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Unusual Bronchopulmonary Foregut Malformation Associated with Pericardial Defect: Bronchogenic Cyst Communicating with Tubular Esophageal Duplication

We report a case of unusual bronchopulmonary foregut malformation composed of a mediastinal bronchogenic cyst with sequestrated lung tissue and communicating tubular esophageal duplication associated with complete pericardial defect. A 18-yr-old man, who had suffered from dry cough and mild dyspnea, was admitted because of an incidentally detected chest mass. A computed tomography scan demonstrated a cystic mass with an air fluid level connected with esophagus in the middle mediastinum. The surgically resected mass was a pleural invested accessory lobe of the lung (8.0 × 7.0 × 4.5 cm) connected with the esophageal wall by a tubular structure (3.0 cm in length and 2.0 cm in diameter). A complete left pericardial defect was also identified. Histologically, the cystic wall was composed of fibrovascular connective tissue with a smooth muscle layer, mixed seromucous glands and cartilage, and the inner surface of the cyst was lined by ciliated pseudostratified columnar epithelium. The inner surface of the tubular structure was lined by non-keratinizing or keratinizing squamous epithelium, and the wall contained submucosal mucous glands, muscularis mucosa, and duplicated muscularis propria. This case is important in understanding the embryological pathogenesis of the variable spectrum of the bronchopulmonary foregut malformation.

Key Words : *Pulmonary Sequestration; Bronchogenic Cyst; Pericardium; Defect*

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INTRODUCTION

Bronchopulmonary foregut malformations are characterized by an isolated portion of lung tissue connection with the upper gastrointestinal tract and may result from the imperfect separation of pulmonary and esophageal anlagen or an accessory lung bud arising in the primitive gastrointestinal tube during early embryogenesis (1). They encompass a great variability of anomalies including not only extra- and intra-lobar pulmonary sequestrations with partial or complete gastrointestinal communication but also foregut diverticula, foregut duplications, tracheoesophageal fistulas, bronchoesophageal fistulas, and congenital cystic adenomatoid malformations of the lung (1).

We have recently experienced a very unusual case of bronchopulmonary foregut malformation presented with an extralobar pulmonary sequestration-like bronchogenic cyst connected with a tubular esophageal duplication associated with a pericardial defect.

CASE REPORT

A 18-yr-old man, who had suffered from dry cough and mild dyspnea, was admitted because of an incidentally detected chest mass. A computed tomography (CT) scan demonstrated a cystic mass with an air fluid level connected with esophagus in the middle mediastinum (Fig. 1). A left pericardial defect was also suspected (Fig. 2). Surgical excision of the mass was undertaken. The mass was a pleural invested accessory lobe of the lung connected with the esophageal wall by a tubular structure in the middle mediastinum. The arterial supply was done from branches of the descending thoracic aorta, and the venous drainage was done into the azygos system. A complete left pericardial defect was identified.

Grossly, the accessory lobe of the lung measured 8.0 × 7.0 × 4.5 cm, and the attached tubular structure measured 3.0 cm in length and 2.0 cm in diameter. The cut surface of the accessory lobe of the lung showed a bronchus-like dilated cyst, which contained soap bubble-like material communicated with the tubular structure. The investing lung parenchyma

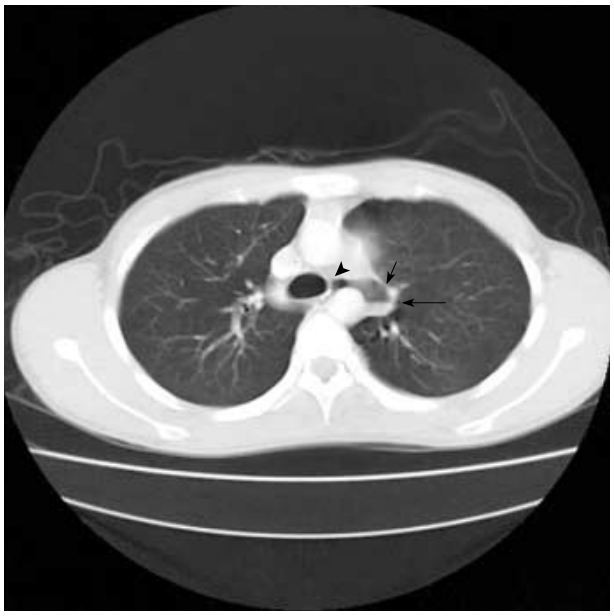

Fig. 1. Axial chest CT on lung setting shows a cyst (short arrow) with an air-fluid level (long arrow) abutting the esophagus by tubular structure (arrow head) in the aortopulmonary window.

