plugs, granulomas, polyps, mucosal folds, bronchial stenosis and foreign bodies.

The presentation of CLE is variable from an acute neonatal respiratory emergency to recurrent respiratory embarrassment or chest infection in the older child.² Antenatal diagnosis is possible with ultrasonography; however, babies with prenatally diagnosed cases are not always symptomatic at birth. Usually the earlier the presentation, the more severe the symptoms.² The main diagnostic dilemma arises when attempting to distinguish

Figure 4 Histology of lung showing distension of alveoli without tissue destruction, in the pattern of lobar overinflation. H&E, scale bar 500 µm.

CLE from more commonly encountered childhood respiratory disease such as pneumonia. Pneumonia is a common misdiagnosis for CLE and although it would be unlikely to result in inappropriate chest tube insertion, it is an important consideration nonetheless.

Clinical diagnosis can be challenging and chest radiograph is often diagnostic. Key findings include: hyperlucency on the affected side with ipsilateral atelectasis, widened rib spaces and diaphragmatic flattening. The mediastinum may be displaced away from the affected side and may herniate into the contralateral hemithorax.^{1–3} As has been previously reported in other cases, it is essential not to misdiagnose a pneumothorax because inappropriate chest drain insertion can have potentially disastrous consequences.^{1–3} The primary distinguishing feature of CLE is the extension of bronchovascular markings into the radiolucent area.²

CT chest is the usual adjunct to chest radiograph in confirming the diagnosis. CT scan can accurately delineate the anatomy of the affected lobe as well as identify other potential aetiological factors such as vascular anomalies and tumours. MRI is an alternative modality, but is not routinely used. Radionuclide ventilation-perfusion scans show diminished ventilation and loss of perfusion in the affected lobe. The technique also establishes whether the lobe affected by CLE is non-functional, and whether the compressed lung is functioning normally.^{1–3}

The role of bronchoscopy is controversial. Although useful in cases of suspected foreign body or other endobronchial causes of CLE, bronchoscopy may in fact aggravate the respiratory distress in patients with CLE and is not usually required for the diagnosis.³

The mainstay of treatment in patients with serious symptoms is lobectomy; however, anaesthetic considerations are paramount to a positive surgical outcome. The greatest risk is if excessive positive pressure ventilation is applied before opening the chest. This may cause rapid inflation of the emphysematous lobe and subsequent cardiopulmonary compromise. There is no means of judging the critical airway pressure at which dangerous overinflation may occur. Spontaneous ventilation, gentle assisted manual ventilation or titrated pressure-support ventilation settings are all feasible.⁶ It is suggested that airway pressures be maintained at <20–25 cm H₂O.^{7–9} However, prolonged preoperative ventilation has been described,¹⁰ as has utilisation of high frequency ventilation with minimal airway pressures.¹¹

The other technique to avoid overinflation of the diseased lung segment is lung isolation to prevent further ventilation of the emphysematous area. This may be achieved by endobronchial intubation; placement of an endobronchial blocker, which is usually positioned extraluminally to the endotracheal tube; or even with a modified double-lumen tube for paediatric patients.^{6 12–15} It is essential to have a surgeon ready to perform an immediate thoracotomy, if required.

Techniques to facilitate thoracotomy in the absence of muscle relaxation include use of regional analgesic approaches, or use of local anaesthesia and a mixed analgesia plan with ketamine and opioids.^{16 17} Maintenance of spontaneous ventilation, usually with inhalational agents, is most commonly described, but this may also be supported by adjunctive ketamine and judicious use of opioids.^{4 5} Once the emphysematous lobe has been delivered through the thoracotomy site it is unlikely there will be any issues with standard positive pressure ventilation. Although in this case the patient remained intubated and ventilated overnight, there is often no immediate impediment to immediate extubation. Conservative management is reserved for asymptomatic or mildly symptomatic cases.¹

The use of video-assisted thoracoscopic lobectomy for other types of congenital lung malformations has been reported in the literature. This surgical approach is difficult in patients with CLE because the pleural cavity is mostly occupied by the affected lobe which usually remains inflated. Lobectomy via thoracotomy is the usual surgical approach.¹⁸

We earlier stated the importance of appreciating a mother's concern given that there had been several encounters with the local medical officer with reports of 'fast breathing' prior to the patient's acute presentation. Furthermore, the patient's weight gain during this period was inadequate, which should have also raised concerns. An earlier chest radiograph could have prevented the need for an afterhours emergency operation and allowed for planning of an elective procedure.

To conclude, CLE is a rare disease with a varied presentation and is often misdiagnosed. Lobectomy is the mainstay of treatment in symptomatic cases, although conservative management is reasonable in infants and older children who have no or minimal symptoms. Prognosis after lobectomy is usually excellent. Children with CLE face a significant anaesthetic risk and require treatment expertise.

Learning points

- ▶ Congenital lobar emphysema (CLE) is a rare disease with a varied presentation.
- ▶ CLE is often misdiagnosed as more commonly encountered childhood respiratory disease such as pneumonia.
- ▶ The possibility of pneumothorax must always be considered.
- ▶ There is a significant anaesthetic risk at induction of anaesthesia and excessive positive pressure ventilation should be avoided.
- ▶ Acknowledging a mother's concern is an important part of paediatric history taking.

Contributors HS had the idea for the case report and is the guarantor for the finished article. JJM wrote the case report. JJM and AW performed the literature search. HS and AW were directly involved in the management of the case. AC provided the commentary and photograph of the histopathology.

Competing interests None declared.

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