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A Case of Localized Persistent Interstitial Pulmonary Emphysema

Interstitial pulmonary emphysema is a well-documented complication of assisted mechanical ventilation in premature infants with respiratory distress syndrome. Localized persistent interstitial pulmonary emphysema (LPIPE) confined to a single lobe was incidentally presented in a 4-day-old female infant. This patient was a normal full-term baby with no respiratory distress symptom and no experience of assisted mechanical ventilation. Chest radiograph showed radiolucent area in right lower lobe zone, which needed differential diagnosis from other congenital lesions such as congenital cystic adenomatoid malformation and congenital lobar emphysema. CT scan showed irregular-shaped air cystic spaces and pathologically, cystic walls primarily consisted of compressed lung parenchyma and loose connective tissue intermittently lined by multinucleated foreign body giant cells.

Key Words: *Emphysema; Pulmonary Emphysema, Interstitial; Localized Persistent Interstitial Pulmonary Emphysema; Cystic Lung Disease; Lung Diseases*

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INTRODUCTION

Interstitial pulmonary emphysema in the newborn is a frequent complication of assisted mechanical ventilation. It most commonly occurs in premature infant with respiratory distress syndrome, but occasionally, it arises spontaneously with no respiratory symptom (1, 2). Localized persistent interstitial pulmonary emphysema (LPIPE) is a rarer form of interstitial emphysema than diffuse form, with a propensity for localized progressive accumulation of air in the interstitium. This accumulation results in cystic air spaces that are typically associated with mediastinal shift and progressive respiratory distress. To our knowledge, this is the first reported case in Korea, and we therefore describe the pathologic characteristics and review of literatures.

CASE REPORT

A 4-day-old female infant (normal full-term spontaneous delivery, 3.25 kg of birth weight) was admitted to the hospital due to neonatal jaundice with total bilirubin level of 18.1 mg/dL. The baby was otherwise in good general condition. She was placed under phototherapy, and jaundice gradually improved to the total bilirubin level of 11.1 mg/dL on her 8th day of hospitalization. Incidentally, radiolucent area was noted in the right lower lung field of chest radiograph (Fig. 1). Mediastinal shift was not evident. On physical examination, the breathing sound over the posterior wall of right lower chest was somewhat low. The rest of the chest was clear to auscul-

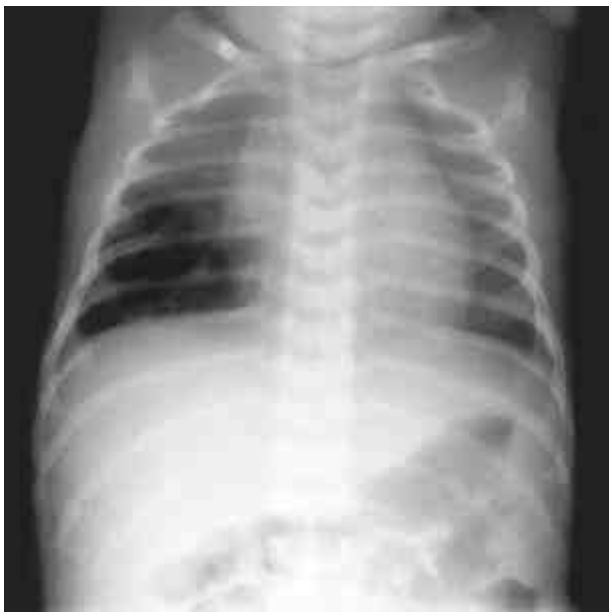

Fig. 1. Chest radiograph shows radiolucent area in the right lower lung.

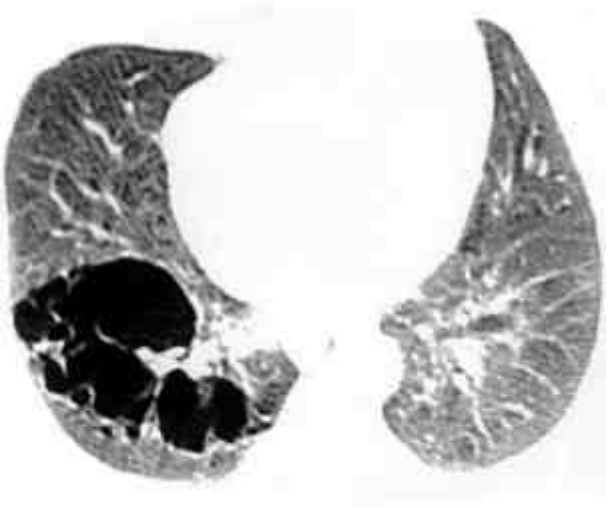

Fig. 2. High resolution CT scan with lung window setting shows irregular-shaped cystic air spaces, separated by fine or coarse septae in the right lower lobe.

