# 2018 RSNA Image Interpretation Session

Neuro

# 10 y.o. old boy with fevers and fatigue x 5 days, admitted due to altered mental status

HR 123, BP 140/90

**Lumbar puncture** 

- Opening pressure 35 cm H20
- 194 nucleated cells/mL, 99% lymphocytes
- protein 234 mg/dL
- glucose 45 mg/dL

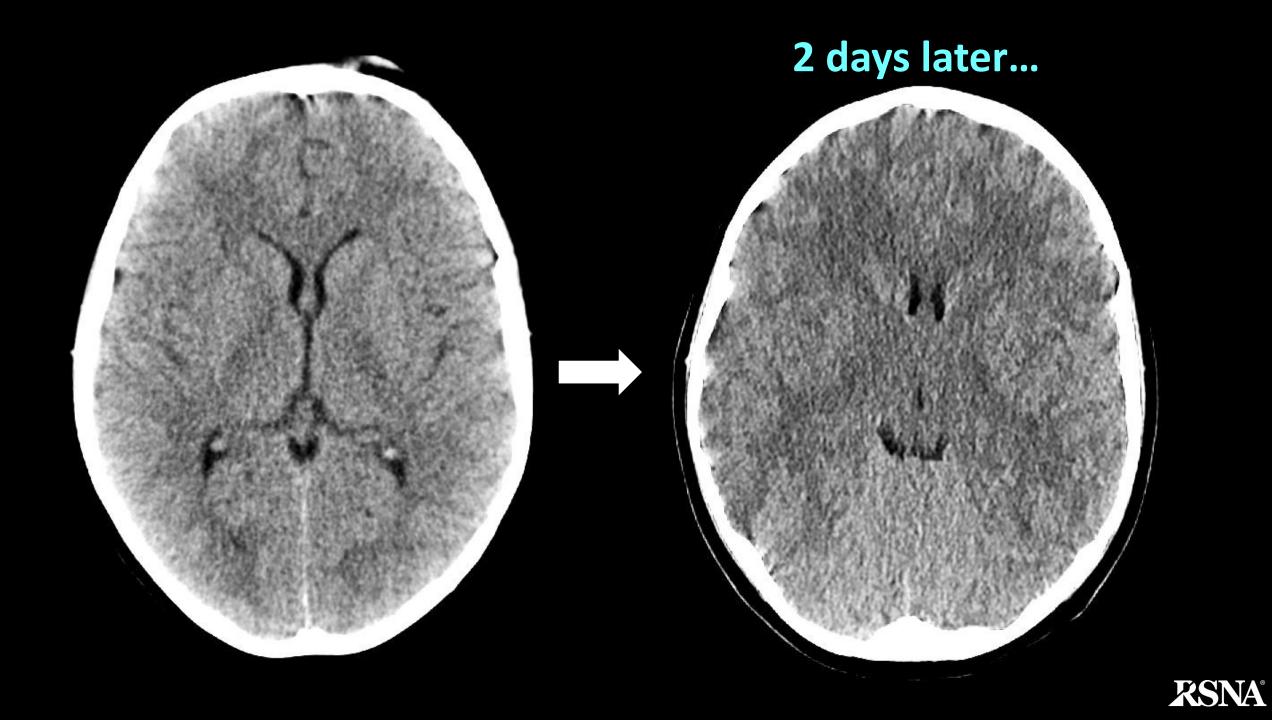
Started acyclovir & broad spectrum antibiotics





