

Pulmonary Specialty Conference: Case 5

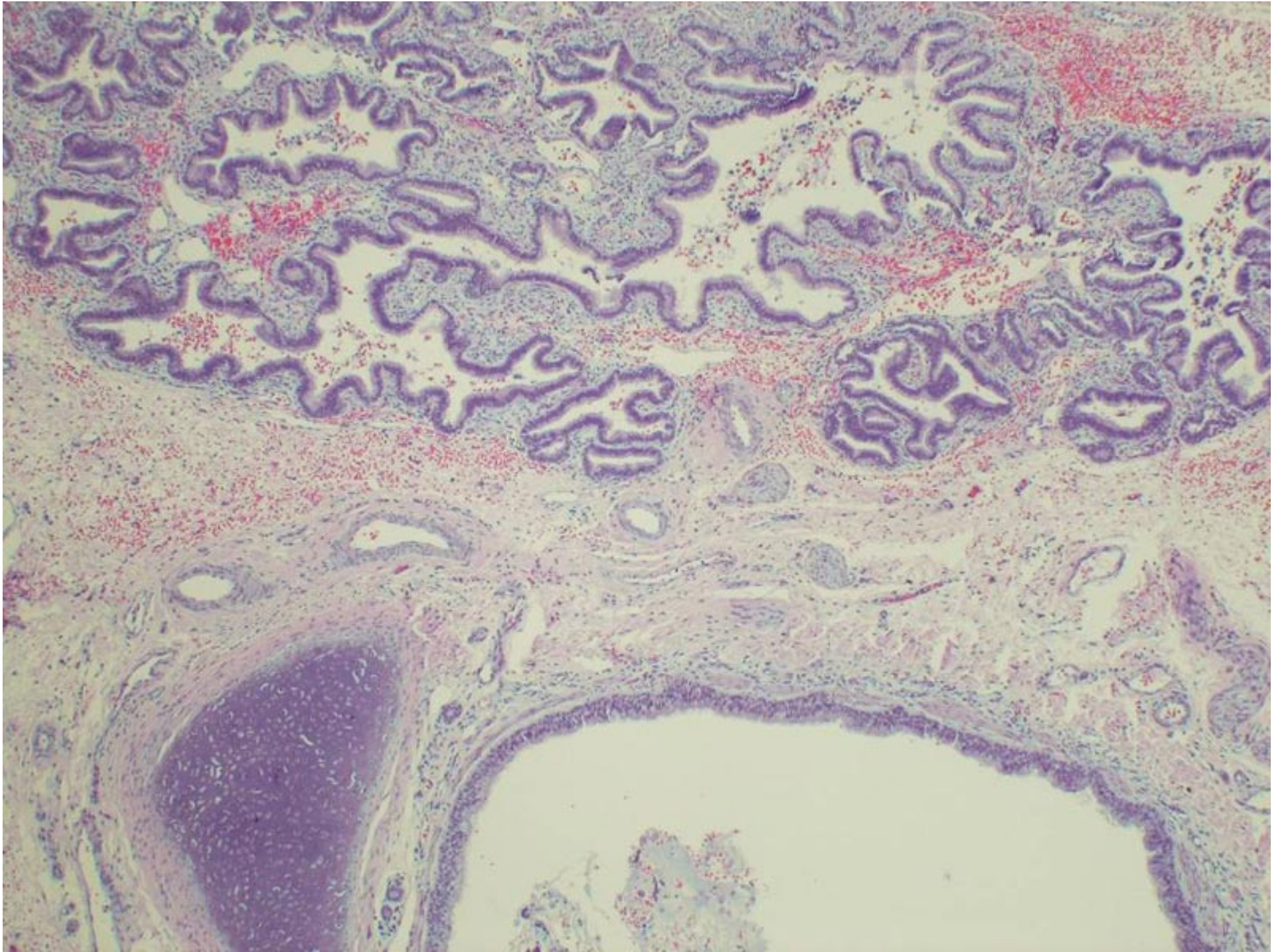
Aliya N. Husain
University of Chicago

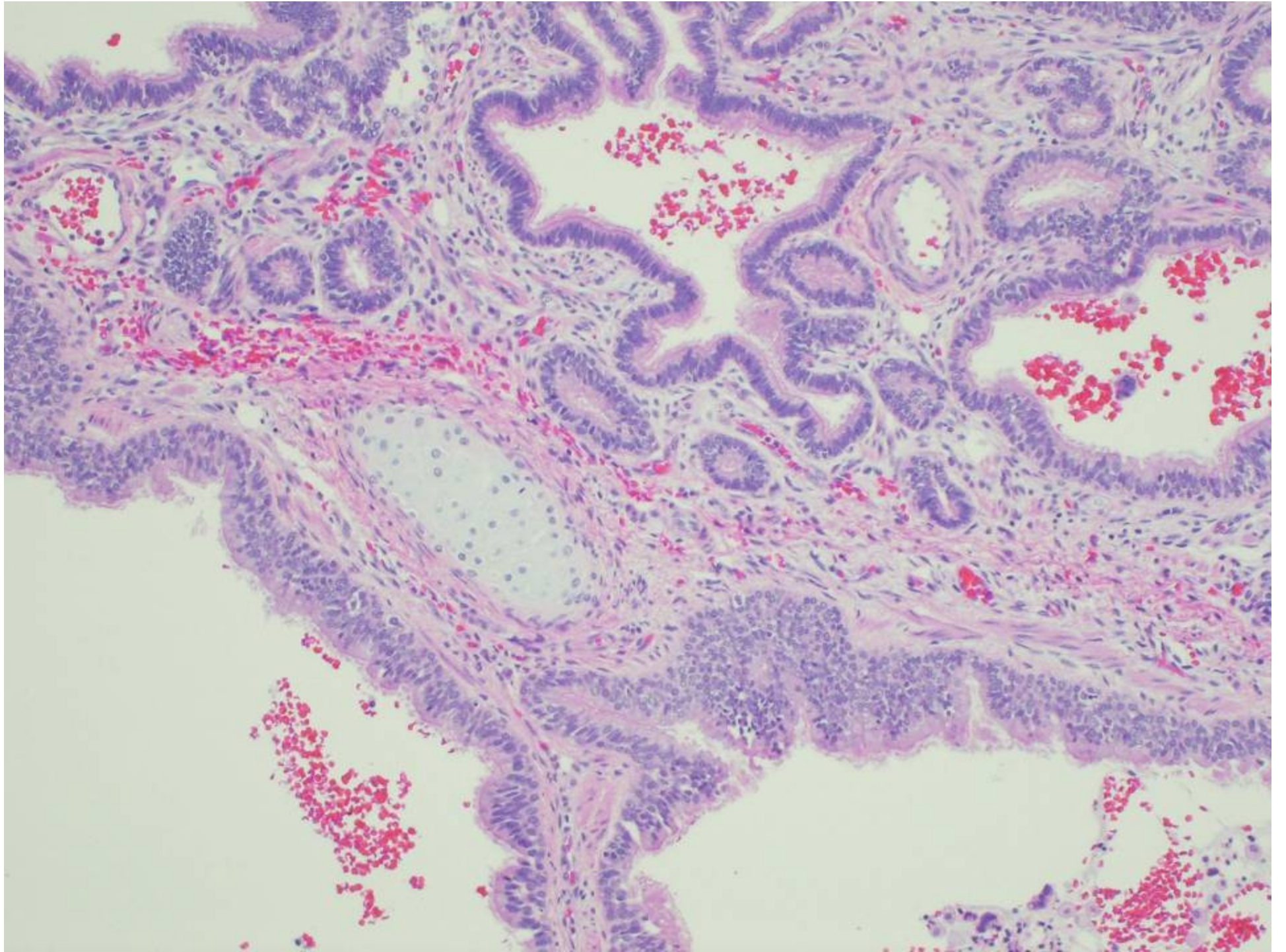


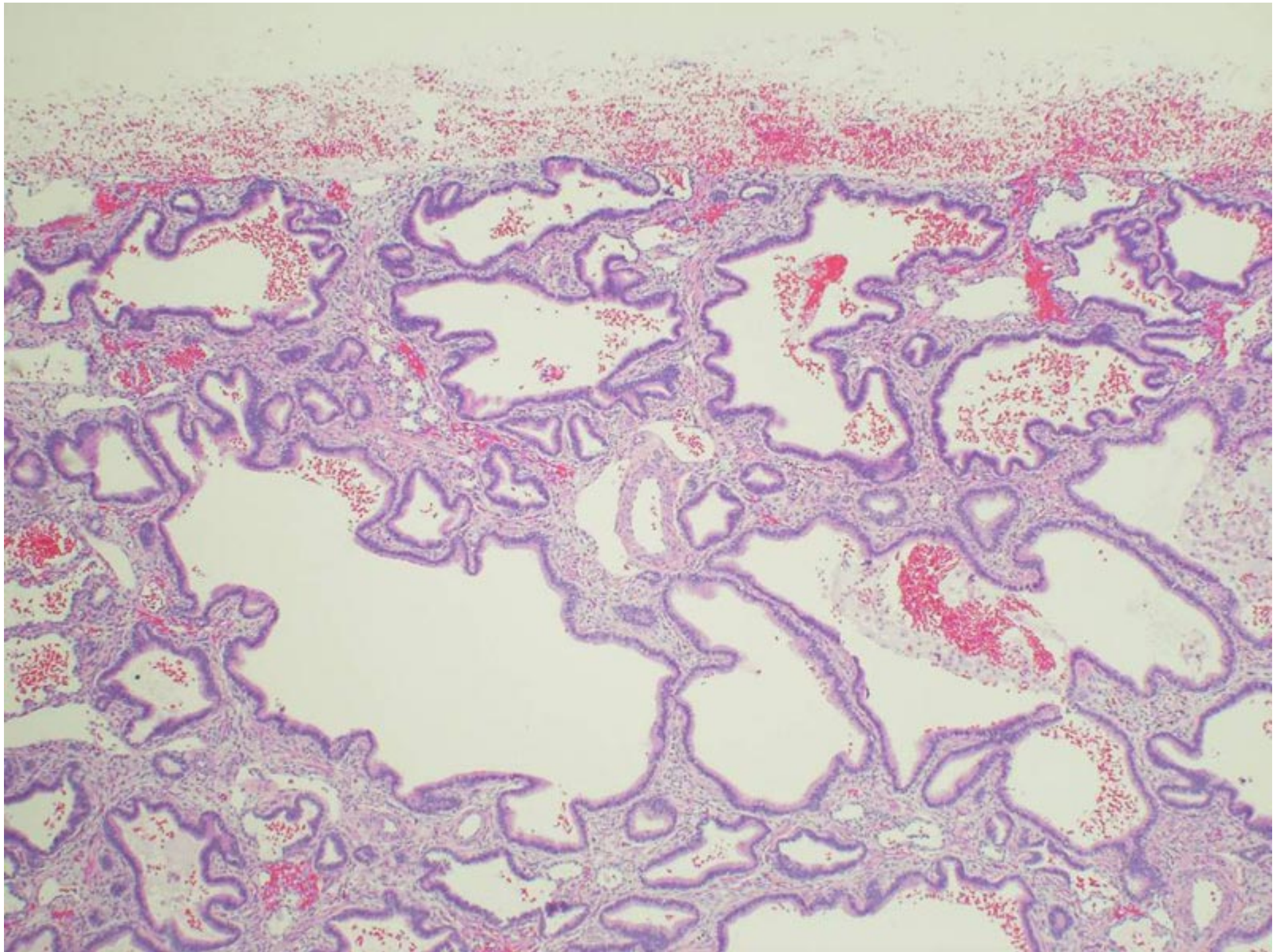
Case 5: History

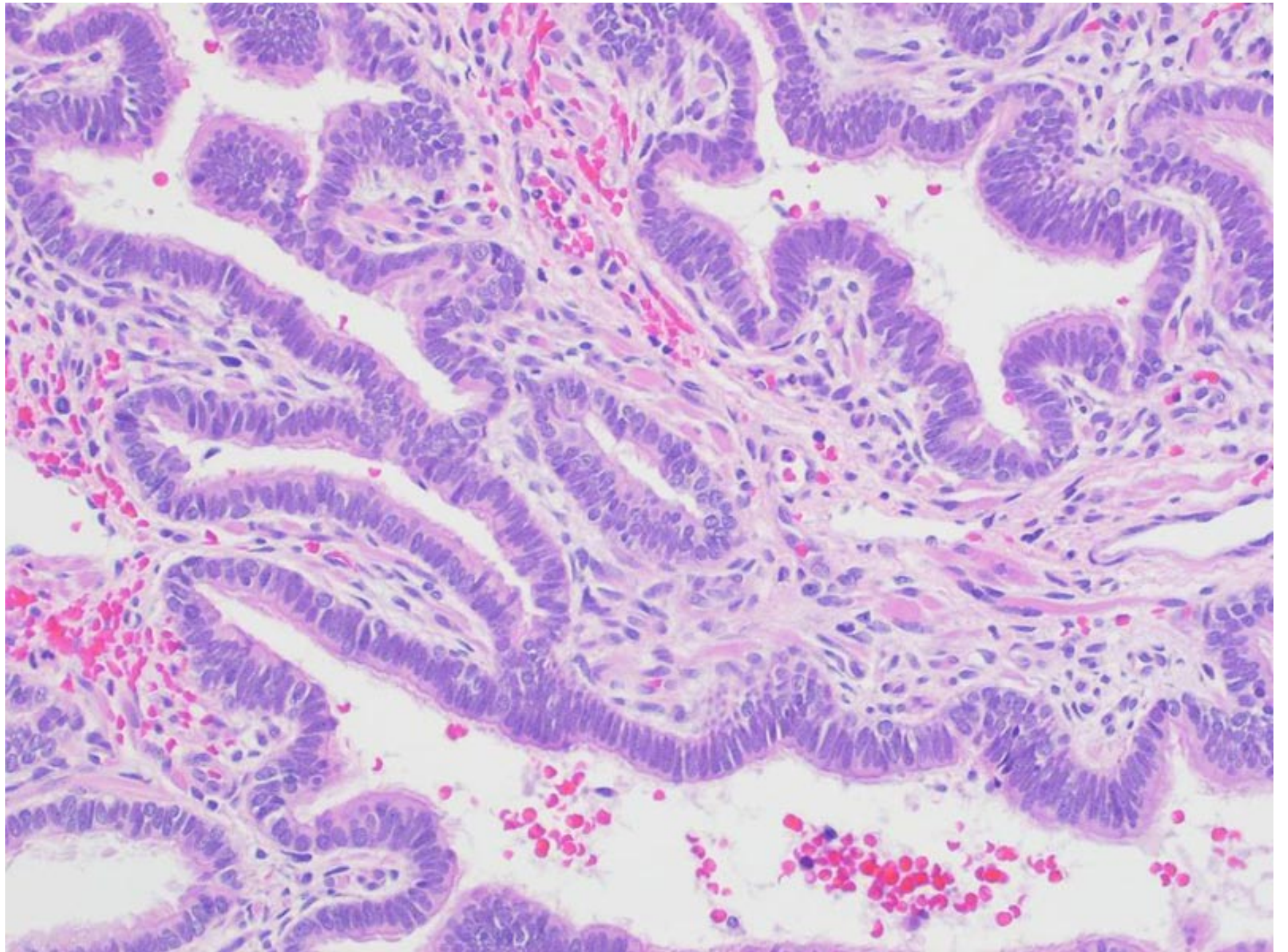
- On prenatal ultrasound, this baby boy was found to have a left lung sequestration/mass. There was no other malformation seen. The lesion was resected at 8 weeks of age. The specimen consisted of tan-pink tissue measuring 3.3 x 2.6 x 1.7 cm. Serial sectioning showed multiple cysts varying in size from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

