Pulmonary Sequestration

Jonathan Shaw, Harvard Medical School Year IV
Gillian Lieberman, MD
What do these two patients have in common?

Patient 1: 50 y.o. non-smoking female with several months cough and hemoptysis;
CXR: posterior left lower lung consolidation

Patient 2: Asymptomatic neonate with incidental chest mass found on prenatal U/S;
CT: posterior left lower lung mass
Patient 1
50 y.o. female with several months of cough and hemoptysis
Patient 1
50 y.o. female with several months of cough and hemoptysis

Findings:

- Ill-defined density in posterior LLL
- several rounded densities, some with air-fluid level
Patient 1
50 y.o. female with several months of cough and hemoptysis

Wide differential for this infiltrate with apparent cysts/cavitations includes:

- Bronchogenic carcinoma
- Metastatic Disease
- Abscess
- Infarction
- TB
- PCP
- Fungal infection
- Wegners
- Pulmonary sequestration

and more…
Patient 2

CT of Neonate with L chest mass on prenatal U/S

 Courtesy of Dr. Fabio Komlos, Children’s Hospital
Patient 2

CT of Neonate with L chest mass on prenatal U/S

Findings:

- Well-circumscribed heterogenous mass
- Posterobasal, above left hemi-diaphragm
- Arterial supply to mass from the aorta (not visible on this slice)

Courtesy of Dr. Fabio Komlos, Children’s Hospital
Patient 2
CT of Neonate with L chest mass on prenatal U/S

Differential of this congenital pulmonary mass includes:

- Dipahragmatic hernia
- Pulmonary sequestration
- CCAM (Congenital cystic adenomatoid malformation)
- Bronchogenic cyst
- Paraspinal lesion

Courtesy of Dr. Fabio Komlos, Children’s Hospital
What do the two patients have in common?

Patient 1 and 2 share a similar diagnosis--admittedly uncommon, but not rare…

The clue: both patient’s lesions are in the posterior left lower lung
What do the two patients have in common?

One of these diagnosis is characteristically found in left lower lung…

Patient 1 DDx:
- Abscess
- Bronchogenic carcinoma
- Fungal infection
- PCP
- TB
- Metastatic
- Infarction
- Wegners
- Pulmonary sequestration

Patient 2 DDx:
- Dipahragmatic hernia
- Pulmonary Sequestration
- CCAM (Congenital cystic adenomatoid malformation)
- Bronchogenic cyst
- Paraspinal lesion
What do the two have in common?

When you encounter a persistent lesion in the left lower lung, add this to your differential: **Pulmonary Sequestration**

Patient 1 DDx:
- Abscess
- Bronchogenic carcinoma
- Fungal infection
- PCP
- TB
- Metastatic
- Infarction
- Wegners
- **Pulmonary sequestration**

Patient 2 DDx:
- Dipahragmatic hernia
- **Pulmonary Sequestration**
- CCAM (Congenital cystic adenomatoid malformation)
- Bronchogenic cyst
- Paraspinal lesion
Pulmonary Sequestration

- A Congenital lung malformation:
  - A mass of abnormal, nonfunctioning, Pulmonary tissue
  - No communication with the tracheobronchial tree
  - Receives blood from an anomalous systemic artery
    (instead of pulmonary arterial system)
  - Usually occurs in left lower lung
Pulmonary Sequestration

• Pathophysiology:
  1. Primitive foregut gives rise to accessory lung bud
  2. Pluripotent tissue migrates caudally with the developing normal lung
  3. Remains connected to aortic blood supply for the primitive foregut
Two Types of Sequestration

• **Intralobar (75%)** = WITHIN visceral pleura of a pulmonary lobe

• **Extralobar (25%)** = “Accessory lung”—tissue in its own pleura
Intralobar vs. Extralobar

WITHIN visceral pleura of a pulmonary lobe

• "Accessory lung": lung tissue in its own pleura

Drawings from CIBA Netter Collection
Intralobar  vs.  Extralobar

Venous Drainage into pulmonary veins (L→L recirculation)

Venous Drainage into systemic (hemiazygous) (L→R shunt)

Drawings from CIBA Netter Collection
Intralobar vs. Extralobar

Cystic, frequently infected—
air gets in thru pores of Kohn between adjacent alveoli