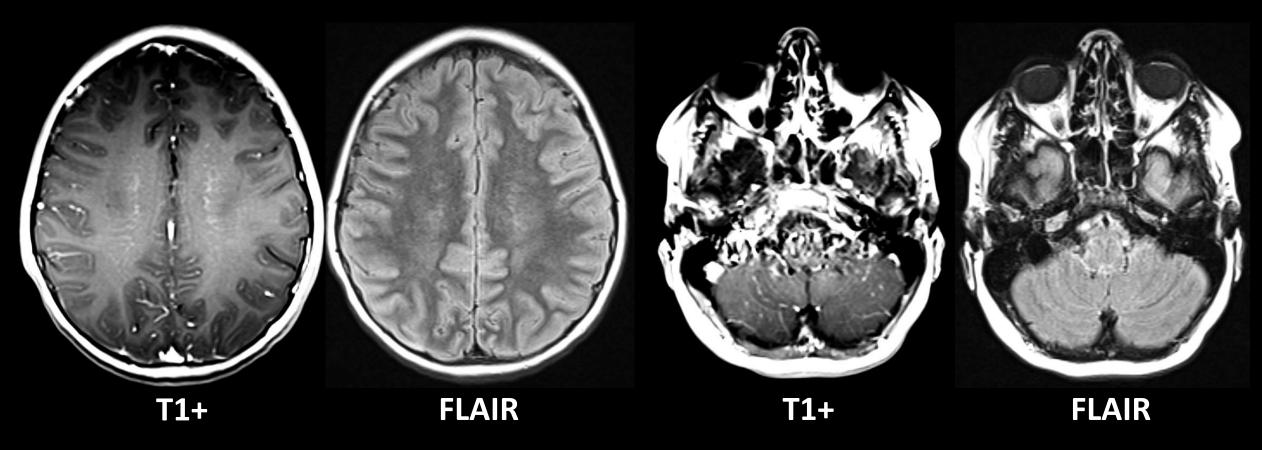
#### **Initial Evaluation**

- History viral prodrome
- Exam Fever, HTN, tachycardia
- LP lymphocytic pleocytosis, high opening pressure
- RX acyclovir, antibiotics



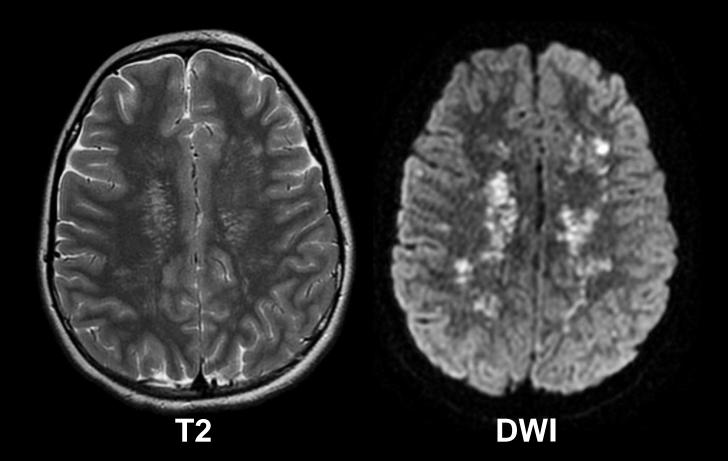
# 1 day later... CSF cultures and PCR come back negative; continued tachycardia and hypertension

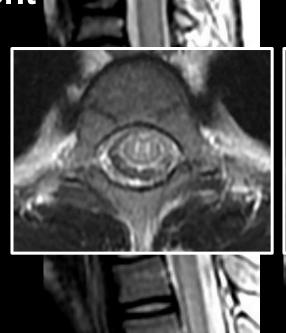
- Subtle central perivascular enhancement
- Leptomeningitis

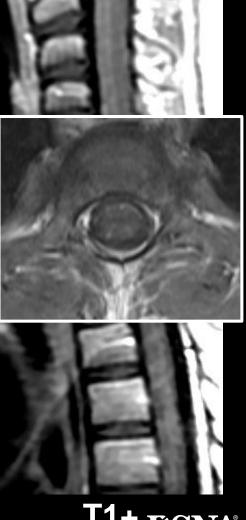


### 3 days later... coma, flaccid paralysis

- Periventricular T2 worse, reduced DWI
- Longitudinally extensive myelitis
- Subtle spinal leptomeningeal enhancement







## 3 days later... coma, flaccid paralysis

- Periventricular T2 worse, reduced DWI
- Longitudinally extensive myelitis
- Subtle spinal leptomeningeal enhancement



Viral, Inflammatory, Vasculitis, Autoimmune

What would you do next to treat this patient?

