Intradiaphragmatic Pulmonary Sequestration
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History
Left suprarenal mass on prenatal ultrasound.

Diagnosis
Intradiaphragmatic Pulmonary Sequestration

Additional Clinical
The patient developed pyloric stenosis at 6 weeks of age. Inspection of the left upper quadrant at laparoscopic pyloromyotomy did not reveal a mass. A repeat MR was then performed.

Discussion
Pulmonary sequestration is the result of an aberrant lung bud. During the fourth week of gestation the normal lung bud develops from the ventral wall of the pharynx at the laryngotracheal groove. From the 5th to 17th week, the developing lung is glandular with epithelial tubes branching into the surrounding mesenchyme to form the airways. Development of an aberrant lung bud from the foregut caudal to the laryngotracheal groove leads to disorganized lung tissue that is never connected with the normal developing lung, tracheobronchial tree or pulmonary arteries. Depending on the timing of the formation of the aberrant lung bud, the aberrant tissue may or may not be covered by visceral pleura, either being intra- or extralobar, respectively. Extralobar sequestrations tend to be associated with other anomalies, while intralobar sequestrations more frequently become infected. An interesting variant is the rare intradiaphragmatic pulmonary sequestration. Development of the diaphragm starts in the 4th week with the septum transversum, which incompletely separates the thoracic and abdominal cavities. The resultant defects on each side are called pleuropertitoneal canals and are closed by the pleuropertitoneal membrane around the 8th week of gestation. If the aberrant developing lung migrates to the level of the pleuropertitoneal canals near the 8th week of gestation, it can be enclosed by the pleuropertitoneal membrane, resulting in an intradiaphragmatic pulmonary sequestration. On imaging all sequestrations are solid masses that often contain cysts. Identifying the systemic arterial supply is key to diagnosis. Most commonly they are in the chest adjacent to the left hemidiaphragm, 10-15% are intraabdominal, and only rarely are they intradiaphragmatic. The challenge arises in locating the unsuspected intradiaphragmatic location before the time of surgery. Neither a transthoracic or intraabdominal approach will directly visualize the sequestration. This can lead to multiple surgeries before correct diagnosis and successful surgical removal. A high index of suspicious is required at initial diagnosis.

Findings
Fetal MR-Sagittal and coronal SSFSE T2 images show a hyperintense extrapulmonary suprarenal mass with a few intralesional cysts. Postnatal US-Performed shortly after birth shows hyperechoic mass with few small cysts bounded by the adrenal gland and kidney. Postnatal MR-Axial T2 images from MR performed after pyloromyotomy shows the lesion embedded
within the diaphragm.

Reference
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