

Evaluation and treatment of emphysema in a preterm infant

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Citation

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Abstract

Infants delivered prematurely often have respiratory distress related to surfactant deficiency or pneumonia. The current case describes a premature neonate who presented with increased work of breathing and recurrent left pneumothorax. Although chest x-ray showed left upper lobe changes consistent with pulmonary interstitial emphysema, subsequent CT scan performed due to recurrent pneumothoraces suggested possible congenital lobar emphysema. This case demonstrates the importance of CT imaging in obtaining an accurate diagnosis in a premature infant with respiratory distress and atypical x-ray findings.

INTRODUCTION

Pulmonary interstitial emphysema (PIE) is a well-recognized complication of premature infants with respiratory distress syndrome (RDS), pneumonia, and mechanical ventilation. Occasionally, it arises spontaneously with no respiratory symptoms or mechanical ventilation (1).

PIE is a form of air leak with accumulation of air in the interstitium. (1). Infants with PIE are usually normal at birth, with symptoms developing over the first few days of life. PIE may be diagnosed by chest x-ray, but in cases where there is uncertainty; CT scan is more sensitive in determining the extent and etiology of the hyperinflation. Congenital Lobar Emphysema (CLE) is a rare pathological condition resulting in hyperinflation of a pulmonary lobe, air trapping, and frequently a mediastinal shift.

CASE

A 2465g male infant born at 34 2/7 weeks of gestation after preterm labor, with Apgar scores of 8 and 8 at 1 and 5 minutes respectively. By 6 minutes of life he showed signs of respiratory distress with grunting and retractions. The infant was transferred to the special care nursery for further evaluation. On examination, he was pink in room air with intercostal and subcostal retractions and oxygen saturation 80%. Initially he was placed in an oxyhood to maintain oxygen saturations > 92%. Arterial blood gas was within normal limits. Chest x-ray was performed and interpreted as normal (Figure 1). Four hours after birth he had worsening symptoms, and was switched to nasal CPAP with marked improvement. A subsequent chest x-ray was interpreted as consistent with pneumonia. Chest x-ray performed after

recurrent respiratory distress at 20 hours after birth revealed a left tension pneumothorax (Figure 2). A needle thoracentesis was performed and 370 cc of air was evacuated from the left side of the chest. The infant was intubated, and a second thoracentesis drained 400cc air followed by chest tube placement with continuous air leak. After 2 days the air leak resolved. The infant was extubated and the chest tube removed.

The infant progressed to ad lib feeding on room air. Serial chest x-rays demonstrated progression of left upper lobe cystic changes. Seven days after the chest tube was removed he developed acute severe increased work of breathing. Chest x-ray revealed a tension left pneumothorax. A needle thoracentesis was performed, followed by a chest tube, and intubation (Figure 3). A CT scan showed a cystic lesion consistent with PIE (Figure 4). The patient underwent a left thoracostomy with resection of the congenital emphysematous segment. Pathology was consistent with pulmonary interstitial emphysema (Figure 5). He tolerated the procedure well, had an uneventful postoperative course, and was discharged from the hospital on post-operative day 7.

Figure 1

Figure 1. AP chest radiograph of the neonate done after birth. Interpreted as normal

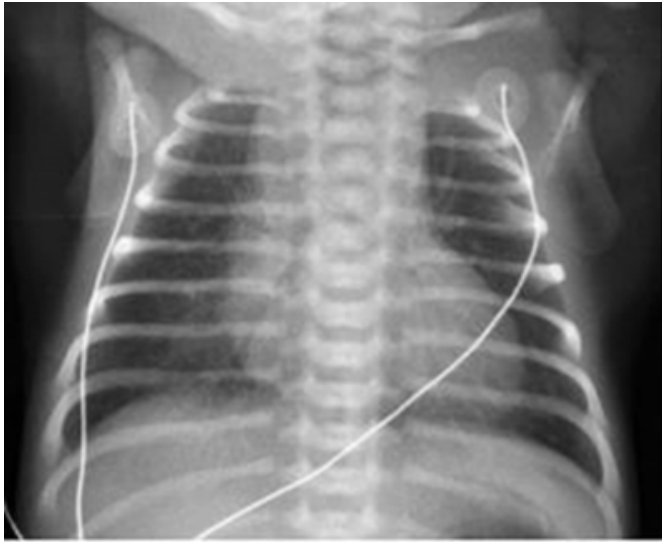


Figure 3

Figure 3: AP chest radiograph with left chest tube in place, with minimal left



Figure 2

Figure 2. . AP chest radiograph interpreted as left pneumothorax

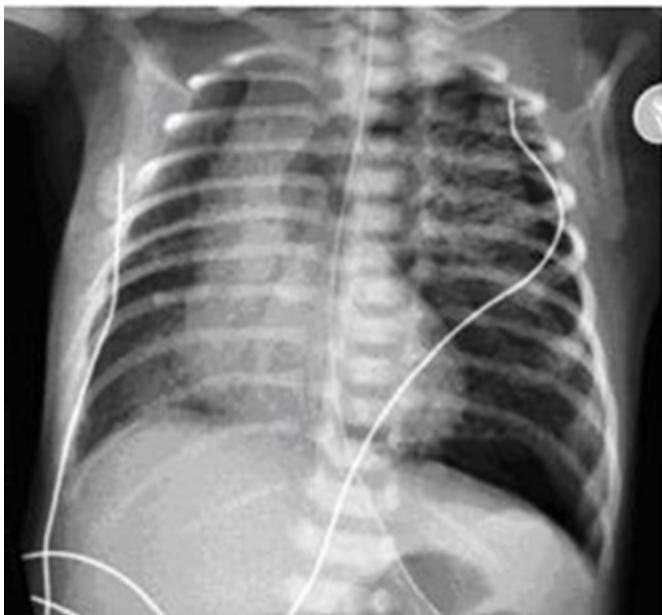


Figure 4

Figure 4: CT scan of the chest shows left cystic changes consistent with lobar emphysema.