**T2** 

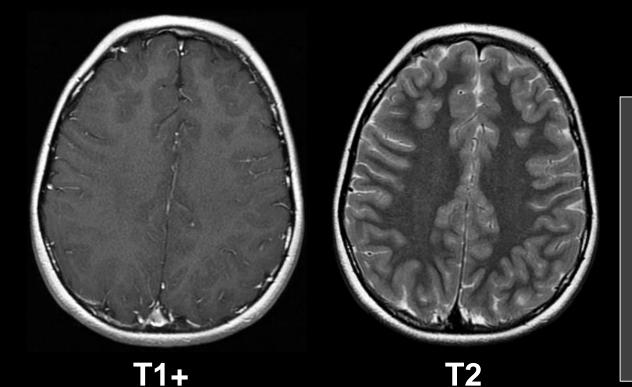
DWI

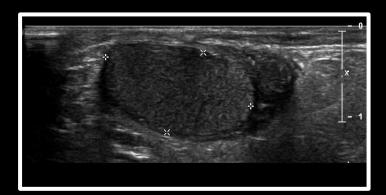
**T2** 

T1+ RSNA

#### Steroids, IVIG, Plasmapheresis... dramatic improvement!

- Walking with assistance 2 weeks later
- Maintained on IV steroids deficits resolve (mild difficulty concentrating)
- Brain MRI normal on 6 month follow-up
- Normal CAP CT, scrotal US