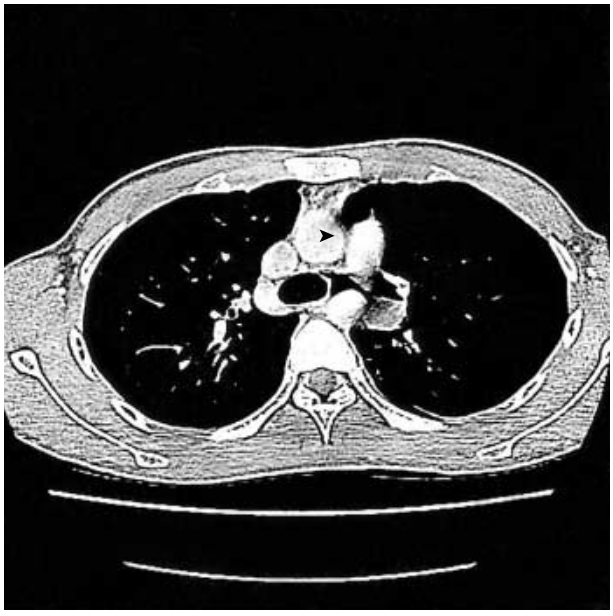

Fig. 2. Axial contrast enhanced CT shows an abnormal interposition of the lung tissue (arrowhead) between the aorta and the main segment of the pulmonary artery, indicating the absence of the pericardium.

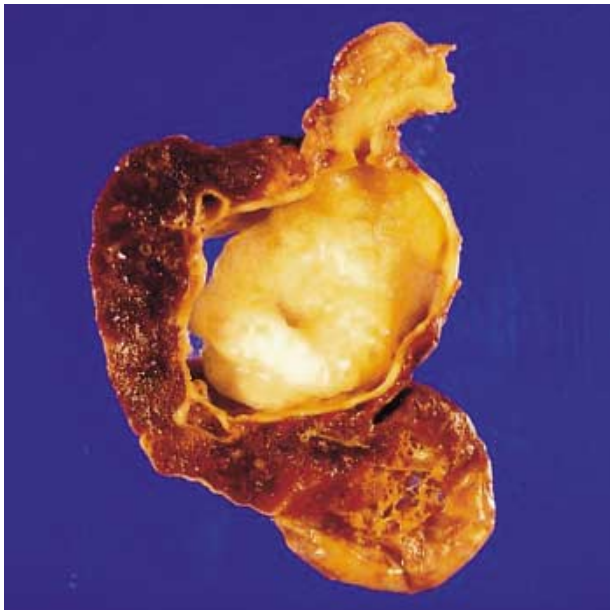

Fig. 3. The cut surface of the mass shows a unilouclar cyst containing soap bubble-like material, which is connected with tubular structure. The investing lung parenchyma shows a subpleural bullous formation.

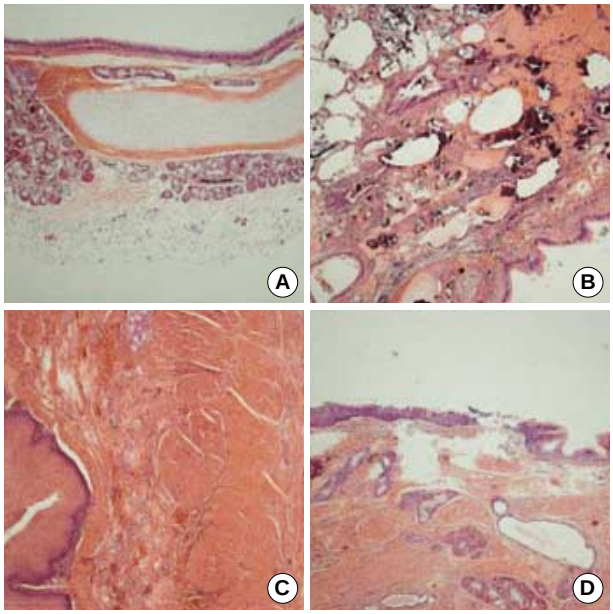

Fig. 4. Microscopic features of the specimen (H&E ×40). (A) The wall of tubular structure lined by squamous epithelium contains duplicated muscular layer and submucosal mucous glands. (B) The wall of bonchogenic cyst lined by ciliated pseudostratified columnar epithelium is composed of fibroconnective tissue, hyaline cartilage, and mucous glands. (C) The investing lung parenchyma shows of mature alveolar spaces with an emphysematous change. Some alveolar spaces are filled with proteinaceous fluid. (D) A transition from squamous epithelium to columnar epithelium is noted in the junction of the bronchogenic cyst and tubular esophageal duplication.