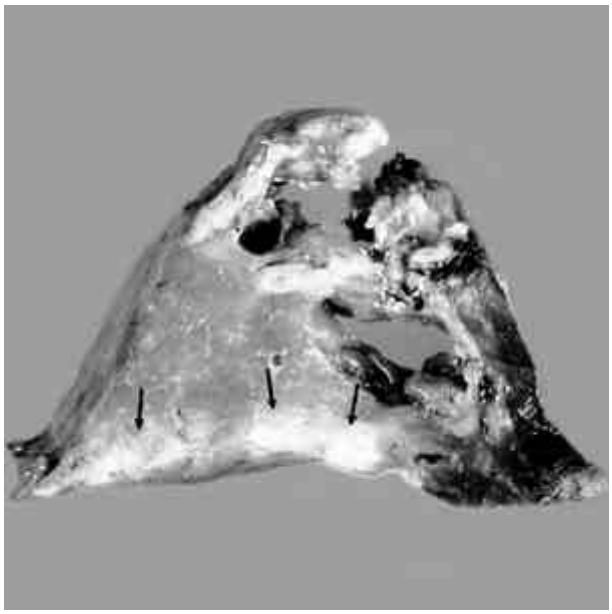

Fig. 3. Formalin-fixed resection specimen of right lower lobe shows intercommunicating, irregular-shaped air space with yellowish patchy areas of atelectasis (arrows).

tation. High resolution computed tomogram (CT) and spiral chest CT angiogram were done. High resolution CT revealed air cysts of irregular shapes, ranging in size from few millimeters to 2.5 cm in the right lower lobe (Fig. 2). The radiologic differential diagnoses of congenital cystic adenomatoid malformation and segmental bronchial atresia with lung overinflation were made. The diagnosis of LPIPE was not considered preoperatively because of no symptom of respiratory distress and no use of assisted mechanical ventilation. The baby was discharged after jaundice was improved. On follow-up chest radiograph, the lesion became larger. The right lower lobectomy was done on the 36th day of life. During operation, the cyst partially collapsed after air leaked out from the cystic lesion. Pathologic examination of the right lower lobe showed irregular-shaped, multiloculated air cysts, measuring up to 2.5×1.5 cm with patchy areas of mild atelectasis (Fig. 3). Microscopically, the striking abnormality was observed with the presence of large air cysts. The cystic wall primarily consisted of compressed parenchyma and loose connective tissue intermittently lined by multinucleated foreign body giant cells (Fig. 4). These air spaces commonly surrounded the bronchovascular bundles (Fig. 5). Some portions of cystic wall showed fibrotic change with variable thickness. No cyst was lined by anything resembling bronchial or bronchiolar epithelium. Small cysts were mostly located in interlobular septa (Fig. 6). These pathologic findings were consistent with LPIPE. After surgery, she has been doing well without any problem.

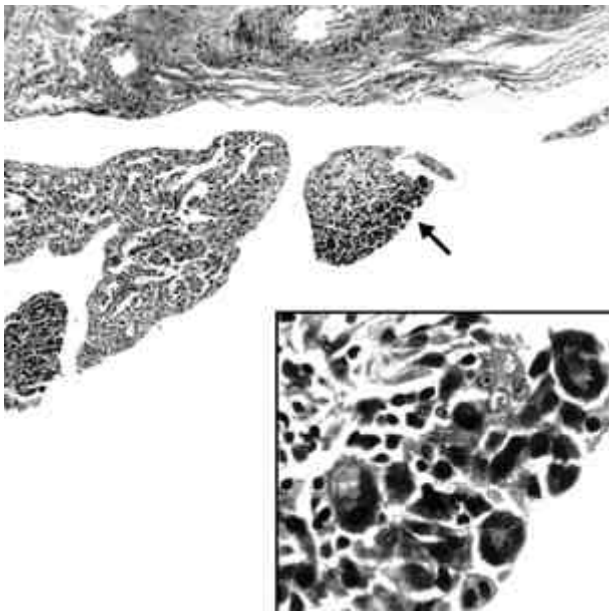

Fig. 4. The cystic walls are primarily consisted of compressed parenchyma and loose connective tissue, intermittently lined with multinucleated foreign body giant cells (arrow) (H&E, × 100). Inset: magnification of multinucleated foreign body giant cells (H&E, ×400).

DISCUSSION

Macklin and Macklin (3) demonstrated in the animal experiment that an increase in the alveolar pressure over

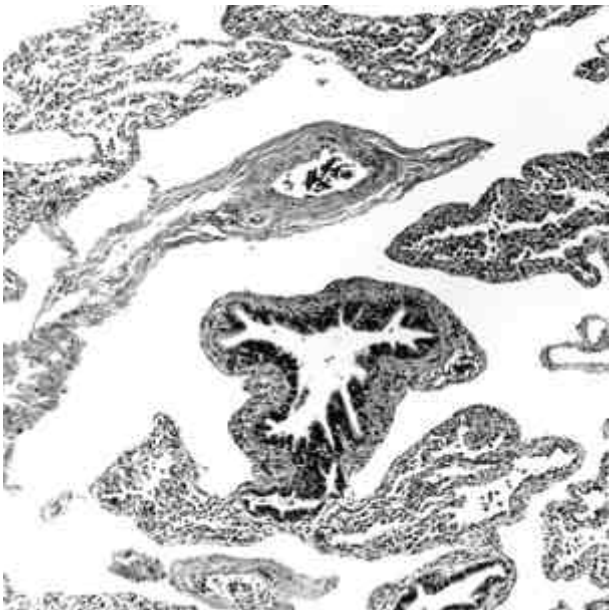

Fig. 5. Air cysts are seen around the bronchovascular bundles (H&E, ×40).

