Title: Sequestering the Diagnosis of Extralobar Bronchopulmonary Sequestration: A Case Report

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Background/Synopsis: Congenital abnormalities of the lower respiratory tract are rare, encountered in approximately 1 in 10,000 to 35,000 live births. Within this category of anomalies are congenital cystic adenomatoid malformations, teratomas, diaphragmatic hernias and bronchopulmonary sequestration. Bronchopulmonary sequestrations (BPS) are particularly rare, comprising only 0.15 - 6% of all respiratory tract abnormalities. BPS is a non-functioning mass of lung tissue that contains airway and alveolar air elements but lacks normal communication with the tracheobronchial tree. This non-functioning lung tissue receives blood supply from systemic circulation. There are two categories of bronchopulmonary sequestration, intralobar and extralobar. Intralobar sequestrations are located *within* the same pleural cover as the normal lung within the thorax, while extralobar sequestrations are located *outside* the pleural covering of the lung and may be intrathoracic or extrathoracic. The differential diagnoses for extrathoracic (i.e. intra-abdominal) BPS include mesoblastic nephromas and neuroblastomas.

Objective/Purpose: According to the literature, tertiary care referral centers may identify and treat less than 1 case of bronchopulmonary sequestration per year. This case of extralobar BPS offers an opportunity to discuss its antenatal and early postnatal course and recognize the wide range of differential diagnoses when evaluating a fetal pulmonary or upper abdominal mass.

Case Report: A 19-year-old obese female, gravida 2 para 1001 at 20 weeks of gestational age was referred for maternal-fetal-medicine consultation for evaluation of a suspected fetal intra-abdominal abnormality. AFP, NIPT and gene carrier screening were normal. Ultrasound revealed a fetal heterogeneous mass measuring 2.0 x 1.9 x 1.8 cm, located superior to and contiguous with an otherwise normal appearing left kidney. There was no visible systemic blood supply to this mass. A single umbilical artery was also appreciated. No other fetal abnormalities were identified. The fetus had a male phenotype. The fetus was growing normally.

The differential diagnoses for the abdominal mass included adrenal neuroblastoma, adrenal hemorrhage, simple adrenal cyst, renal malformations, mesoblastic nephroma and extralobar, extrathoracic pulmonary sequestration, with neuroblastoma seemingly most likely. Serial ultrasound evaluations were performed monthly. At 30 weeks EGA, an adjacent 1.1 x 1.1 x 1.1 cm simple cyst was identified. Imaging findings remained relatively stable until 38 weeks EGA when echogenic bowel was appreciated, with a heightened suspicion for metastasis. Delivery was recommended and the patient underwent a successful induction of labor and uncomplicated vaginal delivery. The neonate weighed 3400 oz., with Apgar scores of 8/9.

The male infant was evaluated by the neonatology and pediatric surgery teams. The neonate appeared clinically well and the newborn physical examination was unremarkable. Neonatal abdominal ultrasound was significant for a complex solid and cystic mass in the left upper quadrant posterior to the spleen with the solid echogenic component consistent with fetal lung tissue. A prominent systemic vessel was seen arising from the abdominal aorta feeding the solid component. These ultrasound findings were consistent with extralobar sequestration. The infant and mother had an uncomplicated post-delivery course and were discharged home on post-delivery day 2. The infant was scheduled for

pediatric surgery reevaluation at 3 months of age to examine the lesion and plan for elective surgical excision if the mass persists.

Conclusion: Bronchopulmonary sequestration is a rare occurrence that may present in both intralobar and extralobar forms, making antenatal diagnosis challenging. As in this case, extralobar pulmonary sequestration may present in a retroperitoneal location, mimicking renal neuroblastoma. This case allows the opportunity to review contrasting ultrasound findings between pulmonary sequestration, neuroblastoma, and other intra-abdominal masses. Neonates with pulmonary sequestration may have be quiescent course or suffer complications ranging from massive hemothorax, recurrent infections and hemoptysis. If BPS is suspected, delivery should occur at a tertiary care center.