Persistent Pulmonary Interstitial Emphysema in a Preterm Infant

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Abstract

Persistent pulmonary interstitial emphysema is a chronic disease reported in mechanically ventilated premature newborns. We describe a case of localized persistent pulmonary interstitial emphysema in a preterm infant without mechanical ventilation but on continuous positive airway pressure using nasal prongs. The condition resolved without surgery.

Key words: *Persistent pulmonary interstitial emphysema, Preterm, Respiratory distress syndrome.*

Pneumothorax, pneumomediastinum, and pulmonary interstitial emphysema (PIE) are the most common causes of air leak syndromes in the newborn period(1). Air escapes from intraalveolar space to interpleural area in pneumothorax and to mediastinal area in pneumomediastinum(1). In PIE, air accumulates in the interstitial tissues of the lung along the bronchovascular bundles and the interstitial septa leading to the formation of cysts(2).

Advanced use of mechanical ventilation and the increased survival of premature infants, has led to a rise in the incidence of PIE. Pulmonary interstitial emphysema may develop in preterm newborns treated with assisted ventilation such as nasal continuous positive airway pressure (CPAP). Air leaks, as well as pneumothorax do occur on CPAP, which may be related to overdistention of alveoli(2).

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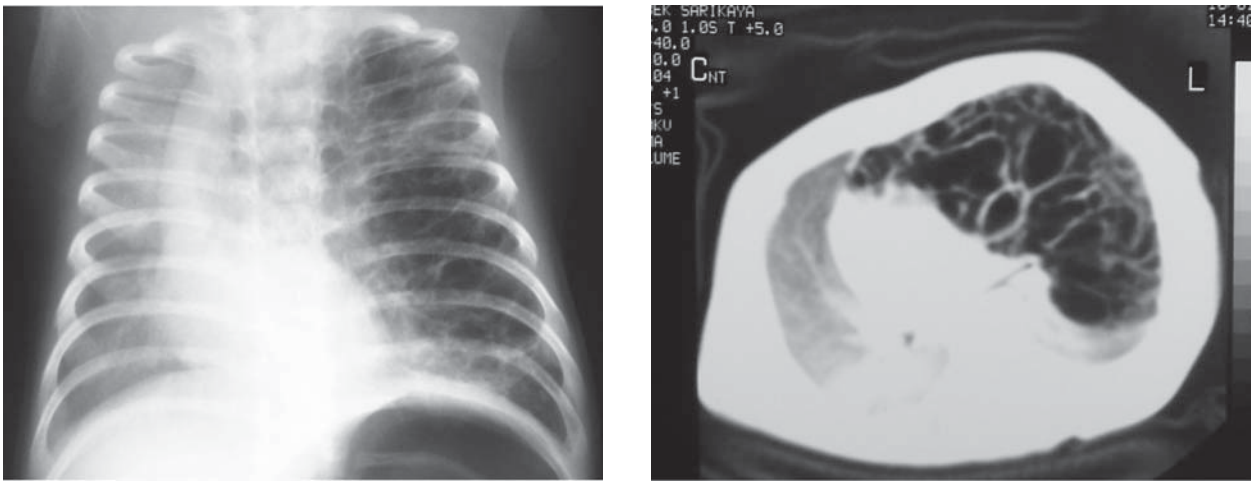
CASE REPORT

A male infant was born to a 32 years old multigravida mother via spontaneous vaginal delivery at 33 weeks of gestation with a birthweight of 2440 grams. The Apgar scores were 7 and 8 at 1 and 5 minutes. Because of mild respiratory distress, nasal CPAP was instituted at 5 cm H_2O (Hudson nasal prongs). The initial findings on chest radiography were compatible with mild respiratory distress syndrome (RDS). The child improved and required nasal CPAP only until his third day of life. The child was discharged at 7 days of age.

On 24th postnatal day, the infant was admitted to our children's hospital with respiratory distress. Physical examination revealed tachypnea, intercostal retractions and mild cyanosis. Lung fields were clear, and there was no cardiac murmur. Initial laboratory studies, blood gases, complete blood count and C-reactive protein were normal. Chest X-ray showed paracardiac pulmonary densities and normal heart size and pulmonary vasculature.