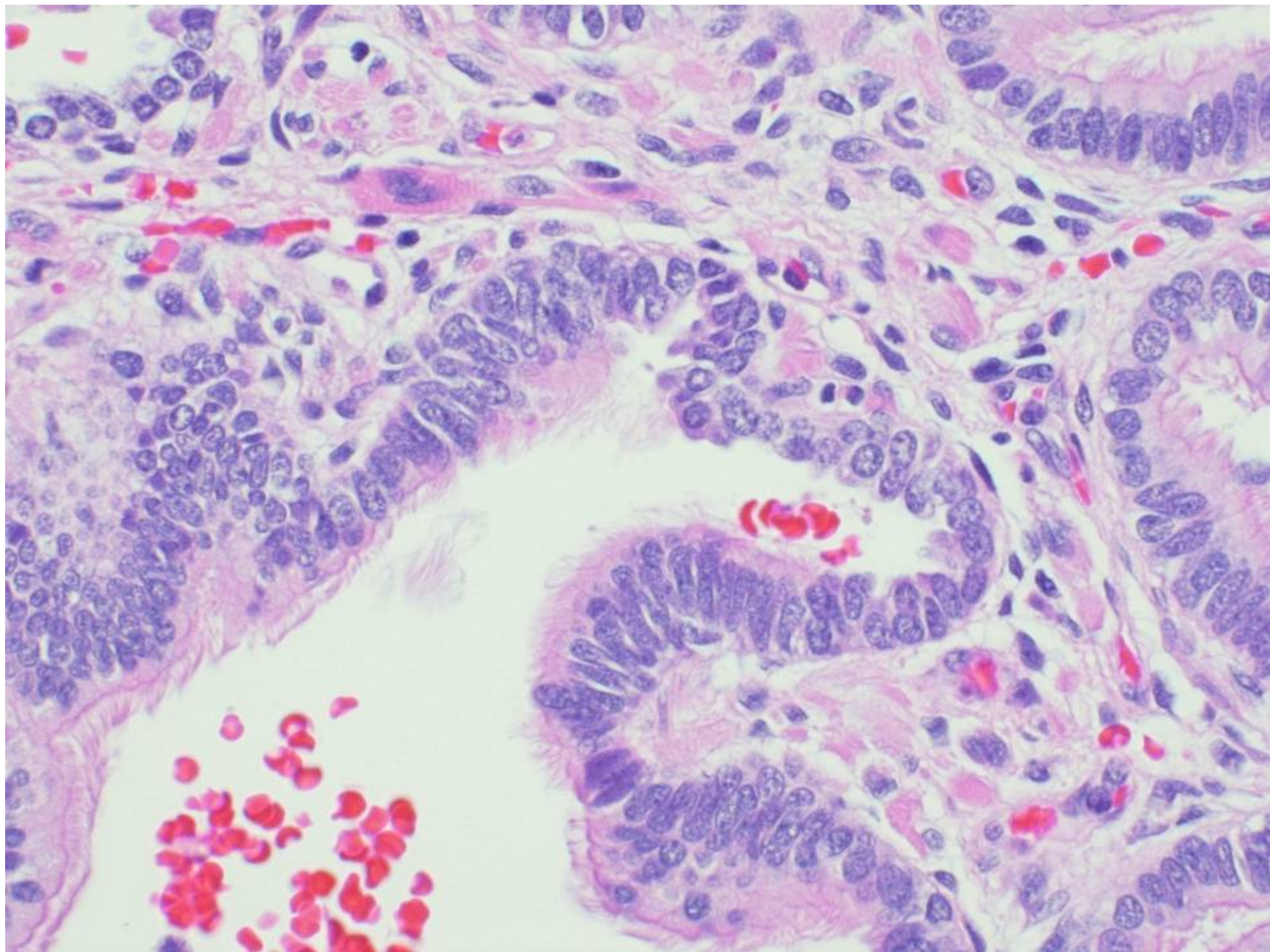
Courtesy of Dr. Chejfec, University of Illinois











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

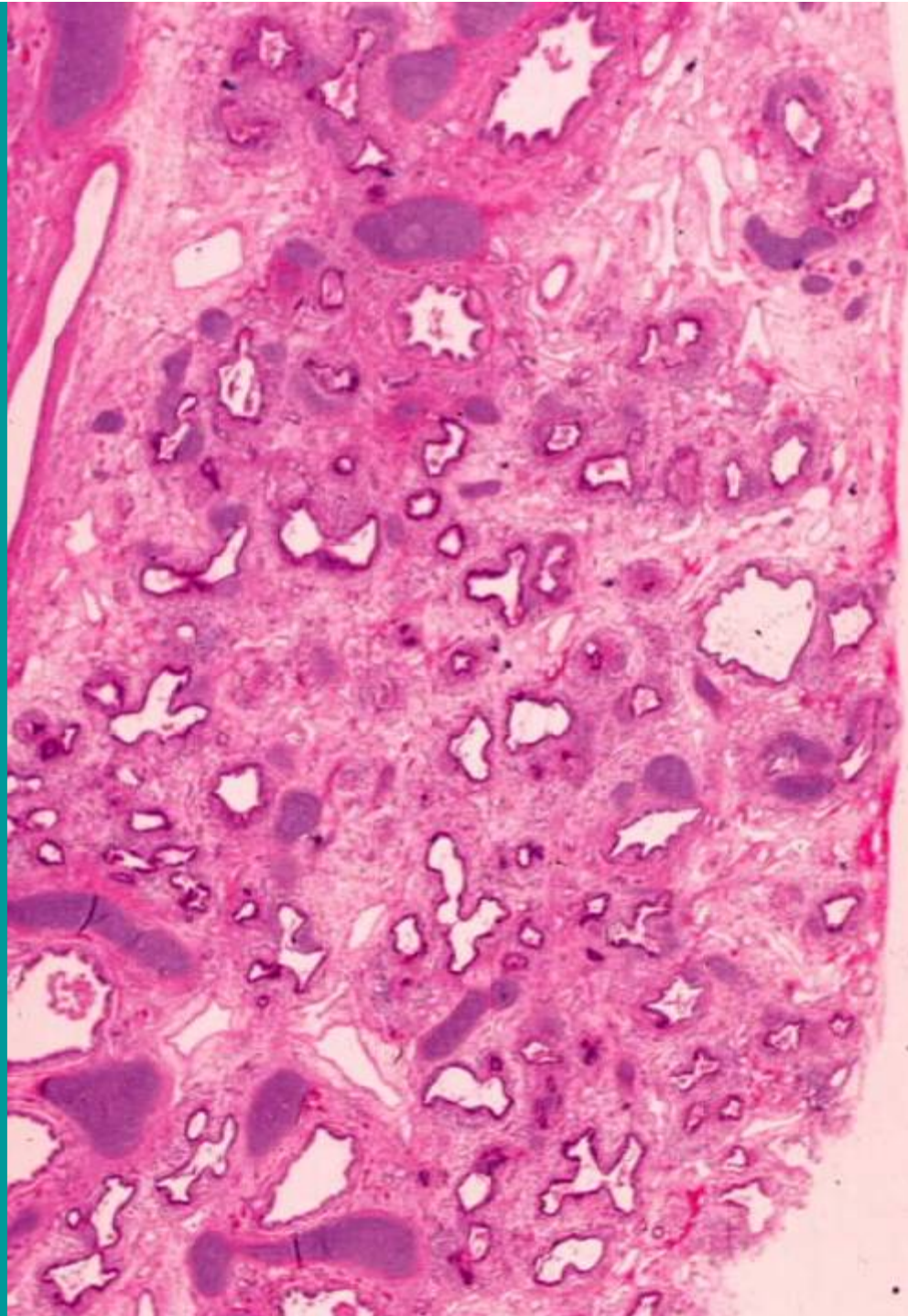
Congenital pulmonary airway malformation (CPAM)

