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A Case of Intrapericardial Extralobar Pulmonary Sequestration

— First Case in Korea —

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Extralobar pulmonary sequestration, known as accessory lung, is a rare congenital anomaly and intrapericardial lung sequestration is extremely rare.

To the best of our knowledge, only four reported cases were intrapericardial. We report a case with intrapericardial extralobar pulmonary sequestration confirmed by operation and morphologic basis.

Key Words: *Intrapericardial. Extralobar pulmonary sequestration*

INTRODUCTION

Extralobar pulmonary sequestration is defined as a mass of pulmonary parenchyma anatomically separate from the normal lung.

In extralobar pulmonary sequestration, the tissue has a distinct pleural covering and may be supplied by an aberrant artery from the aorta or its branch. in rare instances, extralobar pulmonary sequestration communicates with the esophagus or stomach[1~4].

The pulmonary sequestration is found lying mostly at the left lung base but can be found in the right lung and in the mediastinum at any level from the neck to below the diaphragm[1,2,5~8].

Extralobar pulmonary sequestration is often discovered at the time of routine roentgenogram of the chest or at the time of repair of associated congenital anomalies such as diaphragmatic hernia and fistulous communication with the gastrointestinal tract because extralobar sequestration rarely presents like the intralobar form with recurrent pulmonary infection[1,4,6,8].

CASE REPORT

A 27-year-old man was admitted to Yong Dong Severance Hospital, Yonsei University Medical Center, for evaluation and management of mediastinal mass.

Three years prior to admission, he found mediastinal mass on computerized tomographic (CT) scan performed in another institution.

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At admission, he was noted to be with generalized weakness and right chest pain, but he denied cough, sputum and dyspnea.

The temperature was 38.6°C, the pulse rate 72/min and the blood pressure 110/70 mmHg.

On examination, he appeared chronically ill. the skin was warm and dry. the head and neck were normal and cervical lymphadenopathy was not found. the lungs were clear and the heart was normal.

The abdomen and extremities were normal.

The urine was normal and the hemoglobin was 13.5 gm/dl; hematocrit 41.9%; white cell count 5,500/mm^3 with 64 percent neutrophils, 35 percent lymphocytes and 1 percent eosinophils. the serum electrolytes and liver function tests were normal. T_3: 150 ng/dl, T_4: 7.1 ug/dl, FT_4: 1.6 ng/dl and TSH was 1.5 uU/ml.

The electrocardiogram showed right axis deviation and tall R wave in right precordial leads. and the sputum exanination did not suggest malignancy or tuberculosis.

Chest roentgenogram (Fig. 1) revealed well defined mediastinal mass on the right paratracheal area.

Chest CT scan revealed large multi-cystic mass lesion on the right paratracheal area which was attached to the lower pole of right thyroid gland, displaced the superior vena cava anteriorly and extended to the level of the azygous arch vein (Fig. 2).

The lung perfusion scan and whole body bone scan were normal and the thyroid scan revealed cold area in the lower pole of right thyroid gland.

The fiberoptic bronchoscopy revealed extrinsic mass effect without mucosal lesions on right intermediate bronchus.

The pulmonary function studies showed mild restriction with FEV_1:2.85 L/sec and FVC: 3.49 L/sec.

He underwent operation under the impression of mediastinal mass.

8×8 cm sized mass composed of multi-cystic and solid part was located in the pericardium and excised easily from the surrounding structures. and the feeding vessel maybe originated from the subclavian artery.

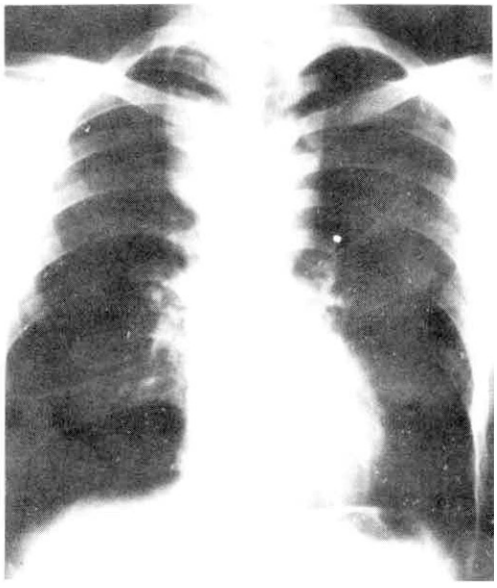

Fig. 1. *Chest PA revealed well defined mediasthinal mass on the right paratracheal area.*

