

Cystic Intrapulmonary Lymphangioma : Cystic Lung Disease Presenting as Pneumothorax in a 6 Month Old Male Baby

(SNUH CPC 94-2)

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CASE HISTORY

This six month-old male baby was brought to Seoul National University Children's Hospital(SNUCH) for the further evaluation of known cystic lung lesion on January 21, 1994. He was delivered at the gestational age of 35 weeks due to maternal preeclampsia on July 16, 1993. The birth weight was 1.85 kg and history of other perinatal problems was denied except incubator care for 13 days.

On January 2, he was brought to a general hospital due to poor oral intake and diarrhea along with associated URI symptoms including noisy respiration. He had been admitted at that hospital and supportive management was performed under the clinical impression of bronchiolitis. During the admission, pneumomediastinum and subcutaneous emphysema developed, and on January 9, thoracotomy was conducted due to left-sided

pneumothorax. Pleural effusion was found on January 15.

The chest radiograph on January 4 revealed multiple small cystic lesions involving the left upper and lower lung field which increased in size in a subsequent radiograph along with subcutaneous emphysema of the neck and chest on January 6. Interval chest radiographs revealed tension pneumothorax on January 8 and pleural effusion.

The chest CT taken on January 20 demonstrated multicystic lesions involving lingular segment of left upper lobe and superior and basal segments of left lower lobe. There were right-sided mediastinal shifting and pleural thickening of left lung.

Under the clinical impression of congenital cystic lung disease, he was transferred to for the further evaluation. On physical examination, the lung sound was coarse and the heart sound was normal. The vital signs were as follows; blood pressure 80/50 mmHg, heart rate 136/min, respiratory rate 54/min, body temperature 37.2°C. There was frequent coughing. Other laboratory findings were within normal range and the arterial blood gas analysis showed pH 7.38, PCO₂ 33mmHg, PO₂ 177 mmHg, and HCO₃⁻ 19mmol/l.

Left pneumonectomy was performed on January 24, and chylous pleural effusion was

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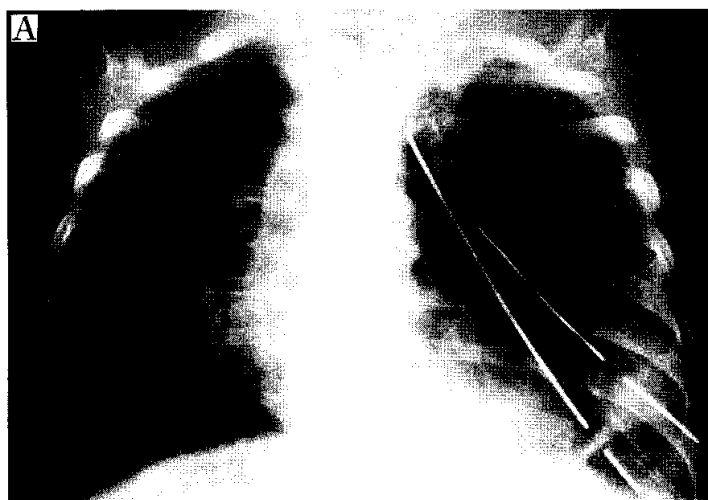


Fig. 1. Cystic lung lesion in a 6-month-old male baby.



A. Chest radiograph obtained at admission shows multiseptated cystic lesion occupying the left upper and middle lung zone. The lesion is located just lateral to the left hilum. Small amount of left pleural effusion after insertion of two chest tubes is also noted.

B. Thin-section (1.5mm collimation) CT scan obtained at level of thoracic inlet shows multiseptated cystic lesion. Also noted are thickened lobular septae (arrows) and bronchovascular bundles (arrowheads).



C. CT scan obtained at ventricular level shows a large multiseptated cystic lesion. Again note the thickened lobular septae and bronchovascular bundles.

found on operation.