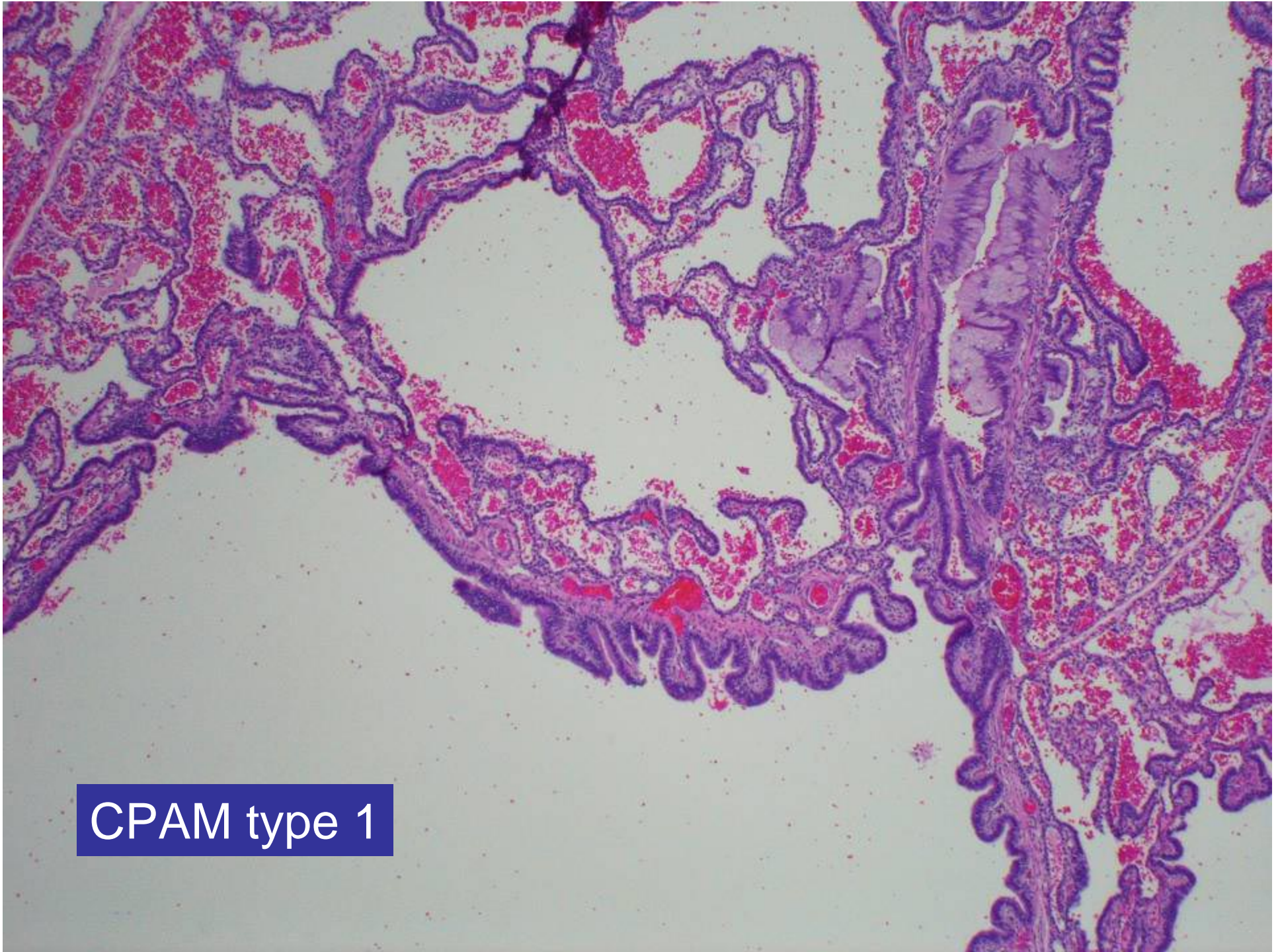
- Classified based on:
 - size of cysts
 - composition of cyst walls

