Case Conference:
Pediatric Lung Masses

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Congenital cystic lung masses

Congenital cystic adenomatoid malformation
Pulmonary sequestration
Congenital lobar emphysema
Bronchogenic cysts

-all present on imaging studies as abnormal air, air-fluid, or fluid filled cysts
-each have different histology, presentation, and treatment
Congenital Cystic Adenomatiod Malformation (CCAM)

- intercommunicating cysts lined by respiratory epithelium
- typically restricted to part of one lung
- communicates with tracheobronchial tree

Classification scheme:
Type 1 - single/multiple cysts, some > 2 cm diameter
Type 2 - multiple macroscopic cysts < 2 cm
Type 3 - solid variant; (cysts so small that gives solid appearance)
CCAM

**Prenatal Presentation**
- may be incidental finding on routine prenatal US
- see cystic or solid lung mass in part of one lung
- polyhydramnios present in ½ cases Dx’d prenatally
  (lung compresses esophagus with decreased fetal swallowing of amniotic fluid)
- fetal hydrops (generalized edema and heart failure) occurs with large masses due to mediastinal shift, vessel obstruction, and impaired cardiac filling
- pulmonary hypoplasia can result from large lung mass impairing normal lung development
CCAM

Postnatal Presentation
- 60% present with neonatal respiratory distress
- 20% present with infectious complications, usually before age 6 (eg. recurrent pneumonia)
- 20% Dx’d as incidental finding on CXR
- CXR shows air-filled cysts of varying size
- CT scan differentiates from other cystic lung masses
CCAM

Prenatal Treatment
-intervention in utero depends on the presence of fetal hydrops (almost 100% mortality if mass left untreated);
Cyst aspiration: for acute decompression (but cyst may reaccumulate)
Percutaneous pulmonary-amniotic shunt placement
In utero resection
-if no hydrops, follow with sequential US, as many CCAM remain stable or regress
CCAM

Postnatal Treatment

- Resection routinely done at time of Dx
  
  1) to ameliorate symptoms - respiratory compromise or recurrent infections
  
  2) malignant association between CCAM and rhabdomyosarcoma and other malignant tumors

- Tx: lobectomy or partial lobectomy when anatomic resection would sacrifice normal lung tissue

- Px: good outcomes unless pulmonary hypoplasia present (extracorporeal membrane oxygenation may be helpful postoperatively)
Pulmonary sequestration

-nonfunctional lung tissue mass that receives blood supply from an anomalous systemic artery and does not communicate with the normal tracheobronchial tree

Intralobar sequestration

Extralobar sequestration
Intralobar pulmonary sequestration

-sequestration contained within visceral pleura of another lobe, usually posterolateral segment of lower lobe

-normally contain air within cystic lung tissue

-frequently become infected (may see air-fluid level)

-arterial supply: 75% by descending/abdominal aorta 
  25% from another thoracic/abdominal vessel

-venous drainage: usually pulmonary veins
Extralobar pulmonary sequestration

- Abnormal lung tissue contained in pleural envelope separate from the normal lung
- Commonly located in posterior lower chest; 5% are located below the diaphragm
- Arterial supply: systemic artery, mostly aorta
  15% blood supply from below diaphragm
- Venous drainage: usually azygos venous system;
  some drain into portal/pulmonary venous system
- Approx 40% incidence of associated congenital anomalies, most commonly diaphragmatic hernia
Pulmonary sequestration

**Diagnosis**

Prenatal US- see hyperechoic mass
   - Doppler can trace anomalous artery

Neonates/Infants
- extralobar Dx’d earlier age during workup for associated abnormalities, or during evaluation of respiratory distress
- intralobar often diagnosed during workup of frequent infections during first few decades of life
Pulmonary sequestration

**Diagnosis**
- CXR, chest CT, US, V-Q scan all helpful
- Angiography is gold standard for Dx, however risks don’t justify additional information obtained; anomalous blood supply is identified at surgery
Pulmonary sequestration

Treatment
- resection done at time of diagnosis to alleviate symptoms, prevent future infections
- arterial supply is first identified and controlled
- extralobar: resection of mass
- intralobar: usually require formal lobectomy
Congenital Lobar Emphysema (CLE)

-massive distention of a pulmonary lobe with varying degree of respiratory distress

-main cause: ball-valve bronchial obstruction that allows inflation, but collapses and prevents exhalation

-identifiable cause of bronchial obstruction in 50%; approx 50% of these due to congenital deficiency of bronchial cartilage; other causes: enlarged blood vessel in congenital heart disease (eg. L to R shunt with enlarged pulmonary artery); bronchial stenosis; mucosal flaps
Congenital Lobar Emphysema (CLE)