Less likely to get infected—
isolated from lung

Drawings from CIBA Netter Collection
Intralobar vs. Extralobar

Diagnosis usually made in adolescence or adulthood…

Presenting with recurrent pneumonia

Diagnosis usually made in neonates or infants…

Often asymptomatic, but discovered during evaluation of other anomalies…

If not diagnosed prenatally/neonatally, usual presents by 6 months with cyanosis or difficulty feeding
Intralobar vs. Extralobar

Not associated with other anomalies

Often associated with:

• Diaphragmatic hernias
• Cardiac malformations
• Foregut anomalies
e.g. Neonate with Dextrocardia and Extralobar Sequestration

Courtesy of Dr. Fabio Komlos, Children’s Hospital
Which type of Sequestration?  

**Intralobar** or **Extralobar**

**Patient 1:**
50 y.o. female with several months cough and hemoptysis

**Patient 2:**
Asymptomatic neonate with mass found on prenatal U/S

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PACS, BIDMC  

Courtesy of Dr. Fabio Komlos, Children’s Hospital
Patient 1 – Intralobar Sequestration
Patient 1 – Intralobar Sequestration
Patient 1 – Intralobar Sequestration
Patient 1 – Intraloblar Sequestration
Patient 1 – Intralobar Sequestration
Patient 1 – Intralobar Sequestration
Patient 1 – Intralobular Sequestration
Patient 1 – Intralobar Sequestration

CT demonstrates typical findings of infected interlobar sequestration

- Dense, heterogeneous opacity within the posterobasal segment of the LLL
- Cystic appearance with multiple fluid-air levels
Patient 1 – Intralobar Sequestration

5 months later, after extended fluoroquinolone therapy

-Resolution of fluid

-persistant cystic spaces and consolidation
Treatment

- Surgical resection for both intralobar and extralobar sequestrations, to avoid/treat
  - Chronic infection
  - Symptoms from compression of normal lung
## Intralobar vs. Extrapulmonary Anatomy

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PACS, BIDMC

Courtesy of Dr. Fabio Komlos, Children’s Hospital
# Intralobar vs. Extralobar Anatomy

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Patient 1 – CT with contrast (vessels)

Twin anomalous arteries
aorta → sequestration
Patient 1 – CT with contrast (vessels)

Twin anomalous arteries
aorta  sequestration
Patient 1 – CT with contrast (vessels)

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Patient 1 – CT with contrast (vessels)

Twin anomalous arteries
aorta $\rightarrow$ sequestration
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<td>Venous Return</td>
<td><strong>Pulmonary Veins</strong></td>
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- Intralobar anatomy is within the visceral pleura and is considered an "accessory lobe".
- Extralobar anatomy has its own pleura and is located on the left hemidiaphragm.
- Intralobar arteries are supplied by the aorta (abdominal/thoracic), while extralobar arteries are usually abdominal.
- Intralobar venous return is through pulmonary veins, while extralobar venous return is through systemic veins.
Patient 1 – CT with contrast (vessels)

- Venous drainage into pulmonary veins
- Characteristic of intralobar sequestration
Imaging Pulmonary Sequestrations

• Identifying anomalous vessels = key to definitive diagnosis

• Useful for planning surgical resection

Variety of arterial patterns:
  » Descending Thoracic aorta (usually)
  » Infradiaphragmatic aorta (20%)
  » Dual arteries (10%)
  » Coronary arteries (rare reports)
Imaging Pulmonary Sequestrations

Various techniques have been used…

Definitive Diagnosis traditionally by

• Invasive Arteriography

Image from: http://www2.hawaii.edu/medicine/pediatrics/pemxray/v5e14.html, Craig T. Nakamura, MD
Imaging Pulmonary Sequestrations

Aortagram:

• Anomalous vessel from infra-diaphragmatic aorta to density at left lung base

• Venous phase (not shown): drainage to the hemiazygous.

