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Complex bronchopulmonary foregut malformation: extralobar pulmonary sequestration associated with a duplication cyst of mixed bronchogenic and oesophageal type

Abstract  We report a 13-year-old girl with an unusual, complex bronchopulmonary foregut malformation. The malformation included extralobar pulmonary sequestration and a duplication cyst of mixed bronchogenic and oesophageal type. Preoperative CT and MRI demonstrated the cystic and solid portions of the mass and indicated an aberrant vascular supply, suggesting the possibility of bronchopulmonary foregut malformation and several other differential diagnoses. A direct communication between the cyst and the bronchus of the sequestrated lung was found on pathological examination. This unusual combination of an extralobar pulmonary sequestration and a foregut cyst points to a common embryological pathogenesis.

Introduction

Bronchopulmonary foregut malformations encompass a great variety of anomalies that may arise from abnormal differentiation of the respiratory and alimentary tracts, abnormal separation of the two systems, or abnormal development of blood supply, perhaps singly or in combination, during early embryogenesis [1, 2]. The presence of accessory lung tissue that arises from the primitive gastrointestinal tube is the common factor in the development of all forms of bronchopulmonary foregut malformation [3].

We have recently experienced a case of complex bronchopulmonary foregut malformation with an unusual combination of an extralobar pulmonary sequestration and a duplication cyst of mixed bronchogenic and oesophageal type.

Case report

A 13-year-old girl was admitted because of a chest mass that had been discovered incidentally. Chest radiography showed a large lobulated and elongated mass in the left paratracheal area which had a sharp border with adjacent lung parenchyma (Fig. 1). Contrast-enhanced CT demonstrated a mixed solid and cystic mass located in the superior mediastinum, just to the left of the trachea. The upper part was composed of an irregularly shaped soft-tissue mass with inhomogeneous enhancement (Fig. 2a). The lower part was composed of a well-defined, round cystic mass with a smooth, thin wall and a small daughter cyst. A small aberrant artery origi-
nating from the thoracic aorta was seen along the medial side of the inferior, cystic component of the complex mass (Fig. 2b). Coro-
nal MRI of the chest clearly demonstrated the topographical relationship between the cystic and solid components. Two daughter
cysts were found additionally. A branching tubular structure with
signal void, considered to be an aberrant artery or feeder of the
complex mass, was shown to originate from the descending thoracic
aorta, identical to that seen on CT (Fig. 3).

Thoracoscopic excision of the mass was undertaken. The mass
consisted of solid and cystic components. The solid component,
which turned out to be composed of lung tissue on frozen biopsy,
comprised the upper portion of the mass and the cystic component,
which contained thick mucin-like material, was located in the low-
er portion. A small feeding artery originating from thoracic aorta
was identified. Gross pathological examination showed a mal-
formed lung invested in its own pleura and a cystic mass communi-
cating with a bronchus of the sequestrated lung (Fig. 4).

Microscopically, cartilage plates were present in the neck portion
of the cyst, lined by respiratory epithelia, in close proximity to the
sequestrated lung (Fig. 5a). The remaining part of the cyst was
lined by squamous epithelium and layers of smooth muscle were
found (Fig. 5b).

The patient’s postoperative course was uncomplicated and she
has done well since then.

Discussion

Communicating bronchopulmonary foregut malforma-
tions (BPFM) are rare tracheobronchial anomalies
characterized by a fistula between an isolated portion of
respiratory tissue and the oesophagus or stomach
[3]. According to a review and analysis of 29 cases of
BPFM by Heithoff et al. [1], the majority of the patients
with BPFM presented in the first 8 months of life, usual-
ly with respiratory distress exacerbated by feeding. In
older children and adults, presentation with recurrent
pneumonia and bronchiectasis, haemoptysis, gastroin-
testinal bleeding and dysphagia have been described, al-
though some patients are totally asymptomatic.

The presence of supernumerary lung buds that arise
from the primitive gastrointestinal tube caudal to the
normal lung buds is considered the most common factor
in the development of all forms of BPFMs [1–4]. The
type of BPFM that develops depends on (1) the stage of
embryological development when the accessory tis-

Fig. 1 Chest X-ray shows a large lobulated and elongated mass (ar-
rows) in the left paratracheal area

Fig. 2a, b Chest CT scans show a well-defined solid and cystic
mass in the superior mediastinum. An inhomogeneously enhanc-

ing solid component is seen in the upper portion of the mass (a).
A round and cystic lesion with a thin, smooth wall is revealed in
its lower portion (b). Note a small aberrant artery (arrow) in the
medial aspect of the cyst
