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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 T2weighted 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 \times 4.1 cm \times 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.

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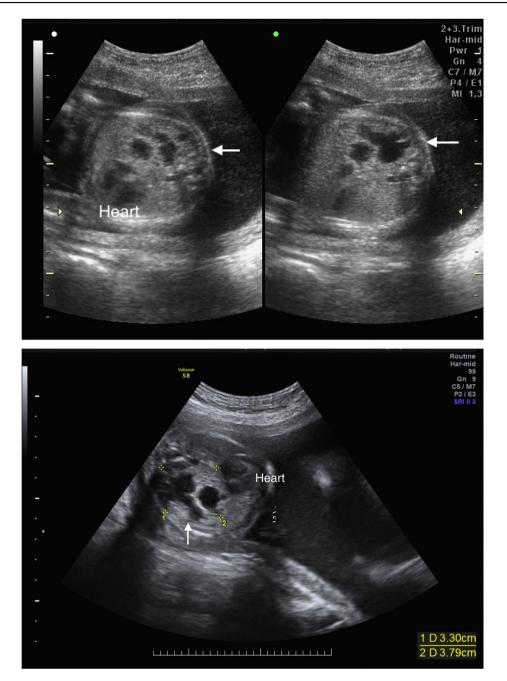


Figure 1 Prenatal sonography demonstrates the axial view of the fetal chest, which shows a multiple cystic lesion (arrow) in the right lung.

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

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