# AMSER Rad Path Case of the Month:

## An 8-month Old Male With a Fever and a Cough


Dakota Hammrich, MS4

PNWU Health Sciences

Julie Kaczmark, MD

Inland Imaging

Sarah Hill, MD

Providence Pediatric Surgery

Sanjay Logani, MD

Pathology, Incyte Diagnostics

# Patient Presentation

**HPI**:

An 8-month old male with a recent diagnosis of pneumonia, presents with a complaint of fever and a cough. The fever is refractory to Tylenol. His cough started two days ago and was been progressively worsening. His mother reports no other symptoms. She describes that the current symptoms are the same in quality and location, of that when he was diagnosed with pneumonia approximately 2 months ago at age 6-months.

Pertinent ROS: fever, cough

**Pertinent History:**

- Rx: Tylenol 15mg/kg q4 prn
- PMHx: none, no surgical history
- Allergies: none
- HCM: immunizations current. Influenza vaccine 10/2019

**Physical Exam:**

- Vitals: T: 38.4, HR: 129, RR: 32, O2 98% on RA
- Playful, smiling, interactive
- Diffuse anterior wheezing and mild wheezing in right posterior lower lung field

# Pertinent Work Up

- Age 6 months:
  - CBC, BMP, UA: unremarkable
  - RSV, Influenza A/B: negative
  - CRP: 1.6 (H)
  - CXR
- Age 8 months:
  - CXR

# ACR Appropriateness Criteria

Date of origin: 1999
Last review date: 2015

**American College of Radiology**
**ACR Appropriateness Criteria®**

**Clinical Condition:** **Fever Without Source or Unknown Origin—Child**

**Variant 1:** **Neonate younger than 1 month of age with fever without source (FWS) and no respiratory symptoms.**

| Radiologic Procedure | Rating | Comments | RRL* |
|---|---|---|---|
| X-ray chest | 5 | This procedure may be appropriate but there was disagreement among panel members on the appropriateness rating as defined by the panel's median rating. | ☢ |
| **Rating Scale:** 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate | | | *Relative Radiation Level |

**Variant 2:** **Neonate younger than 1 month of age with FWS and respiratory symptoms.**

| Radiologic Procedure | Rating | Comments | RRL* |
|---|---|---|---|
| X-ray chest | 8 | | ☢ |
| **Rating Scale:** 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate | | | *Relative Radiation Level |

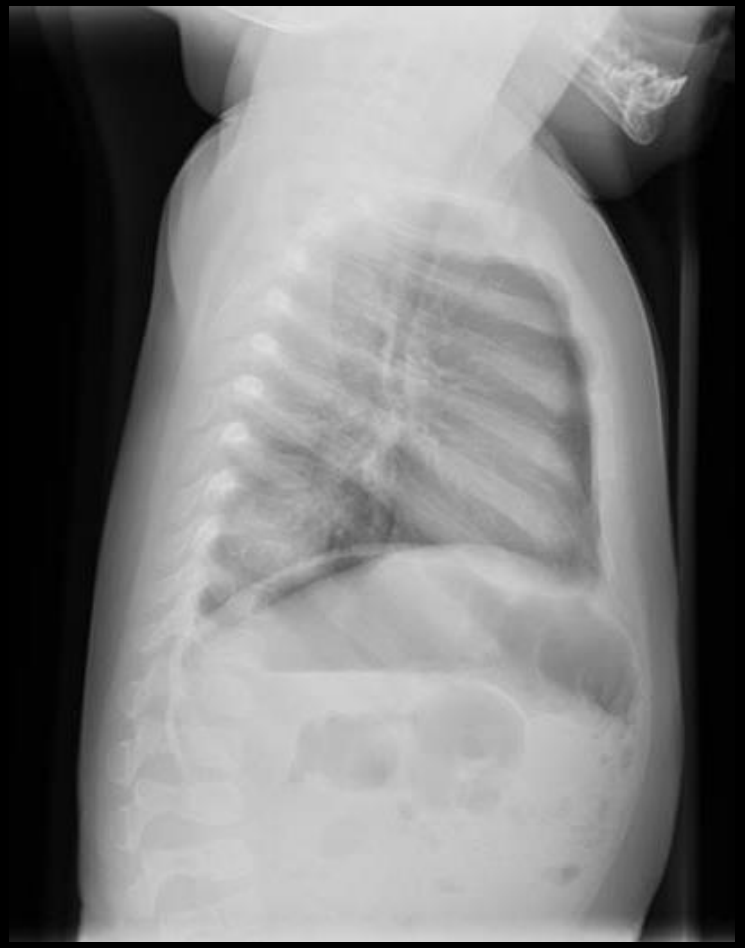
**Variant 3:** **Infant or child aged 1 to 36 months with FWS with low risk for occult pneumonia (no respiratory signs or symptoms, fever <39°C, leukocytosis <20,000/mm$^3$).**

| Radiologic Procedure | Rating | Comments | RRL* |
|---|---|---|---|
| X-ray chest | 2 | | ☢ |
| **Rating Scale:** 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate | | | *Relative Radiation Level |