Congenital pulmonary airway malformation

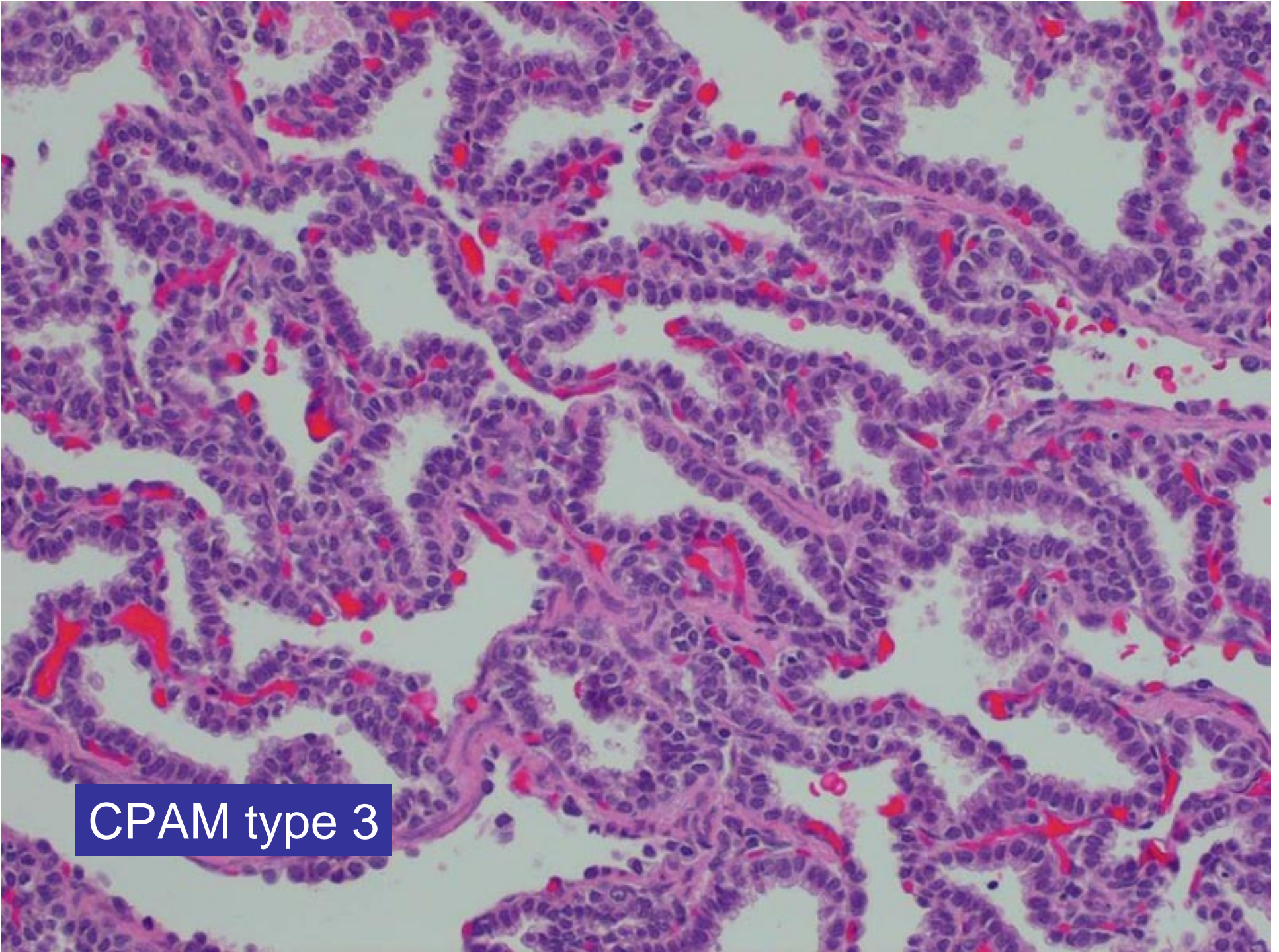
Type	Size of cysts	Cyst wall structure
0	Grossly solid	Bronchi and cartilage
1	3-10 cm	Ciliated, pseudo-stratified with thick muscle, mucinous cells in one-third
2	<2 cm	Ciliated, pseudo-stratified, thin muscle layer
3	Grossly solid	Cuboidal lining
4	Up to 7 cm	Pneumocytes (types 1 & 2)

