Case Based Fetal Lung Masses

Advances in Fetal and Neonatal Imaging Course
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Prenatal Imaging of fetal lung masses

- US primary imaging modality
  - Lungs homogeneous; hyperechoic to liver and increasing echogenicity with gestational age
  - Focal increase in echogenicity, mediastinal shift, cysts can indicate mass

- MRI can help further delineate lung mass
  - Added clinical value debated
  - Levine, et al 2003, MRI changed management in only 8% of cases with fetal thoracic abnormalities; additional information in 38%
Fetal Chest: KEY POINTS

- CLASSIFICATION IS DIFFICULT
- BRONCHIAL ATRESIA IS LIKELY UNDERLYING CAUSE OF MOST LESIONS
- BIG IS BAD
- HYDROPS IS VERY VERY BAD
Fetal Lung Masses

• Bronchopulmonary malformations:
  • Bronchial Atresia
  • Congenital Lobar Overinflation
  • Foregut Duplication Cyst
  • Congenital Pulmonary Airway Malformation
  • Bronchopulmonary Sequestration
  • Hybrid Lesions

• Other Masses:
  • Pleuropulmonary Blastoma
  • Mediastinal Teratoma
  • Congenital High Airway Obstruction
Fetal Lung Masses

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Bronchial Atresia

• Proximal bronchial atresia with mucoid impacted central bronchi and normal distal lung architecture.

• Most common: apicoposterior segment of left upper lobe > RUL > RML.

• Likely occurs between gestational weeks 5 and 15, may be due to loss of arterial supply.

• Associations: CLE, BPS, CPAM ("bronchial obstruction sequence")
Bronchial Atresia
Bronchial Atresia
Fetal Lung Masses

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Congenital Lobar Overinflation

- Progressive over distention from obstruction in at least one segment or lobe.
- Obstruction can be intrinsic or external
- Congenital heart disease 15-40%.
Congenital Lobar Overinflation
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Foregut Duplication Cyst

- Bronchogenic cyst, enteric cyst, neuroenteric cyst.
- Abnormal ventral budding of the tracheobronchial tree.
- Well defined mass, usually middle mediastinum
- Homogeneous, brighter than CSF in T2 and STIR.
Foregut Duplication Cyst
Foregut Duplication Cyst
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Congenital Pulmonary Airway Malformation

• Stocker Classification:
  • Original, 1977, 3 types
  • Current 4 types
  • Controversial, not comprehensive

• Langston Classification: more descriptive, based on pathogenesis
Congenital Pulmonary Airway Malformation
Congenital Pulmonary Airway Malformation

- Type 0
- Acinar dysplasia
- Exceedingly rare
- Usually bilateral
- Incompatible with life
Congenital Pulmonary Airway Malformation

• Type 1
• Large cyst type
• 65%
• Associated with airway obstruction in utero
• Hamartoma vs. neoplasm
Congenital Pulmonary Airway Malformation

- Type 2 - medium cysts
- Bronchial atresia with obstruction
- 10-15%
- Seen in ELS and ILS
Congenital Pulmonary Airway Malformation

- Type 3 - very small cysts
- Pulmonary hyperplasia, solid/adenomatoid type
- 5-8%
- Hamartoma vs. hyperplasia
Congenital Pulmonary Airway Malformation

- Type 4 - Peripheral cyst type
- Pleuropulmonary blastoma: type 1 PPB?
- Regressed type 1 adenomatoid malformation
- 10-15%
- neoplasm
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Bronchopulmonary Sequestration

- Bronchial atresia with systemic vascular connection
- Nonfunctioning pulmonary tissue with no connection to the tracheobronchial tree or pulmonary arteries.
- Well defined wedge shaped mass.
- Most common in posterior segment of left lower lobe. LLL > RLL.
- 90% above and 10% below the diaphragm.
- Homogeneous and solid if above diaphragm.
- Often cystic if below the diaphragm - can be mistaken for an adrenal mass.
Bronchopulmonary Sequestration
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BPS/CPAM hybrid lesions

- Systemic vascular supply with cystic components
- “bronchial atresia malformation sequence”
BPS/CPAM hybrid lesions
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Pleuropulmonary Blastoma

- Malignant embryonal tumor
- 3 types: cystic, mixed cystic and solid, solid
  - Type 1 occurs only in infants
- Current thinking is that Stocker Type 4 CPAM and PPB can be easily mistaken
  - Possible regressed or undersampled
Pleuropulmonary Blastoma
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Mediastinal Teratoma

- Most common mediastinal germ cell tumor.
- Arise from primitive germ cell rests.
Mediastinal Teratoma
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  - **Congenital High Airway Obstruction**
Congenital High Airway Obstruction Syndrome

- Pulmonary hyperplasia resulting from laryngeal atresia
- Hyperechogenic lungs
CHAOS:

day 1

day 2

3 months
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