

Pulmonary Specialty Conference: Case 5

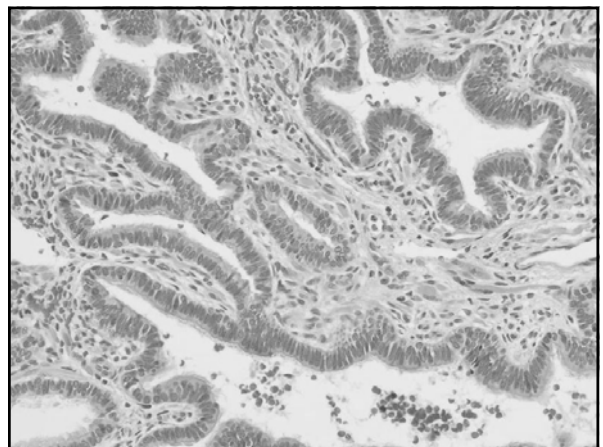
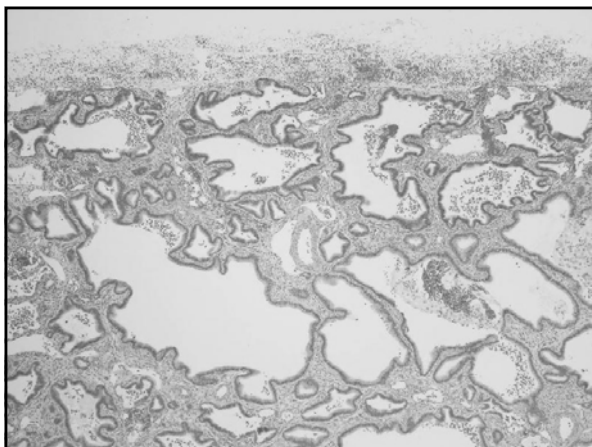
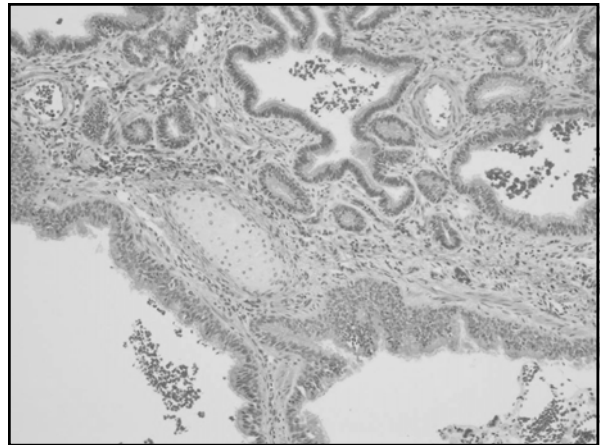
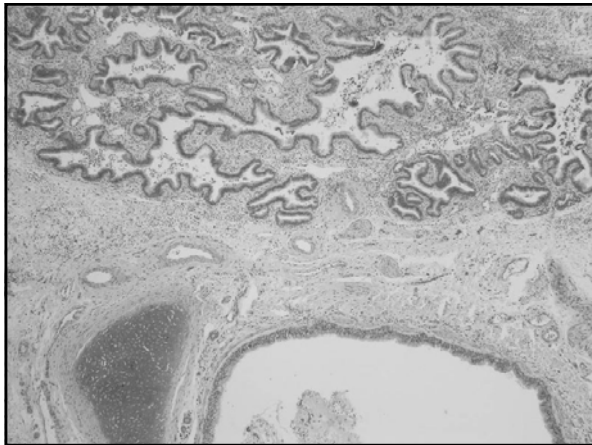
Aliya N. Husain
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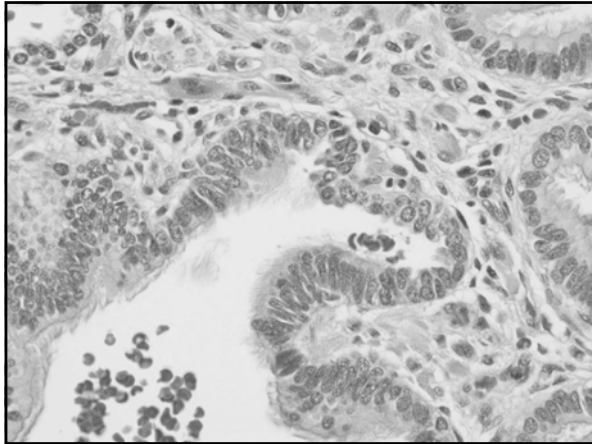


Case 5: History

- On prenatal ultrasound, this baby boy was found to have a left lung sequestration/mass. There was no other malformation seen. The lesion was resected at 8 weeks of age. The specimen consisted of tan-pink tissue measuring 3.3 x 2.6 x 1.7 cm. Serial sectioning showed multiple cysts varying in size from 0.1 to 0.5 cm.

