

IMAGING FOR RESIDENTS

A Hypoechoic Lesion in the Lung of a Fetus at 22 Weeks of Gestation

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Section 2—Answer

A 31-year-old, primigravid woman, without known underlying systemic or obstetrical history, received regular prenatal examinations that had been uneventful. She and her husband were healthy, nonconsanguineous, and without known family history of congenital malformations.

The fetal sonography performed at 22 weeks of gestation revealed a singleton with a hypoechoic lesion in the right lung (Figure 1). Amniotic fluid amount was normal and fetal growth was as up to schedule. No anomalies were found in other internal organs.

What is the diagnosis?

Interpretation

Prenatal sonography showed a multiple cystic lesion in the right lower lobe of the lung (Figure 1), which was consistent with the subsequent fetal magnetic resonance imaging that

revealed a hyperintensive lesion with multiple soap bubbles appearance on T2-weighted images (Figure 2). Coronal T2-weighted images showed intact diaphragm (arrow) and normal location of liver (Figure 3). Macrocystic congenital pulmonary airway malformation (CPAM) in the right lower lobe of lung was suspected. The patient opted to terminate the pregnancy. *Postmortem* examination revealed that the right lower lobe of lung was occupied by a cystic lesion measuring 4.2 cm × 4.1 cm × 3.1 cm in size (Figure 4). The lesion appeared as multiloculated cystic space with septa. Other lung lobes were intact. Microscopically, multiloculated cystic spaces were found in the lung parenchyma (Figure 5), and were lined by single-layered cuboid, ciliated epithelium in the background of alveolar structure (Figure 6). Presence of smooth muscle fibers beneath the respiratory epithelium suggested terminal bronchial origin (Figure 7). Other fetal lung parenchymas were in canalicular phase. No mucous cells were present in the cystic lining. No cartilage or primitive neoplastic components were found. The features were consistent with type 2 CPAM.

Conflicts of interest: The authors declare no conflict of interest.

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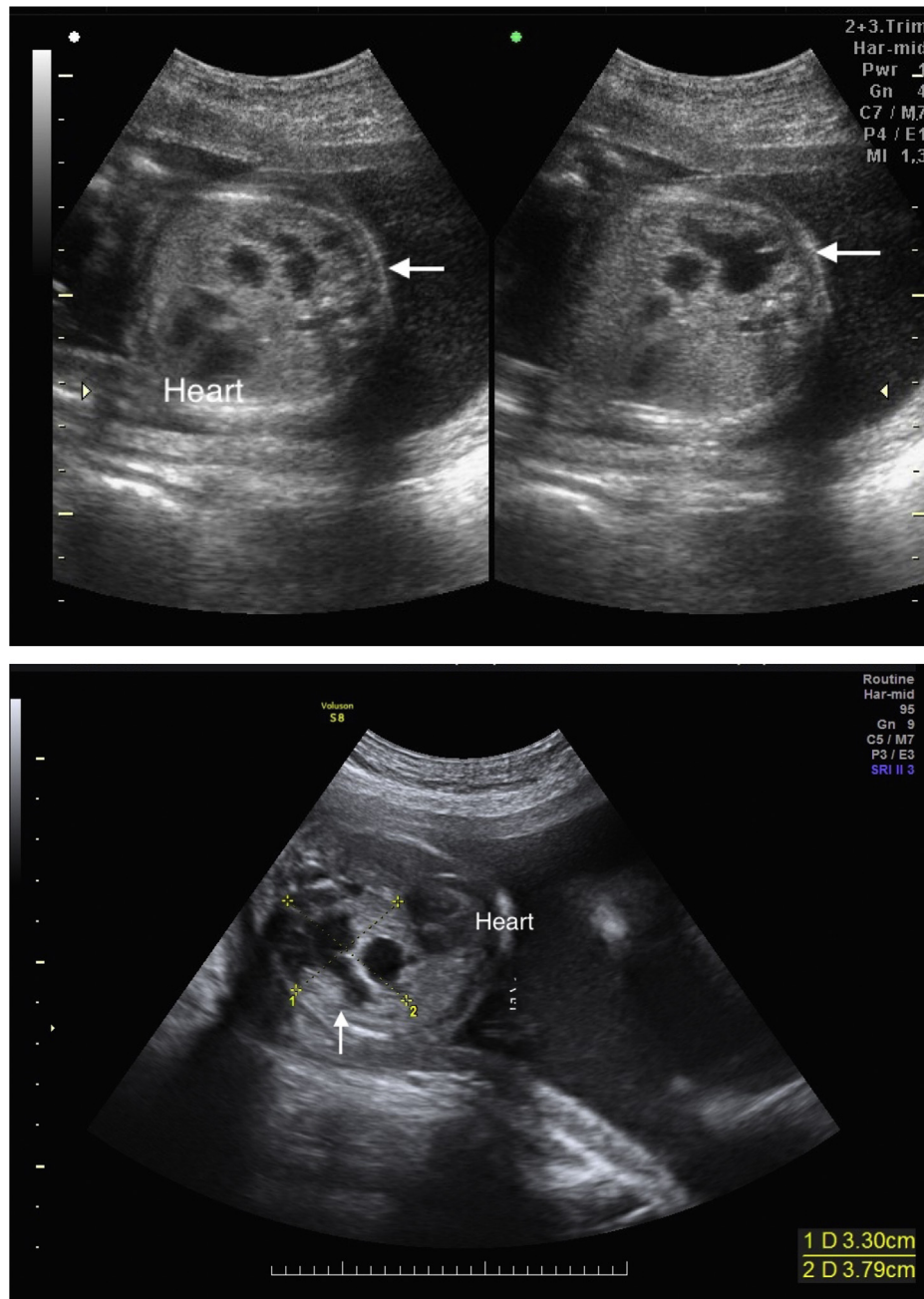