CPAM type 0

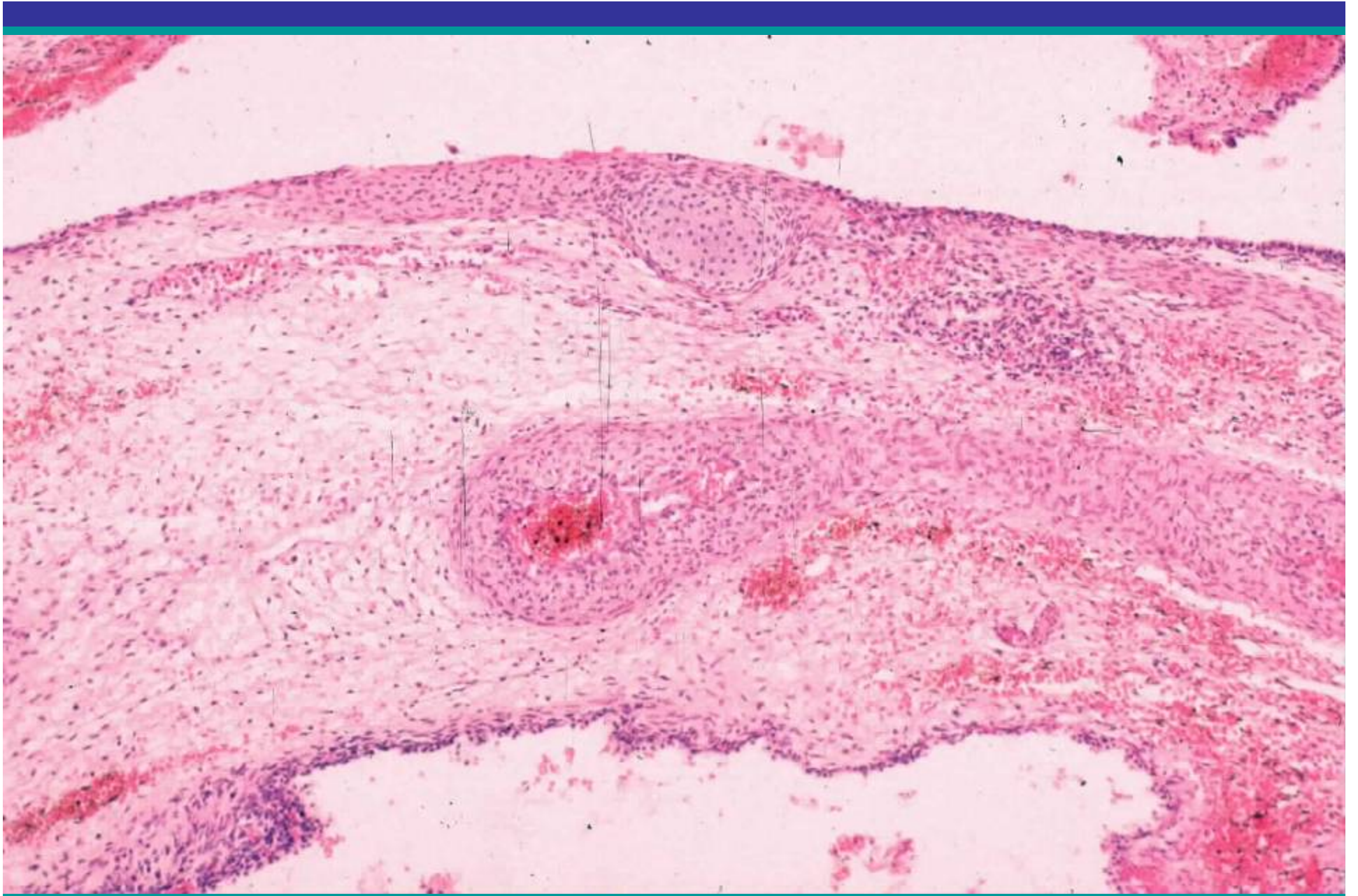




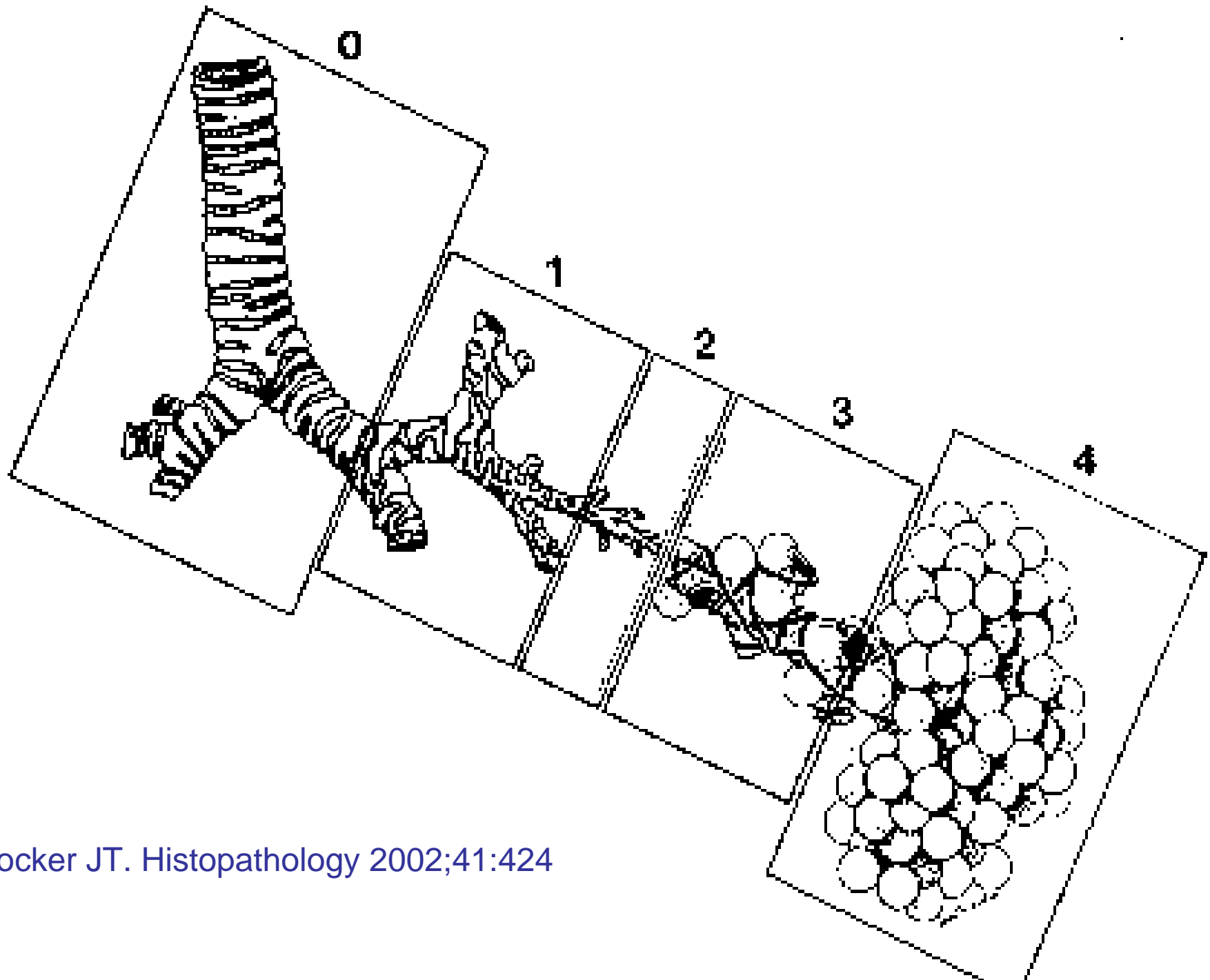
CPAM type 1



CPAM type 3

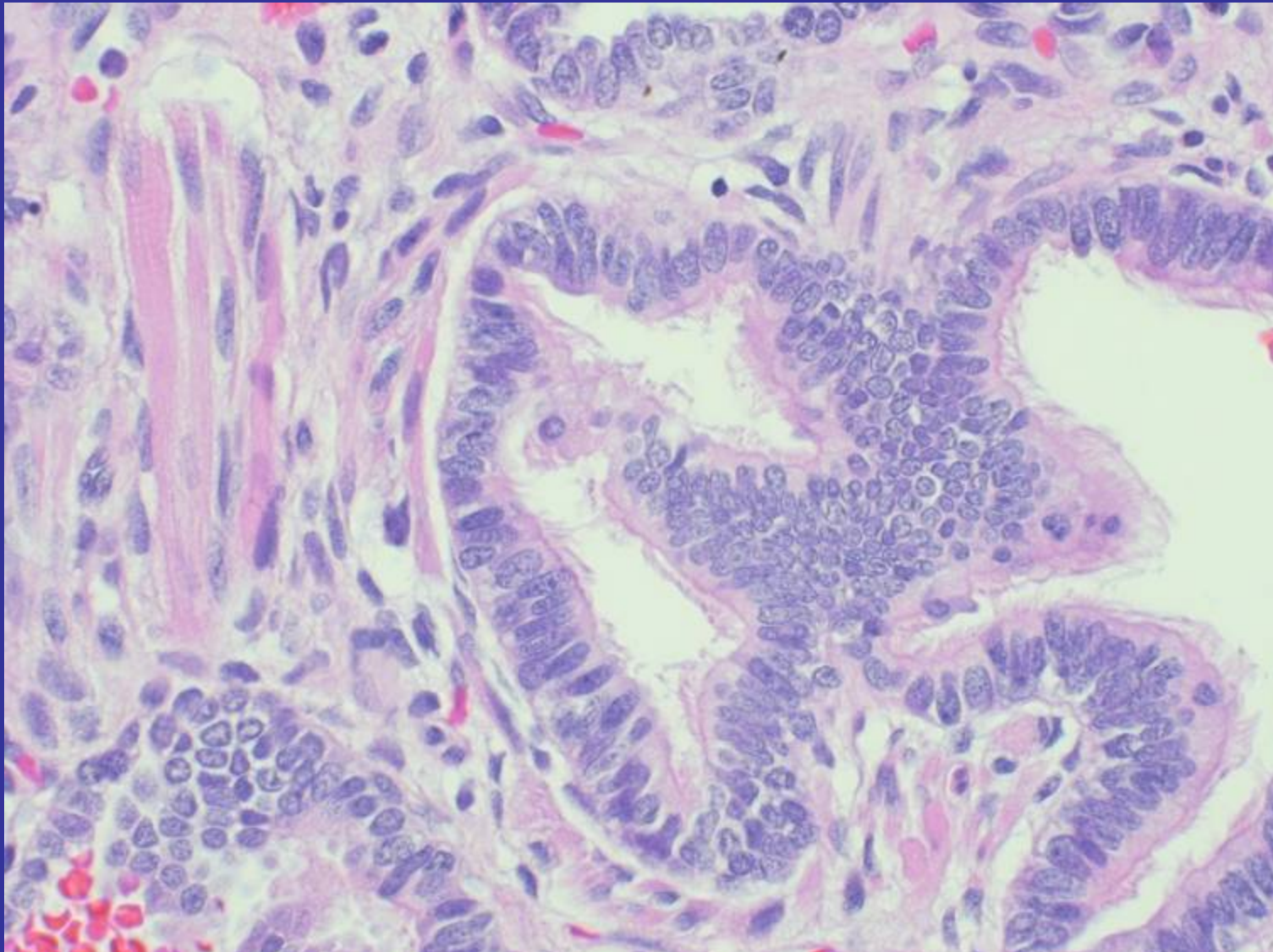


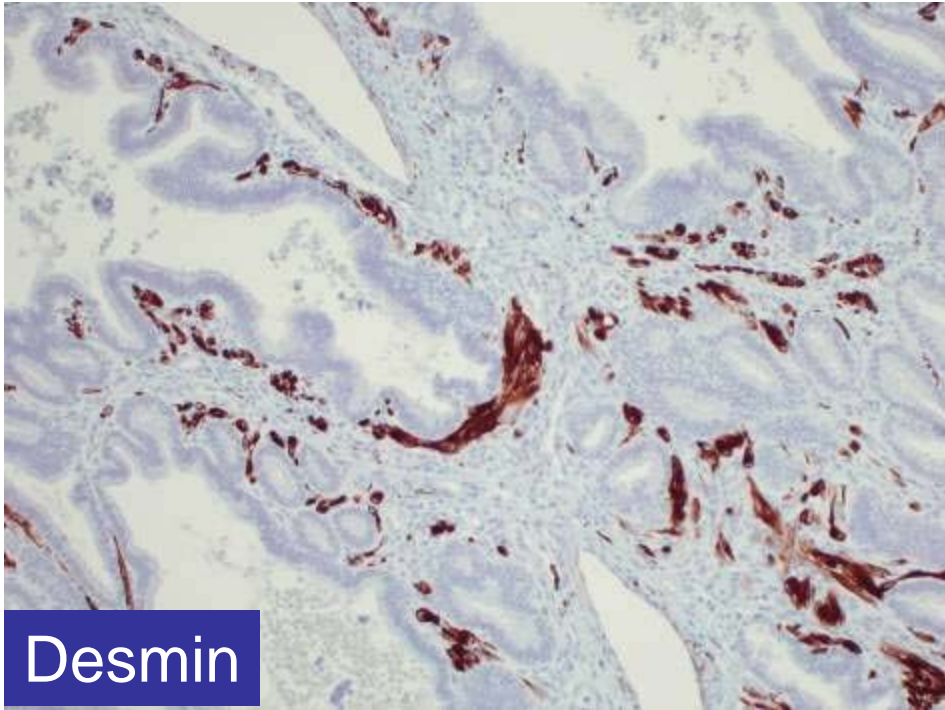
CPAM type 4



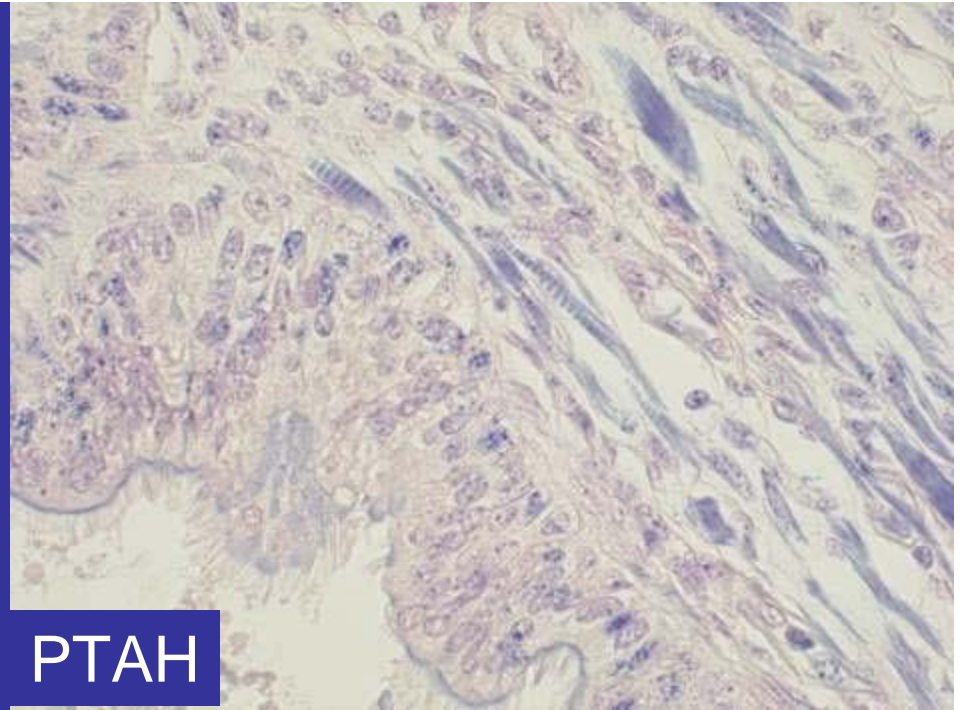
Stocker JT. Histopathology 2002;41:424