Treatment was started with oxygen by hood (5 L/min) and ampicillin and gentamicin. The infant's clinical status stabilized within 10 days, but retractions persisted. On hospital day 13, the infant's retractions increased, and a chest film at this stage revealed diffuse cystic changes in the left lung (***Fig.* 1*a***), which was confirmed on CT (***Fig.* 1*b***). Repeated radiographs of chest revealed continued increase in left lung cystic lesions in the absence of further respiratory support. Given the diagnosis for localized persistent pulmonary interstitial emphysema (LPPIE), pediatric surgery unit offered left upper lobectomy. At this stage, we decided to manage the baby conservatively. Serial radiographs



(*a*) (*b*)

FIG.1(*a*) *Diffuse cystic changes in the left lung.* (*b*) *Thoracic CT revealed multiple cysts of varying sizes occupying the entire of the left upper lobe, atelectasis in the left lower lobe and a mediastinal shift to the right.*

revealed slow resolution of LPPIE. The child recovered and was discharged on day 30 without tachypnea and retractions. There was no respiratory symptoms on follow-up after 1 and 2 month.

DISCUSSION

Pulmonary interstitial emphysema is a peribronchial, perialveolar air-leak syndrome, and can be classified clinically as acute (<7 days duration) or persistent(3). Persistent pulmonary interstitial emphysema (PPIE) is subclassified as localized or diffuse, according to the extent of involvement(4,5). Most cases of air leak syndromes occur in newborns with underlying lung disease, especially if mechanical ventilation is required(6).

In PIE, air enters the pulmonary interstitial tissue following rupture of an overdistended alveolus, and dissects along the periarterial sheath(7). Preterm infants are at increased risk because the perivascular connective tissue is more abundant and less dissectible in preterm than older infants. This predisposes to air trapping in the perivascular space resulting in pulmonary interstitiel emphysema (PIE)(1).

A number of adverse side effects and complications of CPAP have been described(8). It is not possible to conclusively identify that how CPAP led to the development of PIE in our infant. But it is claimed that air leaks, as well as pneumothorax do occur on CPAP. The mechanism may be related to overdistention of the more compliant areas of the lung(8). Localized PPIE may evolve from structural characteristics of the immature lung. This immaturity may explain the vulnerability to alveolar rupture of the preterm lung even in infants who are not mechanically ventilated, as in our case(9).

The differential diagnosis for cystic pulmonary changes includes congenital cystic adenomatoid malformation, lymphangiectasia, bronchogenic cysts, congenital lobar emphysema, cystic lymphangioma, diaphragmatic hernia and sequelae of prior infection(3). It is important to distinguish LPPIE from other cystic lesions because the initial management of LPPIE may be nonoperative.

There is little empirical data available to guide the management of infants suffering from LPPIE. In four of the previously reported LPPIE cases without history of mechanical ventilation, the infants underwent surgical lobectomy or pneumonectomy(3). We tried a conservative approach, maintaining spontaneous breathing by positioning the infant with the involved side down and, antibiotics and oxygen therapy.

Our case indicates that LPPIE may occur in

preterm infants who have been treated with nasal CPAP and these cases can be successfully managed nonsurgically, if the diagnosis can be made by imaging prior to operative intervention.

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Biotinidase Deficiency

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ABSTRACT

A three month old baby presented with refractory seizures, dermatosis and persistent metabolic acidosis. Biotinidase deficiency was diagnosed on enzyme assay. Patient responded dramatically to biotin supplementation.

Key words: *Biotinidase deficiency, Inborn error of metabolism*

Biotinidase deficiency, first described in 1983, can be profound (<10% enzyme level) or partial (10-30% enzyme level). Clinical manifestations include neurological, dermatological, immunological, and ophthalmological abnormalities. We report a case of profound biotinidase deficiency(1).

CASE REPORT

A 3-month old boy, born of nonconsanguineous marriage, presented with sudden onset of breathlessness, lethargy and refusal of feed for twelve hours. He had one episode of multiple seizures at 2 months of age. MRI brain at that time revealed ischemic changes in bilateral parietal lobe

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