#### **Differential Diagnosis**

Viral
Inflammatory
Vasculitis
Autoimmune

**KSNA**°

# 2018 RSNA Image Interpretation Session

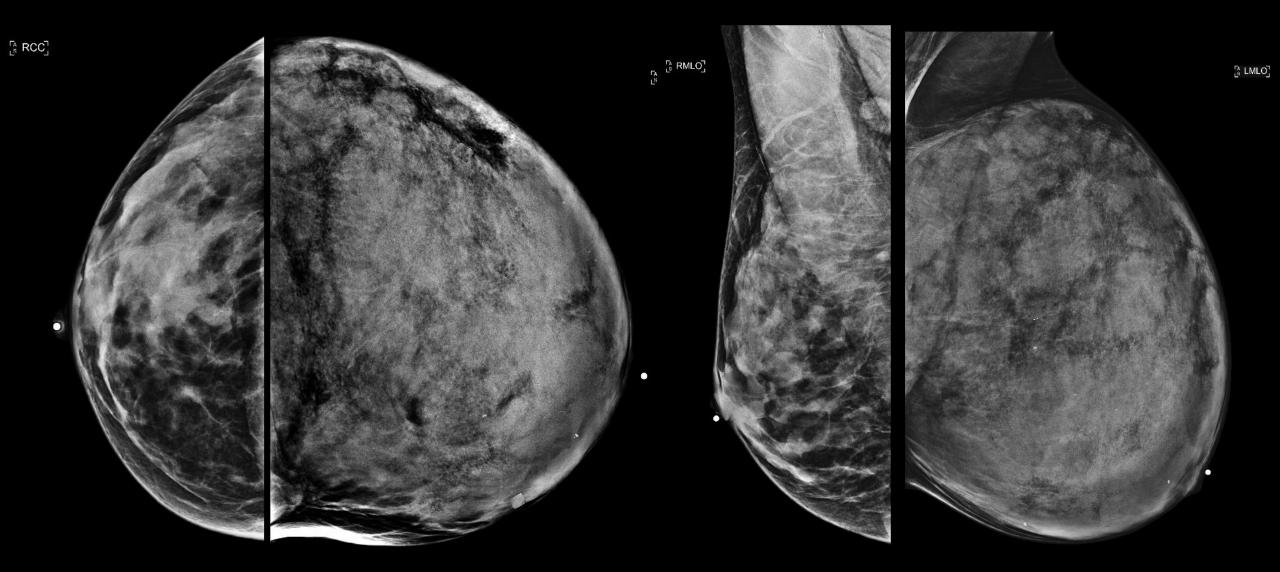
Breast



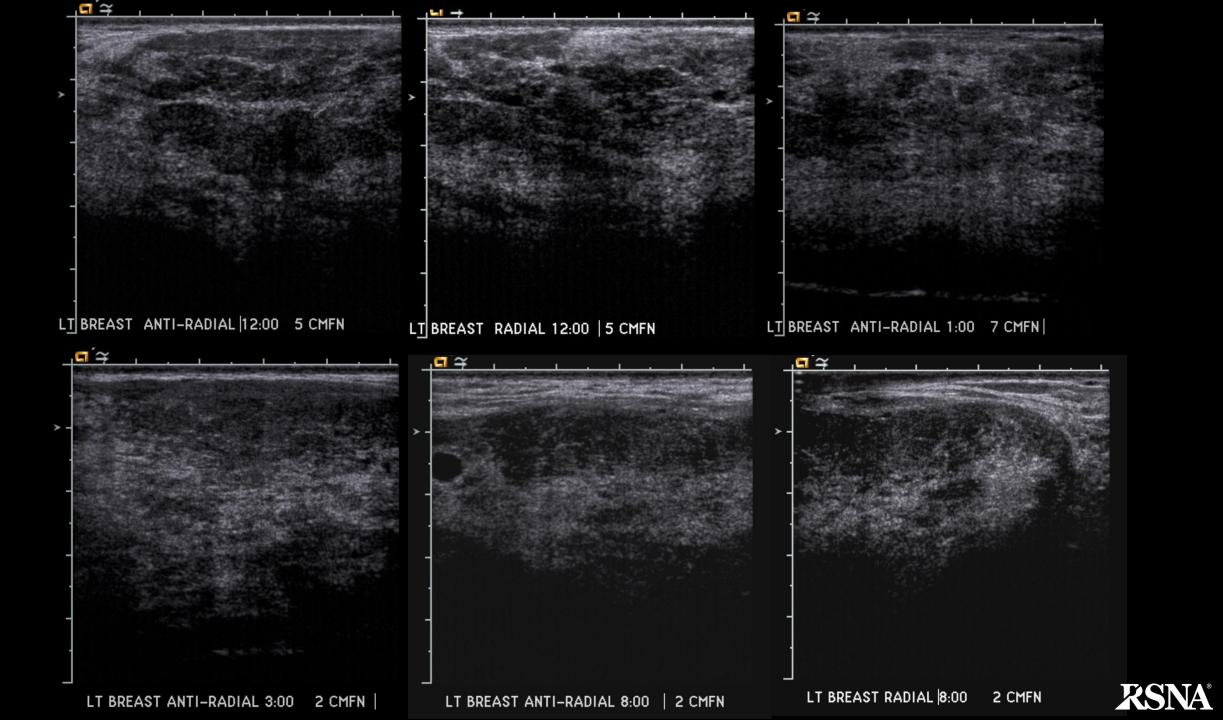
# History

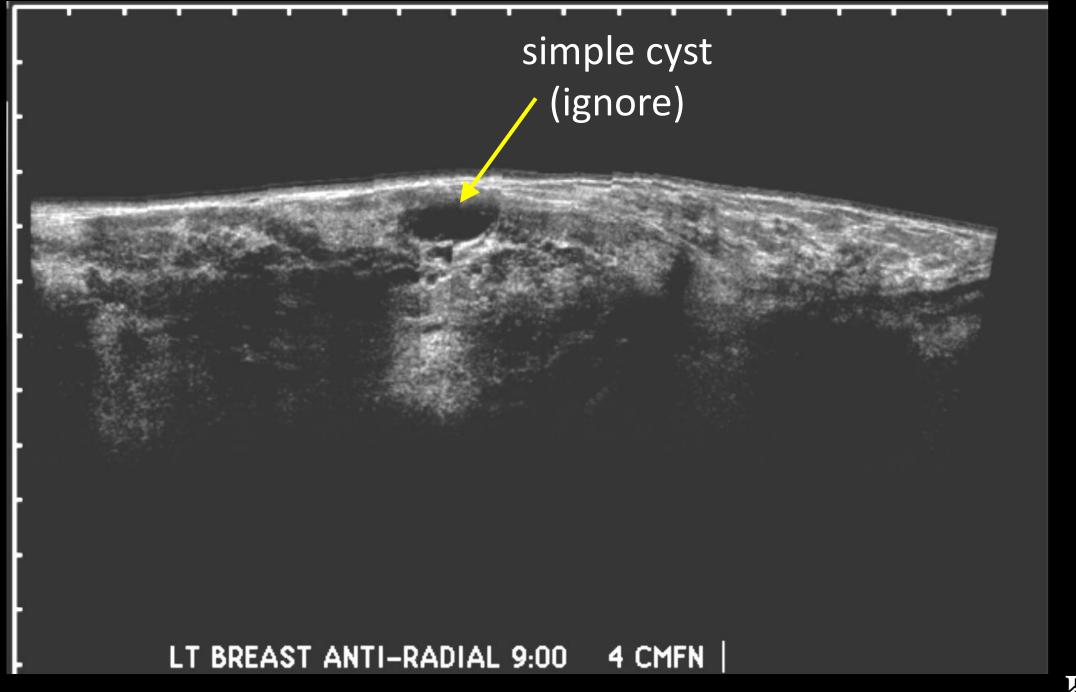
- 37 yo woman with no contributing history
- Presents with painless enlarging left breast over last 12 months. Left now 2 bra sizes asymmetric c/w right
- Physical exam:
  - right breast normal exam
  - left breast diffusely firm, no skin changes, no axillary or supraclavicular adenopathy; prominent veins visible throughout the left breast





