ADULT CONGENITAL LUNG DISEASE

Outline Congenital Lung Disease

- Anomalies associated with lung development and airways
- Anomalies resulting in cysts
- Anomalies associated with pulmonary vessels

Pulmonary Agenesis

- Complete absence of a lung
- Absent bronchi and vascular supply
- Can be incidental finding in adults
- Infant: associated anomalies
  - Congenital heart dx
  - Tracheo-esophageal atresia
  - Renal anomalies

Tracheal Bronchus

- Supernumerary
  - Normal branching of RUL
  - Accessory bronchus
- Displaced
  - One branch of RUL is missing
  - Normal branch from abnormal position
  - Segmental or subsegmental
  - Usually supplies apical segment
- "Pig bronchus"
  - Occasionally subject to disease
    - Infection, bronchiectasis

Accessory Cardiac Bronchus

- Ends blindly
- May be assoc with small amounts of pulm parenchyma
- Possible source of infection or hemoptysis

Bronchial Atresia

- Focal atresia or stenosis of a bronchus
  - Lobar
  - Segmental
  - Subsegmental
- Distal airway patent
- Distal obstructed lung is hyperinflated due to collateral air drift
- Most common LUL > RUL > RML
Bronchial Atresia

- Imaging
  - Mucous plug or mucocele
  - Hyperinflation in the segment distal to the mucocele

Congenital Lobar Overinflation

- Congenital Lobar "Emphysema"
- Findings:
  - Severe overinflation
  - Atelectasis of adjacent lobe
  - Respiratory distress in newborn
- Only 5% present after 6 months of age
- Most treated surgically

CXR and CT: CLO

- Few vessels
- Air trapping with lobar hyperinflation
- Atelectasis of adjacent lung

Congenital Lobar Overinflation

- Thought to be secondary to partial bronchial obstruction
  - Intrinsic
    - Deficient cartilage
    - Mucosal flaps
    - Stenosis/malacia
  - Extrinsic Mass
    - Bronchogenic cyst
    - Vessel eg PDA
- LUL > RML > RUL
### Congenital Pulmonary Airway Malformation (CPAM)

- **Spectrum of airway malformations (hamartomas)**
- **Involve various portions of the tracheobronchial tree**
- **CCAM referred to as CPAM**
  - Some are not cystic
  - Some are not adenomatoid
  - Spectrum follows the microscopic features of the pulmonary airways


### Congenital Pulmonary Airway Malformation (CPAM)

- **Type 0: Tracheobronchial Origin**
- **Type 1: Bronchial/Bronchiolar**
  - The large cyst lesion
- **Type 2: Bronchiolar Origin**
  - The small cyst lesion
- **Type 3: Bronchiolar/Alveolar Duct**
  - The adenomatoid lesion
- **Type 4: Distal Acinar Origin**
  - The “unlined cyst” lesion


### Congenital Pulmonary Airway Malformation (CPAM)

- **Multicystic mass**
- **Cysts may communicate with airways**
- **Usually normal arterial and venous connections.**
- **DDX**
  - Cystic bronchiectasis
  - Sequestration
  - Intrapulmonary bronchogenic cyst
  - Prior infection with pneumatocele

CPAM and CT

- **Cysts larger than 2.5 cm**
  - Mostly Stocker Type 1 CPAM
- **Cysts < 2.5 cm or solid lesion**
  - Difficult to predict Stocker subtype
- **Difficult to differentiate Type 2 CPAM from other types on CT**


### CPAM

- **Most often lower lobes**
- **Can involve entire lobe**
- **Usually present early in life but reported up to the age of 60**
- **Complications**
  - Resp distress in newborn
  - Recurrent pneumonia*
  - Spontaneous PTX
  - BAC (type 1)


**CPAM Type I**
Cysts
- Bronchogenic
- Esophageal duplication cysts

Bronchogenic Cysts
- Abnormal bud of the embryonic foregut and tracheobronchial tree
- Separated focus of tracheal-bronchial tissue does not develop further
- Connection to bronchus is unusual
- Location
  - Mediastinal 2/3
  - Parenchymal 1/3

Bronchogenic Cyst CT
- 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

**Bronchogenic Cysts**
- When infected can look like an abscess
- Rapid increase in size can occur from infection or hemorrhage
- When large can exert mass effect

**Esophageal Duplication Cyst**
- Abnormal esophageal development
- The muscularis propria of the esophagus is contiguous with the muscular layer of the cyst wall
- Often inferior and on the right
- Similar imaging features to bronchogenic cyst

**Anomalies associated with vessels**

**Pulmonary Arteriovenous Fistula**
- Direct communication between artery and vein without intervening capillary bed
- 67% are associated with OWR
- Multiple in 33%
- Most commonly congenital, can be acquired:
  - CHD (Glenn and Fontan)
  - Chronic liver dz
  - Infection (TB and actinomycosis)
- Lower lobe 50-70%

**Pulmonary arteriovenous fistula**
- Simple AVM: most frequent
  - Consists of dilated vascular sac connected to one feeding artery and one draining vein
  - Enlargement of the vascular sac is common, occasionally rapid enlargement
- Complex AVM (20%):
  - Lesions with one or more feeding arteries and/or draining veins
  - More difficult to treat
Pulmonary arteriovenous fistula

- Clinical manifestations
  - None when small
  - Hypoxemia
  - R to L shunt
  - Embolic complications: stroke and cerebral abscess
  - Polycythemia
  - Pulmonary hemorrhage

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:
   - Hypoplasia
   - 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

- Central pulmonary artery completely absent (proximally interrupted)
- Distal pulmonary arteries (in the lung) are intact and are supplied by collaterals:
  - Bronchial artery
  - Transpleural intercostal
  - Internal mammary
  - Phrenic
- Affected lung is hypoplastic

Proximal Interruption of the PA

- “Interrupted” pulmonary artery occurs:
  - Most common on the right
  - Most often opposite the aortic arch
- Left sided “PA interruption” has high incidence of CHD, especially Tetrology of Fallot

Proximal Interruption of the PA

- Clinical findings
  - Dyspnea, exercise intolerance
  - Hemoptysis, bronchiectasis
  - Pulmonary HTN (19-25%)- exacerbated by high altitude or pregnancy

Pulmonary Sling

- Anomalous origin of left pulmonary artery from the right PA
- Left pulmonary crosses between the esophagus and trachea
- Can cause airway obstruction/malacia

Pulmonary Sequestration

- Mass of disorganized pulmonary tissue:
  1. Lacks normal bronchial communication (i.e. "sequestered")
  2. Systemic arterial supply
     - Characteristic feature on imaging
     - One or more arteries

- Intralobar vs Extralobar:
  - Pleural covering
  - Venous drainage

Intralobar Sequestration

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

- More common
- Most often left sided, 2/3 near diaphragm
- Arterial supply thoracic aorta
- May be acquired


Intralobar Sequestration

- Imaging:
  - Homogeneous solid mass
  - Cystic mass
  - Region of hyperlucent hypovascular parenchyma (collateral ventilation and air trapping)
  - Due to collateral ventilation infection can occur

Hx: 39 yo with pleuritic chest pain. Treated for pneumonia

6 weeks later.
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

- Arterial supply:
  - Low thoracic
  - Abdominal aorta
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 in the adult patient is RARE
- These disorders have characteristic imaging features which assist their diagnosis