CPAM type 2 with striated muscle cells

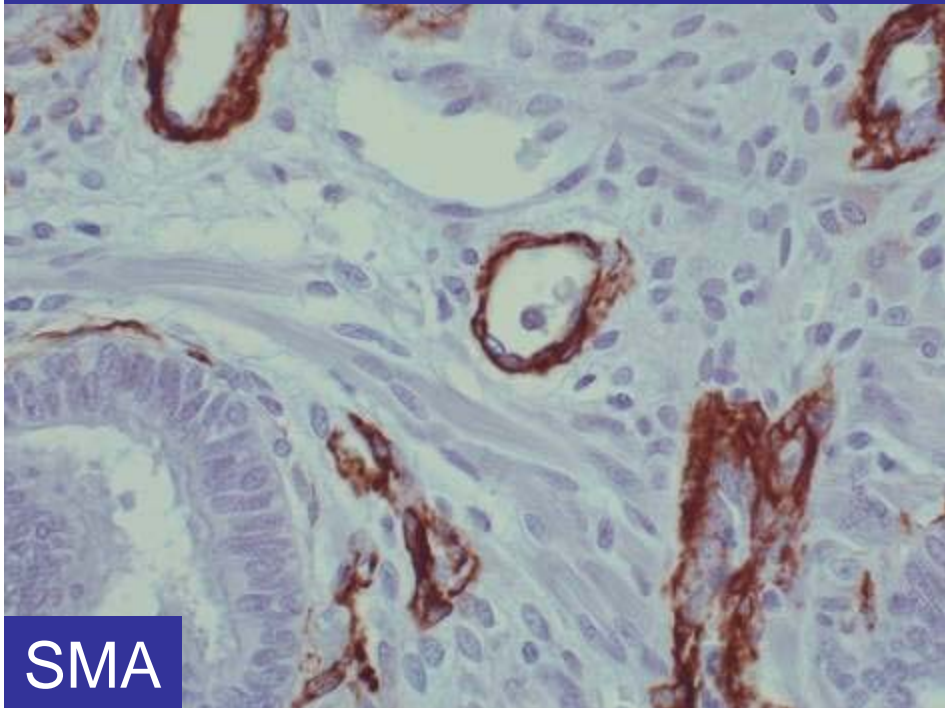




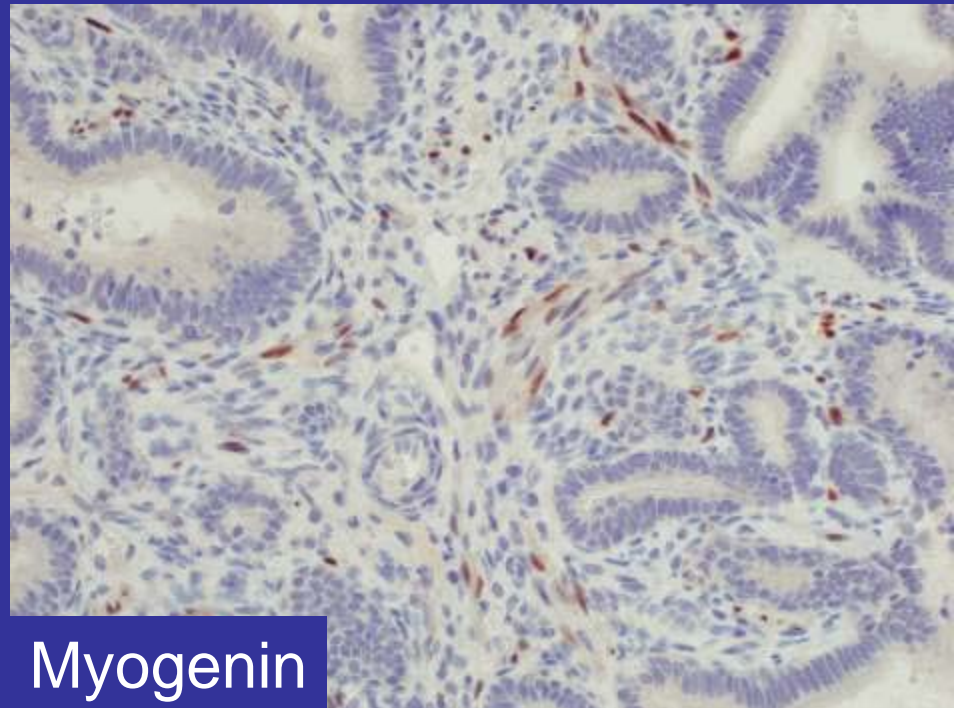
Desmin



PTAH



SMA



Myogenin

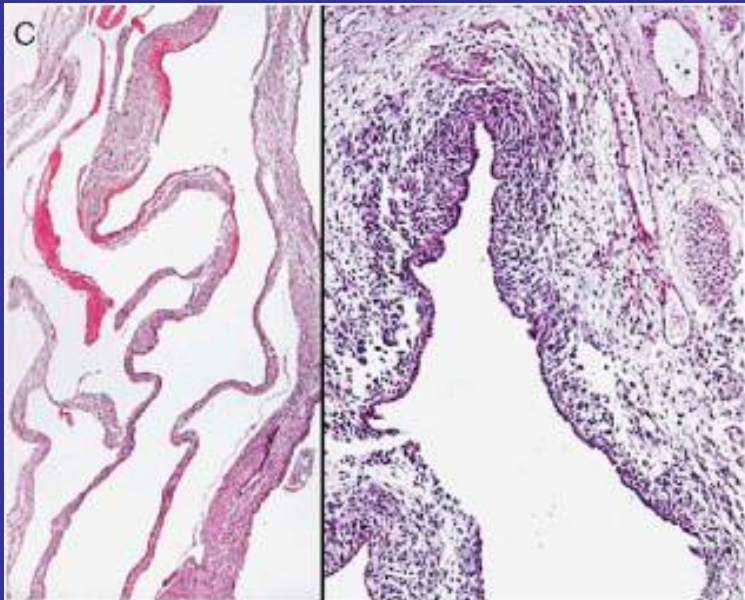
CPAM type 2 with striated muscle cells

- 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

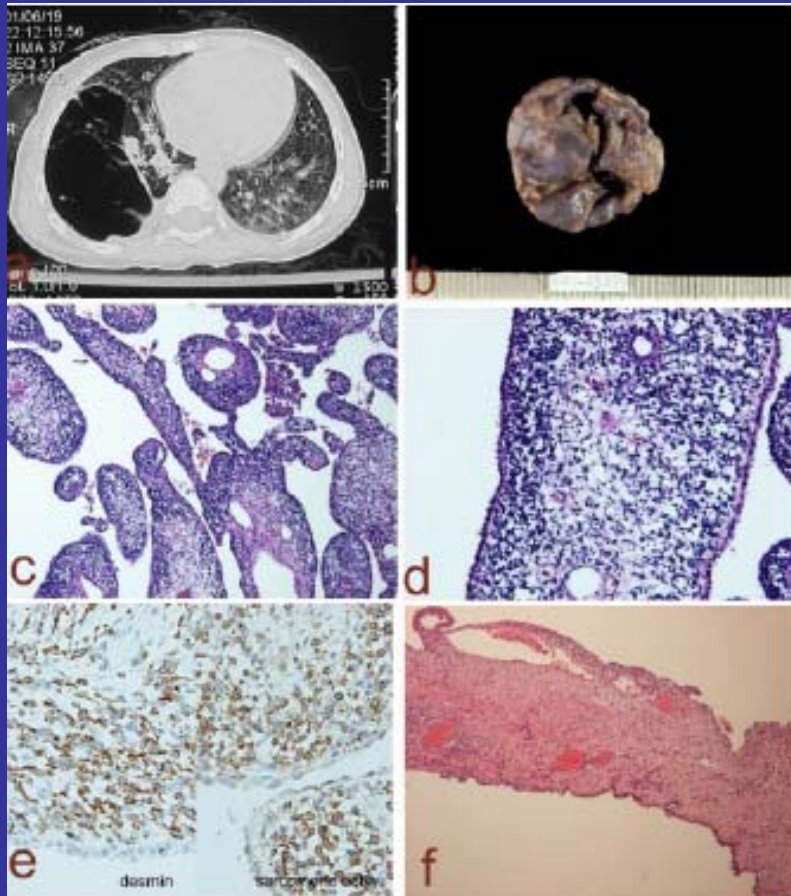
