



Superior Mediastinal Mass Revealed as Bronchopulmonary Sequestration Supplied by a Branch of the Left Pulmonary Artery

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Bronchopulmonary sequestration (BPS) is a rare congenital abnormality of the lower airway, generally characterized by blood supply received from the systemic circulation. We present a rare case of a 19-year-old man with incidentally detected BPS supplied by a branch of a pulmonary artery, rather than a systemic artery. Computed tomography showed a sequestered segment supplied by a branch of the left pulmonary artery and containing an ectopic bronchus. As chest computed tomography revealed necrosis in the sequestered tissue, infection was presumed, and the tissue was surgically removed. This may represent a very unusual occurrence, as such cases have yet to be reported in the literature.

Keywords: Bronchopulmonary sequestration, Pulmonary artery, Thoracoscopy

Case report

Bronchopulmonary sequestration (BPS) is a rare congenital abnormality of the lower airway in which the sequestered tissue receives blood supply from the systemic circulation. If located within a normal lobe, BPS is classified as intralobar sequestration (ILS), while it is classified as extralobar sequestration (ELS) if it is located outside the normal lung with its own visceral pleura. ILS and ELS also differ in the route of venous drainage. Since infection can develop in ELS cases, close follow-up should be considered. We present a rare case of a 19-year-old man with BPS where the sequestered tissue was located in the superior mediastinum, mimicking ELS, and received arterial blood supply from a branch of a pulmonary artery, rather than a systemic artery. Have yet to be reported in the literature.

A 19-year-old man was admitted to Pusan National University Hospital because of a superior mediastinal mass found incidentally on a medical examination. A chest computed tomography (CT) scan revealed a 3.6-cm necrotic mass in the left superior mediastinum. Initially, a dermoid cyst or abscess was strongly suspected; however, the mass was found to be supplied with blood from an unknown large artery. To identify the origin of the blood supply, CT angiography was performed, revealing arterial supply from an accessory branch of the left pulmonary artery instead of

the systemic circulation (Fig. 1). As chest CT showed necrosis in the sequestered tissue with presumed infection, surgical removal was planned.

The sequestered tissue was resected through video-assisted thoracoscopic surgery. Intraoperatively, extralobar lung tissue in the superior mediastinum was observed to be fed by the left main pulmonary artery (Fig. 2), as shown on CT angiography. The feeding branch was divided with a stapler, and the extralobar lung tissue was extracted. The pathologic findings showed bronchocele with focal rupture and mucus spread along adjacent alveoli, mimicking ELS. These findings, along with the results of preoperative imaging, led us to conclude that the mass was an ELS receiving arterial blood supply from an aberrant pulmonary artery. The patient was discharged on the third postoperative day without complications, and no abnormal findings were reported during outpatient follow-up after discharge.

The patient provided written informed consent for the publication of clinical details and images.

Discussion

We present a rare case of a 19-year-old man with incidentally detected BPS that was supplied by a pulmonary artery, rather than the systemic circulation.

As typically defined, BPS consists of sequestered tissue



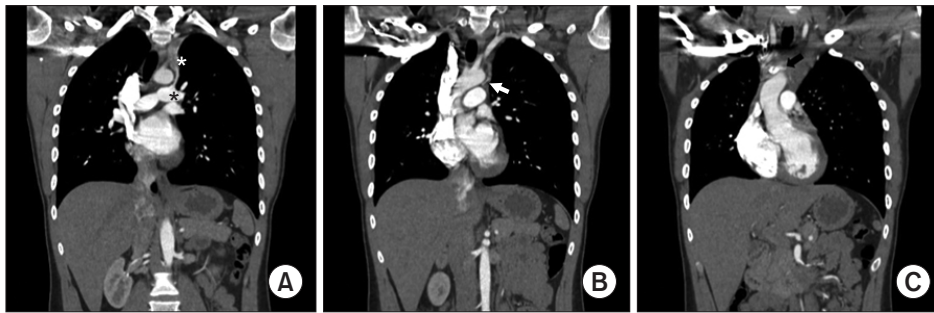


Fig. 1. (A) Necrotic mass (white asterisk) in the left superior mediastinum, determined to be ELS receiving arterial blood supply from a branch (black asterisk) of the left pulmonary artery. (B) Drainage vein (white arrow) from the ELS. (C) The drainage vein from the ELS is connected to the azygos vein (black arrow). ELS, extralobar sequestration.