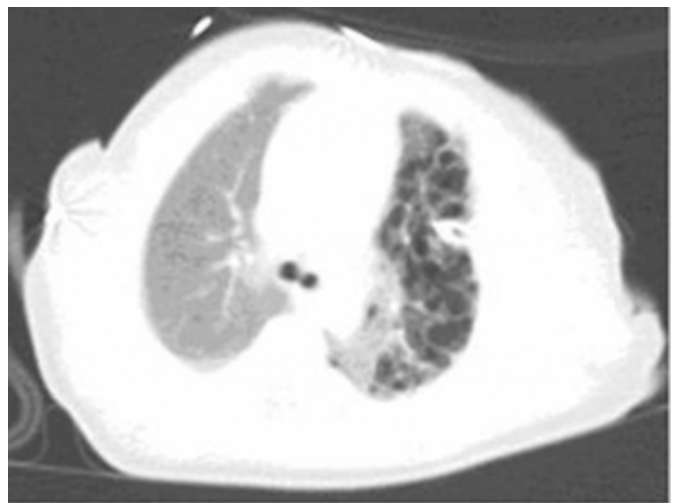
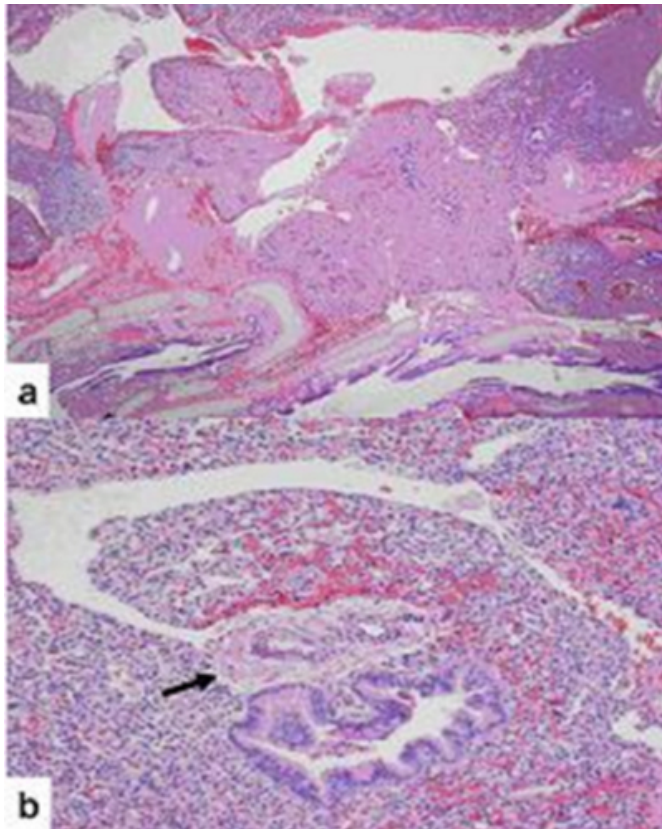


Figure 5

Figure 5: a) Air in intralobular septa creates a lobular appearance, with atelectatic lung parenchyma in this left upper lobectomy specimen. Blood in spaces is probably related to thoracentesis procedures (hematoxylin & eosin, original magnification x12.5). (b) A bronchovascular bundle adjacent to an intralobular septum with air contains a pulmonary vessel with a thickened wall (arrow), consistent with persistent pulmonary hypertension (hematoxylin & eosin, original magnification x100).



DISCUSSION

Air leak syndrome (ALS) has been described among ventilated and non-ventilated infants with RDS (2). The pathogenesis of spontaneous ALS developing immediately after birth has been postulated by Chernik and Avery (3). Pulmonary interstitial emphysema (PIE), a subset of ALS, is relatively common in very low birth weight infants, with a reported incidence of 2–4% (5). While it is associated with mechanical ventilation, there are cases of PIE in non-ventilated and CPAP supported preterm infants (4, 6). PIE has three distinct forms: acute PIE, localized persistent PIE, and diffuse persistent PIE. In localized persistent PIE (LPPIE), air tracks from the interstitium into adjacent spaces. This can cause pneumothorax, pneumomediastinum, or pneumopericardium. (7-9). LPPIE is associated with RDS, but can develop spontaneously. The diagnosis of

LPPIE is usually made by chest x-ray.

The radiographic finding of LPPIE mimics that of a pneumothorax and may prompt the placement of a chest tube. This can result in serious morbidity and mortality, as the placement of a chest tube in a patient with LPPIE often results in lung puncture and development of bronchopleural fistula. The chest x-ray findings are usually diagnostic or suggestive of LPPIE, but there are a number of other diagnoses that share similar radiographic finding, including pneumothorax, tension pneumothorax, atelectasis, congenital lobar emphysema, diaphragmatic hernia, pneumatocele, mucous plug, cystic adenomatoid malformation, or external compression by a large pulmonary artery. In cases in which diagnosis is not completely clear on plain radiographs, CT scan of the chest often provides the definitive diagnosis. (2, 7, 8)

There is no definitive treatment for LPPIE. Management is supportive and depends on whether the disease is localized or diffuse. Supportive methods include decreasing the mean airway pressure as much as possible and placing the infant in the lateral decubitus position with the affected side down (11,8,12). Surgical resection of the involved lobe may be indicated when a relatively large cystic area produces persistent respiratory distress or pneumothoraces. (8)

The present case demonstrates the value of a CT scan of the thorax in making a diagnosis and guiding therapy.

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