Evolution and sequelae of acquired lobar emphysema in a premature infant with chronic lung disease

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ABSTRACT

The evolution of acquired lobar emphysema in a premature infant with chronic pulmonary disease was demonstrated by serial radiological examinations using multiple imaging methods including plain radiography, serial computed tomography and radionuclide studies. Long term clinical and radiological follow up is presented. The value of computed tomography in the evaluation of the patient is emphasized. The literature on the patho-physiological mechanism of the disease is reviewed.

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Acquired lobar emphysema (ALE) is a well-known complication of chronic pulmonary disease of prematurity.[1,2] Pathogenesis of the condition appears to be multifactorial and early detection of the lesion is essential for prompt management. Although plain film radiography is the mainstay of diagnosis, computed tomography (CT) may play an important role in the early detection and confirmation of the lesion. Using sequential follow up, this article highlights the CT appearances of early and later phases of the lesions.

Case Report. We present a 34-week-old boy with an uncomplicated gestation, weighing 2,260 grams at birth. He required endotracheal intubation at 8 hours of age for Respiratory Distress syndrome (RDS). Initial chest radiography showed mild diffuse ground-glass opacity in both lungs indicating alveolar hypo-aeration. He developed right-sided pneumothorax at 20 hours of age, which was treated with chest tube drainage. Subsequently the pneumothorax reduced leaving interstitial air in both lungs. Tension pneumothorax recurred on the third day leading to collapse of the right lung and needing additional chest tube drainage. On day 7 there was a hint of hyper-inflation of the right lower lobe (RLL) although the patient was clinically stable. He was extubated at 8 days of age and appeared to make good progress until 3 weeks of age when he again developed respiratory distress and was given only supplemental oxygen.

Radiographically there was a gradual persistent air trapping in the RLL (**Figure 1a, 1b, 1c & 1d**) with associated mass effect and contra-lateral cardio-mediastinal displacement. Computed tomographic examination at this stage confirmed a grossly expanded RLL showing multiple thin-walled cysts causing compression of the adjacent right lung, displacement of cardio-mediastinal structures and an indication of narrowing of the bronchus of the right lower lobe. The respiratory distress was progressive, requiring oxygen, therefore the bronchoscopy and surgical intervention were considered. Pre-bronchoscopic

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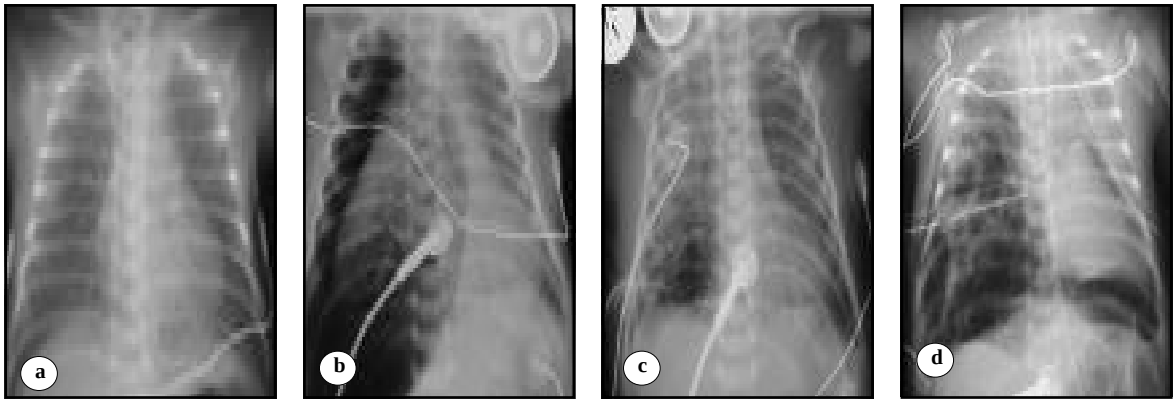

Figure 1 - Chest radiograph at **a)** 8 hours of age demonstrating changes of mild Respiratory Distress syndrome, **b)** 24 hours there is interstitial emphysema at right lower lobe (RLL) leading to tension pneumothorax, **c)** there is persistent interstitial emphysema at RLL after chest tube drainage of pneumothorax and **d)** 3 weeks of age there is progressive air trapping and multicystic changes at RLL.