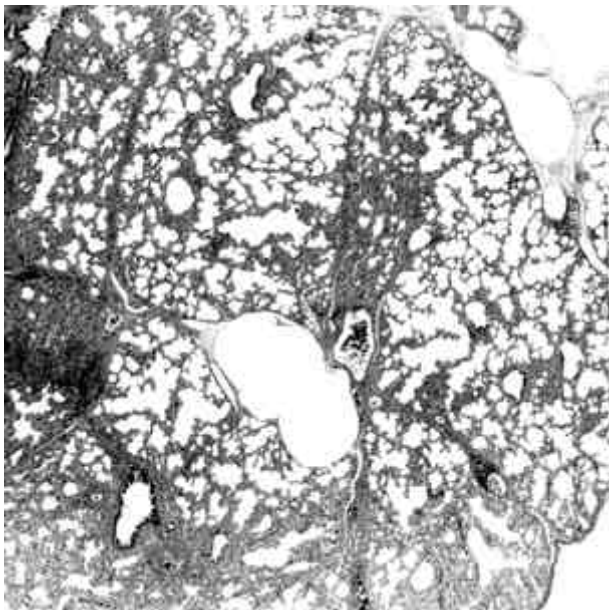

Fig. 6. Small cysts are mostly seen in the interlobular septae (H&E, ×10).

physiological limits can produce a rupture of alveolar base at its junction with the less expandable fluid-rich perivascular connective tissue. After that, air leaks into the perivascular sheath and dissect it, giving rise to interstitial emphysema, pneumomediastinum, and pneumothorax. In newborn, aspiration of foreign material such as meconium, or partial occlusion of bronchi or bronchiole increase intraalveolar pressure and may develop interstitial pulmonary emphysema. It tends to occur during the first few breaths because the alveolar pressure is further increased by the high pressure applied across the lung right after delivery (4).

Interstitial pulmonary emphysema can be acute (less than 7 days duration) or chronic (or persistent) and may be localized to a single or more lobe, or distributed diffusely through all lobes (1). The three forms of intersitial pulmonary emphysema (acute pulmonary interstitial emphysema, LPIPE, diffuse persistent interstitial pulmonary emphysema) have different clinical, radiologic and pathologic features.

Clinically, acute pulmonary interstitial emphysema most commonly develops with hyaline membrane disease. Less commonly it could develop with air being trapped by obstruction due to meconium aspiration or pulmonary hypoplasia, requiring high ventilatory pressure. LPIPE develops usually in infants with hyaline membrane disease but could also occur spontaneously with no apparent lung disease or, as in our case, with no assisted mechanical ventilation (1, 2). Diffuse persistent pulmonary interstitial emphysema is most commonly associated with bronchopulmonay dysplasia (1, 5).

Radiologically, LPIPE can be differentiated from acute pulmonary interstitial emphysema and diffuse persistent interstitial pulmonary emphysema by its expansile multicystic appearance. The cystic air spaces are localized to one or more lobes and are associated with mediastinal shift and mass effect (1, 5). Other localized radiolucent chest lesions including congenital cystic adenomatoid malformation and congenital lobar emphysema show similar findings in simple chest film, and therefore it is necessary to distinguish them from LPIPE. CT can be used for definitive diagnosis (5-7) which may help to avoid unnecessary surgery because the initial treatment of LPIPE can be medical (selective intubation and selective bronchial obstruction or decubitus positioning) as opposed to the surgical treatment required for other congenital cystic lesions (8, 9).

Pathologically, LPIPE appears as irregular-shaped, multiloculated air cysts along the interlobular septa and bronchovascular bundle. These cystic spaces show variable size, measuring up to 3 cm (1). The cystic wall shows variable degrees of fibrosis, possibly related to the duration. Small cysts are mostly located in the interlobular septae, which are considered as early lesions of this disease. Bronchovascular bundles may be seen as projections into the cyst walls. A multinucleated giant cell reaction that may form a partial lining of air spaces is a characteristic histologic feature. The origin of giant cell is unclear, but two possibilities have been considered: 1) giant cells as reaction to the content of alveolar sac or 2) as reaction to air (1, 10). Wright (10) demonstrated the giant cell reaction to the injection of oxygen or

nitrogen into the subcutaneous tissue of animals.

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