

Complex Bronchopulmonary Foregut Malformation: Extralobar Pulmonary Sequestration Communicating with an Esophageal Duplication Cyst – A Case Report –

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We report here on a case of a rare, complex bronchopulmonary foregut malformation (BPFM) that was composed of an extralobar pulmonary sequestration communicating with an esophageal duplication cyst. A 33-year-old female presented with an incidentally detected chest mass. The computed tomography revealed a 7.5 × 4.0 cm sized heterogeneous, solid and cystic lesion in the right superior mediastinum. Surgical resection demonstrated the solid portion to be isolated lung tissue invested in its own pleura. A unilocular cyst was communicating with the bronchus of the sequestered lung, and microscopically the cyst was lined by squamous epithelium overlying the thick layers of smooth muscle. This case is important for understanding the spectrum of BPFMs and for differentiating a mediastinal mass, especially one at the unusual location.

Key Words : Bronchopulmonary sequestration; Esophageal cyst

Bronchopulmonary foregut malformations (BPFMs) encompass a broad spectrum of anomalies that may arise from abnormal differentiation of the respiratory and alimentary tracts during early embryogenesis.¹ We report here on a rare case of complex bronchopulmonary foregut malformation that consisted of an extralobar pulmonary sequestration associated with an esophageal duplication cyst.

CASE REPORT

A 33-year-old female presented with a chest lesion that was incidentally discovered on a chest X-ray during a routine examination. Computed tomography (CT) revealed a 7.5 × 4.0 cm sized heterogeneous mass located in the right superior medi-

astinum and it was abutting the trachea (Fig. 1). The anterior portion of the mass appeared to be solid with vascular structures within the mass, whereas the posterior portion had a cystic appearance, suggesting a mediastinal tumor. The patient underwent an open thoracotomy with excision of the mass.

On gross examination, the solid portion of the lesion was revealed to be isolated lung tissue invested in its own pleura. A unilocular cyst was filled with mucus and it communicated with the bronchus of the sequestered lung (Fig. 2). On microscopic examination, the lung parenchyma was covered by visceral pleura and it showed mature alveolar spaces filled with proteinaceous fluid (Fig. 3A). The cyst was lined by squamous epithelium and layers of smooth muscle with no cartilage. The lining demonstrated an abrupt transition to respiratory epithelium with overlying cartilage plates at the neck portion, where

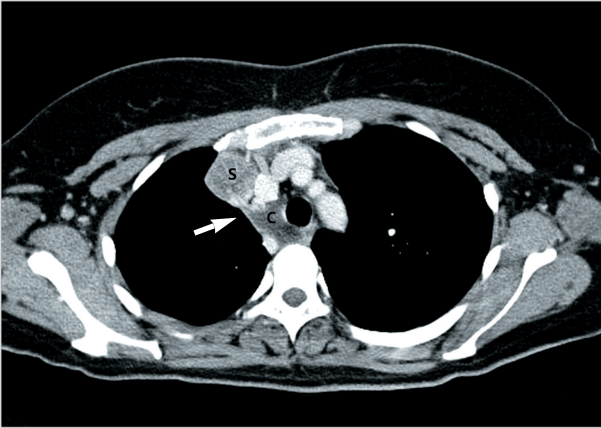


Fig. 1. Chest computed tomography reveals a 7.5 × 4.0 cm sized heterogeneous mass in the right superior mediastinum. The anterior portion of the mass (S) appears to be solid with vascular structures within the mass. The posterior portion (C) has a more cystic appearance. The communication (arrow) between these two portions is noted.

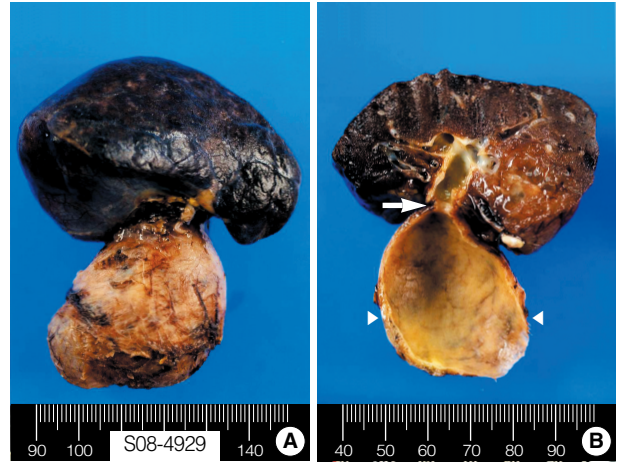


Fig. 2. (A) Gross examination reveals that the solid portion of the mass is isolated lung tissue invested in its own pleura. (B) On the cut section, a unilocular cyst (arrowheads) is communicating with the bronchus (arrow) of the sequestered lung.