Figure 1 Prenatal sonography demonstrates the axial view of the fetal chest, which shows a multiple cystic lesion (arrow) in the right lung.

Discussion

CPAM of the lung is a lesion characterized by a multicystic mass of pulmonary tissue with a proliferative bronchial structures. CPAM is a modified term for congenital cystic adenomatoid malformation, reflecting the developmental disorder of pulmonary airway morphogenesis. CPAM was originally subdivided into three types (Type I, II, and III) based on the pathologic characteristics [1]. Type 0 and Type 4 were later added to the CPAM classification [2]. Type 0 shows grossly solid appearance with histologic features of

bronchi and cartilage; Type 1 shows cysts > 2 cm with presumed bronchial/bronchiolar origin; Type 2 shows cysts < 2 cm with sponge-like appearance resembling bronchioles; Type 3 appears solid or shows very small cysts (< 0.2 cm) with presumed bronchiolar/alveolar origin; and Type 4 are characterized by very large cysts up to 10 cm with presumed distal acinar origin. In fetal imaging the classification of these anomalies is primarily based on cyst size as microcystic (< 5 mm) or macrocystic (> 5 mm) [3,4].

The differential diagnosis of fetal thoracic masses includes congenital diaphragmatic hernia, bronchogenic or

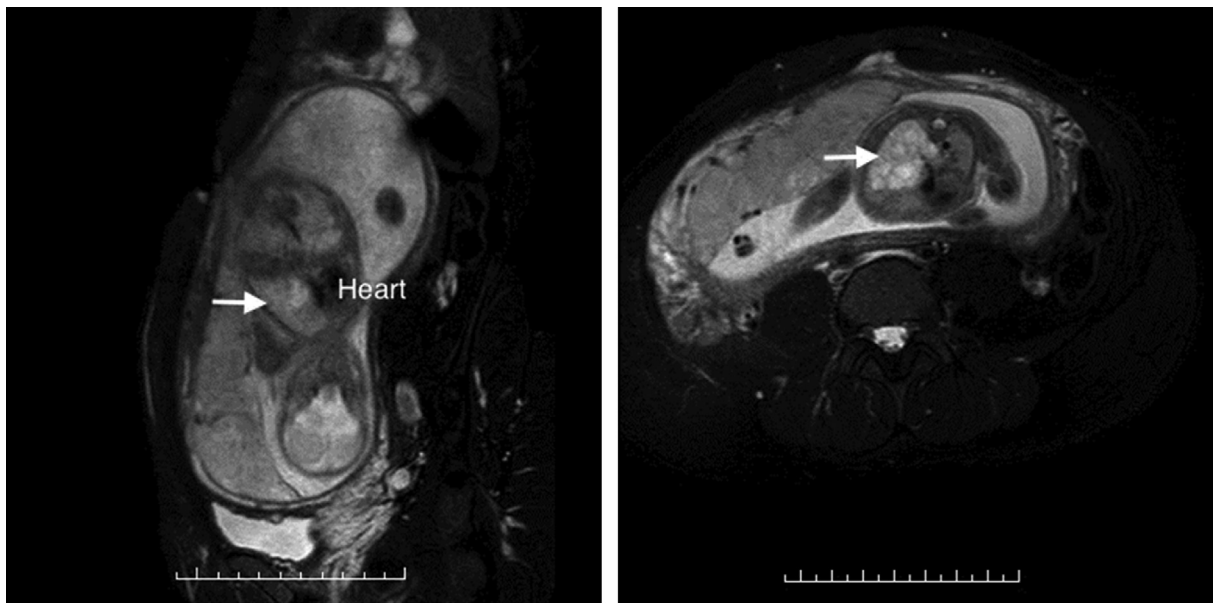


