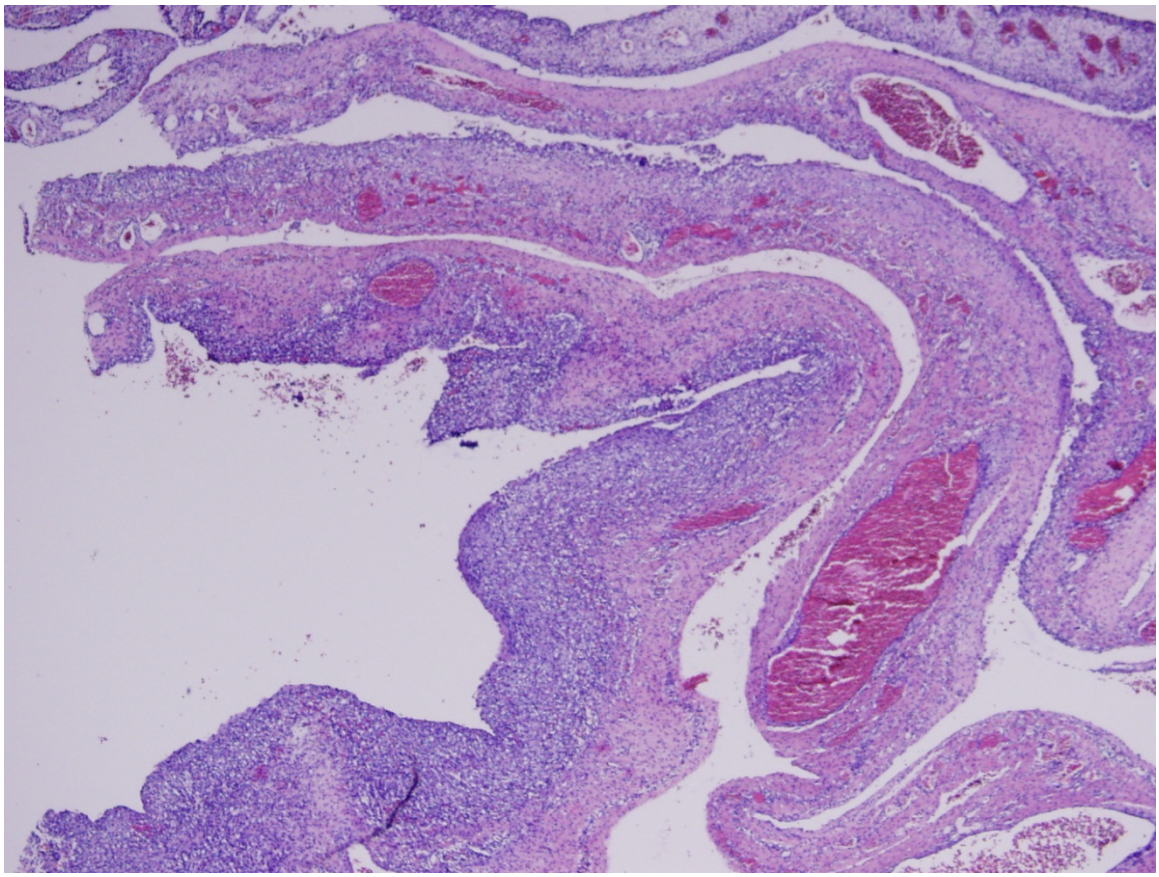
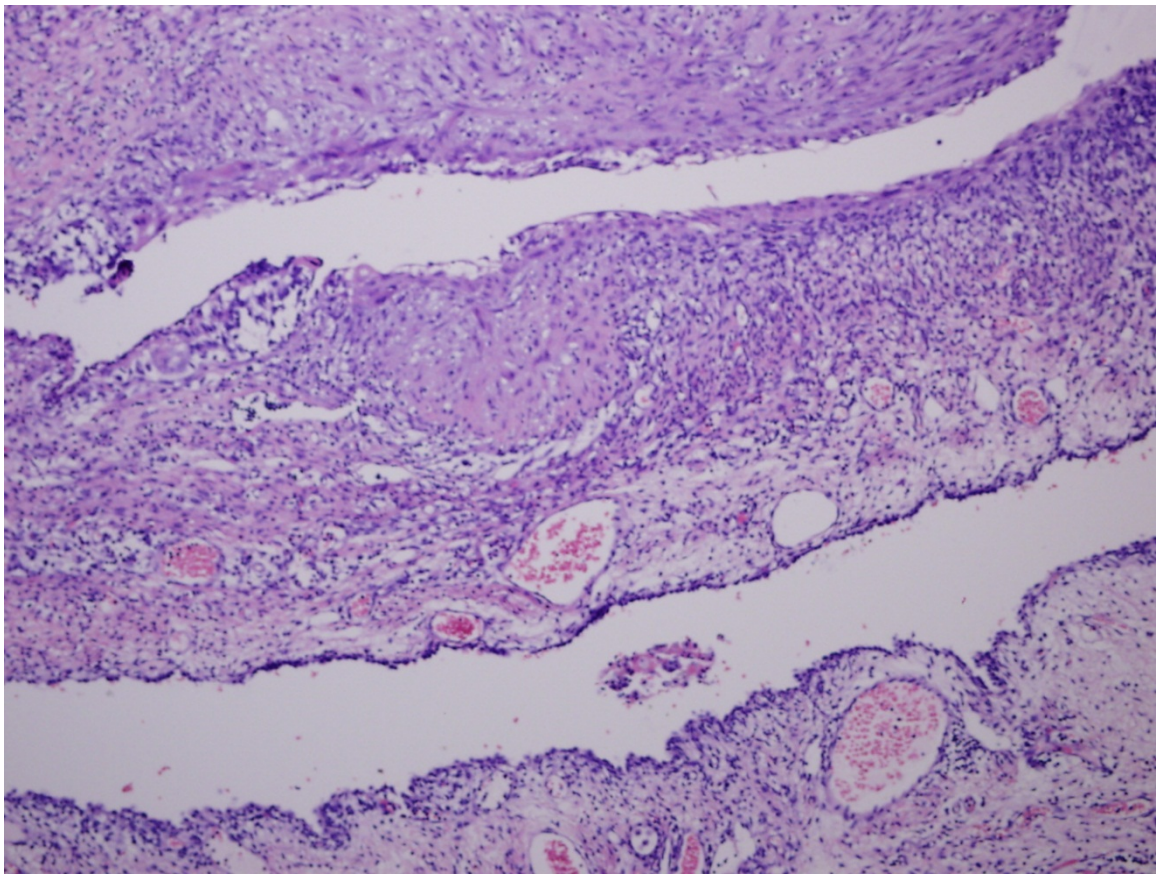
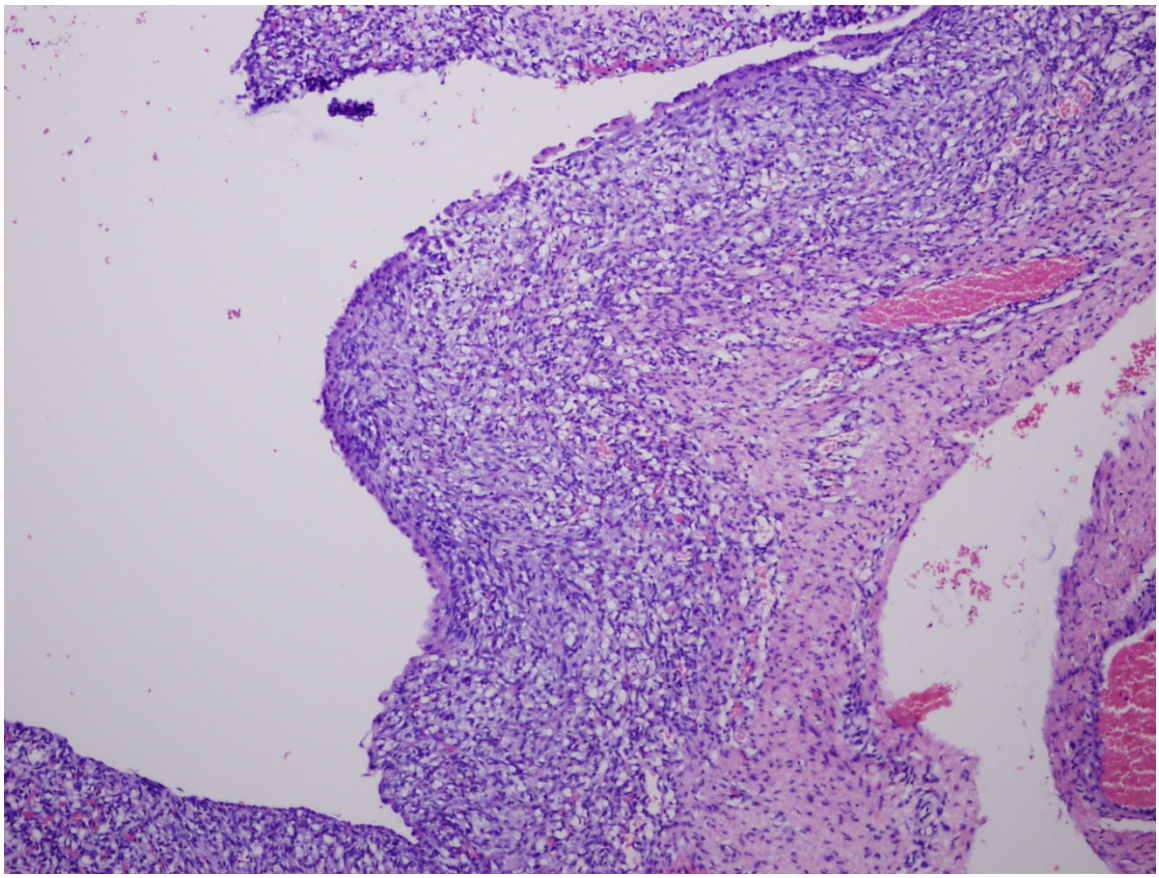
5-month-old child presented with shortness of breath of 2 weeks duration. X-ray chest revealed a cystic emphysematous mass in right lower lobe of lung.