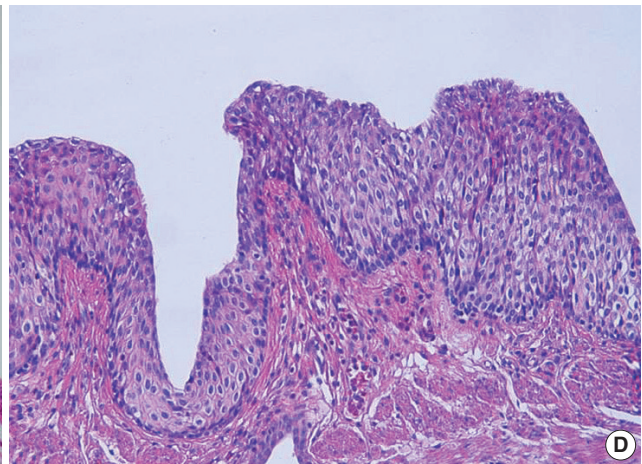
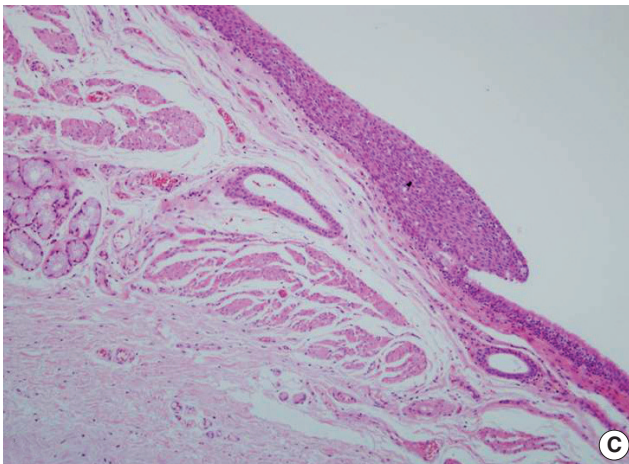
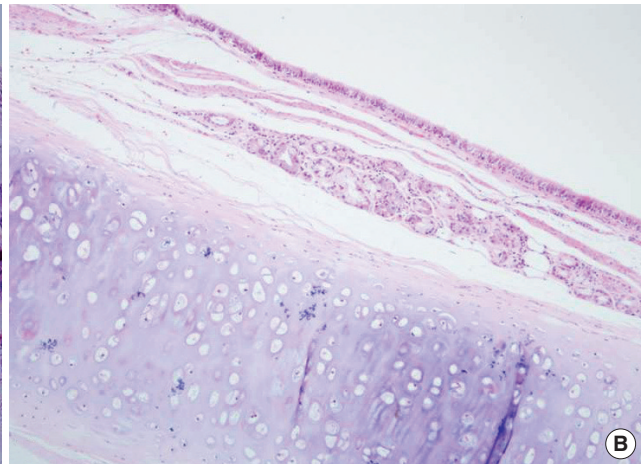
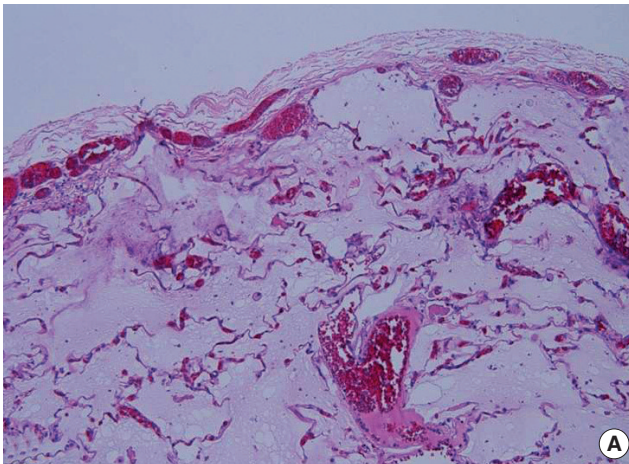


