# AMSER Rad Path Case of the Month August 2018





Synovial Sarcoma

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### **Patient Presentation**

- 26 year old previously healthy male presents to PCP with cough
  - X-ray at the time shows a lung nodule measuring 2.3 x 1.3 cm in the left upper lobe
  - PET/CT negative
- 13 months later comes to urgent care for SOB with blood tinged sputum
  - Prescribed ABX for presumed bronchitis. Symptoms briefly improve.
- In the next two months, SOB returns with night sweats, 30 lb weight loss, and a R abdominal wall mass
  - Also develops R leg pain and low back pain, which prompts him to return to PCP.



### **Patient Presentation**

PMH HTN

PSH None

Hosp None

Meds None

Allergies NKDA, NKA

FMH Father had lymphoma in 50s, now in remission.

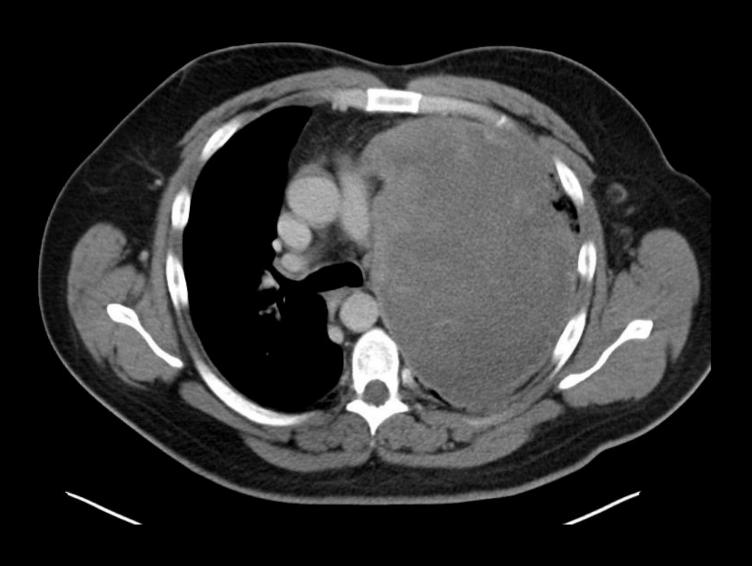
Grandmother had breast cancer.

Social Smoker for 10 years, 6 drinks/week, cannabis use

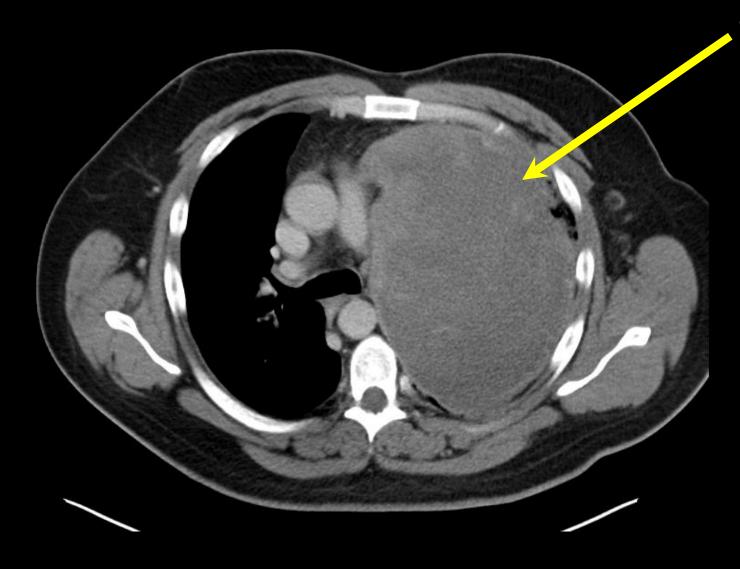
<1/month. Works at a steel mill, previously in plastics for 3 years.

