Case Report

A baby boy was diagnosed prenatally with a right-sided cystic lung mass. A chest CT scan shortly after birth demonstrated this mass involving the right upper lobe. He was asymptomatic, and surgery was planned for when he reached approximately four months of age. A follow-up CT scan done before resection showed the cystic lung mass as well as a right-sided mediastinal cystic mass (Figs. 1 and 2). On a rereview of the first CT scan, the mediastinal mass appeared to be present, though difficult to see because of surrounding atelectasis. He underwent a right thoracotomy and a right upper lobectomy. The mediastinal mass, adherent to the esophagus, was also excised. Pathology revealed a type 1 congenital cystic adenomatoid malformation (CCAM) (right upper lobe mass) and a bronchogenic cyst (mediastinal mass).

Discussion

Congenital cystic disease of the lung and mediastinum encompass a spectrum of anomalies ranging from CCAMs, bronchopulmonary sequestrations (BPSs), congenital lobar emphysema (CLE), bronchogenic cysts, esophageal duplication cysts, and neurenteric cysts. The terminology used to identify these cystic entities has been variable in the medical and surgical literature, with “bronchopulmonary malformations,” “bronchopulmonary foregut malformation,” “foregut duplication cyst,” and “foregut cyst,” often used interchangeably in describing and classifying them. Although these anomalies were once felt to be distinct, reports over the past decade-and-a-half have increasingly highlighted an overlap of these conditions. One example is the hybrid lesion of CCAM and BPS; another is the evidence that histologically, many bronchogenic cysts and esophageal duplication cysts have elements of both. Further, several reports of the coexistence esophageal duplication cysts with CCAM, BPS, and other pulmonary cystic malformations can be found in the literature. In a large series of 105 patients, Tsai et al. reported the concurrent presence of a bronchogenic cyst with CCAM in three of 105 patients with CCAM. MacKenzie et al. described the...
unusual presence of a bronchogenic cyst, BPS, and CCAM in one lesion in a patient and suggests that a common embryologic pathway exists for these diverse lesions.\(^\text{10}\)

Although this common pathway is yet to be identified, bronchial atresia may be the link that unifies these diverse lesions. Bronchial atresia has been increasingly seen and described in the pediatric population.\(^\text{11}\) Imai et al., Kunisaki et al., and Riedlinger et al. have found that bronchial atresia is a common and often unrecognized component of CCAMs, BPS, CLEs, and lesions of mixed pathology.\(^\text{12–14}\) Cases of bronchial atresia associated with bronchogenic cysts have also been reported.\(^\text{15}\) It has been proposed that obstruction from an atretic bronchus could lead to various congenital lung malformations, such as CCAM, BPS, CLE, and bronchogenic cysts.\(^\text{16}\) Another theory is that a bronchogenic cyst could predispose the patient to bronchial atresia.\(^\text{15}\) If this is so, perhaps in this case the bronchogenic cyst in this case led to bronchial atresia, which then led to the development of CCAM.

The current patient had coexisting CCAM and bronchogenic cyst, which emphasizes the need to carefully look for other entities when treating a patient with congenital cystic disease of the lung or mediastinum.

**References**

10. MacKenzie TC, Guttenberg ME, Nisenbaum HL,


12. Imai Y, Mark EJ. Cystic adenomatoid change is common to various forms of cystic lung diseases of children: a clinicopathologic analysis of 10 cases with emphasis on tracing the bronchial tree. *Arch Pathol Lab Med* 2002; 126: 934–40.


