

# AMSER Case of the Month

## November 2025

10-year-old male presenting with cough and chest pain

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AMSER

# Patient Presentation

**HPI:** 10-year-old male with a 2-month history of persistent cough and right sided chest pain since moving to the U.S. from Guam. He developed pneumonia while in Guam and was given an unknown antibiotic at the time, however, his cough did not improve. He also developed swelling on the right side of his chest and was seen by his PCP at an earlier time for asymmetric right sided gynecomastia.

**PMHx:** None

**Allergies:** None

**FamHx:** None

**SocHx:** Lives with 2 dogs, no smokers at home.

**Medications:** Loratadine 5mg/5mL

# Initial Exam and Labs

**Physical Exam:** Decreased breath sounds in right upper hemithorax and absent breath sounds in the mid and lower right hemithorax.  
Otherwise, normal exam.

## Vitals:

- BP: 118/74
- HR: 129
- Temp: 36.7 C
- Resp: 16
- SpO2: 96%

## CBC:

- WBC – 11.21 (normal)
- Hgb – 11.6 (normal)

**TB:** Negative

**Lactate Dehydrogenase:** 289 (elevated)

**Uric acid:** 18 (elevated)

**Viral respiratory panel:** negative

**Bacterial blood cultures:** negative

What Imaging Should We Order?

# ACR Appropriateness Criteria

**Chronic cough lasting more than 8 weeks. Persistent symptoms despite initial clinical evaluation and empiric treatment. Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
Radiography chest	Usually Appropriate	⊕
CT chest with IV contrast	Usually Appropriate	⊕⊕⊕
CT chest without IV contrast	Usually Appropriate	⊕⊕⊕
CT maxillofacial without IV contrast	May Be Appropriate	⊕⊕
Fluoroscopy biphasic esophagram	Usually Not Appropriate	⊕⊕⊕
MRI heart function and morphology without and with IV contrast	Usually Not Appropriate	○
CT maxillofacial with IV contrast	Usually Not Appropriate	⊕⊕
CT chest without and with IV contrast	Usually Not Appropriate	⊕⊕⊕
CT maxillofacial without and with IV contrast	Usually Not Appropriate	⊕⊕⊕
V/Q scan lung	Usually Not Appropriate	⊕⊕⊕
FDG-PET/CT skull base to mid-thigh	Usually Not Appropriate	⊕⊕⊕⊕