**Variant 4:** **Infant or child aged 1 to 36 months with FWS with any of the following: respiratory signs or symptoms, fever ≥39°C, or white blood cell count ≥20,000/mm$^3$.**

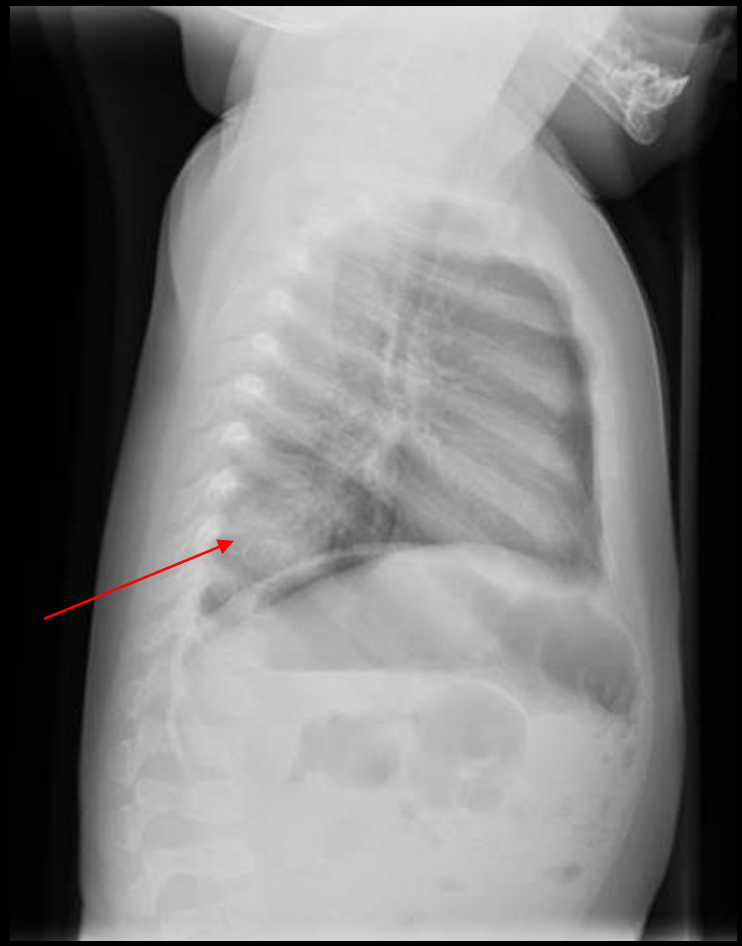
| Radiologic Procedure | Rating | Comments | RRL* |
|---|---|---|---|
| X-ray chest | 9 | | ☢ |
| **Rating Scale:** 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate | | | *Relative Radiation Level |

# Radiology Images (not labeled)

**AP/Lateral Chest Radiograph was obtained at age 8 months of age**

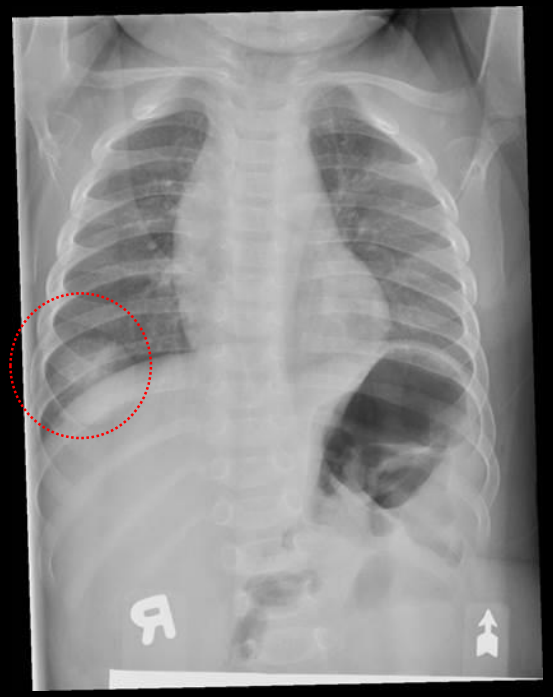
# Radiology Images (labeled)