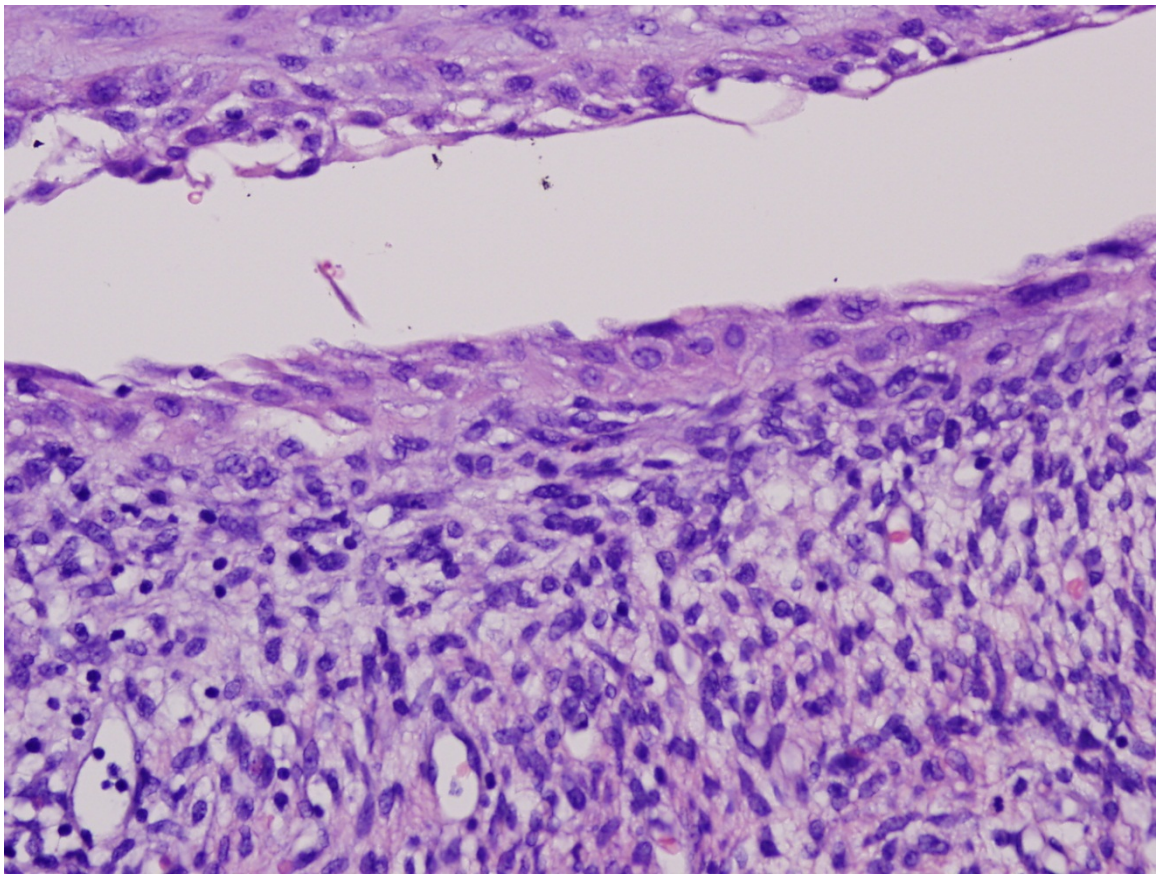
Grossly, a multiloculated cystic mass was received measuring 10x5x2 cms. Cysts had a smooth surface and contained serosanguinous fluid. No solid areas, hemorrhage or necrosis were detected.