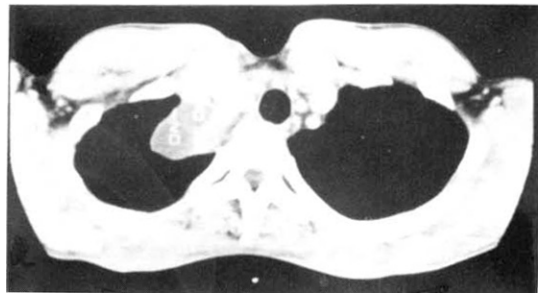

Fig. 2. *Chest CT scan revealed large multi-cystic mass lesion on the right paratracheal area which was attached to the lower pole of the right thyroid gland, displaced the superior vena cava anteriorly and extended to the level of the azygous vein.*

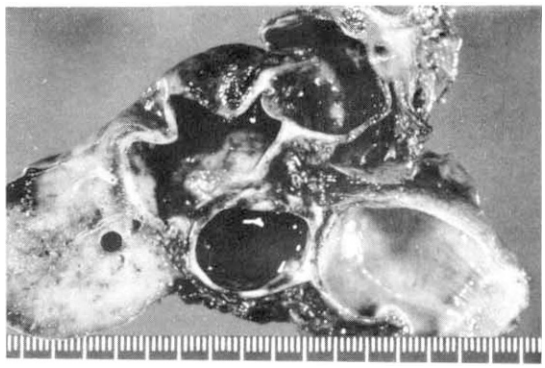

Fig. 3. *The mass is composed of several cystic lesions and yellow solid lesion resembling unexpanded lung tissue.*

Grossly, the mass was composed of several cystic lesions and yellow solid lesion, resembling unexpanded lung tissues (Fig. 3) and the surface of the mass shows a feeding vessel (Fig. 4).

Microscopically, the cyst wall showed a bronchial tissue which was composed of ciliated columanar epithelium, submucosal gland and cartilage (Fig. 5) and solid portion of the mass showed alveolar space filled with inflammatory cells such as macrophages or lymphocytes (Fig. 6)

Discussion

Pulmonary sequestration is an uncommon congenital malformation characterized by the presence of nonfunctioning lung tissue which usually has no communication with the normal bronchial tree and which receives its blood supply from an anomalous systemic artery instead of a pulmonary arterial branch[9].

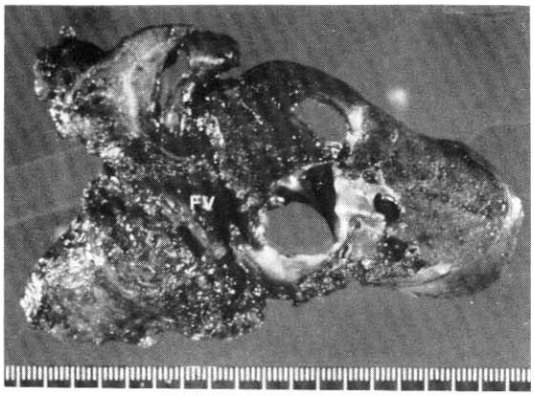

Fig. 4. *The surface of the mass shows a feeding vessel (FV)*

In intralobar sequestration, the abnormal pulmonary tissue is incorporated within the normal lung and shares a common covering of visceral pleura, and in extralobar sequestration, the abnormal pulmonary tissue is separate from the normal lung and has its own pleural covering[1,3,5,7].

"Bronchopulmonary foregut malformation" proposed by Gerle RD and Heithoff as pathogenesis of pulmonary sequestration is generally accepted and their spectrum of bronchopulmonary foregut malformation includes intralobar and extralobar sequestration, esophageal diverticuli, foregut duplication cysts (esophageal cysts) and maybe expanded to include bronchogenic cysts[10~21].

The incidence rate was 1.1 to 1.8 percent of all lobectomied patients by Carter and 0.15 to 6.4 percent of all congenital pulmonary malformation by Savic.

Savic found 133 cases of extralobar sequestration in a review of 547 cases of pulmonary sequestration.