CT evaluation again demonstrated increasing air trapping in the RLL and mass effect and additional atelectatic changes in the remaining right lung and left lung (**Figure 2a and 2b**). Perfusion radionuclide study demonstrated diminished perfusion in the region of RLL. Initially a rigid bronchoscopy was attempted but this was complicated by bilateral pneumothorax and as the patient became unstable and needed resuscitation therefore the procedure was abandoned. Four days later he tolerated extubation and did well subsequently. Two months later he was weaned from oxygen therapy and was discharged from the hospital at 5 months of age when plain chest radiography showed a thin-walled cystic lesion in the RLL without any major signs of air trapping. Observations of plain film were confirmed by CT examination, which showed 3-4 cm thin-walled cavities at RLL, surrounded by normal lung. At 4 years and 6 months of age he was thriving well and has been asymptomatic for 4 years. Plain radiography and CT scans persistently show a small residual cyst in the RLL (**Figure 3a and 3b**).

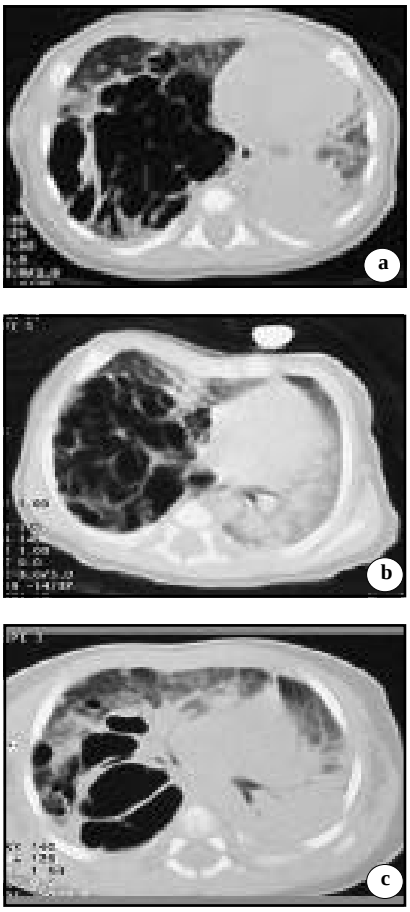

Figure 2 - Computed tomography of chest at **a)** 3 weeks of age revealing multiple irregular air filled spaces in right lower lobe with associated mass effect, **b)** follow up at 8 weeks showing progression of the lesion and **c)** section below the carina demonstrating compression of right lower lobe bronchus.

Discussion. Acquired lobar emphysema, also described as lobar over-inflation,[1] is a well-known complication of chronic pulmonary disease of prematurity. The pathogenesis of ALE is unknown but considered multifactorial; barotrauma, oxygen toxicity and lung immaturity are presumed to play important roles. An affected lobe shows hyperinflation and subsequent destructive emphysema.[1-2] Many patients may have obstructive endoluminal lesions such as endobronchial granulomata. Severe localized bronchomalacia is one of the major anatomical factor. Dynamic hyper-collapsibility at expiration of an affected