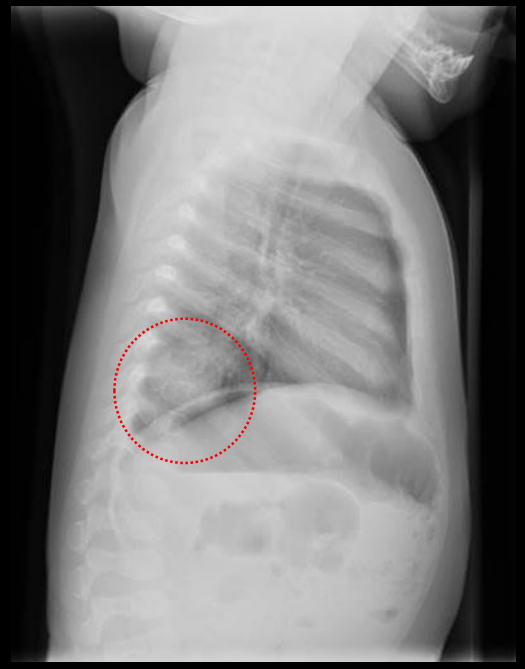


**3.5 cm circumscribed solid mass posterior inferior right lower lobe (red arrows).**

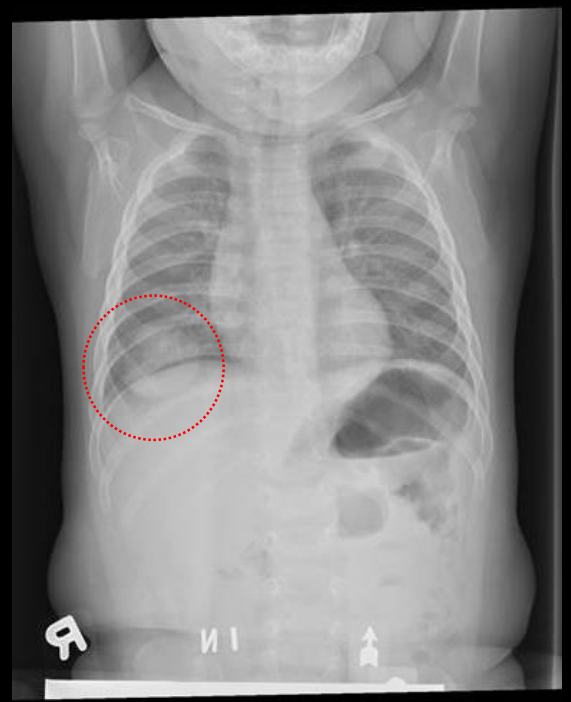
AMSER

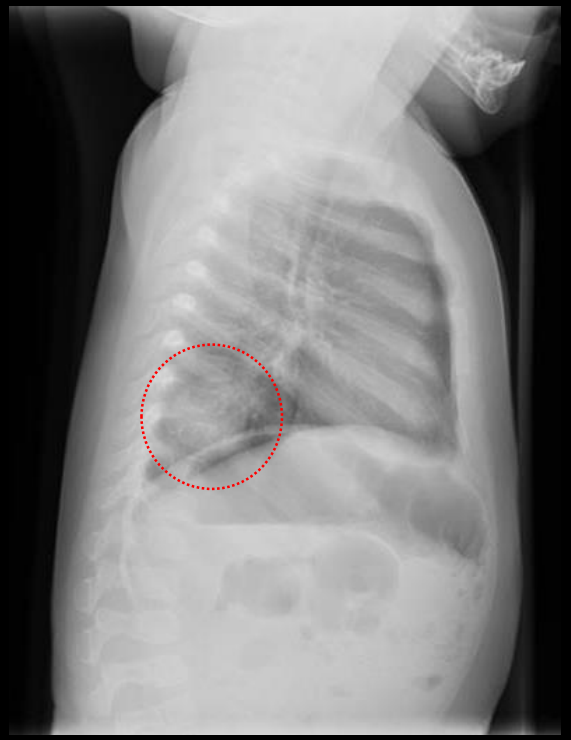
8 months old.





6 months old.