Fig. 3. (A) The lung parenchyma shows mature alveolar spaces filled with proteinaceous material. (B) The bronchus of the sequestered lung demonstrates ciliated pseudostratified columnar epithelium with overlying cartilage plates. (C) There is an abrupt transition of the respiratory epithelium to squamous epithelium at the neck portion where the bronchus of the sequestered lung is connected to the cyst. (D) The unilocular cyst is lined by squamous epithelium overlying thick layers of smooth muscle. No cartilage is found.

the cyst was connected to the bronchus of the sequestered lung (Fig. 3B-D). The lesion was diagnosed as a BPFM, which was composed of an extralobar pulmonary segment that communicated with an esophageal duplication cyst. A retrospective review of the previous CT revealed a small aberrant artery along the medial side of the cystic component and it originated from the thoracic aorta.

The patient's postoperative course was uneventful and she has done well during six months of follow-up.

DISCUSSION

Pulmonary sequestration is defined as a portion of lung tissue that has no identifiable communication with the normal bronchial tree and this lung tissue receives its blood supply from one or more anomalous systemic arteries. Pulmonary sequestrations are divided into two types; extralobar pulmonary sequestration has a distinct pleural covering that maintains complete anatomical separation from the adjacent normal lung tissue, whereas intralobar sequestration is embedded in the normal lung.² There have been many theories for the etiology of sequestration, including vascular insufficiency, vascular traction and post-infectious change.^{2,3} However, it is now widely accepted that pulmonary sequestration results from an embryologic malformation of the lung bud and this may be associated with other congenital anomalies.

BPFM was first introduced by Gerle *et al.*⁴ in 1968 as the pulmonary sequestration with a patent communication to the gastrointestinal tract. Normally, at five weeks of embryonic age, a primitive diverticulum arises from the ventral aspect of the pharynx and grows caudally. Two lateral ridges form between the diverticulum and the dorsal foregut, and they eventually fuse to become the tracheoesophageal septum, which divides the ventral respiratory laryngotracheal tube from the dorsal esophagus. A primitive lung bud develops at the caudal end of the laryngotracheal tube. By seven weeks, it bifurcates into two bronchopulmonary buds, which become the right and left lungs.^{1,5} BPFM occurs during this stage as a variety of anomalies that arise from a supernumerary lung bud from the primitive foregut. Which type of BPFM develops depends on 1) the stage of embryologic development when the accessory tissue arises, 2) the direction in which the aberrant pulmonary tissue grows and 3) the retention or involution of the communication between the accessory lung tissue and the parent viscus.⁶ If the accessory lung bud arises before development of the pleura, then it is

embedded in the adjacent normal lung tissue and it becomes an intralobar sequestration. If it develops late after the pleura has been already formed, then it is invested in its own pleura and forms an extralobar sequestration. Esophageal duplication cysts result from the abnormal budding of the dorsal primitive foregut during the same period of embryogenesis as that for bifurcation of the lung bud, and so esophageal duplication cysts are believed to have a close relationship with extralobar pulmonary sequestration.^{7,8} In the present case, the bronchus of the sequestered lung communicated with the esophageal duplication cyst, but not with the normal tracheobronchial tree or normal gastrointestinal pathway. This explains the absence of any symptoms, and it also supports the theory of common embryogenesis for the spectrum of BPFMs.¹

Extralobar pulmonary sequestration communicating with a foregut cyst is very rare.^{1,6,7,9,10} Only three Korean cases have been reported in the medical literature: two with a duplication cyst of the mixed bronchogenic and esophageal type, and one with a tubular esophageal duplication.^{6,7,9} We have presented a rare case of BPFM that was composed of an extralobar pulmonary sequestration communicating with an esophageal duplication cyst. This lesion was radiologically considered to be a tumorous condition prior to surgery because of its unusual location. Thus, BPFM should be included in the differential diagnosis of a mediastinal mass.

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