Congenital Lung Malformations: Radiologic-Pathologic Correlation

Marilyn J. Siegel, M.D.
Mallinckrodt Institute of Radiology
Washington University School of Medicine
St. Louis, MO

Learning Objectives
• Review clinical & pathologic features of common lung anomalies
• Describe imaging appearances of the common congenital anomalies
• Emphasize imaging differential diagnosis

Congenital Lung Anomalies
• Normal vascularity
  - Congenital lobar hyperinflation
  - Congenital pulmonary airway malformation
  - Bronchogenic cyst
  - Bronchial atresia
  - Parenchymal agenesis, hypoplasia
• Abnormal vascularity
  - Scimitar syndrome
  - Sequestration

Congenital Lobar Overinflation
• Formerly, lobar emphysema-misnomer
• Etiology-partial bronchial obstruction
  - Deficient bronchial cartilage
  - Intraluminal web, stenosis, malacia
• Onset: 90% < 6 months
  - Neonates: dyspnea, cyanosis, cough
  - Older pts: wheezing or incidental
Congenital Lobar Overinflation
Pathologic Features

- Gross path:
  - Sponge-like mass, fails to deflate on resection
  - Compressed normal lung deflates & reexpands

- Histopathology:
  - Alveolar distention 5-10X normal

Lobar Overinflation: Imaging

- Overinflated lobe
- Atelectasis of adjacent lung
- Mass effect
  - Mediastinal shift, attenuated vessels
- Opaque if fluid-filled
- Lobar predilection:
  - LUL (50%), RML (24%), RUL (18%)

Lobar Overinflation
Neonate with respiratory distress

Lobar Overinflation
2-week-old boy with mild dyspnea
**Congenital Lobar Overinflation**

- Usually diagnosed in neonates & infants
- 10% diagnosed in adolescents & adults

**Lobar Overinflation**

**DDX: Swyer-James MacLeod Syndrome**

- Bronchiolitis obliterans
  - viral infection in childhood **
- Imaging findings
  - unilateral lucent (low attenuation) lung
  - small or normal size **
  - bronchiectasis **
  - air-trapping on expiration

**Swyer-James Syndrome**

10-year-old boy, cough

**Helps to differentiate from CLE**
**Swyer-James Syndrome**

- Inspiration
- Expiration

**Adult: Swyer-James Syndrome**

- Unilateral, small lung
- Bronchiectasis

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**Congenital Pulmonary Airway Malformation**

- Formerly, Congenital Cystic Airway Malformation (CCAM)
- 25% of congenital lung lesions
- Result of abnormal airway proliferation
- Normal arterial supply & venous drainage
- Communicates with bronchial tree

**Histology: CPAM**

Cyst-like structures of varying sizes
CPAM: Clinical

- Most symptomatic as neonates
  - Cyanosis, grunting, tachypnea
- May present in older children & adults
  - as recurrent pneumonia
- May be antenatal diagnosis

CPAM: Expanded Classification
Based on origin and histology

- 0 = tracheobronchial (lethal)
- 1 = bronchial/bronchiolar
- 2 = bronchiolar
- 3 = bronchiolar/alveolar (lethal)
- 4 = distal acinar

Stocker JT, Histopathology 2002; 41:424

CPAM: Imaging

- Type 0 (1-3%) perinatal death (not imaged)
- Type I (60-70%) cysts, 3-10 cm
- Type II (15-20%) cysts, 1-3 cm
- Type III (5-10%) microcysts (< 5mm) on cut sections (not imaged, high mortality)
- Type IV (15%) cysts up to 10 cm

Bottom line: 2 basic appearances

Type I CPAM: Large Cysts (3-10 cm)
Type II CPAM: Small Cysts

Type IV Cyst

Type III CPAM / CCAM Antenatal US Diagnosis
- Polyhydramnios
- Fetal hydrops
  - Ascites
  - Anasarca
  - Placental edema
- Solid lung mass
- High perinatal mortality

Spectrum of CPAM
- 10% diagnosed in adolescents & adults
- Infection common
- Imaging studies
  - Thick walled cavities
  - Fluid-fluid levels
  - Pneumonia

VCU med center
Ioanthea E Eur Respir J 2005; 26:1181-1187 (BAC)
DDX: Pleuropulmonary Blastoma

- Children < 3 years
- Malignancy of lung/pleura
  - sarcomatous and embryonal cells
- 3 types-1 (cystic), 2 (cystic/solid), 3 (solid)
- Increasing malignancy 1 to 3
- Survival 80-85% type 1; 40-50% type 3
- Presentation: dyspnea, pneumothorax (clue)
Neonate with respiratory distress

15 year old girl recurrent PTX
Path: cystic PPB, type 1

PBB Associations
- Cystic renal tumor (cystic nephroma)
- Embryonal neoplasms (rhabdomyosarcoma, Wilms tumor, germ cell tumors)

Bronchogenic Cyst
- Failure of lung bud to incorporate into primitive lung
- Lung (30%), mediastinum (70%)
- Asymptomatic & incidental finding
- Symptomatic from mass effect
**Bronchogenic Cyst: Path**

- **Gross**
  - Separate from lung
  - Round or ovoid
  - Clear or turbid contents

- **Histo**
  - Lined by respiratory epithelium
  - Bronchial glands, cartilage, smooth muscle in wall

**Pulmonary Bronchogenic Cyst: Imaging**

- Smooth, rounded, unilocular mass
- Fluid-filled, usually serous fluid
  - Sometimes protein or mucin
- Air or air-fluid levels common
- No mediastinal shift
- Non-enhancing
Bronchogenic Cyst: Adult
- Unilocular mass
- Air-filled with or without fluid

