Benign Pediatric Lung Masses

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Objectives

- Provide a differential diagnosis for pulmonary masses in children
- Briefly review etiology and diagnostic workup
- Discuss management and treatment options of each
Pulmonary Masses

Predominantly cystic
- Bronchogenic cyst
- CPAM
- Lung abscesses
- Hydatid disease
- Post infectious pneumatocele
- Fungal (aspergillosis)
- Congenital lobar emphysema

Predominantly solid
- Pulmonary sequestration
- CPAM
- Pulmonary hamartoma
- Various neoplasms (primary and metastatic)
Variant of bronchomalacia with focal cartilagenous deficiency of tracheobronchial tree
Regional airway collapse with expiration
Rare causes: extrinsic compression from anomalous pulmonary vessel or large PDA
Caucasian males
Clinical presentation:
- Respiratory distress
- Tachypnea
- Cyanosis
- Wheezing
- Retractions
- Cough
- Usually in the first year of life
CLE

Physical findings:
- Decreased breath sounds on the affected side
- Hyperresonance to percussion
- Mediastinal displacement
- Bulging of the chest wall on the affected side
Radiographic findings:
- Overdistention of affected lobe (LUL 42%, RML 35%)
- Wide separation of bronchovascular markings
- Collapse of adjacent lung
- Mediastinal shift
- Depressed diaphragm
Differential diagnosis:

- Pneumothorax
- Pneumatocele
- Must differentiate from regional obstructive emphysema from lobar hyperinflation secondary to other causes
  - Extrinsic compression (bronchogenic cyst, tumor, lymphadenopathy, foreign body, etc)
  - Intrinsic obstruction (infection, inflammation)
In extreme cases mediastinal shift can cause resp and CV collapse requiring emergent thoracotomy
More commonly less dramatic
Expectant management acceptable for completely asymptomatic lesions
Rare to have complete resolution of radiographic findings, concern of retained secretions as nidus for infection
In most cases surgical resection guided by radiographic studies for boundaries of normal lung
Bronchogenic cysts

- Type of foregut duplication cyst
- Lined with bronchial epithelium
- Mucus filled
- Cartilaginous walls in some
- Can be diagnosed prenatally, perinatally or in older kids
- Presentation varies from resp distress from compression, to recurrent infections, to incidental radiographic findings
Bronchogenic cysts

- All should be excised
  - Risk of expansion, infection, hemorrhage
- May be in the mediastinum near hilum or in parenchyma
- Resection can usually be accomplished thoracoscopically
Pulmonary hamartomas

- Benign neoplasm, no malignant potential
- Grow slowly over time
- Disorganized native lung tissue
- Often contain fat, epithelial tissue, connective tissue and fibrous tissue
- 90% are peripheral lesions
- Popcorn-like appearance on CXR
- CT can be diagnostic if see fat and calcifications in well marginated mass (70%)
- Occasionally need biopsy to prove not malignancy
Bronchopulmonary Sequestration

- Cystic mass of nonfunctioning lung parenchyma with no connection to tracheobronchial tree
- Rare, 0.15-6.4% of all congenital pulmonary malformations
- Receives its blood supply from anomalous origin
- Largely solid lesions
- Subdivided into
  - Intralobar
  - Extralobar
  - Bronchopulmonary-Foregut Malformation
Poorly understood pathophysiology

Proposed mechanisms include:
- Portion of developing lung separated by rest of organ
- Compression from cardiovascular structures
- Traction by aberrant systemic vessels
- Inadequate pulmonary flow
BPS

- May have anomalous connections to other bronchi, lung parenchyma or GI tract
- Recurrent infections due to hematogenous seeding or anomalous connections
- Most often occurs in the lower lobes regardless of subtype
Intralobar

- Contained within the same investing pleura as normal lung
- Most common form (75-90%)
- Equal distribution between males and females
- Found on the left in 60% of cases
- Arterial supply usually via lower thoracic or upper abdominal aorta
- Venous drainage often still via pulmonary venous drainage
Extralobar

- Found outside the normal lung and covered with separate pleura
- Almost exclusively found on the left
- Most often identified in the fetal/neonatal period
- More common in males
- Rarely may present as subdiaphragmatic or retroperitoneal mass
BPS

- Extralobar
  - Blood supply usually comes from thoracic aorta
  - Venous drainage usually via systemic or portal venous systems
  - High incidence (>50%) of other associated anomalies
    - CDH
    - Vertebral anomalies
    - Pulmonary hypoplasia
    - Colonic duplication
BPS