X-ray of back ordered that reveals large lung mass CT of chest and abdomen ordered



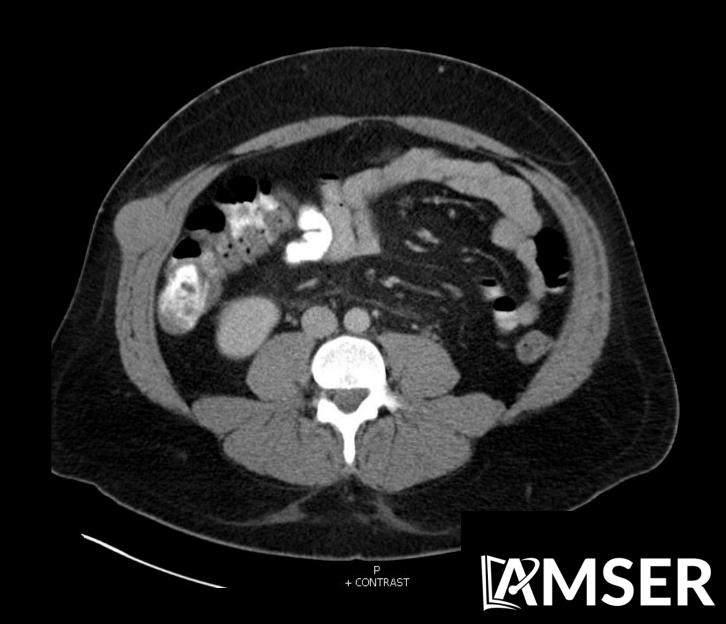




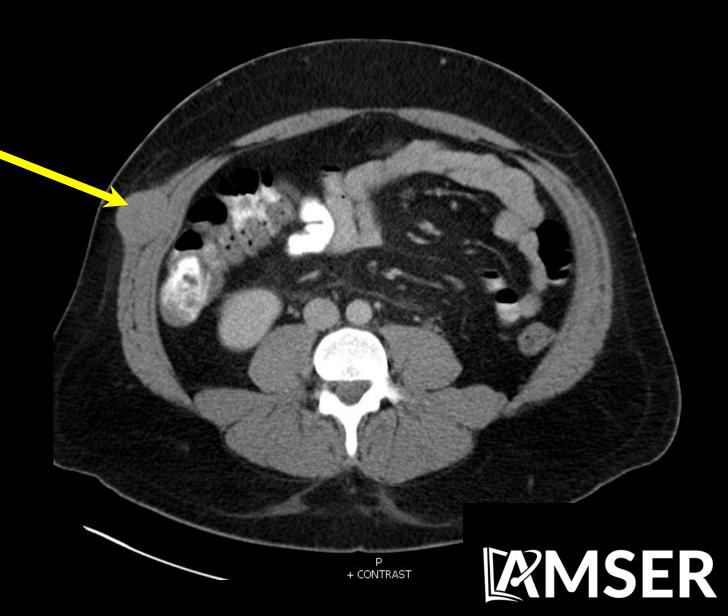


- Lung mass
  - 17.8 x 13.4 x 14.6 cm
  - Heterogeneous enhancing
- Mediastinum displaced right
- Partial encasement of the left main pulmonary artery and the left upper lobe bronchus
  - Not visible on this image





- Abdominal mass
  - 3.0 x 3.4 cm
  - Homogenous mass in the external oblique muscle

