

# Bronchopulmonary foregut malformation presenting as extralobar pulmonary sequestration associated with a bronchogenic cyst: an unusual clinical and radiological feature in an adolescent patient

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**Abstract:** We present a case of symptomatic complex bronchopulmonary foregut malformation (BPFM), including extralobar pulmonary sequestration and a bronchogenic cyst, in the left anterior mediastinum of a 15-year-old boy. Preoperative computed tomography showed a cystic mass with heterogeneous enhancement of adjacent soft tissue components and pleural effusion. We suggested the infected bronchogenic cyst as the first impression. However, pathological examination after surgical resection revealed extralobar pulmonary sequestration and a bronchogenic cyst with unusual manifestation, which was located in the left upper hemithorax and supplied by the pulmonary artery. In patients presenting with a cystic mass with features of inflammation or infection and collateral vasculature, the possibility of a complex bronchopulmonary foregut malformation should be considered in the differential diagnosis.

**Keywords:** Extralobar pulmonary sequestration; bronchogenic cyst; bronchopulmonary foregut malformation (BPFM); thoracoscopic surgery

Submitted Feb 26, 2017. Accepted for publication Jun 16, 2017.

doi: 10.21037/jtd.2017.06.101

**View this article at:** <http://dx.doi.org/10.21037/jtd.2017.06.101>

## Introduction

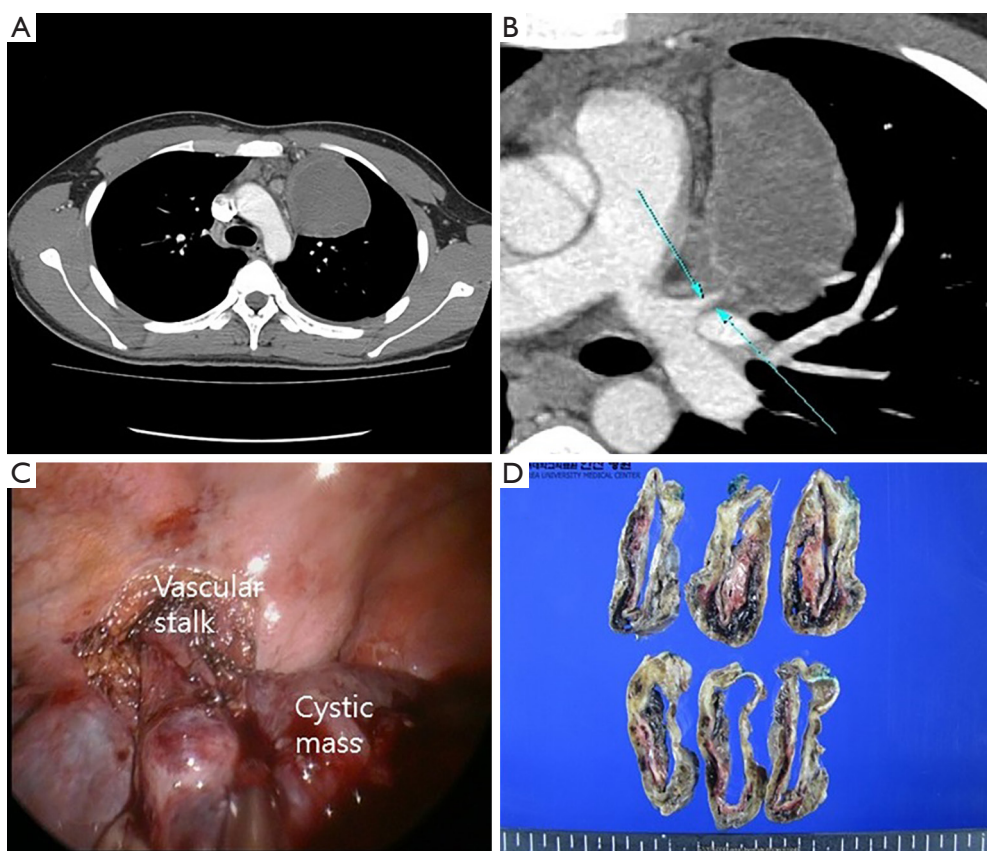
Bronchopulmonary foregut malformations (BPFMs) include congenital anomalies associated with the development of the foregut, pulmonary, airway, and vascular components (1). Among them, extralobar pulmonary sequestration is known as dysplastic or nonfunctioning lung tissue with its own pleura and without communication with the normal bronchial tree. It is usually located in the left lower hemithorax and receives its blood supply from one or more systemic arteries (2). Bronchogenic cysts originate from an abnormal budding of the tracheobronchial tree and are typically solitary cysts lined with either squamous or ciliated columnar epithelium, which is similar to bronchi (3).