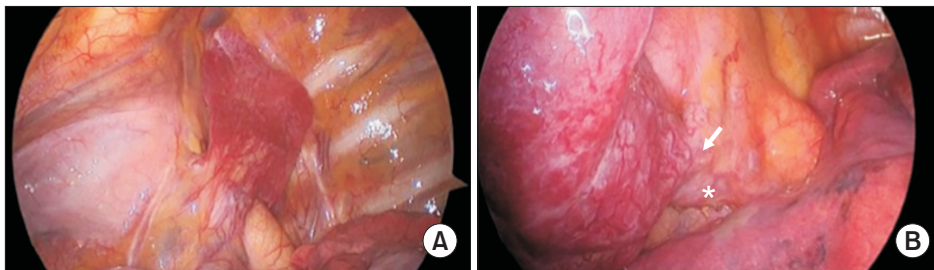


Fig. 2. Thoracoscopic findings. (A) Intraoperative findings showed sequestered lung tissue in the superior mediastinum. (B) Intraoperative findings also showed tissue receiving arterial blood supply from a branch (white arrow) of the left pulmonary artery and draining into the azygos vein (white asterisk).

supplied by the systemic circulation. The tissue does not communicate normally with the tracheobronchial tree [1,2]. BPS can be classified as ELS or ILS, which differ in their routes of venous drainage. Generally, ELS presents as a subdiaphragmatic mass with its own visceral pleura located outside the normal lung and rarely becomes infected, presenting as homogeneous tissue on chest CT [1-3].

In this case, a superior mediastinal mass was detected with central necrosis, suggesting a dermoid tumor. However, arterial supply from an aberrant pulmonary artery was found on CT angiography, indicating sequestration. CT angiography showed 2 major components: (1) a sequestered segment with an ectopic bronchus that lacked communication with the tracheobronchial tree and (2) arterial supply from an aberrant pulmonary artery extending from the left pulmonary artery.

Histologically, ELS is characterized by a bronchiole-like structure composed of bronchial epithelium in immature pulmonary parenchyma instead of the typical alveolar structure. In the present case, pathologic findings indicated a dilated bronchus filled with mucoid material, confirming that this mass was ELS, since the immature lung lacks a normal connection to the bronchus.

The typical definition of BPS would not seem to include sequestered tissue supplied by the pulmonary artery. However, a previous study used the term “sequestration spectrum” to indicate a continuum of congenital bronchopulmonary malformations [4]. This concept includes both general BPS and its modified forms; therefore, our case might also be regarded as a part of the sequestration spectrum [4].

Generally, the treatment of BPS depends on the presence of symptoms [5]. In symptomatic cases, surgical treatment is recommended, while elective surgical treatment or follow-up imaging can be considered in asymptomatic cases [5]. In ILS, which is associated with higher susceptibility to infection than ELS, surgical treatment is typically preferred [5]. Although the sequestered tissue in the present case resembled ELS, it was already shown to be infected on chest CT; for this reason, surgical resection was planned.

Even after surgery, the final diagnosis was ambiguous because the definition of BPS had not been satisfied. Finally, in consultation with a pathologist, we classified this case as ELS receiving arterial blood supply from an aberrant pulmonary artery. Although a few cases of conventional pulmonary sequestration receiving arterial blood supply

from a systemic artery had been seen previously at our center, this case was rare in that the lung tissue was located in the superior mediastinum, mimicking ELS, but received arterial blood supply from a branch of the left pulmonary artery.

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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