Presentation

Prenatal

- Seen on routine ultrasound
- May regress as gestation progresses
- Rarely can cause hydrops and fetal demise from vascular compression
- Appearance of echogenic mass in fetal thorax, with or without mediastinal shift
- Pathognomonic if see systemic artery from aorta to lesion on Doppler
BPS

Presentation
  Postnatal
    Variable age
    ELS usually presents in neonatal period, sometimes with neonatal respiratory distress
    ILS may present late in childhood with recurrent pulmonary infections
    Both can be incidentally identified as part of evaluation for another problem
Radiographic workup:

- Ultrasonography - esp in prenatal identification
- CXR - uniformly dense mass, may see cystic areas, air fluid levels
- CT scan - characteristic emphysematous change at margin, can use contrast to identify aberrant blood supply
Treatment

- Surgical excision curative
- Performed urgently in the face of resp distress
- May be done electively in older kids with recurrent infections
- Asymptomatic pts with ILS still recommend resection to prevent recurrent infections
- Asymptomatic ELS- serial monitoring with CT scanning
- Careful preoperative planning to identify and control blood supply…may come from abdomen!!
- Rarely, fetal intervention for hydrops
Congenital Pulmonary Airway Malformation

Also commonly referred to as congenital cystic adenomatoid malformation (CCAM)

- Multicystic lung mass resulting from a proliferation of terminal bronchiolar structures with associated suppression of alveolar growth
- Histologically- cysts lined by cuboidal or columnar epithelium
- Most common congenital lung lesion
- 1/8300-35000 live births
Type 0
- Rarest form
- Originates from tracheal or bronchial tissue
- Microcystic (cysts <0.5cm)
- Lined with pseudostratified epithelium
- Mucus cells and cartilage present, skeletal muscle absent
- Diffuse malformation involving entire lung
- Severely impaired gas exchange
- Infants typically die at birth
**CPAM**

- **Type 1**
  - Most common
  - Originates from distal bronchi or proximal bronchioles
  - Well differentiated
  - Macrocystic (2-10 cm in diameter)
  - Usually single cysts, but may be multiloculated
  - Lined with pseudostratified columnar epithelium
  - Walls contain smooth muscle and elastin
  - 95% of cases involve only one lobe
  - Risk of malignant transformation
CPAM

- Type 2
  - 15-20% of CPAMs
  - Multiple cysts 0.5-2cm in diameter
  - Cysts resemble dilated terminal bronchioles
  - Lined with cuboidal or columnar epithelium
  - Mucus secreting cells and cartilage NOT PRESENT
  - Little to no mass effect on adjacent lung
  - Up to 60% have other congenital anomalies
    - TEF, renal agenesis, intestinal atresias, CDH, cardiac anomalies
CPAM

- Type 3
  - 5-10%
  - Very large
  - Can involve multiple lobes
  - Acinar in origin, not well differentiated cells
  - Microcystic, lined with nonciliated cuboidal epithelium
  - Thick fibromuscular wall with excess elastin
  - Mucus secreting cells
  - No cartilage present
CPAM

- Type 4
  - 10-15%
  - Cysts with maximum diameter of 7cm
  - Nonciliated, flattened, alveolar lining cells
  - No mucus or skeletal muscle
  - May present at birth or childhood as tension PTX
  - Frequently presents with infection
  - Strong malignant potential (pleuropulmonary blastoma)
Multiple other classification systems
Most clinically relevant system simply divides them into microcystic and macrocystic subtypes
- Macrocytic: generally favorable prognosis, often asymptomatic at birth
- Microcystic: generally unfavorable, more likely to be solid appearing on imaging, more frequently associated with other anomalies
Clinical features

Prenatal presentation
- Ranges from incidental finding on u/s to fetal hydrops
- Peak size occurs at 25 weeks gestation. Many regress.
- If large enough, can compress IVC and heart
- Resultant increased CVP leads to heart failure
- Fetal hydrops and demise in severe cases
Neonatal presentation

- Majority of CPAMs presenting at birth are type 1
- Tachypnea, retractions, grunting, cyanosis
- Type 4 may present as PTX. Strong suspicion for malignancy
- Type 3 can also present with severe sx at birth
  - Often with associated pulmonary hypoplasia
  - Can involve entire lung
  - Severe progressive respiratory distress/failure
  - More common in males
Treatment
- Surgical resection for all symptomatic patients with respiratory distress
- Surgical resection is also offered and recommended to older children with less acute symptoms
- Prevention of recurrent infection
- Eliminate risk of degeneration into malignancy
- Thoracoscopic lobectomy most often treatment of choice
Prenatal Diagnosis and Therapy