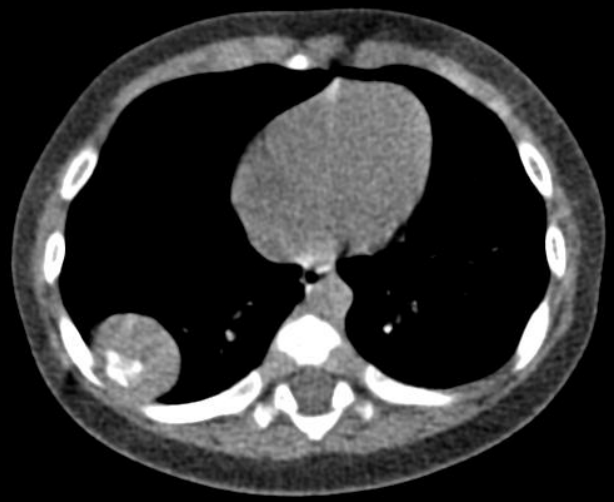


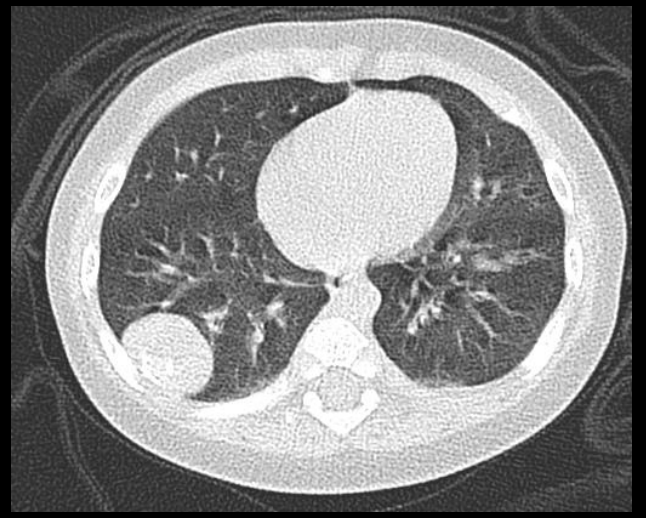
Chest radiographs were compared to a prior study obtained 2 months prior (patient was 6 months old). The mass was unchanged since prior study. Round pneumonia, as diagnosed at 6-months old, was unlikely due to lack of interval change.

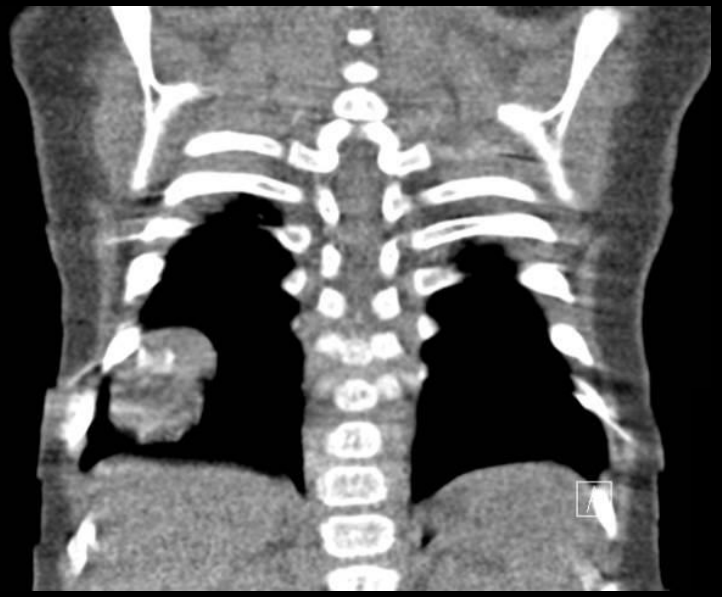
CT chest exam was recommended by pediatric oncology for further evaluation.

# Radiology Images (not labeled)

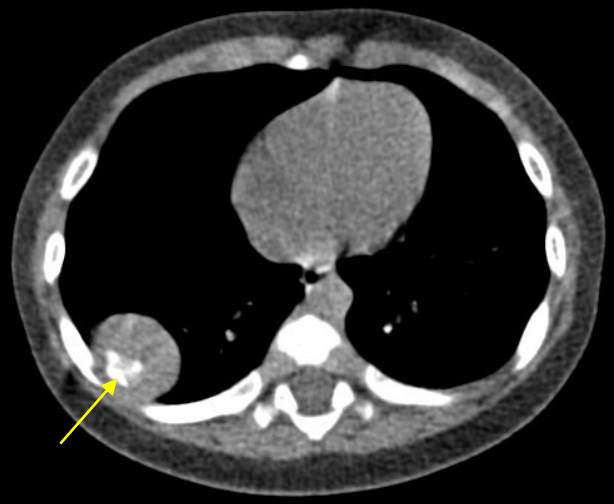
**Sagittal, Coronal and Axial Chest CT Without Contrast, obtained at 8-months old**