We recently encountered a case of BPFM presenting as extralobar pulmonary sequestration associated with a bronchogenic cyst. The patient had an anterior mediastinal cystic mass and suffered from fever and pleural effusion,

a presentation that mimics an infected bronchogenic cyst with perforation. Furthermore, computed tomography (CT) demonstrated the lesion was located in the upper hemithorax and was supplied from the pulmonary artery. We present a case of a complex BPFM consisting of extralobar sequestration associated with a bronchogenic cyst presenting as an unusual clinical and radiological feature.

## Case presentation

A 15-year-old boy was admitted to the emergency room for left pleuritic chest pain and fever (38.0 °C) of 1-day duration. The chest radiograph showed a large lobulated mass in the left hilar area that shared a sharp border with the adjacent lung parenchyma. A chest CT scan showed a cystic mass in the left mediastinal area that formed an acute angle with the adjacent parenchyma of the left upper lobe,

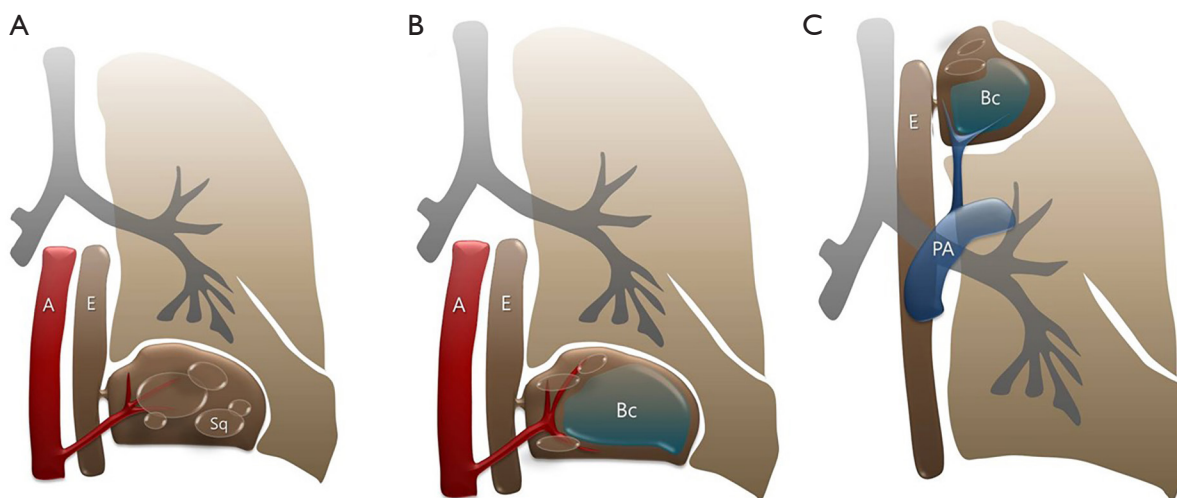


**Figure 1** Cystic mediastinal mass with vessel revealed extralobar pulmonary sequestration associated with a bronchogenic cyst. (A) Contrast-enhanced computed tomography (CT) scan showed a 7-cm cystic mass located in the left anterior mediastinal area, having an acute angle with the adjacent left upper lobe lung parenchyma. Enhancing wall, white arrow; pleural effusion, black arrow; (B) thin-section CT image with 1-mm section thickness revealed a feeding artery originating from the left pulmonary artery (arrows), which entered the cystic mass; (C) thoracoscopic exploration and mediastinal dissection revealed the accessory cystic pulmonary lobe has vascular stalk from hilum (right: cranial; left: caudal; head: anterior mediastinum; bottom: hilum); (D) pathological examination revealed that the large cyst included bronchial cartilage and epithelial lining within lung parenchyma and a relatively large muscular artery along the hilum of the supernumerary lobe invested with visceral pleura. This confirmed the incidence of extrapulmonary sequestration associated with a bronchogenic cyst with acute inflammation and fibrosis.

suggesting an extrapulmonary origin. The mass contained a non-enhanced homogenous low-attenuated fluid component (*Figure 1A*). There was a scanty amount of pleural fluid in the left hemithorax, suggesting perforation of the cystic mass (*Figure 1A*). A small artery originating from the left pulmonary artery and entering the medial side of the cystic mass was visualized (*Figure 1B*). We therefore suggested the presence of a complicated cystic mass, such as a bronchogenic cyst or dermoid cyst, with infection.

