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CASE REPORT

Bronchopulmonary foregut malformation

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Abstract Bronchopulmonary foregut malformation (BPFM) is a rare anomaly of accessory pulmonary tissue that usually arises from esophagus or stomach. We present a case of extralobar pulmonary sequestration (ELS) connecting with the esophagus by a cyst, the inner wall of which is lined with squamous epithelium or respiratory epithelium. BPFM is sometimes used to group a number of ventral anomalies of accessory pulmonary tissue. The term currently refers specifically to those lesions composed of sequestrations that retain communication with the gastrointestinal tract. Usually the communication is a well-formed muscular tube lined with stratified squamous or columnar epithelium. The presence of both epithelia in a communication that is a component of the BPFM suggests embryogenesis. We describe an adult with BPFM composed of ELS and a connecting stalk to the esophagus by a foregut cyst that contains both squamous epithelium and respiratory epithelium.

Key words Bronchopulmonary foregut malformation · Extralobar pulmonary sequestration

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Introduction

Bronchopulmonary foregut malformation (BPFM) is rare congenital anomaly composed of an isolated portion of respiratory tissue and a connecting tissue between an isolated portion of respiratory tissue and the gastrointestinal tract. The ventral foregut budding theory is the best concept to explain the embryogenesis of BPFM.[1–3] The existence of a communicating tissue that contains squamous epithelium and respiratory epithelium between the pulmonary sequestration and the gastrointestinal tract strongly supports the ventral foregut budding theory.

In this report, we describe a case of extralobar pulmonary sequestration (ELS) that is connected to the esophagus by a stalk. That stalk was composed of a cyst that contained squamous epithelium and respiratory epithelium, suggesting its embryogenesis.

Case

A 20-year-old healthy woman was referred to the hospital because of an abnormal mediastinal shadow on chest roentgenography (Fig. 1). Physical examination showed blood pressure of 132/86 mmHg, respiratory rate of 14/min, and pulse rate of 78/min. The lungs were clear to auscultation and percussion. Computed tomography (CT) of the chest revealed a mediastinal mass next to the right side of T6 with a reticular structure and a stalk that exhibited enhancement (Fig. 2). Enhanced magnetic resonance image (MRI) of the chest revealed a flow void pattern in the mass and triangular shape on the coronal view.

Video-assisted thoracoscopic surgery was performed under the diagnosis of either neurogenic tumor or ELS.

A pyramidal solid mass was found at the space between the posterior part of the right upper lobe and the posterior part of the right lower lobe. The mass was connected to the esophagus by a stalk. An aberrant small systemic artery that fed the mass was found beside the stalk. That vessel was ligated, and the mass was resected with the stalk.

The pathological findings consisted of a pea-like mass 4 cm in length with a stalk that contained a cystic structure (Fig. 3). Microscopic examination of the inside wall of the cyst revealed transition from squamous epithelium with smooth muscle to ciliated columnar respiratory epithelium with cartilage and smooth muscle (Fig. 4). Uniformly dilated bronchioles, alveolar ducts, and

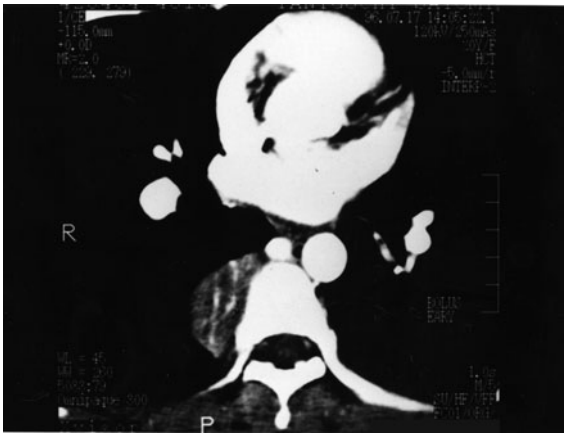

Fig. 2 Computed tomography of the chest reveals a mediastinal mass next to the right side of T6 with a reticular structure and a stalk that shows enhancement