Presentation
-infrequently found on prenatal US, likely because no overdistention while liquid breathing in utero
-typically affects infants, rarely presents > age 6 mo.
-symptoms of respiratory distress usually develop in first few days/wks of life
-upper or middle lobe almost always involved; overdistention causes atelectasis of adjacent lobes;
-with further distention, the mediastinum shifts, causing compression of opposite lung, decreased venous return and shock
Congenital Lobar Emphysema (CLE)

**Diagnosis**
CXR: DD PTX, CCAM
CT and V/Q scans confirm Dx
-No bronchoscopy with CLE, as air trapping can acutely worsen respiratory status
-foreign body aspiration causes lobar distention, usually affecting the lower lobes (CLE affects upper lobes); presents after age 6 mos when infants begin to crawl; Tx is bronchoscopy
Congenital Lobar Emphysema (CLE)

**Treatment**

All symptomatic pts should undergo lobectomy at time of Dx

-surgical team should be prepared for emergent thoracotomy upon induction of anesthesia and positive pressure ventilation, as emphysematous lobe may acutely enlarge, compressing normal lung and heart

Asymptomatic pts unlikely to benefit from resection (no difference in PFTs of asymptomatic CLE and pts tested post resection)
Bronchogenic cyst

-arise from abnormal budding of the tracheobronchial tree during airway development

-depending on timing and orientation of budding, they can be located in mediastinum (2/3), pulmonary parenchyma (1/3), or lower neck (rare)

-cysts lined by ciliated columnar epithelium and surrounded by cartilage, smooth muscle, and mucous glands

-often have connection to the tracheobronchial tree
Bronchogenic cyst

Presentation
Symptoms due to mass effects- compression of the trachea or bronchi impairs ventilation and lung drainage, causing cough, wheezing, stridor, dyspnea, and pneumonia
Bronchogenic cyst

Diagnosis
Prenatal US- see simple cyst, usually in lower lobes
CXR during exacerbation may not show cyst clearly due to inflammation, causing delayed Dx
Routine CT scan decreases diagnostic interval
Bronchogenic cyst

**Treatment**

Resection by partial or total lobectomy is performed on symptomatic and asymptomatic lesions.
- There are reports of malignant degeneration of bronchogenic cysts.
- Asymptomatic cysts that have been followed often become symptomatic later in childhood.

- Retrospective review of 22 congenital lung cyst resections over 10 yrs at Texas Children’s Hosp
- 1051 thoracic operations, 115 pulmonary resections
- 7 CCAM: Dx b/w 27 wks gestation to 6 yrs
  4 symptomatic; 5 operated on age < 60 days;
  6 lobectomy, 1 partial
- 7 pulmonary sequestration: 1 extra-, 6 intralobar;
  3 Dx’d age < 3 mos with multiple anomalies;
  4 age 2 – 11 yrs, presenting with recurrent infections; all had lobectomy

-6 CLE: average age Dx 29 days; 5 pts operated on for respiratory distress

-2 intrapulmonary bronchogenic cysts, both Dx’d on prenatal US; one pt had intrauterine cyst aspiration twice to relieve mediastinal shift, then born via NSVD, had respiratory distress 1st day of life and had Left Upper Lobectomy

- retrospectively review of 175 fetal lung lesions diagnosed by prenatal US at 2 institutions
- 134 CCAM: 14 women had elective abortions
  101 managed expectantly: 25 with fetal hydrops all died, all without hydrops survived
  13 had fetal surgical lobectomy for hydrops at 21 to 29 wks gestation: 5 died intraop/1st day of life;
  8 had hydrops resolution and neonatal survival
  6 had placement of thoracoamniotic shunt, 5 survived

- 41 extralobar pulmonary sequestration cases:
  28 regressed on prenatal US, were asymptomatic after birth, and did not require resection
  3 had tension PTX with secondary hydrops, successfully treated by fetal thoracenteses or shunt followed by postnatal resection
  1 hydropic fetus died, 7 symptomatic lesions resected after birth
Summary

-Congenital lung cysts are a rare, but clinically significant, group of anomalies
-occasionally prenatal intervention is required because of detrimental effects on heart and lungs
-delay in diagnosis avoided by high index of suspicion and low threshold for CT scan
-resection by segmentectomy or lobectomy done for intrapulmonary lesions and simple excision for extralobar sequestrations and bronchogenic cysts
Questions

1) Which is true about congenital lobar emphysema?

a) Most common cause is maternal smoking
b) Diagnosis should be confirmed with bronchoscopy
c) Rarely diagnosed before age 6 months
d) Rarely causes fetal distress
Questions

2) Which most commonly requires fetal intervention?

a) congenital cystic adenomatoid malformation
b) congenital lobar emphysema
c) bronchogenic cyst
d) pulmonary sequestration
Questions

3) Which is not resected when asymptomatic?

a) congenital cystic adenomatoid malformation  
b) congenital lobar emphysema  
c) bronchogenic cyst  
d) pulmonary sequestration