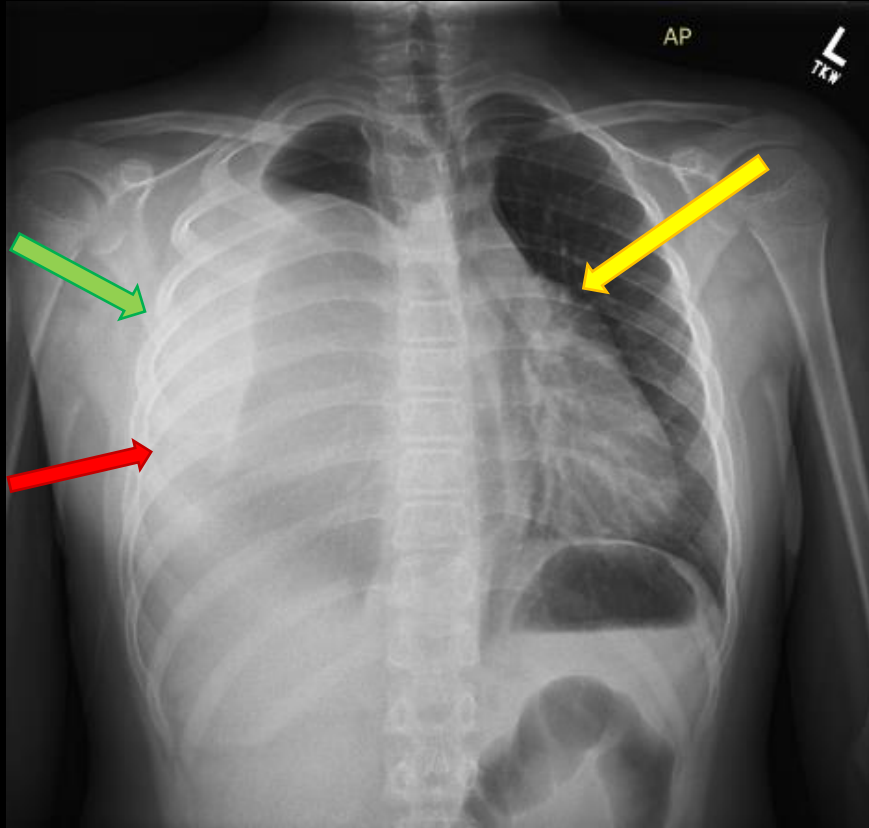


This imaging modality was ordered by the primary care physician

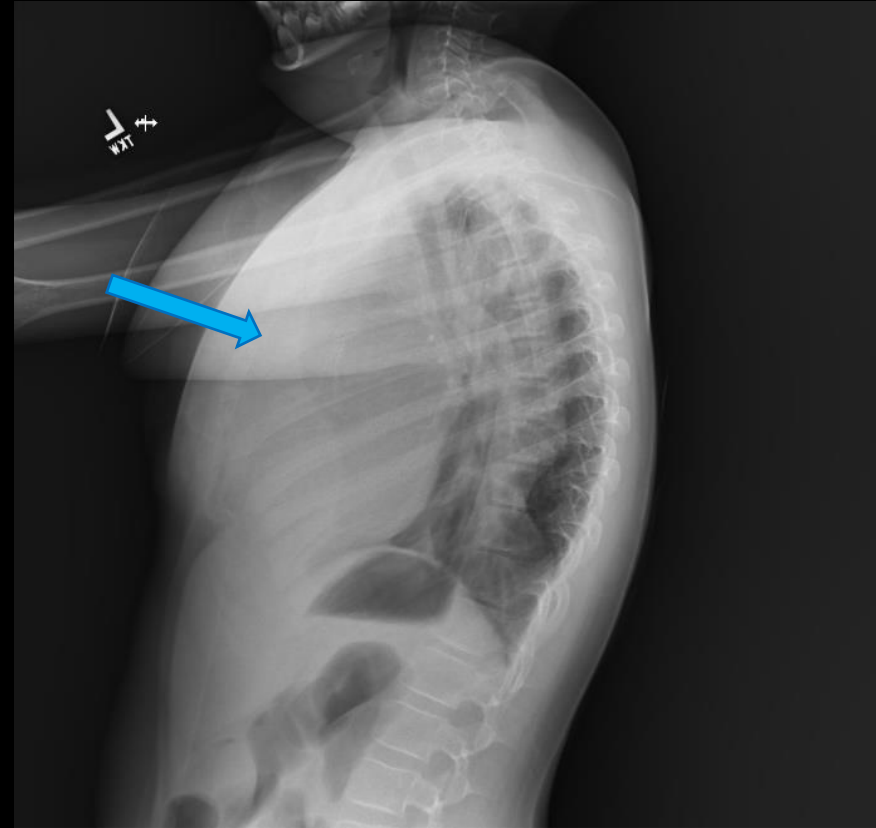
# Findings chest radiographs: (unlabeled)



# Findings chest radiographs: (labeled)



Chest x-ray showing a **large right sided opacity with pleural effusion** causing **leftward mediastinal shift**. The **right anterior 4<sup>th</sup> rib** is not well visualized.



Lateral chest x-ray showing **increased opacity mostly in the anterior mediastinum** with mass effect.

# ACR Appropriateness Criteria

**Indeterminate mediastinal mass on radiography. Next imaging study.**

Procedure	Appropriateness Category	Relative Radiation Level
MRI chest without and with IV contrast	Usually Appropriate	○
MRI chest without IV contrast	Usually Appropriate	○
CT chest with IV contrast	Usually Appropriate	☼☼☼
CT chest without IV contrast	Usually Appropriate	☼☼☼
US chest	Usually Not Appropriate	○
Image-guided transthoracic needle biopsy	Usually Not Appropriate	Varies
CT chest without and with IV contrast	Usually Not Appropriate	☼☼☼
FDG-PET/CT skull base to mid-thigh	Usually Not Appropriate	☼☼☼☼