Courtesy of Dr. Chejfec, University of Illinois





Differential diagnosis of congenital cystic lung lesions

- Congenital pulmonary airway malformation (types 0-4)
- Pulmonary sequestration with CPAM 2
- Bronchogenic cyst
- Infantile lobar overinflation
- Congenital pulmonary lymphangiectasia
- Post-infarction cysts

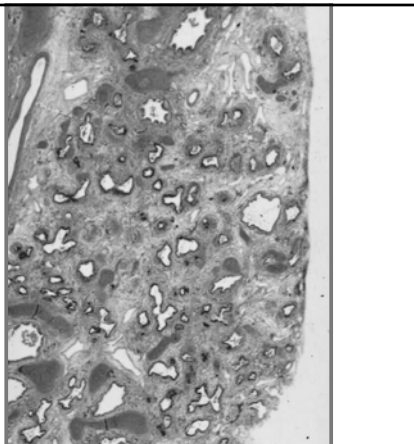
Congenital pulmonary airway malformation (CPAM)

- Classified based on:
 - size of cysts
 - composition of cyst walls

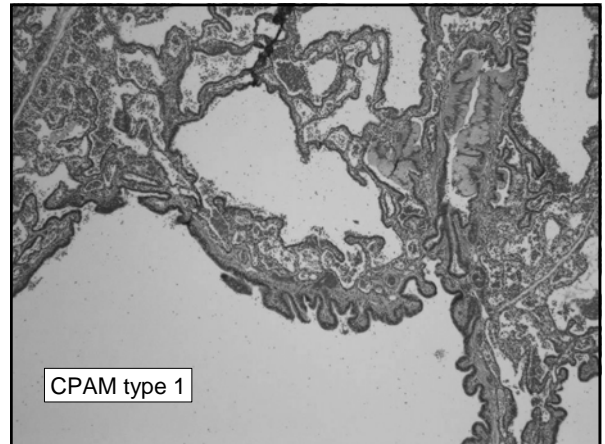
Congenital pulmonary airway malformation

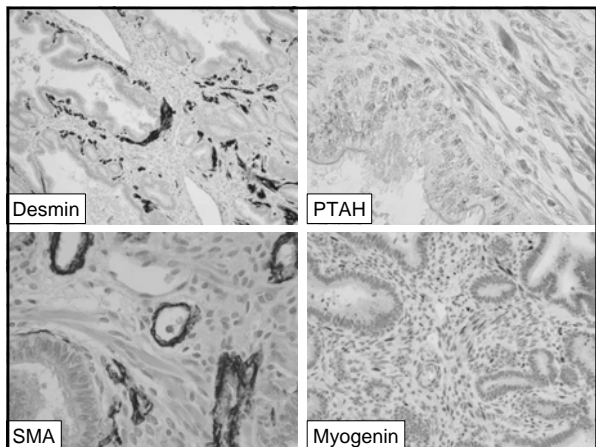
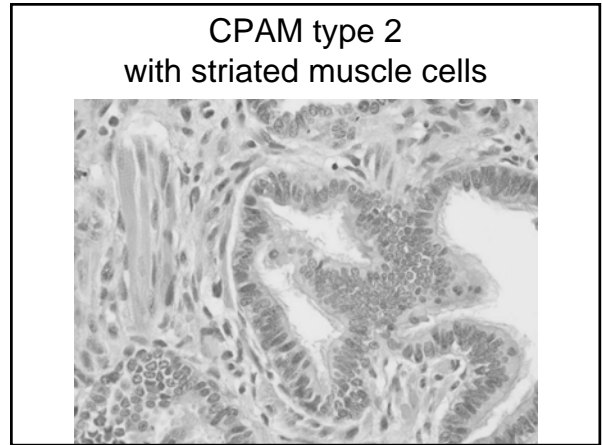
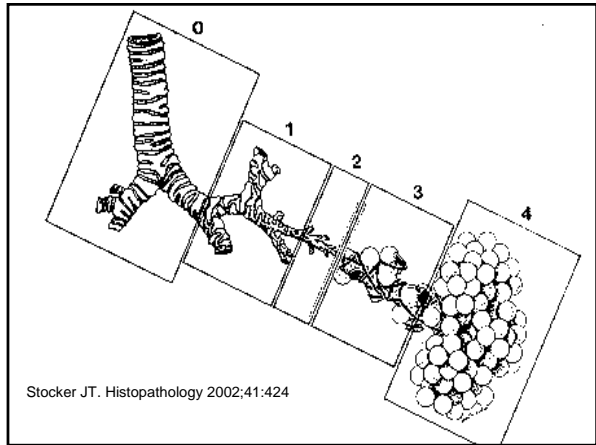
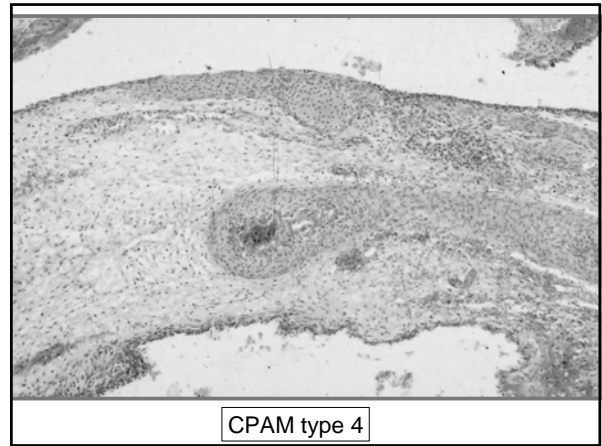
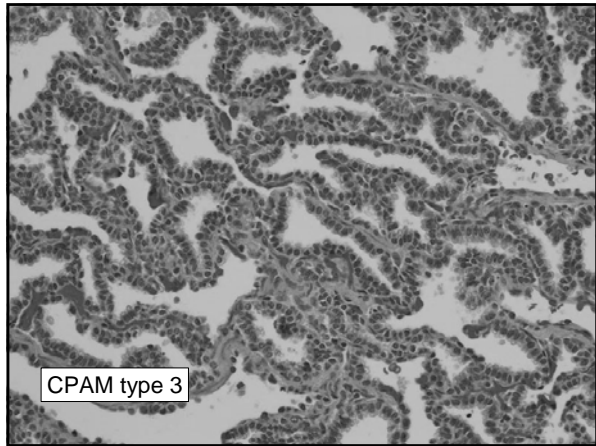
Type	Size of cysts	Cyst wall structure
0	Grossly solid	Bronchi and cartilage
1	3-10 cm	Ciliated, pseudo-stratified with thick muscle, mucinous cells in one-third
2	<2 cm	Ciliated, pseudo-stratified, thin muscle layer
3	Grossly solid	Cuboidal lining
4	Up to 7 cm	Pneumocytes (types 1 & 2)

