CASE REPORT

Congenital Lobar Emphysema: a Diagnostic Challenge

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ABSTRACT

Congenital lobar emphysema is an abnormality of the lung of unknown aetiology that can present a diagnostic and therapeutic dilemma. This report describes a neonate with respiratory distress for whom an initial diagnosis of meconium aspiration syndrome was made. Congenital lobar emphysema of the upper lobe of the left lung with collapse and consolidation of the upper lobe of the right lung was eventually diagnosed at age 3 months following X-ray, computed tomography, and scintigraphy examinations. A left thoracotomy and left upper lobectomy was performed at age 3.5 months. The patient improved dramatically and the postoperative period was uneventful; postoperative chest X-rays showed resolution of the right upper lobe changes. Histological examination of the resected lobe showed normal bronchial cartilage of the major airways and distended alveoli. A thorough evaluation of the patient’s history, consideration of the differential diagnosis of respiratory distress, re-evaluation of chest radiographs, and computed tomography are helpful in difficult cases or for early diagnosis of congenital lobar emphysema.

Key Words: Differential diagnosis; Infant, newborn; Pulmonary emphysema/congenital; Radiography, thoracic; Tomography, X-ray computed

INTRODUCTION

Congenital lobar emphysema (CLE) is an abnormality of the lung of unknown aetiology that can present a diagnostic and therapeutic dilemma. CLE is an important cause of respiratory distress in neonates and often requires surgery for definitive treatment. The diagnosis is usually based on the clinical and plain radiological findings, but it may not always be straightforward. In cases in which diagnosis is difficult, computed tomography (CT) and ventilation and perfusion (V/Q) scintigraphy may be helpful. This report describes a neonate with respiratory distress for whom an initial diagnosis of meconium aspiration syndrome was made. CLE was eventually diagnosed at age 3 months.

CASE REPORT

A newborn Indian boy with a birthweight of 3.9 kg was delivered at full-term by Caesarian section at the University of Malaya Medical Centre, Kuala Lumpur, Malaysia, in October 2003 because of foetal distress secondary to the wrapping of the umbilical cord around the baby’s neck during delivery. The 1-minute APGAR score was 5/10 and the 5-minute APGAR score was 7/10. There was marked respiratory distress at birth and copious meconium-stained liquor was aspirated from the pharynx, following which the baby was intubated.

The initial chest X-ray that was taken shortly after birth showed diffuse granular shadowing in both lung fields, and a diagnosis of meconium aspiration syndrome was made (Figure 1). The X-ray changes resolved at day 5 of life, but on day 8, the baby developed a nosocomial chest infection due to methicillin-resistant Staphylococcus aureus. Further chest X-rays revealed consolidation of the upper lobe of the right lung and lower lobe of the left lung, which resolved following antibiotic therapy.

On day 15 of life, the baby developed atelectasis of the right upper and left lower lobes (Figure 2) and a shift of the trachea to the right. These changes were presumed to be secondary to secretions following extubation. His
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The condition improved after ventilation with nasal continuous positive airway pressure was started. He was discharged in a satisfactory condition, but he continued to develop repeated episodes of respiratory distress in spite of treatment with antibiotics and corticosteroids. (Corticosteroids were given to treat the asthmatic component of the patient’s symptoms.)

Successive chest radiographs revealed persistence of the right upper lobe collapse and worsening left lower lobe collapse. The left upper lobe had become hyperexpanded and there was herniation of the lobe across the mediastinum (Figure 3). Bronchoscopy was performed in week 8 of life and revealed tenacious secretions from the left lower lobe bronchus and clear secretions from the right side. The left upper lobe bronchus was slit-like and it was not possible to pass the bronchoscope through it.

CT of the thorax supported the plain X-ray and bronchoscopic findings — namely, a narrowed bronchus of the left upper lobe but no obvious extrinsic mass. The left upper lobe was hyperexpanded, herniated across the mediastinum, and compressing the right lung (Figure 4). Perfusion scintigraphy performed at 10 weeks revealed no perfusion to the left upper lobe; the left lower lobe still well perfused. The right upper lobe was poorly perfused but the rest of the right lung was well perfused.

At age 3 months, a final diagnosis of CLE involving the left upper lobe was eventually established, together with collapse and consolidation of the right upper lobe. A left thoracotomy and left upper lobectomy was performed at 3.5 months of life. The patient’s condition improved dramatically after surgery and the postoperative period was uneventful. The patient made a full recovery, and postoperative chest X-rays showed resolution of the right upper lobe changes. Histological examination of the resected lobe showed normal bronchial cartilage of the major airways and distended alveoli.