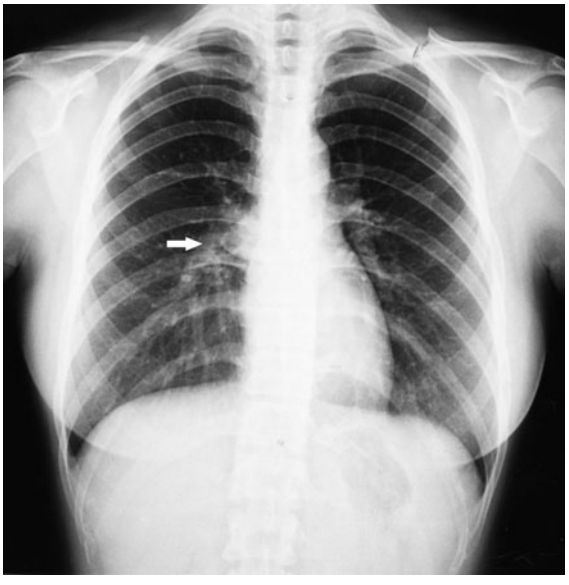

Fig. 1 Chest roentgenography shows a solitary mass (*arrow*) in the right middle mediastinum

Fig. 3 Half section of the tumor. The tumor connects to the esophagus with a stalk that contained a cystic structure

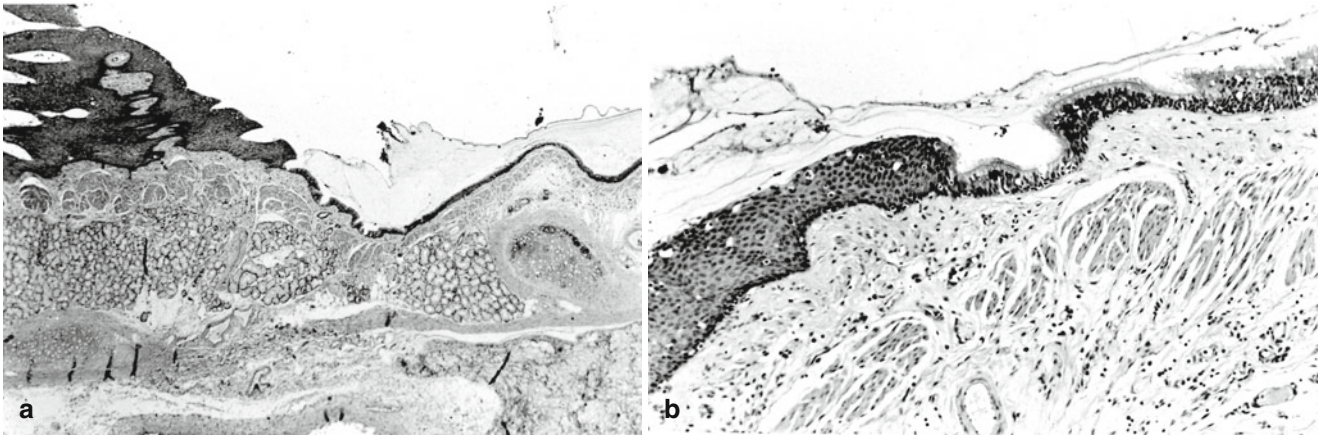

Fig. 4 Microscopic examination demonstrates a transition in a smooth muscle cyst wall that is lined with stratified squamous epithelium to a smooth muscle cyst wall containing cartilage lined with ciliated columnar epithelium. **a** At low magnification. **b** At high magnification. (**a**, **b** H&E)

Table 1 Characteristics of the cases of BPFM with a mixed-type communicating channel

Study (year)	Age	Sex	Location of malformation	Type of bronchopulmonary malformation	Arterial supply	Location of communicating channel
Thomson[7] (1962)	2 days	M	Entire right thorax	Extralobar	Pulmonary artery	Lower esophagus
Lewis[8] (1968)	4 months	F	Entire right thorax	Extralobar	Systemic artery	Lower esophagus
Gerle[2] (1968)	8 months	F	Left and right lower thorax	Intralobar	Thoracic aorta, abdominal aorta	Fundus of stomach
	48 years	F	Left lower thorax	Intralobar	Thoracic aorta	Lower esophagus
Graves[9] (1975)	5 months	M	Left lower thorax	Extralobar	Pulmonary artery	Lower esophagus
Heithoff[1] (1976)	10 weeks	M	Entire right thorax	Extralobar	Innominate artery	Upper esophagus
Crawford[10] (1978)	6 months	M	Right upper thorax	Extralobar	Thoracic aorta	Upper esophagus
Fowler[11] (1988)	1 day	F	Left and right lower thorax	Extralobar	Pulmonary artery	Lower esophagus
Srikanth[4] (1992)	15 years	F	Right lower thorax		Aorta	Gastroesophageal junction
Present case (2010)	20 years	M	Right middle thorax	Extralobar	Thoracic aorta	Mid esophagus