DDX: Congenital Pulmonary Airway Malformation
- Septations and multiple cysts favor CPAM

Review Lung “Cysts”
- Lobar overinflation
- Bronchogenic cyst
- CPAM

Bronchial Atresia (Congenital Mucocele)
- Failure of bronchial bud to maintain communication with airway
  - atresia of central segmental bronchus
- Wheezing, cough or incidental detection
- Imaging:
  - nodular opacity
  - fluid-filled dilated bronchi
  - surrounding overinflation (air drift)
**Bronchial Atresia**

22 yo woman with acute left sided chest pain

Dilated mucus-filled bronchi & overinflated lung

**Acquired vs Congenital Mucocele**

Acquired

Congenital

Distal air drift

**Pulmonary Underdevelopment**

- **Agenesis**: Complete absence of lung tissue, artery, & small or absent bronchus
- **Hypoplasia**: Small lung & bronchus (artery may or may not develop)
Lung Agenesis

- Small bronchus
- No lung or PA

Companion Case Adult: Lung Agenesis

Arrested Pulmonary Development

- Pulmonary hypoplasia
  - Small lung (hypoplasia)
  - Small bronchus
  - Absent or small pulmonary artery
  - Mediastinal shift to side of hypoplasia

Adult: Lung Agenesis
Pulmonary Hypoplasia
2-month-old boy, mild dyspnea

Small lung & bronchus No PA
Bilateral Pulmonary Hypoplasia
Potter Syndrome

- Neonatal respiratory distress
- Flattened nose, small chin, low-set ears
- Small lungs due to compression of fetal thorax from oligohydramnios
- Pneumothorax & no other lung disease
- Cause: renal agenesis

Potters Syndrome

- Bell shaped chest
- Small chest, clear lungs & pneumothorax

Pulmonary Hypoplasia

- Newborn term female with respiratory distress and abdominal mass

Congenital Anomalies with Abnormal Vasculature

- Hypoplasia with anomalous venous return
  - Scimitar syndrome
- Pulmonary sequestration
Hypogenetic Lung Syndrome

- AKA scimitar syndrome
- Lung hypoplasia with PAPVR
  - RLL vein returns to IVC, portal or hepatic vein or RA
- May be symptomatic or incidental finding

Hypogenetic Lung Syndrome

- 10 year old girl
- Pneumonia suspected

Hypogenetic Lung Syndrome

10 year old girl

Scimitar Adult
**Pulmonary Sequestration**

- No normal connection with bronchial tree or pulmonary arteries
- **Systemic** blood supply
- 2 types
  - Intralobar
  - Extralobar

**Pathogenesis**

- Arises from supernumerary lung bud
- If the lung bud arises before the pleura develops, it shares pleura of adjacent lung and becomes ILS
- If the lung bud arises after pleura formation, it acquires its own pleural covering becomes ELS

**Intralobar Sequestration: Path**

- **Gross**
  - No pleural covering
  - Blends with normal lung
- **Microscopic**
  - Chronic inflammation
  - Bronchopneumonia
Extralobar Sequestration: Path

• Separate from normal lung
  - Pleural covering
• Microscopic
  - Dilated bronchioles, alveoli & subpleural lymphatics
  - Well-formed bronchus near edge of lesion (50%)

Sequestration: Clinical

• ELS
  - Asymptomatic 90%
  - Dyspnea, cyanosis (10%)
• ILS
  - Symptomatic
  - Cough, recurrent pneumonia

Sequestration: US Features

• Gray-scale
  - Echogenic mass
    » Above diaphragm-likely ILS
    » Below diaphragm (suprarenal)-ELS
• Doppler US
  - Feeding artery off aorta
  - Draining vein usually not identified

Intralobar Sequestration:
Extralobar Sequestration

Sequestration: CT Features

- Anomalous systemic feeding artery
- Drains to pulmonary or systemic veins
- Parenchymal findings
  - Intralobar:
    - Infiltrate &/or abscess
  - Extralobar
    - Solid mass

Intralobar Sequestration: Infiltrate

5-year-old boy, recurrent pneumonia

Intralobar sequestration: Abscess

6-year-old girl, fever and cough
Neonate Extralobar Sequestration: Solid Mass Systemic Drainage

Adult Extralobar Sequestration: Solid Lower Lobe Mass

Sequestration: MRI

Review: Which Sequestration?
**Arterial Sequestration**  
AKA Pseudosequestration

- Systemic arterial supply to normal lung
- May be dual supply from pulmonary artery
- Usually asymptomatic
- Hemoptysis most frequent clinical symptom, related to high pressure flow
- Surgery if aberrant vessel is only supply
- Embolization if there is dual supply


**Arterial Sequestration**  
Adult male, hemoptysis

- Systemic artery from descending thoracic aorta
- Normal lung parenchyma

**Arteriovenous Malformations**

- Artery joins vein without intervening capillary
- 80% Hereditary telangiectasia (OWR)
  - 15% sporadic
  - 5% cardiac surgeries (Glenn or Fontan)
- Cyanosis, brain/liver abscess
- 80-90% are simple AVMs
  - single feeding and draining vessel

**HTT: Multiple AVMs**

- 19 yo man
- Family history HTT
Arteriovenous Malformation
50 yo seizure

Pulmonary AVM
Simple architecture

AVM-Brain Abscess
Review

Congenital Lung Anomalies

ABNORMAL LUNG
NORMAL VASCULATURE

CLE     Cyst     CPAM     BA     Hypoplasia     Sequestration     Scimitar     AVM

NORMAL LUNG
ABNORMAL VASCULATURE