The patient underwent thoracoscopic surgical intervention for lesion resection. During surgery, we found

a supernumerary lobe of the left lung, with a vascular hilum, between the upper and lower lobes. This lobe consisted of cystic and solid components with hilar vasculature without perforation. The cystic component, which contained thick mucus-like material, included the hilum, and the solid component, which turned out to be composed of lung tissue, was an inclusion of the cystic mass (*Figure 1C*). A small feeding vessel originating from the hilum was identified and divided with an endoscopic stapler. Gross pathological examination showed a unilocular cystic mass within the inflamed sequestered lung (*Figure 1D*) with a wall showing fibrotic change. Microscopically, a bronchial



**Figure 2** Schematic pictures of extralobar type pulmonary sequestrations. (A) Extralobar pulmonary sequestration was composed of nonfunctioning lung tissue and had its own pleura. It was supplied by a systemic artery and was located in the left lower hemithorax area; (B) rarely, extralobar pulmonary sequestration can develop in conjunction with a bronchogenic cyst with systemic blood supply and left lower location; (C) in our case of complex bronchopulmonary foregut malformations (BPFM), a bronchogenic cyst was encircled by sequestered lung tissue with independent pleura and located in the left upper hemithorax, not the lower area. It was also supplied from the pulmonary artery, not the systemic artery. A, aorta; E, esophagus; Bc, bronchogenic cyst; PA, pulmonary artery.

epithelial lining was seen in the large cyst, and inflamed sequestered lung tissue covered the cyst. A relatively large muscular artery was present at the hilum of the sequestered lung. No infectious organisms were present in the culture of pleural fluid. The patient's postoperative course was uneventful and his symptoms disappeared.

## Discussion

BPFMs have been described by numerous classifications and terminologies incorporating a common origin and association. These include congenital anomalies associated with the development of the foregut (bronchogenic cyst, esophageal/neurenteric cyst, tracheoesophageal fistula/diverticula/stenosis), pulmonary [pulmonary agenesis/hypoplasia, congenital lobar emphysema, congenital pulmonary airway malformation (CPAM)], airway (tracheal/bronchial atresia, tracheal bronchus), and vascular (pulmonary sling, alveolar capillary dysplasia) components (1,4,5).

Bronchogenic cysts generally demonstrate an enhanced capsular wall and a homogenous consistency on enhanced CT because they have a bronchial epithelial lining and are filled with mucus (6). In the case of complicated bronchogenic cysts, a thick, contrast-enhanced wall with or without internal high attenuation may be demonstrated (7).

According to the patient's demographics and symptoms, dermoid cyst, necrotic tumor, esophageal duplication cyst, and other congenital cystic lesions can be considered as the differential diagnosis for relatively enhanced, thick walled cystic lesions on CT (8). Furthermore, as collateral neovascularization from an infected lesion can be encountered as a consequence of chronic inflammation (3), we suggested infected bronchogenic cyst as the first impression, despite the presence of a supplying vascular structure.

Among BPFMs, extralobar pulmonary sequestration is composed of non-functioning lung tissue and has its own pleura supplied by a systemic artery (1) (*Figure 2A*). However, BPFMs can be supplied from the pulmonary artery or pulmonary and systemic arteries in extremely rare cases (2). A few reports have demonstrated that pulmonary sequestrations have been described in conjunction with other lung anomalies such as bronchogenic cysts, bronchial atresia, and CPAM, among others (1,8). According to the reports, extralobar pulmonary sequestrations are usually located within the left lower hemithorax region, with systemic artery supply from the descending aorta or intercostal artery and a coexisting bronchogenic cyst noted adjacent to the sequestered lung tissue (*Figure 2B*).

However, in our case of complex BPFM, the radiological

and clinical findings were unusual. On CT images, a bronchogenic cyst was encircled by sequestered lung tissue with independent pleura and was located in the left upper hemithorax, not the lower area. In addition, it was supplied from the pulmonary artery, not the systemic artery (*Figure 2C*). Pleural effusion was also noted. According to a review of BPFM by Heithoff *et al.* (4), in older children and adults, presentations with recurrent infections such as pneumonia and pleurisy have been reported, although almost all of the patients are totally asymptomatic. Conversely, in this case, the patient presented with acute-onset pain and fever. Thus, we anticipated a thick capsular wall as the sequelae of infection in the cyst or an abscess in a malformed mass, together with symptoms such as fever and pain as a result of perforation of the cystic lesion.

We have described an unusual case of a complex BPFM consisting of extralobar sequestration associated with a bronchogenic cyst, which mimicked an infected mediastinal bronchogenic cyst with perforation.

### Acknowledgements

None.

### Footnote

*Conflicts of Interest:* The authors have no conflicts of interest to declare.

**Cite this article as:** Choo JY, Hwang J, Lee JH, Lee KY. Bronchopulmonary foregut malformation presenting as extralobar pulmonary sequestration associated with a bronchogenic cyst: an unusual clinical and radiological feature in an adolescent patient. *J Thorac Dis* 2017;9(7):E632-E635. doi: 10.21037/jtd.2017.06.101

*Informed Consent:* Written informed consent was obtained from the patient for publication of this manuscript and any accompanying images.

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