Figure 2 A hyperintense lesion (arrow) with multiple soap bubbles appearance on T2-weighted magnetic resonance images is noted in the right lower lobe of lung.

enteric cysts, bronchopulmonary sequestration (BPS), bronchial atresia or stenosis, congenital lobar obstruction, and hybrid lesion (CPAM + BPS). The appearance under sonography of different types of CPAM can lead to varied differential diagnosis. Macrocystic CPAM appears as cystic or echolucent pulmonary mass and is more likely to be

confused with a congenital diaphragmatic hernia. In contrast, microcystic CPAM typically appears as a large hyperechogenic mass and the main differential diagnosis is BPS. Fetal magnetic resonance imaging is also a useful tool for the diagnosis and characterization of CPAM to differentiate from other fetal thoracic masses [5].



Figure 3 Coronal T2-weighted magnetic resonance images showed intact diaphragm (arrow) and normal location of liver.

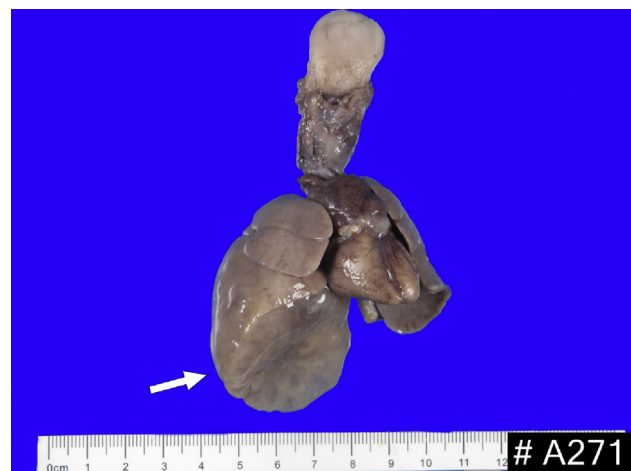


Figure 4 The specimen consists of tongue, larynx, thymus, heart, and bilateral lungs. Macroscopically, an enlarged right lower lobe of the lung measuring 4.2 cm × 4.1 cm × 3.1 cm in size is identified. The lesion is composed of multiple small cystic spaces with mucoid contents. Other lung lobes are intact.

