Objectives

• Review the etiology and anatomic basis of the major congenital lung diseases
• Review their radiographic and cross sectional imaging features
• Review their pertinent clinical features and complications

• No financial disclosures

Outline

• Anomalies associated with airway and lung development
• Resulting in cysts
• Associated with pulmonary vessels

Bronchial Atresia

• Focal atresia or stenosis of a bronchus
• Distal airway patent and accumulates a mucocele
• Location
  – Segmental
  – Lobar
  – Subsegmental
• Most common LUL > RUL > RML

CXR Findings

• Mucocele
• Hyperinflation distal to point of atresia
Congenital Lobar Overinflation

• Congenital Lobar "Emphysema"
• CXR Findings:
  – Severe overinflation
  – Atelectasis of adjacent lobe
• LUL > RML > RUL
• Respiratory distress in newborn
• Only 5% present after 6 months of age
• Surgery is the definitive treatment

Etiology
• Partial bronchial obstruction
  – Intrinsic
    • Deficient cartilage
    • Mucosal flaps
    • Smooth muscle
  – Extrinsic Mass
    • Bronchogenic cyst
    • Vascular e.g. PDA
• 50-55% Cause Not Known


Congenital Pulmonary Airway Malformation (CPAM)

• CCAM now referred to as CPAM
  – Some are not cystic
  – Some are not adenomatoid
  – The group of malformations follow the microscopic features of the pulmonary airways

**CPAM**

**Imaging**
- Multicystic lesion
- May be solid
- Usually normal arterial and venous connections.

**CPAM and CT**
- Cyst larger than 2.5 cm
  - Mostly Stocker Type 1 CPAM
- Cysts < 2.5 cm or solid lesion
  - Difficult to predict Stocker subtype


**CPAM**

**Clinical**
- Usually present early in life
- Reported up to the age of 60
- Most often lower lobes
- Dx in adult: careful
- Complications
  - Resp distress in newborn
  - Recurrent pneumonia*
  - PTX
  - BAC (type 1 CPAM)


**Bronchogenic Cyst**

**CT Findings**
- Bud of embryonic foregut and tracheobronchial tree becomes separated
- Separated tissue fails to develop further
- Location
  - Mediastinal 2/3
  - Parenchymal 1/3
- Connection to bronchus is unusual

**Bronchogenic Cyst**

**CT Findings**
- HU usually 0-20
- Occasionally have higher HU due to proteinaceous contents, hemorrhage or calcium

**HU = 8**

**T1 pre Gad**

**T1 post Gad**

**T2**
**Pulmonary Bronchogenic Cyst**
- Most common in lower lobes, usually in medial lung
- Eventually become infected in 20%
- When infected may contain air or air fluid level, resembling abscess

**Esophageal Duplication Cyst**
- Abnormal esophageal development
- Often contact the inferior esophagus on the right
- Similar imaging features to bronchogenic cyst

**Bronchogenic Cysts**

**Scimitar Syndrome**
- Group of disorders of lung and vascular development
- Anomalous pulmonary venous return
  - Complete
  - Partial
- Pulmonary hypoplasia
  - Varying degrees

**Additional Disorders:**

1. **Lung Development:**
   - Bilateral left sided bronchi
   - Horseshoe lung
   - Diverticula, bronchiectasis
   - Sequestration

2. **Arterial Supply:**
   - 50% PA is reduced in size
   - Systemic arterial supply

3. **Cardiac malformations 25%**
   - ASD
   - VSD, TOF, coarct, left SVC
Proximal Interruption of the PA

• A central pulmonary artery
  • Completely absent or
  • Terminates w/in 1cm of its origin
• Distal pulmonary arteries (in the lung) are intact
• Collaterals:
  – Bronchial artery
  – Transpleural intercostal
  – Internal mammary
  – Phrenic

CXR Findings

• Small hemithorax
• Decreased vascularity
• No identifiable pulmonary artery
• Rib notching
• Similar to Scimitar

“Interrupted” pulmonary artery
occurs:
  – Most common on the right
  – Most often opposite the aortic arch
• Left sided “PA interruption” has higher incidence of CHD, especially Tetrology of Fallot
**Proximal Interruption of the PA**

- Clinical findings
  - Asymptomatic
  - Dyspnea, exercise intolerance
  - Hemoptysis
  - Bronchiectasis (recurrent infection)
  - Pulmonary HTN (19-25%) – most important prognostic feature

**Pulmonary Sequestration**

Mass of pulmonary tissue:
1) "Sequestered" from the bronchial tree
2) Systemic arterial supply
- The diagnostic feature on imaging
- Intralobar vs Extralobar:
  - Pleural covering
  - Venous drainage

**Intralobar Sequestration**

- The "sequestered" lung tissue:
  - Within the visceral pleura of the affected lobe
  - Pulmonary venous drainage
- May be acquired*


**Intralobar Sequestration**

- Imaging May Vary:
  - Homogeneous solid mass
  - Cystic mass
  - Region of hyperlucent hypovascular parenchyma
- Due to collateral ventilation
  - Infection can occur

**Hx:** 39 yo with fever & pleuritic left sided chest pain.
Extralobar Sequestration

- The “sequestered” lung tissue is:
  - Within its own pleural envelope
  - Not subject to collateral ventilation
  - Does NOT contain air
  - Rarely infects because it is enclosed
  - Venous drainage to systemic veins

Extralobar Sequestration

- Imaging:
  - Solid vascular mass
  - Most often left base near diaphragm

Sequestration: Goals of Imaging

1) Delineate the extent of the lesion
2) Identify artery or arteries
3) Identify venous drainage
4) Evaluate for involvement below the diaphragm

CONCLUSION

- Congenital lung disease is RARE
- These disorders have characteristic imaging features