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**Figure 1.** Chest X-ray taken 3 hours after birth showing a diffuse appearance in the lungs due to meconium aspiration.

**Figure 2.** Chest X-ray taken 15 days after birth showing collapse and consolidation of the upper lobe of the right lung, and consolidation in the lower lobe of the left lung.

**Figure 3.** Chest X-ray taken at age 6 weeks showing overexpansion of the upper lobe of the left lung, herniation of this lobe into the right chest, and collapse of the lower lobe of the left lung.
DISCUSSION

Respiratory distress is a common neonatal problem. Many conditions can result in the same clinical presentation: tachypnoea, chest wall retraction, cyanosis, and poor feeding. The physical examination is difficult to perform and may be unrevealing. Laboratory investigations may indicate the severity of the respiratory distress but not the aetiology.

In this case report, a neonate presented with non-specific features of respiratory distress. In keeping with the clinical scenario and plain-film radiographic evidence, the initial diagnosis was meconium aspiration syndrome. Although meconium aspiration was probably the initial diagnosis, subsequent chest X-rays did not conform to this diagnosis, and only later did serial radiographs reveal the characteristic appearances of CLE — that is, progressive overinflation of a lobe and compression of the remainder of the lung.

Lobar emphysema is a rare abnormality and is usually congenital. Its incidence is 1 per 20,000 to 30,000 live births and is characterised by marked overinflation of a single pulmonary lobe — usually 1 of the upper lobes, more often the left, and less commonly the right middle lobe. The lower lobes are rarely affected. Initially, the affected lobe may sometimes appear opaque because it fills with fluid, before becoming hyper translucent. There is gross overinflation of the affected lobe, compression of the remainder of the ipsilateral lung, mediastinal shift, and herniation of the emphysematous lobe into the opposite side of the chest. Severe respiratory distress may be present, which requires urgent lobectomy. In some cases, the respiratory distress is mild, so surgery can be delayed and spontaneous resolution may even occur. In this case, because of the copious secretions, it was conceivable that the lobar emphysema was spontaneous, but the progressive changes meant that it was reasonable to assume that the disease was of the congenital type and surgery was hence undertaken.

Most cases of CLE become symptomatic; respiratory distress in the neonatal period may be life-threatening. In as many as 25% of cases, presentation is delayed until after the first month of life. Various suggestions have been put forward as to the cause of CLE but approximately half of the cases are of unknown aetiology, as in the case described. A further 25% of cases are due to a bronchial cartilage defect, which can range from hypoplasia or absence of tissue, such that the bronchus collapses during expiration. As a consequence, there is air trapping and overinflation of that lobe. The characteristic changes are bronchopulmonary dysplasia and obstructive intra-alveolar emphysema. In addition, 13% of cases are due to endobronchial obstruction, such as a fold, web, or mucous plug. Other causes include bronchial compression by a vascular structure such as a patent ductus arteriosus or aberrant left pulmonary artery. Finally, there is an association of CLE with congenital heart disease, notably ventricular septal defect and patent ductus arteriosus in 15% of cases.

CT was an important diagnostic procedure in this case because it not only showed the abnormally narrowed bronchus, the hyperinflated left upper lobe, and the collapsed lower lobe, but it also showed that although the right lung was compressed, it consisted of 3 lobes, thereby excluding a hypoplastic right lung as the cause of the appearances. CT can also exclude a vascular abnormality and other conditions that might be confused with CLE such as pneumothorax, pneumatocele, diaphragmatic herniation, or cystic adenomatoid malformation.

V/Q scintigraphy is helpful in showing absent ventilation and perfusion to the affected lobe and normal ventilation and perfusion indicating unimpaired blood flow and gas exchange to the rest of the lung. Nevertheless, in the case described in this report, perfusion scintigraphy alone did not aid the diagnosis and the changes of CLE were evident on serial chest radiographs and the CT scans. Bronchoscopy may be of value in ruling out the presence of an intraluminal foreign body that...
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causes either obstructive emphysema or atelectasis with compensatory emphysema.\textsuperscript{3,6}

CLE usually follows a progressive course and, if not recognised and treated, it may be fatal. Surgery is usually undertaken and a simple lobectomy is highly effective.\textsuperscript{7} Thorough evaluation of the history, consideration of the differential diagnosis of respiratory distress, and re-evaluation of the chest radiographs, together with CT, are helpful in difficult cases or for early diagnosis of CLE.

REFERENCES