DISCUSSION

Dr Lee: The patient was admitted to SNUCH with known cystic lung disease. On radiographs the main lesion showed air-filled multiseptated cystic appearance (Fig. 1A). Associated findings were pneumothorax and pleural effusion. In addition to these findings, the thickened interlobular septa and bronchovascular bundles were observed in the peripheral lung on CT scans (Fig. 1 B and C). The main cystic lesion occupied the central lung near the pulmonary hilum (Fig 1 C). In the following discussion, I would like to introduce the pulmonary diseases manifesting as a cystic lung lesion in infancy or childhood. After then, I will differentiate the diseases with emphases on the radiological findings. The cystic lung lesions in infancy or childhood are summarized in Table 1 (Hernanz-Schulman, 1993). First of all, I would like to begin with congenital cystic adenomatoid malformation (CCAM).

CCAM represents an abnormal proliferation of mesenchymal elements and failure of maturation of bronchiolar structures, occurring at approximately the fifth to sixth week of gestation. The lesions have been divided into three types reflecting both anatomic and clinical differences. The type 1 lesion comprises approximately 50% of cases, and consists of few, large cysts, usually one to four in number, ranging in diameter between 3 and 10cm. Type 2, accounting for approximately 40% of cases, consists of more numerous cysts of small to medium size, generally ranging between 0.5 and 3cm in diameter, diffusely distributed through the affected area. Type 3, which accounts for approximately 10% of cases, are solid to the naked eye, and consist of multiple minute cysts, usually 2 mm in diameter (Stocker *et al.*, 1977). This type tends to involve an entire lobe or an entire lung. The chest roentgenogram in the newborn presents a solid, space-occupying mass, which, in cases of type 1 or 2 malformations, becomes air-filled over

the course of days, weeks, or hours. The chest radiograph illustrates the internal cystic architecture of the mass and its lobar distribution. computed tomography (CT) details this anatomy, but care should be taken in localization of the anomaly when associated with lobar overexpansion.

Whereas CCAM originates in an intrapulmonary overgrowth and immaturity of bronchiolar tissue, sequestration is believed to result from an abnormally caudal accessory budding of the foregut, which dissociates from the tracheobronchial trees and retains its embryonic systemic arterial connection. The sequestration is usually subcategorized into extralobar and intralobar type. Current thinking favors the theory that intralobar sequestration occurs earlier, resulting in incorporation of the sequestered lung within the main pulmonary pleural envelope, whereas the extralobar variety occurs later, with resultant separation of the sequestered tissue within its own visceral pleural envelope (Shanji *et al.*, 1988). Extralobar sequestration is the disease of the infant whereas intralobar sequestration is seldom found in the infant and it does not achieve a comparable statistical representation in autopsy series of neonates (Stocker and Dehner, 1988). The presence of internal cystic spaces in utero may be the initial clue to the existence of this malformation. The initial chest roentgenogram will demonstrate a water density mass. When the cystic mass communicates with the airways mostly often due to bronchopulmonary infection, the lesion may appear with air-filled cyst. On CT, the lesion may show fluid or air-filled cysts with surrounding air-trapping. Solid portion may co-exist. The systemic supply arises from the thoracic aorta in most cases and from the supraceliac infradiaphragmatic aorta in a significant number, extended caudally by differential growth during embryonic life. Owing to the small size of the vessels and their generally coronal orientation, CT scanning is not as successful in demonstrating the vascular anomaly and therefore is not the procedure of choice in most cases. Arteriography is rarely necessary in

the preoperative evaluation of sequestration due to high diagnostic accuracy of Doppler and magnetic resonance(MR) imaging.

Cystic intrapulmonary lymphangiomas are extremely rare. They are composed of cystic spaces containing both fluid and air and in the neonate closely resemble CCAM, thus joining their differential diagnosis(Milovic & Oluic, 1992). Lymphangiomas are considered to be of developmental origin but are known to undergo cystic enlargement in response to localized or systemic infection.