Micro images:







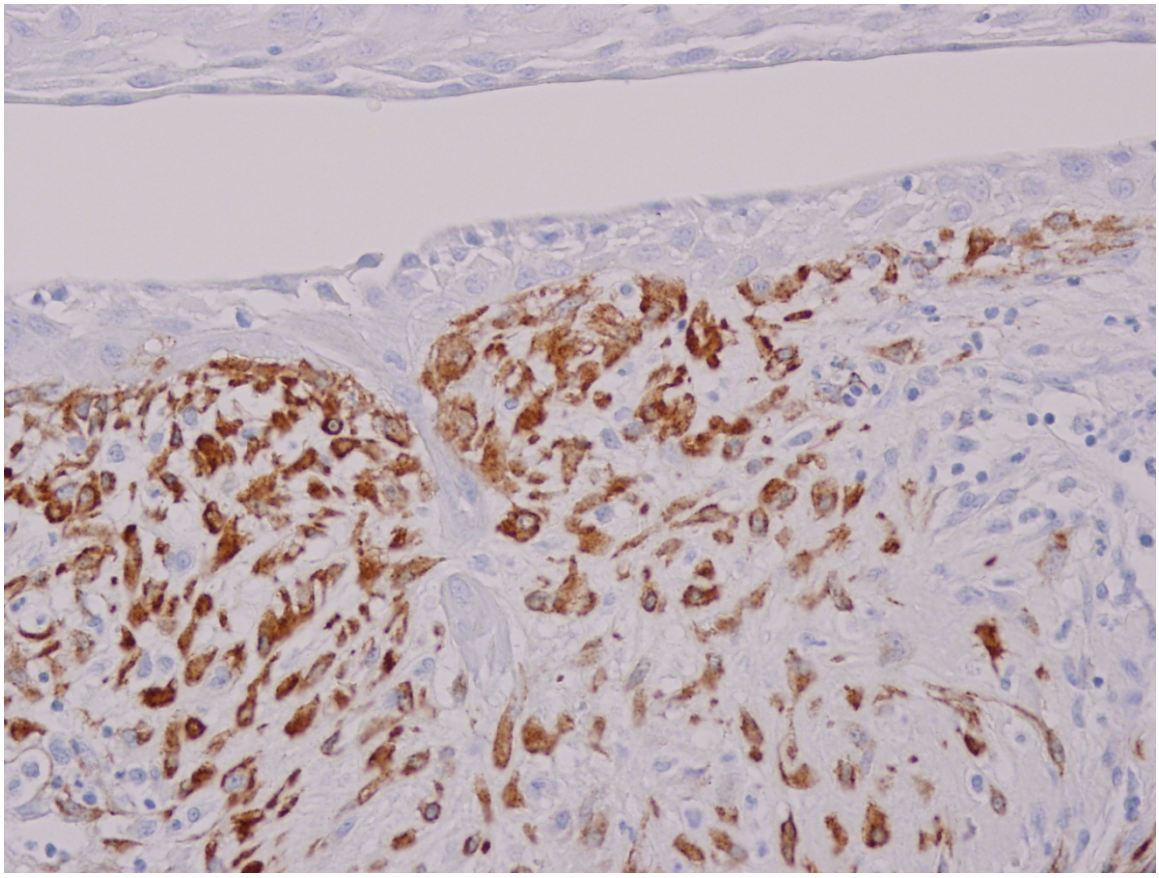


What is your diagnosis?

Diagnosis: Pleuropulmonary blastoma, Type I

Discussion:

Immunostains were obtained:



SMA

Cysts were separated by thin-walled septa and lined by ciliated columnar epithelium. Subepithelial condensation of neoplastic primitive mesenchymal tumour cells was noted. Some cysts showed mature cartilage, and smooth and skeletal muscle in their wall.

Immunohistochemically, neoplastic mesenchymal cells stained positive for vimentin. Smooth and skeletal muscle stained positive for SMA and MyoD1.

This dysontogenetic neoplasm is classified with the "mesenchymal neoplasms" in the WHO Classification of Lung Tumors. (Pathology and genetics of tumours of the lung, pleura, thymus and heart. World Health Organization Classification of Tumours. Lyon, France, IARC Press, 2004)

It is postulated to arise from primitive mesenchymal cell. Abnormally regulated mesenchymal cells proliferate and secrete growth factors that promote proliferation of proximate epithelial cells, leading to formation of cysts (Am J Surg Pathol 2008;32:282).

Etiology is not known. DICER mutations have been recently identified (Science 2009;325:965).

Type I PPB does not have any distinctive features to differentiate it from a developmental lung cyst on imaging. Pathologic examination is the only way to establish the diagnosis, so high clinical suspicion is paramount in correct diagnosis

Children presenting with pneumothorax, multifocality, family history of lung cysts, PPB, rhabdomyosarcoma, nodular hyperplasia or carcinoma of thyroid gland, hamartomatous intestinal polyps, cystic nephroma, or any childhood malignancy should raise the possibility of PPB and encourage removal of lung cysts for examination.

Surgical resection is the treatment of choice. Adjuvant chemotherapy has been recommended by PPB registry since 2003. Close surveillance is required for recurrence or progression to type II or III PPB (Journal of Clinical Oncology 2006;vol24 no.27:4492).