BPFM, bronchopulmonary foregut malformation

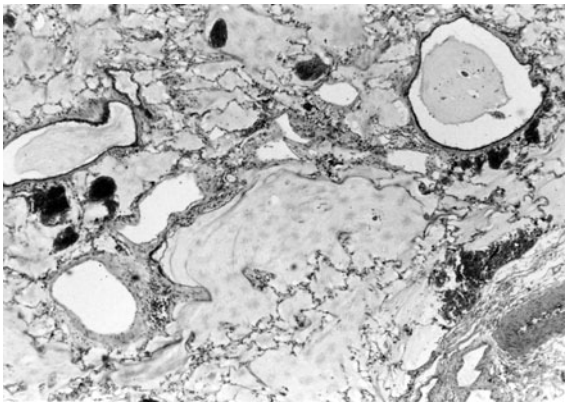

Fig. 5 Histopathological examination shows uniformly dilated bronchioles, alveolar ducts, and alveoli throughout the lesion. (H&E, at high magnification)

alveoli were seen throughout the resected solid mass (Fig. 5). The histological features established the diagnosis of ELS with foregut communication.

Postoperative recovery was uneventful. The patient was discharged from the hospital 5 days after the operation.

Discussion

Bronchopulmonary foregut malformation is an uncommon congenital entity. The term BPFM is sometimes used to group a number of abnormalities of ventral foregut budding.[1] Currently, it refers specifically to pulmonary sequestrations that retain communication with the gastrointestinal tract.[2–4] There are several theories of the embryogenesis of pulmonary sequestration.[5] The concept of ventral foregut budding best explains the entire sequence of BPFM, including intralobar and extralobar sequestration, bronchogenic cyst, and esophageal cyst.[2] The existence of a communication between a sequestration and the gastrointestinal tract strongly supports the foregut budding theory.[6]

The nature of the communicating channel is either bronchus-like, esophagus-like, or mixed. According to Srikanth et al., 9 of 36 cases were reported to have a mixed-type communication. 24 had bronchus-like communication, and 3 cases had esophagus-like communication.[1,2,4,7–11] The characteristics of the previously reported nine mixed-type cases and the present case of BPFM with a mixed-type communicating channel are summarized in Table 1. These 10 cases consist of 4 men and 6 women. In 3 of the 10 cases, the BPFM occupied the entire right thorax; in 2 it occupied the left and right lower thorax; and in 2 it was in the left lower thorax. Among the other three cases, there was one case each in which the BPFM was in the right upper thorax, the right middle thorax, and the right lower thorax. Thus 5 of 10 of the BPFMs appeared in the lower thorax. As for the type of BPFM, seven cases were extralobar and two were intralobar (one report did not give the type of BPFM). The arterial supply was from a systemic artery in seven cases and from the pulmonary artery in three cases. The lower esophagus or gastroesophageal junction was the most common location of the communicating channel—in 6 of 10 cases. In two cases, the upper esophagus communicated with the BPFM. The fundus of the stomach communicated with the BPFM in one case, and in our case the middle esophagus communicated with the BPFM.

The present case showed a communication between ELS and the esophagus by a stalk that contained a cyst. Microscopic examination of the inner wall of the cyst revealed a transition from squamous epithelium to ciliated columnar respiratory epithelium. The existence of

both respiratory and squamous epithelia in the cyst led to the diagnosis of a developmental foregut cyst of mixed type. Our case showed findings indicating that the BPFM was composed of an ELS and a developmental foregut cyst. This unusual combination supports a common embryological pathogenesis and the foregut budding theory.

There are no prospective data on the therapeutic modality concerning ELS. In most cases the BPFMs were resected to make a diagnosis pathologically because it was not easy to distinguish ELS from an intralobular pulmonary sequestration or a neurogenic tumor.[12] Although a case of ELS was reported to manifest as a massive hemothorax in an adult,[13] asymptomatic cases might be followed up without surgery.[14]

Conclusion

An adult with ELS that was connected to the esophagus by a cyst that contained both squamous and respiratory epithelia is reported. The existence of both of these epithelia in a communication suggests two-direction development from its origin and strongly supports the concept that abnormal ventral foregut budding is the cause of BPFM.

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