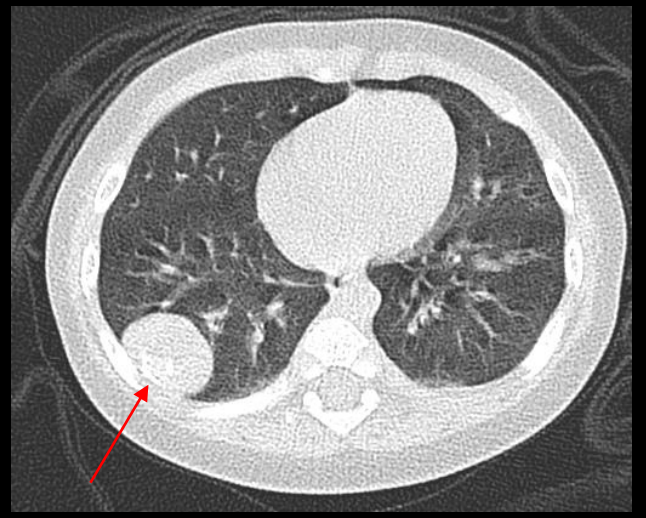






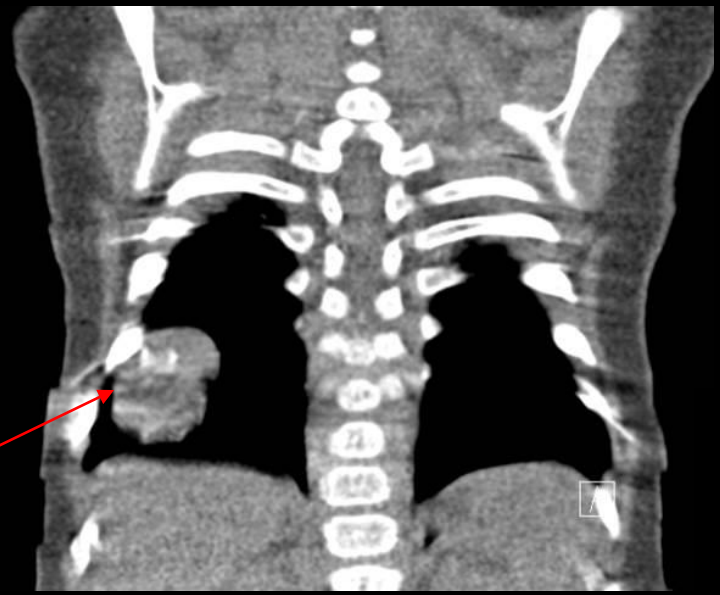
# Radiology Images (labeled)





**Sagittal, Coronal and Axial Chest CT Without Contrast, obtained at 8-months old**

This study showed a sharply circumscribed 3.3 cm diameter soft tissue lung mass in the right lower lobe, posterolateral aspect (red arrows). The lung mass shows coarse intratumoral calcification (yellow arrow).