Striated muscle cells in lung

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

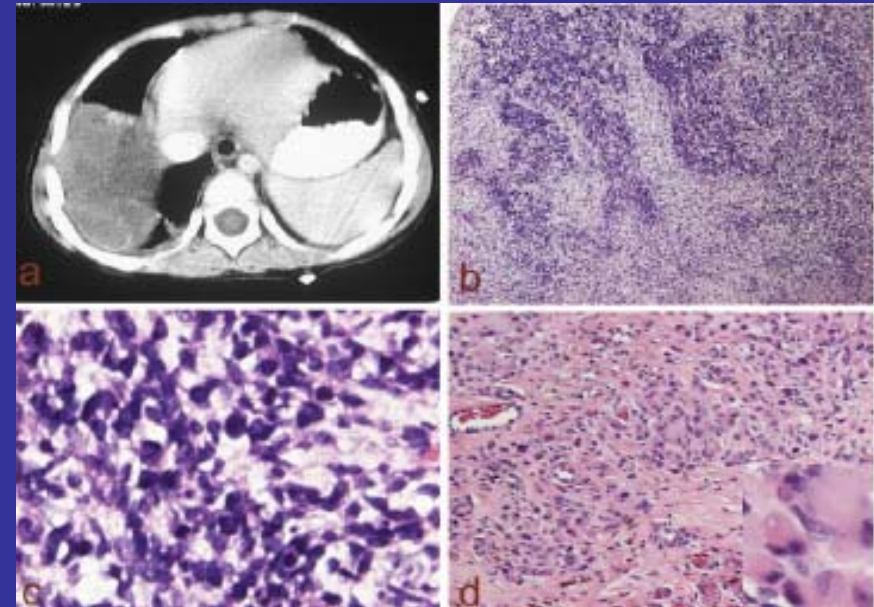


Hill et al. Type 1 Pleuropulmonary Blastoma: 51 cases from the PPB registry. Am J Surg Pathol 2008 32;282-295

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