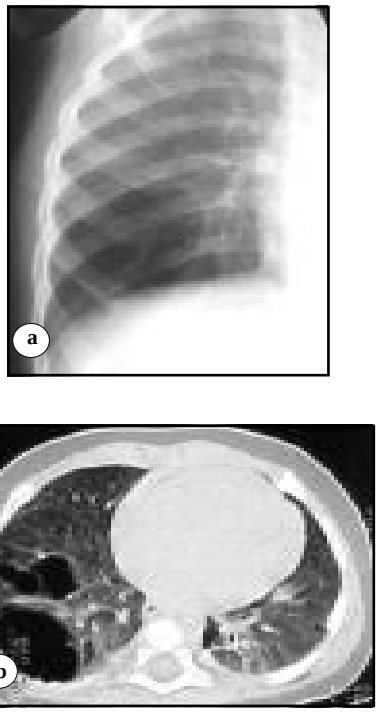

Figure 3 - **a)** Plain radiography and **b)** computerized tomography examination at 24 months demonstrating residual cystic lesions at right lower lobe.

bronchus is considered an etiological factor for bronchial obstruction. The obstruction is aggravated by mucus secretions, hyperplasia of bronchial mucosa and peribronchial fibroplasia. Endoscopic confirmation of such an event is often demonstrated.[1] It is not clear whether collapsibility of the bronchus is primarily due to a decrease in bronchial cartilage, a qualitative defect in the cartilage or a decrease in the bronchial luminal area. Morphometric features correspond poorly to demonstrable airway collapse and studies with age-matched autopsy controls[1] have shown no appreciable differences for bronchial cartilage, internal luminal area or mean cartilage per wall area. Also, there is no evidence of quantitative differences of bronchial cartilage and diminished luminal area. Imaging of ALE has been limited mainly to plain radiography until recently when CT and radionuclide studies became common place. Serial radiographic assessment of the radiograph remains the prime evaluation tool in these critically ill patients but there is an increasing contribution from cross sectional studies[2] although published CT evaluations of lung changes in ALE are few. In the evaluation of acquired lobar emphysema CT offers several advantages over plain radiography; in the early stage of assessment focal hyperinflation is detected earlier by CT scanning, although this finding may not have a clear predictive value regarding the progression, CT is superior in defining the extent of total involvement and in differentiating hyperinflation versus emphysema which might have prognostic value; also there is demonstration of bronchial collapse that can be shown by multiplanar reconstruction and 3D imaging of the proximal bronchi. Virtual endoscopic views and real time volume rendered images may be of great value in this situation.[3] In our case CT study and serial follow up aided in the precise delineation of the extent of the lesion and evolution of the pathology over period of time. Demonstration of residual changes in the involved lobe was far superior in the CT examination. Bronchial collapse and focal hyperinflation were demonstrated clearly by CT (**Figure 2c**). Major limitations of CT in such patients are the possible adverse effects of transporting and performing the examination in severely ill, unstable patients plus the additional radiation burden, which can be minimized to some extent by performing a tailored examination utilizing a low mAs technique.[4-5] In our view, CT examination and serial follow up is more precise in managing patients with ALE. Careful attention to technique and tailored examination, especially on follow up will limit the radiation dose to a great extent. Ventilation and perfusion scans (V/Q scan) demonstrate the expected photon defect in the affected area. The non-ventilated, non-perfused state of the involved lung may support a decision to perform surgery.[6-7] Dynamic magnetic resonance imaging may have a future role in the evaluation of proximal bronchi by showing expiratory airway collapse.[6] Any decision regarding surgical intervention is based mainly on clinical grounds and on failure of non-operative management. Often bronchoscopy is performed before considering lobectomy. Failure of medical therapy, progressive disease, recurrent respiratory distress, persistent atelectasis, hyperinflation and stridor are some of the common indications for bronchoscopy. Computed tomography examination complements patient evaluation at this stage. Due to the complex nature of the disease and its unpredictable course, long-term outlook is poor. There are no well-documented studies in the literature showing residual pulmonary parenchymal changes in the survivors. Our patient had a plain film and CT examination 24 months after the onset of ALE, showing a thin-walled cavity in the RLL, minimal incidental changes in the upper lobe and left lung in the form of subsegmental atelectasis and hyperinflation. The extent of residual lung changes has a bearing on the clinical symptomatology. Pulmonary function studies may further complement the assessment of a patient's respiratory status.

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