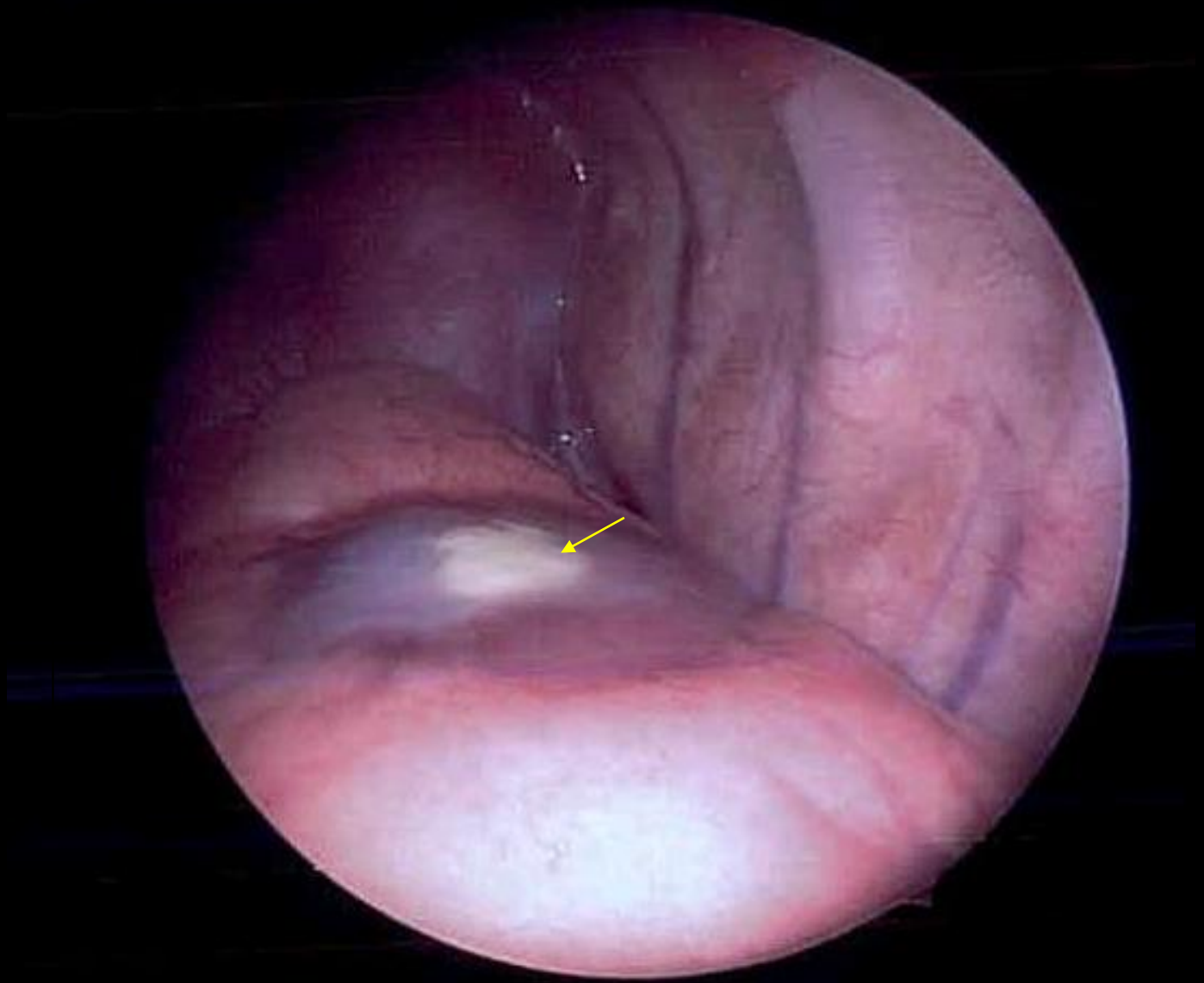


AMSER

# Infantile Pulmonary Mass DDx

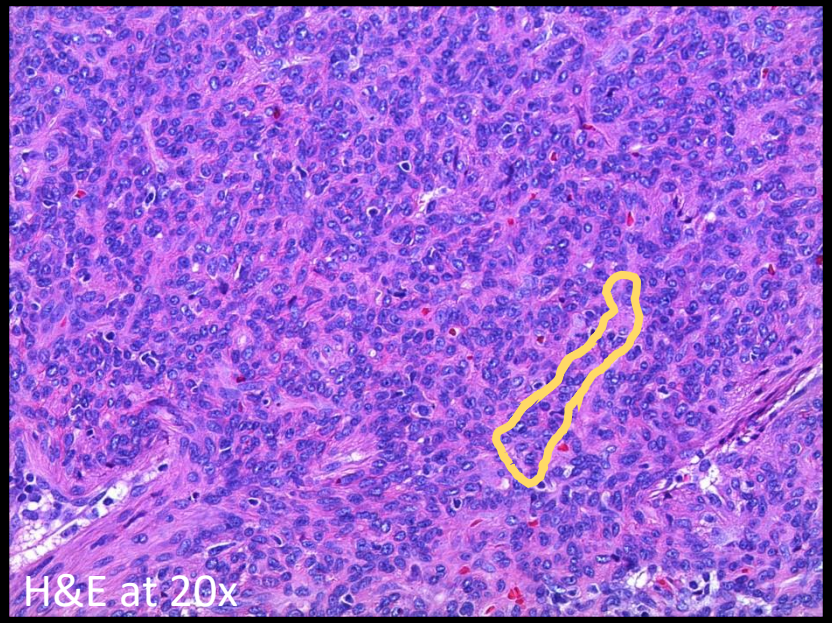
- Congenital
  - Bronchogenic Cyst
  - Congenital Pulmonary Airway Malformation
  - Pulmonary Sequestration
  - Congenital Lobar Emphysema
- Inflammatory
  - Histiocytosis
- Infectious
  - Fungal Infection
  - Pulmonary Tuberculosis
- Neoplastic
  - Pulmonary Hamartoma
  - Pulmonary Myofibroma
  - Metastatic Disease
  - Pulmonary Hemangioma
  - Pulmonary Blastoma
- Vasculopathy
  - Arteriovenous Malformation
  - Pulmonary Infarction