• Typical for an extralobar sequestration

Image from: http://www2.hawaii.edu/medicine/pediatrics/pemxray/v5c14.html, Craig T. Nakamura, MD
Imaging Pulmonary Sequestrations

Conventional angiography is largely being replaced by non-invasive techniques: **CT, U/S, and MRI**

*Image from: [http://www2.hawaii.edu/medicine/pediatrics/pemxray/v5e14.html](http://www2.hawaii.edu/medicine/pediatrics/pemxray/v5e14.html), Craig T. Nakamura, MD*
CXR for Sequestrations

- Remains an important screening tool
- But doesn’t show the vessels
Radiographic Appearance of Intralobar Sequestrations

- **CXR** typically shows:
  - Lower lobe paraspinal opacity
  - Often cavitary or cystic

**DDx**
- Pneumonia
- Lung abscess
- CCAM

Intrapulmonary sequestration with large air-fluid level

*Courtesy of Dr. Fabio Komlos, Children’s Hospital*
Radiographic Appearance of Extralobar Sequestrations

- **CXR** typically shows:
  - Solid retrocardiac opacity
  - rounded or triangular
  - supra or sub-diaphragmatic

**DDx (in neonate)**
- Diaphragmatic hernia
- Loculated pleural effusion
- Esophageal duplication cyst
- Neuroblastoma
- Adrenal hemorrhage

Triangular extralobar sequestration in a 13 month-old p/w 1 month of wheezing, coughing and rhinorrhea.

Image from: [http://www2.hawaii.edu/medicine/pediatrics/pemxray/v5c14.html](http://www2.hawaii.edu/medicine/pediatrics/pemxray/v5c14.html), Craig T. Nakamura, MD
CT Appearance of Sequestrations

- **CT/CTA**
  - provides the best display of parenchyma

  - Intralobar: mixed cystic/solid lesion or homogenous soft tissue.
  - Extralobar: well circumscribed homogenous lesion.

Patient 1 - Intralobar Sequestration
CT Appearance of Sequestrations

- **CT/CTA**
  - typically can show the anomalous systemic arterial supply

*PACS, BIDMC*

Patient 1 – Arterial supply
CT Appearance of Sequestrations

- Newer MultiDetector CT combined with 3D reconstruction can consistently identify both the arterial and venous anatomy.
Patient 2

Neonate with extralobar sequestration

(left antero-lateral perspective)

Courtesy of Dr. Fabio Komlos, Children’s Hospital
3D CT Reconstruction
Patient 2 – Neonate with extralobar sequestration

Not easily seen on axial scan
a 3-D reconstruction reveals:

- Typical arterial supply from the aorta

- Unusual systemic venous drainage via the internal mammary vein

Courtesy of Dr. Fabio Komlos, Children’s Hospital
3D CT Reconstruction
A 6 month old girl with recurrent LLL PNA – Intralobar Sequestration

(not recognized on axial CT)
Anomalous venous drainage to LA

Anomalous arterial supply from descending aorta

Courtesy of Dr. Fabio Komlos, Children’s Hospital
Sonogram of Sequestration

- May show anomalous vessel to the sequestration

Courtesy of Dr. Fabio Komlos, Children’s Hospital
Sonogram of Sequestration

- Can also image prenatally

Axial sonogram of chest obtained at 22 weeks gestation in fetus with extralobar sequestration (*asterisk*). Cysts (arrows) are also visible.

_from Raipal Dhingal et al. AJR 2003; 180:433-437_
Sonogram of Sequestration

- Prenatal use of Doppler
- Distinguish sequestration from other congenital thoracic lesions

Sagital sonogram of chest obtained at 32 weeks' gestation in fetus with extralobar sequestration (mass).

from Raipal Dhillon et al. AJR 2003; 180:433-437
MRI of Sequestration

- MRI can also show precise anatomy of sequestrations (CT still better)
- MRI and U/S safe prenatally
- Accuracy still unknown

Coronal T2-weighted MRI obtained at 23 weeks' gestation, showing extralobar sequestration (asterix).

from Raipal Dhingal et al. AJR 2003; 180:433-437
Recap

**Intralobar**
1. Adults/adolescents
2. Pulmonary veins
3. No anomalies

**Extralobar**
1. Neonates/infants
2. Systemic veins
3. Often other anomalies
Acknowledgements

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References


• Scheung-Fat K *et al*: Noninvasive Imaging of Bronchopulmonary Sequestration. *AJR* 175:1005-1012, 2000