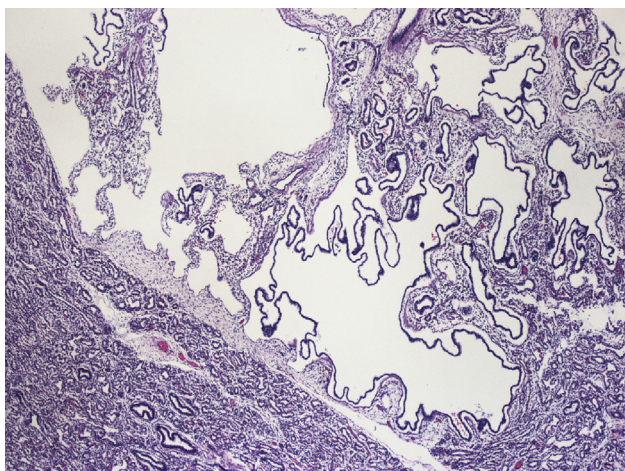


Figure 5 Multiloculated cystic spaces of varied sizes are lined by ciliated respiratory epithelium. Left lower field shows fetal lung parenchyma in canalicular phase (hematoxylin–eosin, original magnification $\times 40$).

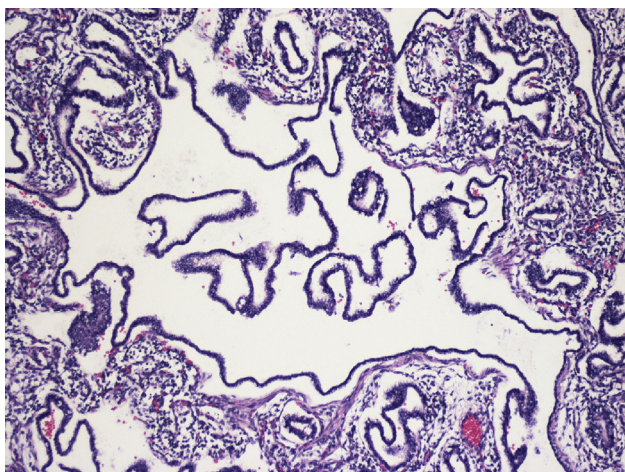


Figure 6 Multiloculated spaces are lined by single-layered cuboid, ciliated epithelium in the background of alveolar structure (hematoxylin–eosin, original magnification $\times 40$).

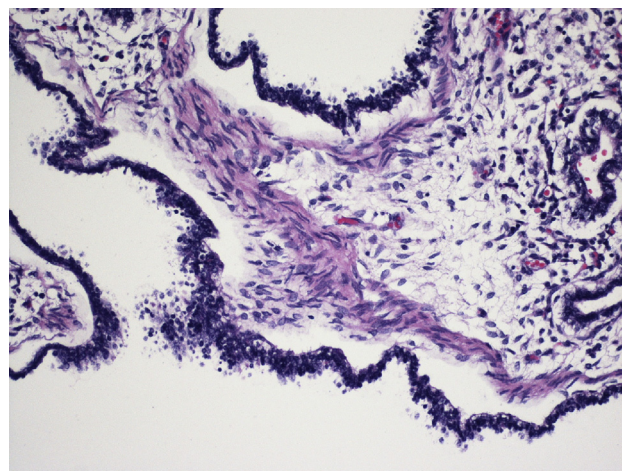


Figure 7 The congenital pulmonary airway malformation cyst is lined by ciliated respiratory epithelium (hematoxylin–eosin, original magnification $\times 400$).

References

- [1] Stocker JT, Madewell JE, Drake RM. Congenital cystic adenomatoid malformation of the lung. Classification and morphologic spectrum. *Hum Pathol* 1977;8:155–71.
- [2] Stocker JT. Congenital pulmonary airway malformation: a new name and an expanded classification of congenital cystic adenomatoid malformation of the lung. *Histopathology* 2002;41:424–31.
- [3] Adzick NS, Harrison MR, Glick PL, et al. Fetal cystic adenomatoid malformation: prenatal diagnosis and natural history. *J Pediatr Surg* 1985;20:483–8.
- [4] Adzick NS. Management of fetal lung lesions. *Clin Perinatol* 2009;36:363–76.
- [5] Levine D, Barnewolt CE, Mehta TS, et al. Fetal thoracic abnormalities: MR imaging. *Radiology* 2003;228:379–88.