The male predominance of patients with extralobar pulmonary sequestration (3:1) was described by Carter and Savic described that

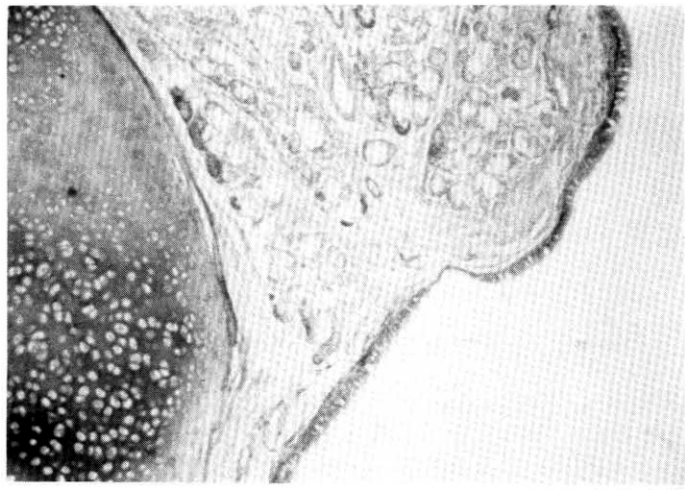

Fig. 5. *The cyst wall shows a bronchial tissue which is composed of ciliated columnar epithelium, submucosal glands and cartilage (×100, H-E).*

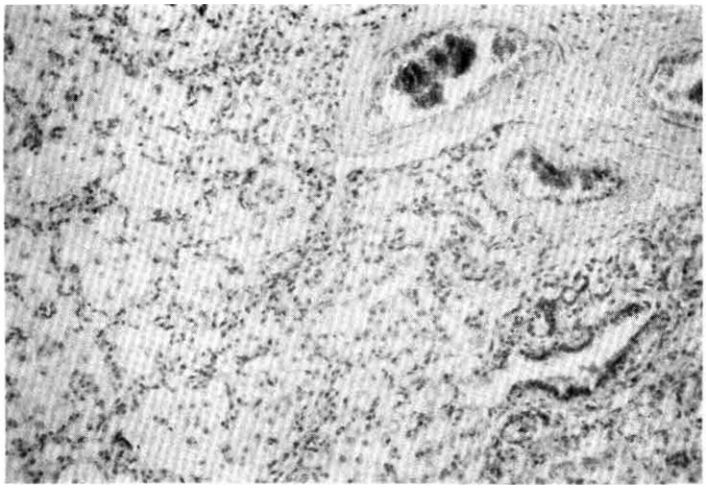

Fig. 6. *The solid portion of the mass shows alveolar spaces filled with inflammatory cells (×200, H-E).*

extralobar sequstration was located between the diaphragm and lower lobe in 77.4% and on left lung in 78.9% of the cases[8].

Most of the published cases of extralobar pulmonary sequestration were located outside the pericardium, only four reported cases were intrapericardial[15,13,14].

Since an extralobar sequestered segment is enveloped in its own pleural sac, the chance of becoming infected are very small, unless there is communication with the gastrointestinal tract[8].

The extralobar sequestration (42%) is more often associated with other anomalies than intralobar type (14%) such as diaphragmatic hernia and fistulous communication with the gastrointestinal tract[1,5,15].

Consequently, extralobar sequestration is usually an incidental finding on routine roentgenograms of the chest or during management of some other congenital anomaly. and infrequently, extralobar sequestration presents with symptoms similar to those seen with the intralobar form and with a picture of congestive heart failure or pulmonary overcirculation[1,5,7,16],

In all cases of intrapericardial extralobar pulmonary sequestration, a large quantity of pericardial fluid was found, but no sign of congestive heart failure or cardiac tamponade was noticed[13,14].

A correct preoperative diagnosis is made in 39 percent of intralobar lung sequestration and only in 9 percent of extralobar lung sequstration by Savic and correct preoperative diagnosis is more difficult in the cases of unusual location.

Preoperative angiography is important to confirm the diagnosis and to afford a safe operative approach[1,6,7].

The sequestratered segment is typically made up of multiple cysts or noncystic mass containing branching bronchi which run in the direction of the aberrant artery. The cysts contain mucus or mucopurulent material or air.

Histologically, the sequestration consists of normal lung elements in an abnormal and disorderly arrangement with a variable presence of cartilage, bronchial glands and alveolar parenchyma. The cysts are lined with ciliated respiratory epithelium[7,16].

Inflammatory changes and fibrosis are common in those patients who have had previous infection[7,16].

Extralobar pulmonary sequestration with symptoms or discovered at operation for another problem should be excised and it is feasible to observe asymptomatic and definite extralobar pulmonary sequestration without operation[1,7,8].

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