The Association of Esophageal Duplication Cyst with Esophageal Atresia

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Abstract. Bronchopulmonary foregut malformations are a group of related disorders with a common embryologic pathogenesis. Two bronchopulmonary foregut malformations may occur concomitantly in the same patient.

Key words: Bronchogenic cyst – Bronchopulmonary foregut malformation – Enteric cyst – Esophageal atresia – Esophageal duplication cyst – Foregut cyst – Tracheoesophageal fistula

Bronchopulmonary foregut malformations include a number of congenital abnormalities of the primitive foregut. A case of an esophageal duplication cyst occurring in a patient with known esophageal atresia and tracheo-esophageal fistula is reported. Due to the related embryogenesis of bronchopulmonary foregut malformations, it is not surprising that two or more of these congenital anomalies may occur in combination.

Case Report
J. H., a 4 month old white female, was referred to Duke University Medical Center for evaluation of known esophageal atresia and recurrent pneumonia. Esophageal atresia without a lower tracheo-esophageal fistula was diagnosed at birth. An initial chest radiograph showed minimal anterior bowing of the trachea by the air-filled proximal pouch but no soft tissue mediastinal mass. A gastrostomy and cervical esophagostomy were performed at 2 days of age at an outside hospital. Chest radiography at 4 months of age showed a large mediastinal mass on the right which displaced the trachea anteriorly. A polyethylene catheter readily passed from the proximal esophageal pouch into the trachea at the level of C6. Barium confirmed the presence of a fistula between the proximal esophagus and the trachea. Moreover, the proximal pouch was small and separate from the mediastinal mass. The proximal pouch tracheoesophageal fistula was cannulated at bronchoscopy and subsequently ligated and divided via a transcervical approach. A chest radiograph after repair of the proximal pouch fistula again demonstrated a middle mediastinal mass with tracheal displacement (Fig. 1).

At 4½ months of age, the mediastinal mass was excised via a right thoracotomy. This cyst was attached by a fibrous stalk to the membranous portion of the trachea just above the carina. There was at least 5 cm between the two atretic ends of the esophagus. The excised cyst was 4 cm in diameter (Fig. 2a), was shiny pink in color, and contained viscid, turbid white fluid. Microscopic sections of the cyst wall showed stratified squamous epithelium and smooth muscle tissue consistent with an esophageal duplication cyst (Fig. 2b). Mild chronic inflammatory changes were present. A colonic interposition procedure is planned at an older age.

Discussion
Two patients with associated esophageal atresia and intrathoracic foregut cysts have recently been reported in the surgical literature [2]. An awareness of this association of bronchopulmonary foregut malformations led to the correct preoperative diagnosis in our patient with esophageal atresia, tracheoesophageal fistula, and foregut cyst (esophageal duplication cyst).

The differential diagnosis of a middle mediastinal mass in a patient with known esophageal atresia includes bronchogenic-enteric cyst [2], distention of the proximal pouch of the esophagus, mediastinal abscess due to leakage after surgical repair, and esophageal distention related to circular myotomy [4]. Since the esophageal atresia had not been repaired, the latter two postoperative complications were excluded. Contrast opacification of the proximal pouch in our patient demonstrated adequate decompression by the cervical esophagostomy as well as a separate mediastinal mass.

Bronchopulmonary foregut malformations are a related group of congenital abnormalities that
Fig. 1 a and b Radiology: a AP chest radiograph shows large mediastinal mass (M) on the right. b This middle mediastinal mass (M) displaces the trachea anteriorly (arrows)

Fig. 2 a and b Pathology: a Gross pathology. The esophageal duplication cyst is 4 cm in diameter. b Microscopic section (× 200). The lumen (L), stratified squamous epithelium (E) and submucosa of the cyst are consistent with an esophageal duplication

includes bronchogenic cyst (parenchymal and mediastinal), pulmonary sequestration (intralobar and extralobar), systemic arterial supply to normal lung, esophageal atresia, tracheo-esophageal fistula, enteric duplication, neurenteric cyst, esophageal diverticulum, and bronchial rests in the esophagus. Esophageal atresia and/or tracheo-esophageal fistula are presumably due to a faulty separation of the ventral laryngotracheal tube from the dorsal esophagus [3]. Esophageal duplications, neurenteric cysts, and