was partly consolidated or partly emphysematous with a subpleural bullous change (Fig. 3). Histologically, the cystic wall was composed of fibrovascular connective tissue with a smooth muscle layer, mixed seromucous glands, and hyaline cartilage, and the cysts were lined by pseudostratified ciliated

columnar epithelium (Fig. 4A). The investing lung parenchyma was composed of mature alveolar spaces filled with proteinaceous fluid or red blood cells, and bronchial or bronchiolar structures were randomly distributed among these mature alveoli (Fig. 4B). Focal emphysematous changes with a subpleural bulla were also noted. The inner surface of the tubular structure was lined by ciliated non-keratinizing or keratinizing non-ciliated squamous epitheliums and the wall contained submucosal mucous glands, muscularis mucosa, and duplicated muscularis propria (Fig. 4C) No ganglion cells were noted. The mass was diagnosed as a bronchopulmonary foregut malformation composed of an extra-lobar pulmonary sequestration-like bronchogenic cyst communicated with tubular esophageal duplication.

DISCUSSION

Bronchogenic cysts presumably represent supernumerary lung buds from the primitive foregut, but in contrast to sequestrations, they only rarely contain distal lung parenchyma (2). The cystic wall lined with ciliated columnar epithelium generally contains smooth muscle, cartilage, and bronchial mucous glands. Our patient's lesion revealed a unilocular cyst lined with a ciliated columnar epithelium with smooth muscles, hyaline cartilages, and bronchial mucous glands and well developed lung parenchyma composed of mature alveoli with randomly distributed bronchi and bronchioles. It possessed characteristics of both bronchopulmonary sequestration and bronchogenic cyst. Bronchogenic cyst and extralobar pulmonary sequestration are believed to originate form the embryonal foregut and have a close embryologic relationship. There are a few reports of bronchogenic cyst arising in pulmonary sequestration or bronchogenic cyst combined with pulmonary sequestration-like features (3, 4).

Bronchopulmonary foregut malformation was first described by Gerle et al. who reported 13 rare cases of pulmonary sequestration that communicated with the gastrointestinal tract (5). Since then, this terminology has been used for variable congenital malformations arising from a supernumerary lung bud from the primitive foregut showing a variable histopathologic spectrum, from the variably developed bronchopulmonary malformation to the foregut cyst without pulmonary structures (1). The type of bronchopulmonary foregut malformation depends on the stage of embryological development when the accessory tissue arises and the direction in which the aberrant pulmonary tissue grows.

The primitive foregut gives rise to the pharynx and lower respiratory tract as well as upper gastrointestinal tract. If the accessory lung bud from the primitive foregut arises before development of the pleura, it is invested by adjacent normal lung parenchyma and becomes an intralobar pulmonary sequestration. If it develops late, after the pleura has already formed, it grows separate from the adjacent lung, is invested by its own pleura, and becomes and extralobar pulmonary sequestration (1). The most common foregut cysts, the bronchogenic cyst and the esophageal duplication representing abnormal budding of the vertebral and dorsal primitive foregut, respectively, indicating their common origin from the primitive foregut and close embryologic relationship (6).

Congenital pericardial defect, a relatively rare condition, can vary from a small communication between the prericardium and pleural spaces to a complete pericardial absence. The most common variation of this anomaly (found in 65% of patients) is the deficiency of the left pericardium, and it is widely accepted that pericardial defect represent persistence of the embryonic pleuro-pericardial foramen, perhaps due to inadequate blood supply following premature atrophy of the left common cardinal vein (7). Congenital pericardial defect may be associated with cardiothoracic diseases including patent ductus arteriosus, atrial septal defect, and rarely bronchopulmonary foregut malformation (7).

During the fifth week of embryonic development the bronchial buds develop at the caudal end of the laryngeotracheal tube and grow into the pericardial-peritoneal canals, the future pleural cavities (7). At this stage the primitive heart and lungs share a common coelomic cavity and are closely related each other (8). As the future pleural cavity enlarges to accommodate the growing lung buds, a fold called the pericardio-pleural membrane is produced in the lateral coelemic cavity between the future pericardial and plural cavities, and they fuse in the midline, separating the pleural and pericardial cavities. Considering the close relationship between the branchial pouches, foregut primordium, pleuropericardial folds, and pharyngeal pouches in embryogenesis, various masses may result from abnormal buddings of the ventral diverticulum of the foregut or tracheobronchial tree, including bronchogenic cysts, and esophageal duplications could be associated with pericardial defect. The incidence of congenital pericardial defect associated with bronchopulmonary foregut malformation is extremely rare (9). To our knowledge, this is the first case in Korea.

In summary, our case showed findings of communicating bronchopulmonary foregut malformation composed of a bronchopumonary cyst having the characteristics of both extralobar pulmonary sequestrations and bronchogenic cysts and a tubular esophageal duplication associated with a complete congenital pericardial defect of the same side. This case is important in understanding the embryological pathogenesis of the bronchopulmonary foregut malformations, which range from pulmonary sequestrations to bronchogenic cysts and foregut duplication cyst.

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