# Thoracoscopic photograph

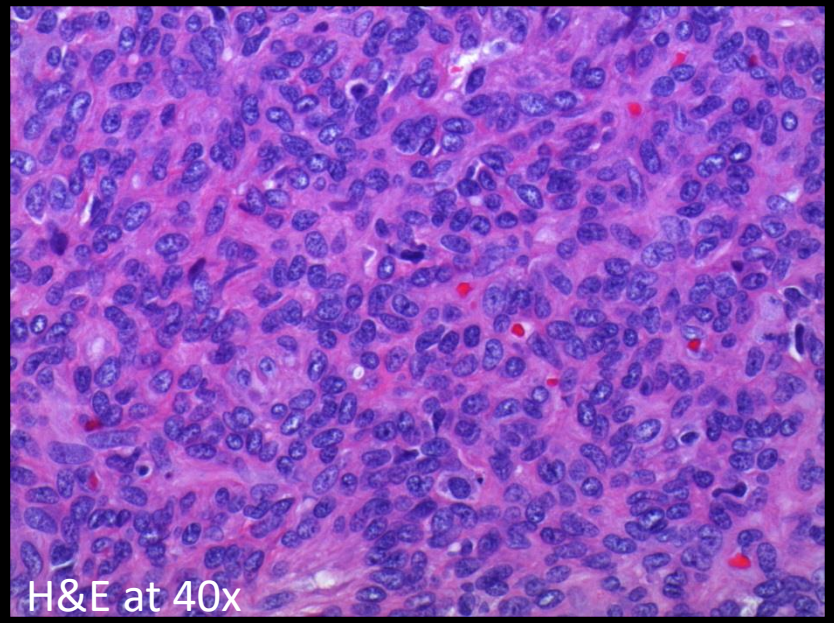


Pulmonary mass in the right lower lobe of the lung with a central necrotic calcified region (yellow arrow).

# Micro Path (labeled)

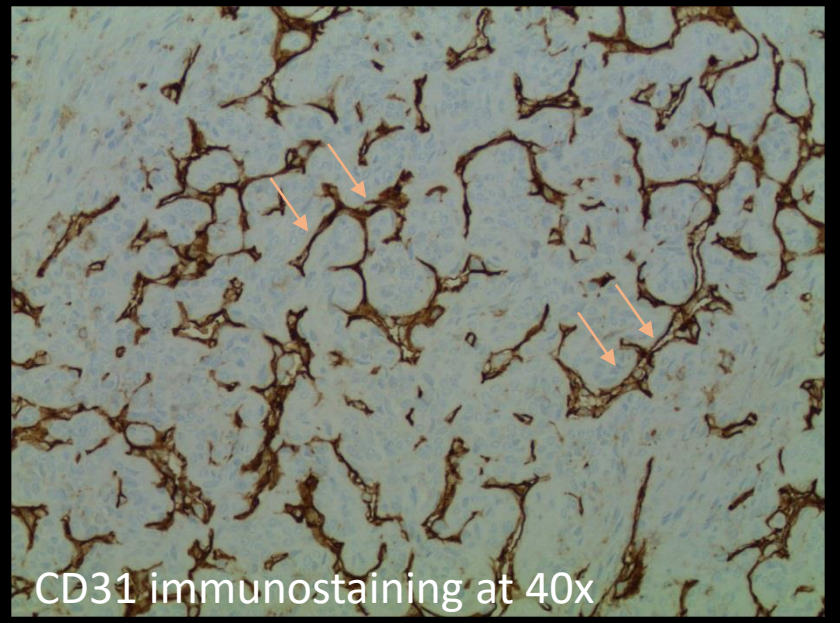



Round to oval tumor cells in a sheet like growth pattern forming round a around complex vasculature.




40x magnification demonstrating the nuclear characteristics of the tumor cells. Minimal mitotic activity is identified in the tumor.

Some cells adjacent to vessels have a more spindled or myoid phenotype.




CD31 immunohistochemical stain at 40x magnification highlights the delicate native vasculature of the tumor.

AMSER

# Final Diagnosis:

Pulmonary Myofibroma

# Myofibroma Discussion

- Background
  - Myofibromas are a mesenchymal neoplasm composed of myofibroblast, spindle cells.
  - They are the most common fibrous tumor seen in infancy, typically present in patients less than 2 years old and are most commonly sporadic.
  - Solitary lesions account for approximately 50-80% of cases, are typically benign and have a male predominance.
  - Solitary lesions may be seen anywhere in the body but most commonly occur in the dermis of the head and neck.
  - Multi-centric or lesions with visceral involvement are more likely to be malignant and carry higher morbidity and mortality. These lesions are more commonly seen in females.
  - If operable, solitary lesions which are completely removed carry an excellent prognosis with low recurrence risk.
  - Infantile myofibromatosis is a more severe form of multiple visceral myofibromas involved and has a female predominance.

# Myofibroma Discussion

Presentation:

- Clinical presentation depends on the location of the lesion.
- The most common presentation is a non-painful mass in the head or neck that is subdermal.
- Presentation will differ if a patient has visceral involvement.
- The most common sites of visceral lesions are the lungs, heart, gastrointestinal tract and pancreas.
- Visceral lesions are typically congenital.