Congenital cystic lesions usually presenting after newborn period include bronchogenic cysts, pulmonary parenchymal cysts and mesenchymal cystic hamartomas. Bronchogenic cyst results from an anomalous supernumerary budding of the ventral or tracheal diverticulum of the foregut during the sixth week of gestation and is thus part of the spectrum of bronchopulmonary foregut malformations(Rodgers *et al.*, 1986). The cysts have a fibrous wall containing cartilage and are lined by ciliated columnar epithelium. In most series, the most frequent location is mediastinum. In the typical case, intrapulmonary bronchogenic cyst is fluid-filled without communication with bronchial trees; repeated infection can cause erosion into a neighboring airway and an air-fluid level may become evident. Chest roentgenogram will reveal a round, water-density mass, in a perihilar location; an air-fluid level and/or thickened wall will be present in association with previous or current infection. A CT scan should be done to define the exact location of the cyst, assess its wall thickness and internal contents, and perhaps more importantly, determine whether an associated anomaly, such as sequestration, is present.

Pulmonary parenchymal cysts, usually located in the periphery of the lung, are believed to represent a disorder of bronchial growth, with separation of the distal bronchial generations from the main branches(Shanji *et al.*, 1988). Patent bronchial communication typically occurs, resulting in recurrent infection and/or air trapping, with development of a ten-

sion cyst. Rupture of a peripheral cyst into the pleural space can result in a spontaneous pneumothorax. When asymptomatic, they can be found incidentally. The cysts can be single or multiple. These cysts appear with thin-walled cysts with internal air filling or air-fluid level both on chest roentgenogram and CT scan.

Mesenchymal cystic hamartoma(MCH) can occur at any age. In the classic case, the lesion begins as a nodule that develops central cystic cavitation and progresses to involve both lungs. It occurs less frequently in children in contradistinction to CCAM, which it can superficially resemble radiographically(Mark,1986). Pathologically they are distinguished by the presence of primitive mesenchymal stromal cells in mesenchymal cystic hamartoma, whereas the stroma in CCAM is composed of fibrous tissue and other mature mesenchymal elements. On chest radiographs and CT scans, MCH appears with multiple bilateral cystic lesions containing possible some mural nodules.

Acquired cystic lesions presenting in children or young adults include inflammatory pneumatoceles and traumatic pneumatoceles. Inflammatory pneumatoceles result from a rapidly progressive inflammatory process. There is plugging of the smaller airways with destruction of dependent alveolar walls and concomitant cystic hyperexpansion of their air space. As the surrounding consolidative process resolves, the pneumatoceles concurrently become more apparent and/or enlarge; thus, they are usually seen during the resolution stage of the pneumonia. Radiographically, they appear as rounded lucent areas interspersed within densely consolidated background and when necessary, are best seen on CT scans. Pneumatoceles are found more often in children than in adults, perhaps as a result of greater susceptibility of the alveolar walls to tear.

Traumatic pneumatoceles are the infrequent accompaniment of blunt chest trauma and pulmonary contusion and are believed to result from tears within the pulmonary parenchyma caused by its sudden elastic recoil after

Table 1. Cystic lesions of the lung in pediatric chest

Congenital lesions usually presenting in the newborn period
Congenital cystic adenomatoid malformation
Sequestration
Cystic intrapulmonary lymphangioma
Congenital lesions usually presenting after the newborn period
Bronchogenic cyst
Pulmonary parenchymal cyst
Mesenchymal cystic hamartoma
Acquired lesions presenting in children or young adults
Inflammatory pneumatocele
Traumatic pneumatocele
Pulmonary cyst in systemic disease
Langerhans' cell granulomatosis

the rapid application of a compressive force. Traumatic pneumatoceles usually present within 12 to 24 hours of injury, although their appearance on the radiograph may await clearing of the enveloping pulmonary contusion, which begins to dissipate within 24 to 72 hours from the acute event. They may be multiple or multilocular (Fagan & Swischuk, 1976), a characteristic best disclosed on CT scanning. They may contain an air-fluid level, the latter representing extravasated blood. Paralleling the inflammatory pneumatoceles, traumatic pneumatoceles are self-limited, with a chronology analogous to that of pulmonary hematoma. Complete resolution occurs within 2 to 16 weeks.