- Widespread use of prenatal u/s has increased identification of the most common bronchopulmonary malformations
- Sometimes confused with CDH based on u/s
- MRI occasionally used in fetal diagnosis
- Cystic lung lesion remains the most commonly identified pulmonary anomaly
Ultrasound may have difficulty differentiating between CPAM and BPS

If aberrant vascular supply is present may help

MRI can be used if confusion

If no in-utero complications, distinction may be irrelevant
Prenatal diagnosis

- Overall prognosis relates mostly to size
- Small cystic masses often incidental findings, asymptomatic in utero and postnatally
- Very large masses important physiologic sequelae
  - Esophgeal compression can cause polyhydramnios
  - Mass effect on unaffected lung causes pulm hypoplasia
  - IVC/cardiac compression causes low output CHF and fetal hydrops
Prenatal diagnosis

- Fetal hydrops
  - May exhibit ascites, pericardial and pleural effusions
  - Edema of the skin/scalp
  - Heart failure
  - Historically viewed as a harbinger of fetal or neonatal death
  - Led to the development of fetal intervention for such lesions
Prenatal Treatment

- Various methods of fetal intervention
  - Corticosteroid treatment
  - Fetal endoscopic intervention
    - Thoracoamniotic shunt
  - Open fetal surgery
    - Open resection of cystic masses
  - EXIT procedures
Prenatal treatment

Corticosteroids
- Case reports of hydrops resolving and fetal survival after administration of steroids
- Not clear why
- Other cases of some shrinkage of cystic lesions with steroid administration
- Still highly controversial and experimental
Prenatal therapy

- Fetal thoracentesis/thoracoamniotic shunting
- Minimally invasive
- Effective in large, space occupying effusions in BPS or dominant macrocytic cysts
- In one study 9 fetuses with hydrops were successfully treated with shunting and survived
- All require definitive resection after delivery
- Tolerated well by gravid uterus but still requires tocolytics
The fetoscope

The balloon used in fetoscopic procedure
Prenatal therapy

- Open fetal surgery
  - Extensively studied in non human primates
    - Surgical technique
    - Anesthetics
    - Tocolytic regimen
  - Offered to select fetuses with life threatening lesions
  - Only performed at few select centers
  - Extensive prenatal evaluation to exclude other associated anomalies
Prenatal therapy

- Open fetal surgery
  - If meets all other criteria, massive multicystic or solid CPAM with hydrops can be resected prior to 32 weeks gestation
  - Largest reported series
    - 22 cases
    - 11 healthy survivors
    - All resected from 21-31 weeks gestation
    - 16 with single lobectomy, 4 double, 2 pneumonectomies
    - All survivors had resolution of hydrops within 1-2 wks
Open fetal surgery

- Deaths often due to preterm labor and resultant demise
- For fetuses reaching 32 weeks, prefer early elective delivery with ex-uterro resection
  - EXIT procedure with resection prior to division of umbilical cord or placement onto ECMO at the time of delivery
- Prognosis still poor even with ECMO support
Predicting who will progress to hydrops

- CVR (CPAM volume ratio) = CPAM volume/head circumference
- CVR >1.6 is suggestive of progression to hydrops
- Most CPAMs don’t increase in size relative to fetal size after 28 weeks
- 2x/week u/s for fetuses with ration >1.6, weekly for those <1.6
- Once show signs of developing hydrops, consideration of fetal interventions
Typically presence of CPAM or BPS is an indication for resection regardless of symptoms

In asymptomatic newborns often wait until 3-6 mo

Pleuropulmonary blastoma and brochoalveolar carcinoma both reported in pts with known CPAM

May need preoperative imaging for sequestration

Depending on location or size, most commonly requires lobectomy
Postnatal resection

- Increasing use of thoracoscopic resection
  - Shorter hospitalization
  - Reduced pain
  - Improved cosmesis
  - Less overall morbidity than thoracotomy

- Limiting factors:
  - Instability
  - Lesions so large they limit working space
Postnatal resection

Special considerations
- Lesions so large requiring pneumonectomy may result in temporary HTN or resp insufficiency from hypoplasia
- May need to make plans for ECMO support temporarily
Questions?