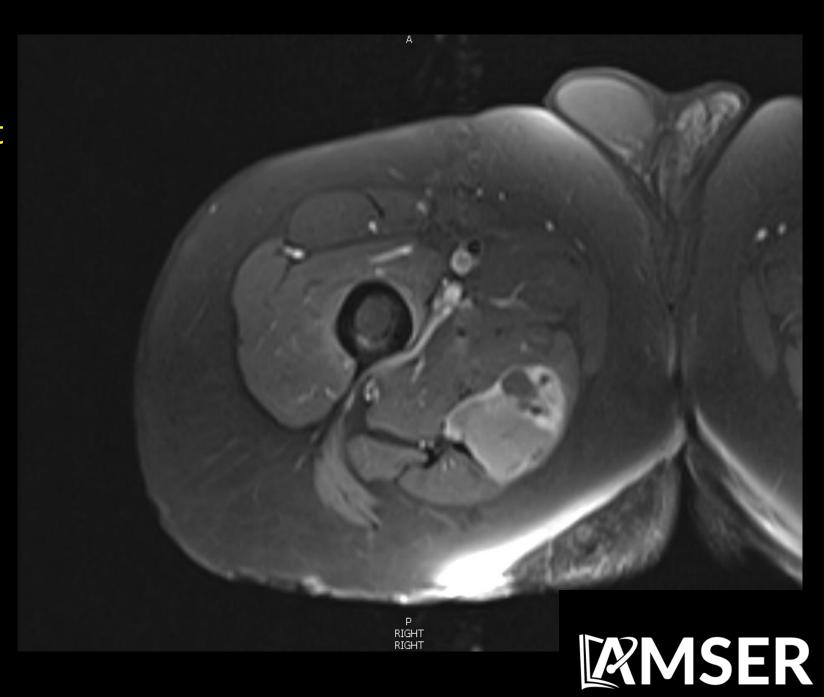




- Thigh mass
  - 5 x 4.5 x 5.8 cm
  - Intimately associated with the adductor magnus
  - Peripheral enhancing and centrally necrotic mass



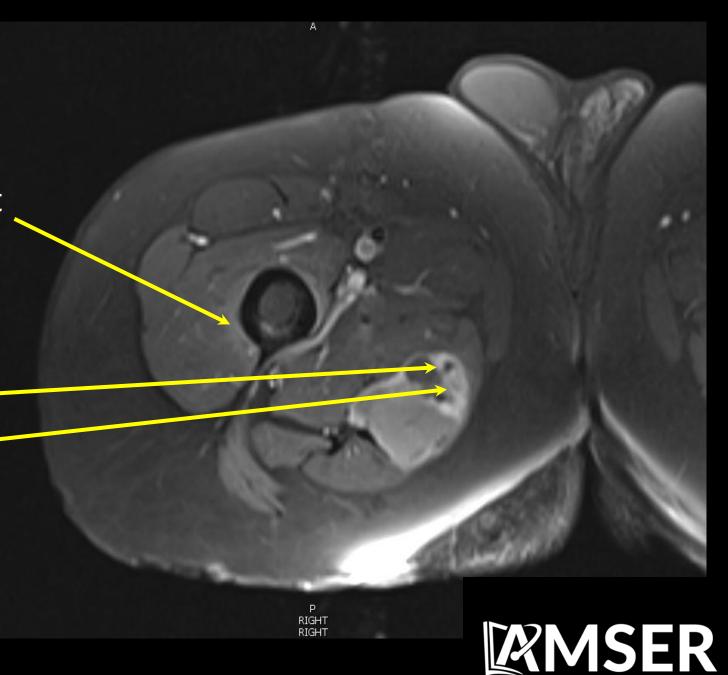
### MRI T1 Fat Sat with Contrast



#### MRI T1 Fat Sat with Contrast

• Enhancing periosteum consistent with periostitis

- Thigh mass
  - Peripheral enhancement
    - Intrinsic hyperintensity suggests blood products and/or proteinaceous material