Pulmonary cysts in systemic disease includes Langerhans' cell granulomatosis (previous histiocytosis X). It is a multisystemic proliferative disorder of unknown cause. Pulmonary involvement consists of granulomas encompassing alveoli and bronchioles, producing denudation of alveolar basement membranes, fibrosis, and destruction of pulmonary elastic fibers, leading to cavitation and emphysematous dilatation of air spaces (Fukuda *et al.*, 1990). Confluent involvement results in a honeycomb lung. These lesions are best evaluated with CT scans.

Among the above-mentioned entities, most probable diagnostic choices are CCAM, extr-

alobar pulmonary sequestration and cystic intrapulmonary lymphangiomas. Unilaterality of the lesion may preclude the possibility of mesenchymal cystic hamartomas and Langerhans' cell granulomatosis. Irregularity of the cystic wall with uneven thickness is the finding against the possibility of bronchogenic cyst or pulmonary parenchymal cyst. The patient did not have a history of chest trauma or overt staphylococcal pneumonia. These facts are also against the possibility of posttraumatic or postinfectious pneumatocele.

The presented case herein showed irregular multiseptated cystic mass along with the thickened interlobular septae and bronchovascular bundles on CT scans. The cystic mass was central in location and left upper lobar in distribution. The findings of the thickened interlobular septae and bronchovascular bundles are usually present in the conditions of lymphatic dilatation, malignant cell infiltration, pulmonary edema or pulmonary fibrosis. In this case, the findings were probably due to lymphatic dilatation. The lymphatic dilatation might have been caused by central cystic mass resulting in obstruction of lymphatic drainage or by the disease process per se. Usually CCAM appears as a lobar lesion and is located more commonly in the lower lobe. In extralobar pulmonary sequestration, the lesion is most commonly located in the left lower lobe. There is a

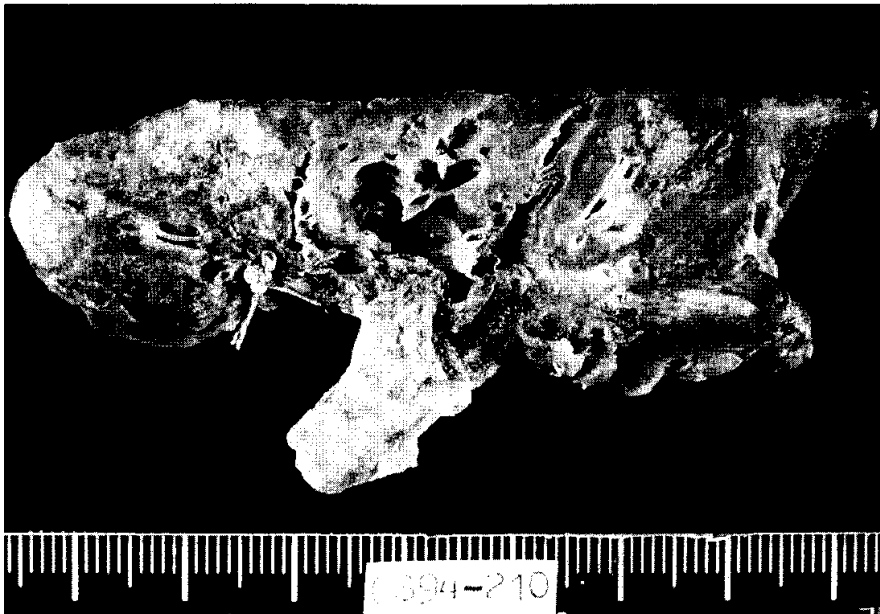


Fig. 2. Cut section of the removed lung shows multiple saccular lesions in the lung, that are particularly crowded in the hilar portion.

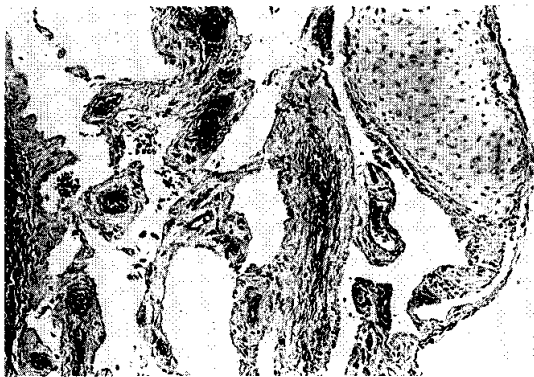


Fig. 3. Photomicrograph of the lung shows irregular dilated spaces involving bronchial wall (note cartilage island). These spaces are lined by a single layer of endothelial cells.