CPAM type 0



CPAM type 1





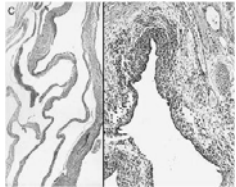
CPAM type 2
with striated muscle cells

- Described in 5-16% of malformations
- Skeletal muscle not seen in normal fetal lung from 8-15 weeks gestation

Fraggetta et al. Striated Muscle Cells in Non-Neoplastic Lung Tissue: A Clinicopathologic Study Hum Pathol 2000;31:1477-81

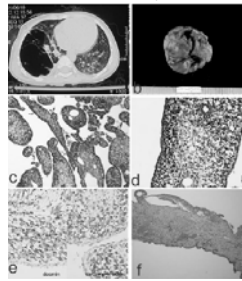
Striated muscle cells in lung

- Neoplasia
 - Pleuropulmonary blastoma
 - Primary rhabdomyosarcoma of lung
 - Rhabdomyosarcoma in CPAM 1/4 ??????

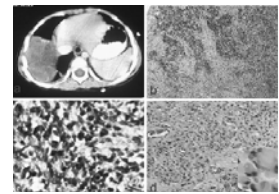


Hill et al. Type 1 Pleuropulmonary Blastoma: 51 cases from the PPB registry. Am J Surg Pathol 2008 32:282-295

First resection of cystic lesion



Recurrence one year later



Pai et al. Rhabdomyosarcoma arising within congenital cystic adenomatoid malformation. Pediatr Blood Cancer 2005;45:841-845

Pai et al. Correction: Pleuropulmonary Blastoma, not rhabdomyosarcoma In a Congenital Lung Cyst. Pediatr Blood Cancer 2007; 48:370-371

Why and how do skeletal muscle cells come to be in the lung?

- Both yolk sac and lung contain progenitor cells, which express endothelial markers and are endowed with a skeletal muscle potential that they reveal when in the presence of differentiating myoblasts, in vitro, and regenerating muscle, in vivo
- Number and potency decreases rapidly with age and is very low in mature mice

Angelis et al. Skeletal myogenic progenitors in the endothelium of lung and yolk sac. Experimental cell research 2003;290:207-216

Why and how do skeletal muscle cells come to be in the lung?

- *Adamtsl2* mRNA is found most abundantly in adult mouse liver, lung and spleen
- IHC localization of ADAMTSL2 protein was similar to mRNA expression
- Induction of *Adamtsl2* mRNA is an integral feature of myogenesis

Koo et al. ADAMTS-like 2 is a secreted glycoprotein that is widely expressed during mouse embryogenesis and is regulated during skeletal myogenesis. Matrix Biology 2007;26:431-441