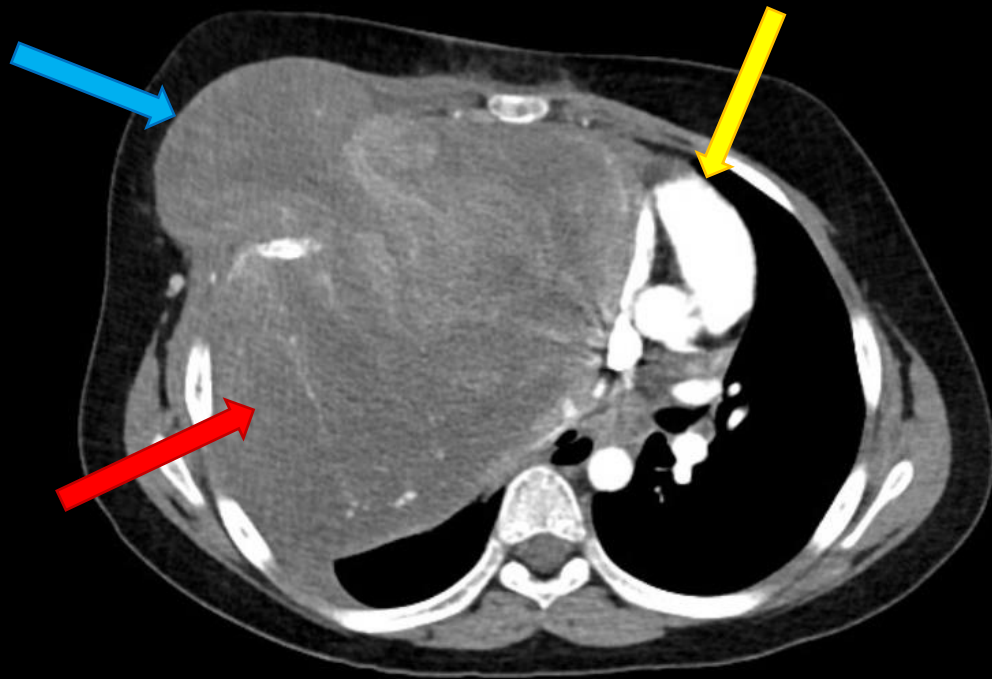


This imaging modality was ordered by the ED physician

# Findings CT chest w/ IV contrast: (unlabeled)



# Findings CT chest w/ IV contrast: (labeled)

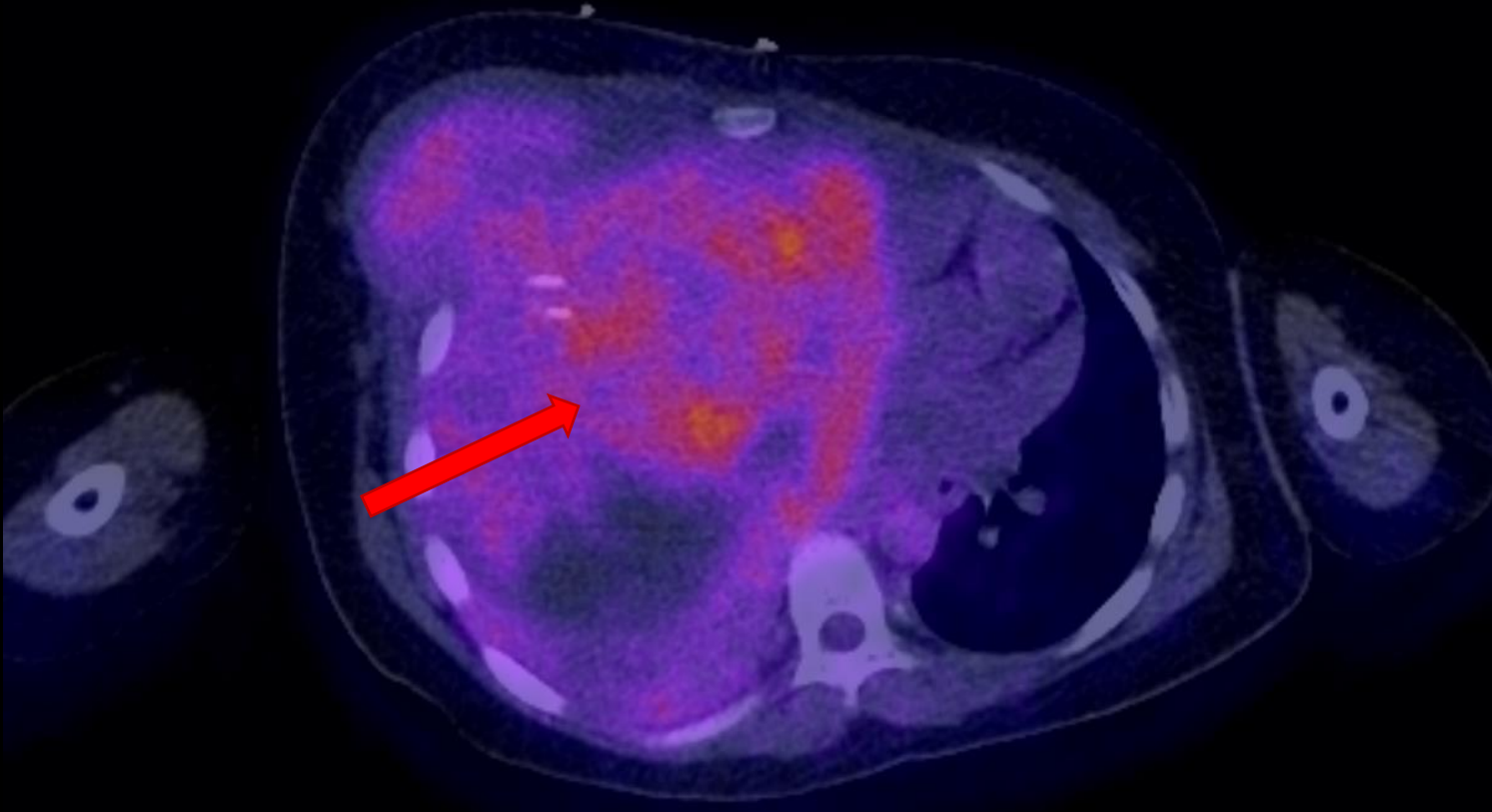


Axial contrast enhanced chest CT showing a large heterogenous pleural based mass in the right hemithorax measuring 16.2 cm x 15.0 cm with extension through the chest wall and rib destruction, and severe mass effect on the midline structures with leftward mediastinal shift.



Sagittal image showing involvement and destruction of the anterior 4<sup>th</sup> rib

# Findings PET/CT: (labeled)



Axial PET/CT showing a **large hypermetabolic mass in the right hemithorax and chest wall.**

## Final Diagnosis:

Ewing-like Sarcoma of the chest wall with  
BCOR:CCNB3 fusion

# Case Discussion: Overview

## Ewing-like sarcoma of the chest wall with BCOR:CCNB3 fusion

- An undifferentiated round cell sarcoma similar to Ewing sarcoma without the characteristic EWSR1-ETS translocation (Ewing-like)<sup>3,4</sup>.
- This specific Ewing-like sarcoma has a fusion of the BCOR and CCNB3 genes, leading to the overexpression of CCNB3, promoting abnormal cell cycle progression and proliferation<sup>3,4</sup>.

## Epidemiology

- Extremely rare tumor constituting only 4% of Ewing-like sarcomas.
  - Exact number not well known, but well under 1 case per million<sup>3</sup>.
- Approximately 80-90% of patients are male<sup>4</sup>.
- The median age tends to be early to mid teens<sup>4</sup>.

# Case Discussion: Presentation and Differential

## Clinical presentation

- Generally, these tend to arise slightly more commonly in bone than in soft tissue.
  - This will often present as pain or swelling over the affected site<sup>5</sup>.
- For those arising in the chest wall, patients will often have unilateral chest pain, shortness of breath and cough due to lung compression or pleural effusion.
  - Patients can also experience constitutional symptoms such as fever, weight loss and night sweats<sup>6</sup>.