### MRI T2 Fat Sat

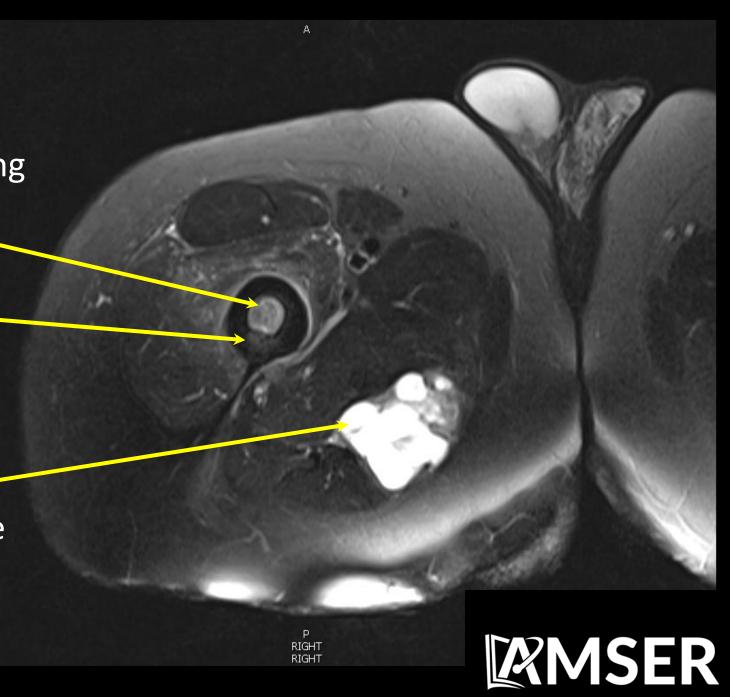


#### MRI T2 Fat Sat

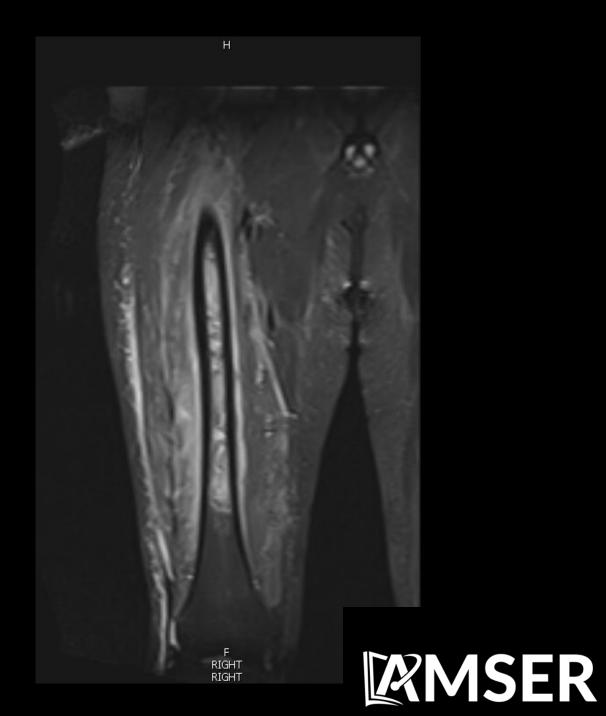
• T2 hyperintense signal concerning for marrow involvement

• T2 hyperintense intracortical signal

- Thigh mass
  - Hyperintense centrally
    - Likely necrosis
  - "Bowl of Grapes" appearance

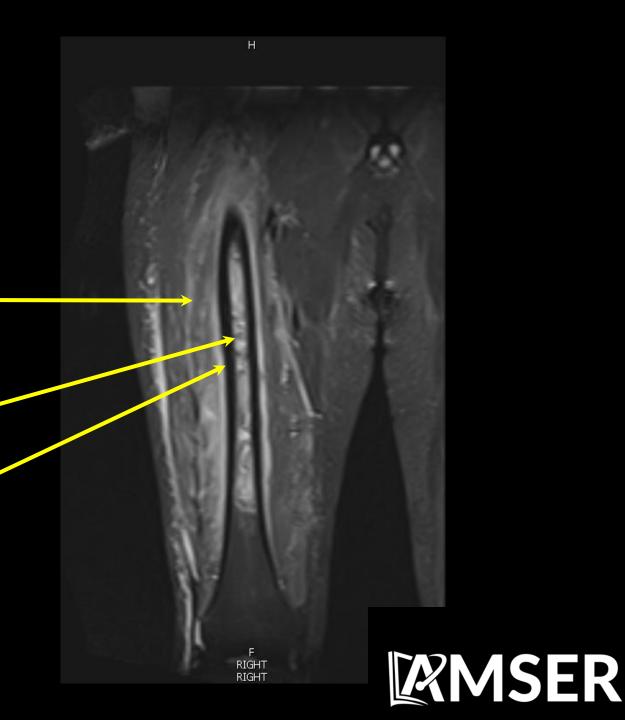


### MRI STIR



#### **MRI STIR**

- Femur
  - Surrounding musculature shows some edema, likely reactive
  - Hyperintense marrow signal abnormalities
    - No focal mass seen
  - Periosteal edema



### Differential Diagnosis Based on Imaging

Primary lung cancer with metastases to other organs
Sarcoma with metastases to the lung
Lymphoma