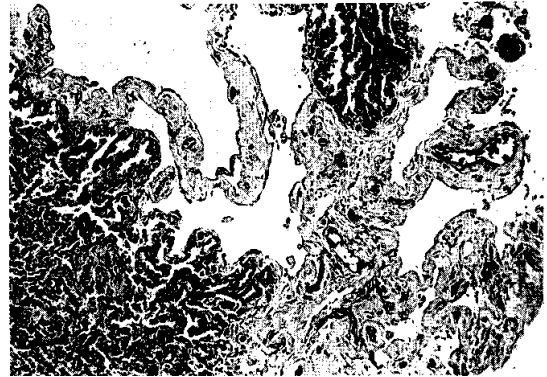


Fig. 4. Cystic lymphangioma extends to the pleura (right lower) and is associated with atelectasis of the lung parenchyma.

feeding vessel arising from the descending aorta. We could not find the feeding vessel in this case. The findings of the thickened interlobular septae and bronchovascular bundles suggesting lymphatic dilatation are not com-

mon accompaniments in CCAM or extralobar pulmonary sequestration. These findings are against the diagnosis of CCAM or extralobar pulmonary sequestration. Furthermore chylous pleural effusion strongly suggests that the lesion originated from the lymphatic tissue. The chylothorax may have been caused by central

cystic mass causing lymphatic obstruction and flooding. Although this is a long and time-consuming discussion, my first-choice diagnosis is cystic intrapulmonary lymphangioma with moderate degree of confidence.

Pathologic findings (Dr. Chong Jai Kim)

This is the pneumonectomy specimen of the patient. The main lesion is mainly confined to hilar portion of the left upper lobe and is composed of conglomerations of empty cystic portions of varying size ranging from 0.3cm to 1.5cm in diameter (Fig 2). The size of the lesion seems to be relatively small when compared with that of radiologic imaging, and this would be mainly due to collapse of air-filled spaces. The approximate size of the lesion is 3cm in diameter. The peripheral portion of the lesion is relatively well circumscribed from surrounding normal lung parenchyma. Histologically, the lesion showed characteristic features of cystic lymphangioma lined by flat endothelial cells, and it encroached bronchi and major vessels (Fig 3). Areas of immediate contact with bronchial epithelium and lymphangioma were frequently found. Associated secondary lymphangiectasia was found along the bronchovascular bundles and pleura (Fig 4).

The differential diagnosis of congenital lymphatic disorders on histological findings alone is very difficult or almost impossible in certain situations. The main differential diagnosis of the present case is a localized form of congenital pulmonary lymphangiectasia is a distinct clinicopathologic entity whose clinical course is mostly grave and the involvement is bilateral. Thus we think that the localized lymphatic lesions with rather benign clinical course like our case and previously reported cases of localized pulmonary lymphangiectasia around bronchovascular bundles and pleura seem to be a secondary phenomenon. In addition, frequent chylous pleural effusion in this patient is also not a feature of congenital pulmonary lymphangiectasia. The location of intrathoracic lymphangioma is usually mediastinum, and the primary intrapulmonary lymphangioma is an ex-

tremely rare occasion. The hitherto reported primary intrapulmonary lymphangioma under various headings in worldwide literature is less than ten cases. The lesion may be potential differential diagnosis of congenital cystic lung disease such as congenital cystic adenomatoid malformation or lobar emphysema.

The dilated lymphatics of the present case were very closely apposed with bronchial epithelium, making direct communication with airway by even trivial trauma. This finding seems to be the basic mechanism of progressive air-trapping in the main lesion. The marked increase in size of the main lesion since 6 months of age in this patient should have been related to increased air content.

Pathological Diagnosis :

Cystic intrapulmonary lymphangioma

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