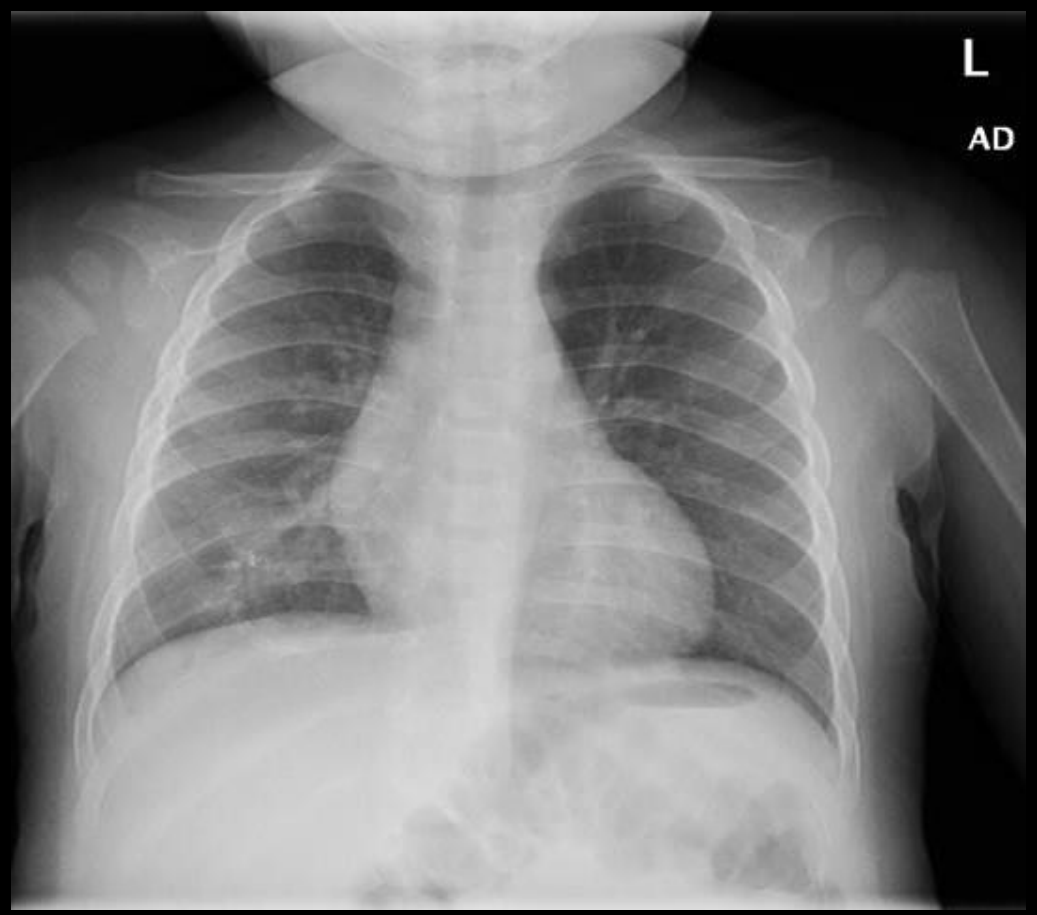
# Myofibroma Discussion

- Radiology Findings:
  - With ultrasound imaging, findings may range from anechoic to hyperechoic. The central portion of the mass may be anechoic due to central necrosis. There may be visible wall thickening. These lesions are typically hypovascular.
  - With CT imaging, a rim of enhancement may be seen peripherally with central necrosis. The remaining enhancement is near or less than that of muscle.
  - On MR imaging, these lesions most commonly appear with bright signal on T2 and low signal on T1 weighted imaging.

# Myofibroma Discussion

- Pathologic Features
  - Histologically, bundles of spindle cells may be seen with an eosinophilic cytoplasm.
  - A biphasic pattern is commonly seen composed of fascicles of spindle cells that have basophilic nuclei.
  - Scattered mitotic figures may be seen with zones of undifferentiated cells.
  - Cells typically stain positive for smooth muscle actin, muscle specific actin and vimentin.
  - Staining is typically negative for S100, desmin and epithelial membrane antigen.
  - Differentiating between benign and malignant lesions has been difficult, but these lesions typically resolve without recurrence if resected completely.
  - Rare genetic involvement has been reported with mutations at NOTCH3 and PDGFR8 loci.

# Radiology Images – Post-operative





**AP/Lateral CXR at 10 months of age after surgical resection**

Surgical staples right posterior lower lung with adjacent mild parenchymal opacity remaining consistent with postsurgical change.

AMSER

# References:


Sargar, K., Khuu, J, C., Curtin, P, T., Rosenberg, Folpe AL. (2016, July 11). Pediatric Fibroblastic and Myofibroblastic Tumors: A Pictorial Review. Retrieved May 16, 2020, from https://pubs.rsna.org/doi/full/10.1148/rg.2016150191

Venkatesh, V., Kumar, B., Kumar, K., & Mohan, A. (2015, March). Myofibroma-a rare entity with unique clinical presentation. Retrieved May 16, 2020, from https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4379250/

Sjirk J. Westra, MD ; Boaz K. Karmazyn, MD ; Adina L Alazraki, MD, et al. Pediatric Fever Without Source. Available at: https://acsearch.acr.org/docs/69438/Narrative/

American College of Radiology. (Accessed on May 16, 2020)


AMSER