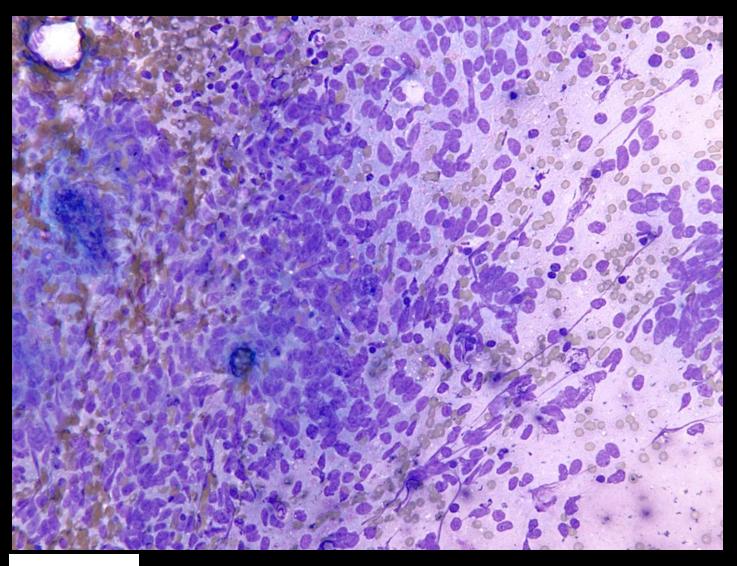
### CT-guided Biopsy of the Left **Lung Mass**

- Necrotic surroundings
- No nucleoli seen in cells
- Poorly organized sheets





**High Power** 



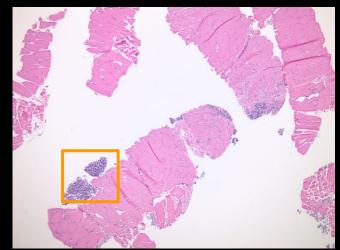
### **Lung Cytology**

- Uniform population of small round blue cells
- "Spindle cells" seen are likely smeared cells from plating



# US-guided biopsy of the Right Abdominal Mass

- Loosely cohesive sheets
- Focal areas with
  - Spindle cells
  - Small round blue cells with medium amount of cytoplasm
  - No nucleoli



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#### CD99 Stain

- Used to distinguish Ewing's sarcoma, mesenchymal chondrosarcoma, solitary fibrous tumors, and synovial sarcoma
- Cytoplasm here shows strongly positive result for CD99
- FISH for t(X;18)(p11;q11) is specific to synovial sarcoma returned positive



### Final Dx:

Synovial Sarcoma



## **Epidemiology**

- Presents in adolescents and young adults (15-40 years of age)
- Mild male predilection (1.2:1)
- Uncommon, 2.5-10% of soft tissue sarcomas

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•	Extremities	80-95%
	<ul> <li>Lower Limb</li> </ul>	60-70%
	<ul> <li>Upper Limb</li> </ul>	15-25%
•	Head and neck	5%
•	Chest wall	Rare
•	Viscera	Rare



### Presentation

- X-ray
  - May show calcifications (30% of cases)
- US
  - Non-specific, may show heterogeneous predominantly hypoechoic mass
- CT
  - Non-specific soft tissue mass of heterogeneous density and enhancement
  - Sensitive to calcifications, if present

- MRI-modality of choice
  - T1
    - Iso or slightly hyperintense to muscle
    - Heterogeneous
  - T2
    - Mostly hyperintense
    - Necrosis and bleeding may cause "bowl of grapes" appearance (10-25% of cases)
  - T1 Contrast
    - Enhancement is prominent

•	Diffuse	40%
•	Heterogeneous	40%
•	Peripheral	20%



### Synovial Sarcoma Histology

- Not derived from synovium
- Microscopically resembles normal synovium
  - Stains for epithelial markers while true synovium does not
  - Usually biphasic appearance with epithelial cells and spindle cells
  - Can be monophasic
- Macroscopically appears as heterogeneous masses with areas of hemorrhage and necrosis originating within soft tissues near large joints



### **Treatment**

- Surgery
  - Curative in 20-70% of patients
  - Used on smaller tumors with no evidence of metastasis
- Chemotherapy
  - Benefit of treatment still unclear
  - Some evidence supports use of doxorubicin/ifosfamide in advanced disease
- Radiotherapy
  - Benefit is less clear than for chemotherapy
  - Used to reduce the chance of local recurrence
- This patient received combination chemo/radiotherapy



### **Prognosis**

- 5 year survival is 36-76% depending on the stage of disease
- Local recurrence is common (30-50% overall)
  - 14% recurrence over 10 years after diagnosis
- Distant metastases is frequent (40-70%) within 5 years of diagnosis
- Common locations of metastasis

•	Lung	80%
•	Bones	15%
•	Lymph Nodes	10%
•	Chest wall/Abdomen	7.5%



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