## Differential Diagnosis

- **Ewing Sarcoma<sup>7</sup>**: Distinguished by the classic EWSR1:ETS translocation and is generally diffusely CD99 positive on immunohistochemistry. Generally considered more aggressive.
  - **Askin tumors** are part of the Ewing sarcoma family and are characterized by its chest wall involvement.
- **Rhabdomyosarcoma<sup>8</sup>**: Can be differentiated on immunohistochemistry by its strong positivity for Desmin, Myogenin, and MyoD1.

# Case Discussion: Diagnosis and Management

## Diagnosis – Imaging<sup>6</sup>

- **Radiography:** large soft tissue mass that can cause rib destruction or pleural effusion.
- **CT:** Heterogeneous mass arising from the pleura or rib with invasion into the thoracic cavity.
- **PET-CT:** Obtained to check for metastases.

## Management<sup>9</sup>

- **Neoadjuvant chemotherapy:** Vincristine, Doxorubicin, Cyclophosphamide (VDC)/Ifosfamide, Etoposide (IE) cycles are used as the initial step in treatment.
  - This is also the Ewing sarcoma chemotherapy protocol.
- **Radiotherapy**
- **Surgical resection**

## Diagnosis – Histopathology and Molecular Tools<sup>3,6</sup>

- **Biopsy:** Malignant small round cell tumor
  - BCOR:CCNB3 can have spindle cells or unusual stroma, however, it is not required or specific for diagnosis.
  - Around 50-60% of BCOR:CCNB3 fusion sarcomas stain weakly positive for CD99 compared to Ewing sarcomas which stain stronger and more diffusely.
- **FISH or RT-PCR:** Detects the absence of the EWSR1 translocation and presence of BCOR:CCNB3 fusion.
  - Molecular confirmation is the **gold standard**.

# Follow-up Imaging



Initial axial contrast CT with mass measuring 16.2 cm x 15.0 cm



Axial non-contrast CT 10 months later after 14 cycles of VDC/IE chemotherapy. Mass measures 14.0 cm x 8.6 cm

# References:

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4. Kao YC, Owosho AA, Sung YS, et al. BCOR-CCNB3 Fusion Positive Sarcomas: A Clinicopathologic and Molecular Analysis of 36 Cases With Comparison to Morphologic Spectrum and Clinical Behavior of Other Round Cell Sarcomas. *Am J Surg Pathol*. 2018;42(5):604-615. doi:10.1097/PAS.0000000000000965
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