Extralobar Pulmonary Sequestration with An Associated Cyst of Mixed Bronchogenic and Esophageal Type
- A Case Report -

We report an unusual case of extralobar pulmonary sequestration (ELS) with an associated cyst of mixed bronchogenic and esophageal type. A 58-year-old woman was incidentally found to have a 6×6×5 cm sized mass in the right superior mediastinum. The mass consisted of sequestrated pulmonary tissue and an unilocular cyst with a direct communication. The cyst could not be easily classified because it was lined by squamous or respiratory epithelium with two distinct muscle layers and bronchial glands. Bronchial cartilage was present in close proximity to the ELS. This unusual combination of ELS with a foregut cyst might be a part of bronchopulmonary foregut malformation, attributed to a common embryologic pathogenesis. *(JKMS 1997; 12 : 567~9)*

Key Words : *Bronchopulmonary sequestration; Bronchogenic cyst; Esophageal cyst*

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INTRODUCTION

Extralobar pulmonary sequestration (ELS) may represent a form of bronchopulmonary foregut malformation, and be associated with other congenital anomalies, including gastroesophageal communication, diverticulum and esophageal or bronchogenic cyst. We have recently experienced a case of ELS with an associated cyst of mixed bronchogenic and esophageal type in an adult patient.

CASE REPORT

A 57-year-old woman was admitted to Kyung Hee University Hospital, for surgical excision of a lung mass which had been found incidentally. She had visited another hospital for an appendectomy. A homogeneous radiopaque mass of 6×6×5 cm, located in the right superior mediastinum on a chest X-ray and on a computed tomography scan. She was otherwise well and had no past history of repeated respiratory infection. Her breathing sound was clear. At operation, the mass consisted of a spherically shaped cystic lesion and underlying small dark consolidated accessory lung with a separate pleural investment from the remaining normal lung. The mass was located in the superior mediastinum just right lateral to the trachea and was fed by small systemic blood vessels.

PATHOLOGIC EXAMINATIONS

Gross examination showed a malformed lung invested in pleura, measuring 5×3×2.5 cm and weighing 32 g. On section, the lumen of the cyst, measuring 5 cm in diameter, communicated with the bronchus of the

Fig. 1. Extralobar pulmonary sequestration, attached to the unilocular cyst. The cyst communicated with the bronchus of the sequestration.

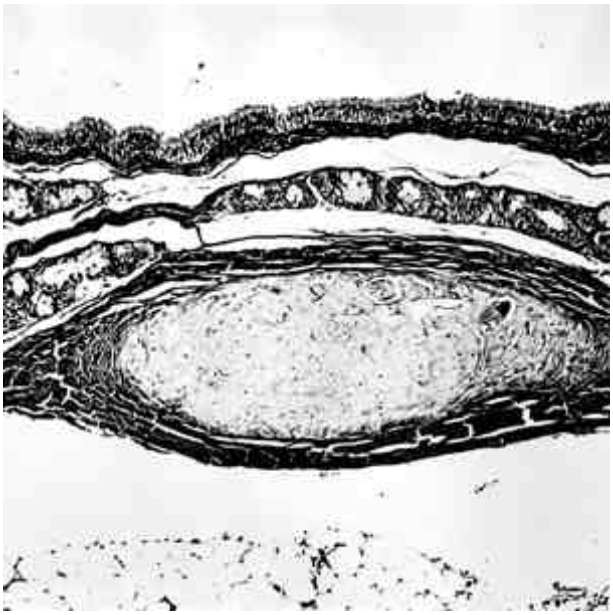

Fig. 2. The cyst wall of bronchogenic type showing respiratory epithelium, underlying hyaline cartilage plate and mucosal glands.

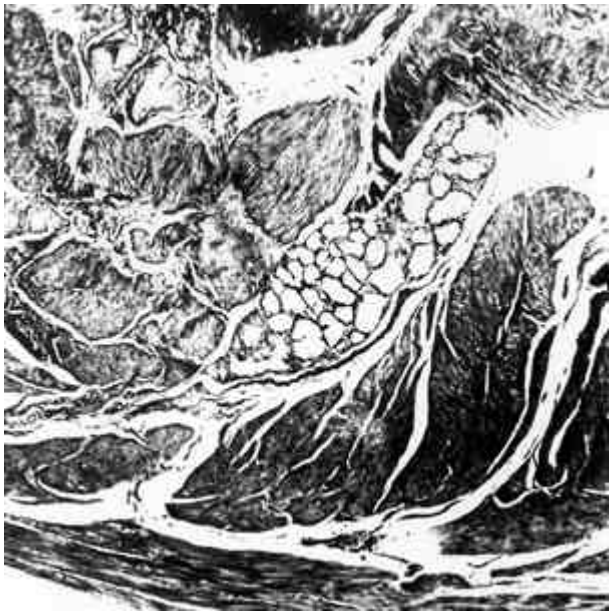

Fig. 3. The cyst wall of esophageal type showing squamous epithelium of immature type and underlying thick smooth muscle bundles.

sequestrated lung by a small stalk (Fig. 1). The cyst wall was thick with smooth outer and inner surface. The sequestrated lung showed a spongy-like appearance, due to cystic dilatation of bronchioles and alveoli.

Microscopically, the extralobar sequestrated lung was composed of a diffuse proliferation of bronchioles, dilatation of alveoli filled with foamy alveolar macrophages, and immature scattered bronchial hyaline cartilage plates. No significant inflammatory cell infiltrates were present. The cyst was lined by squamous epithelium of immature type and respiratory epithelium half in half (Fig. 2). The cartilage plates were present in the neck portion of the cyst, in close proximity to the ELS. However, in the remaining part of the cyst wall, two distinct layers of thick smooth muscles with scattered mucous glands were found (Fig. 3). A diagnosis of ELS with an associated developmental cyst of mixed bronchogenic and esophageal type was made.

DISCUSSION

This is a very unusual and interesting case of ELS with a developmental cyst. The combination of ELS with an associated bronchogenic cyst has been described in children (1). ELS probably develops from a separate outpouching of the foregut in which the connection between the ELS and the foregut disappears. In some cases, a segment of the primitive epithelium remains at the site, subsequently developing into a bronchogenic cyst over a period of years. Interestingly our patient was an adult and the cases reported in the literature were children (7 months old, 5 years old and 10 years old), suggesting that time might be required for such cysts to develop. The cysts reported were closely attached to the ELS with no communication between them. However, our case revealed communication between the ELS and the cyst grossly (Fig. 1). Doctor Travis, W.D., has also seen cases, in which aberrant bronchi at the edge of the ELS became dilated and filled with the mucous produced by the submucosal glands (personal communication), very much similar to ours.

However, the cyst in our case could not be easily classified as either bronchogenic or esophageal cyst. It showed findings of both bronchogenic and eosophageal cyst. Respiratory epithelium, scattered submucosal glands and islands of cartilage are characteristics of bronchogenic cyst (as shown in Fig. 2). Immature squamous epithelium and thick smooth muscle coats are compatible with esophageal cyst (8) (Fig. 3). A developmental foregut cyst of mixed type might be considered. Bronchogenic cyst and esophageal cyst are regarded as foregut cysts originated early in lung bud develpment before the bronchi are formed (2). The combination of ELS and foregut cyst as in this case offers evidence of an early embryologic insult. Heithoff et al. (3) suggested a unified concept, where bronchopulmonary foregut malformation arises from abnormal supernumerary lung bud development.

The spectrum of bronchopulmonary foregut malformation includes intralobar and extralobar sequestration, esophageal or gastric diverticula, and esophageal or bronchogenic duplication cysts. Benign developmental cysts may coexist with ELS (1, 4, 7).

Most cases of ELS have been found in pediatric patients. Age at the time of diagnosis is related to the symptoms (1, 5). ELS is incidentally found or is associated with respiratory symptoms. The ELS is commonly located between the diaphragm and lower lobe (6). In contrast, this ELS with a foregut cyst was located in the superior mediastinum lateral to the tracheobronchial tree. We have presented a unique case of ELS with an associated foregut cyst of mixed bronchogenic and esophageal type in regard to histogenesis.

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