## **KSNA**°

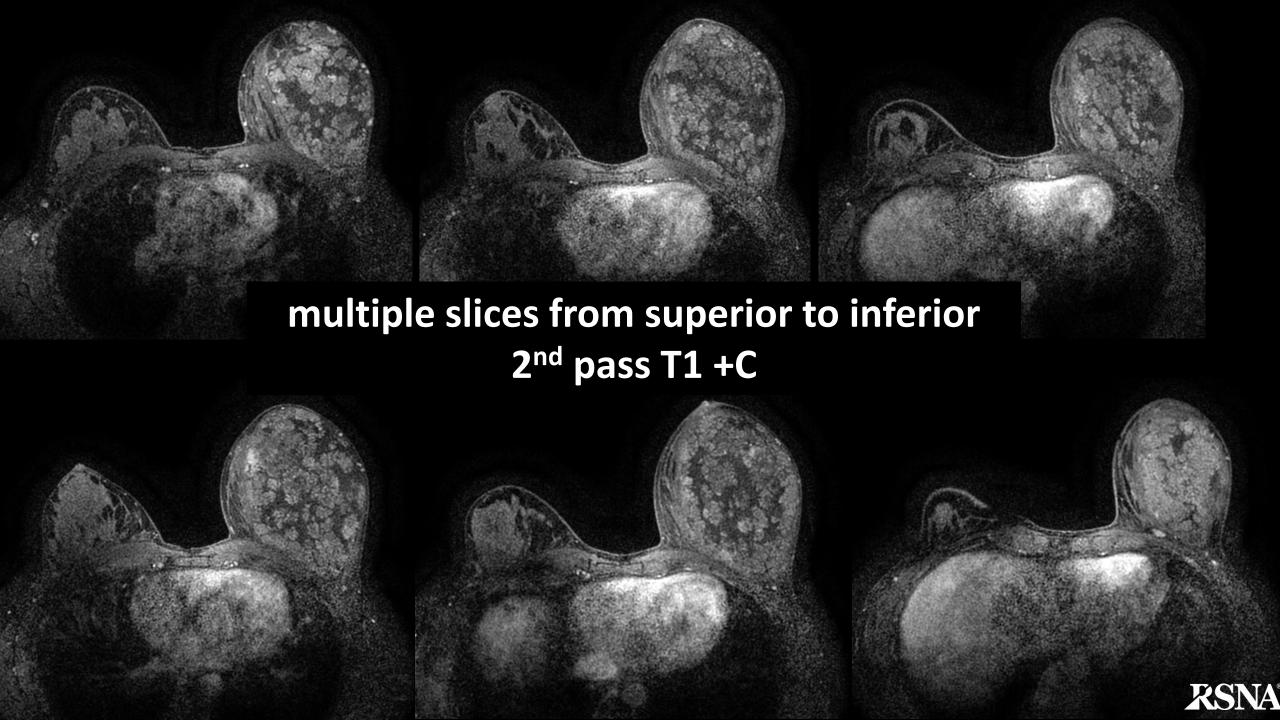


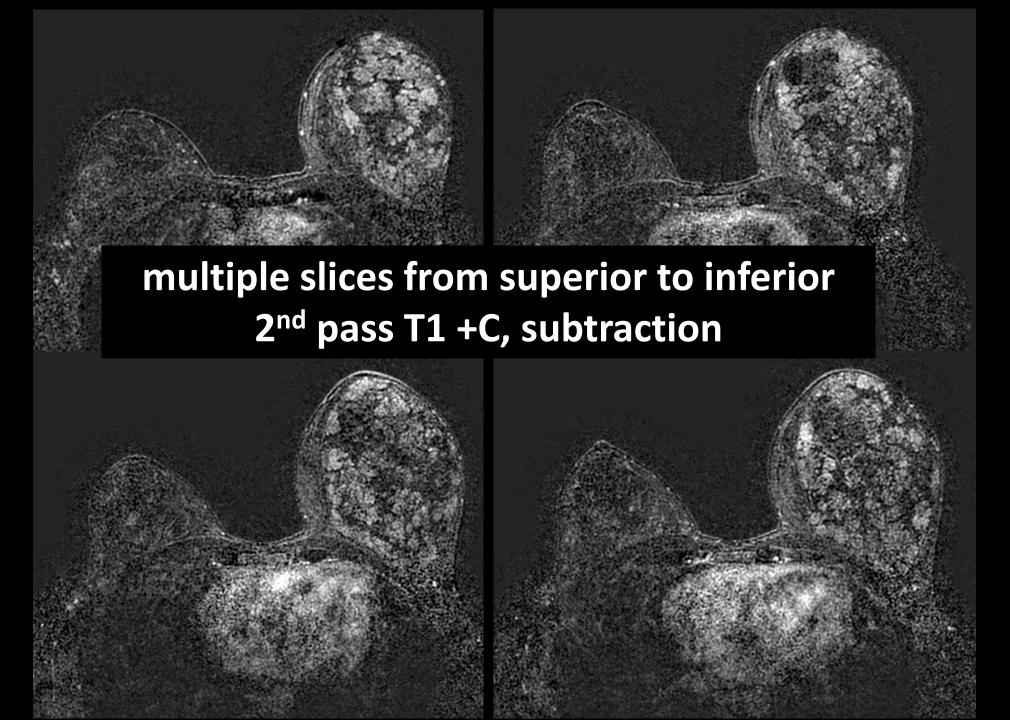


# Large painless mass in a young woman without adenopathy/skin changes growing over a year

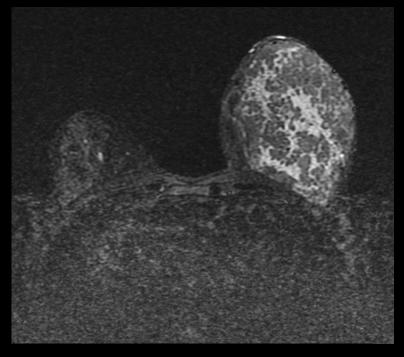
- Lipofibroadenoma (hamartoma)
- Giant Fibroadenoma / Phyllodes Tumor
- Cancer / Angiosarcoma
- Lymphangiomatosis
- Giant Pseudoangiomatous Stromal Hyperplasia (PASH)

# Biopsy performed at this point









T1 fat-sat, pre-contrast

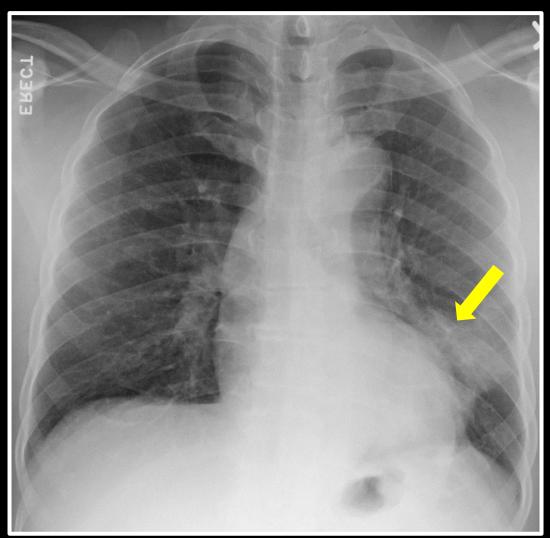
T1 non fat-sat

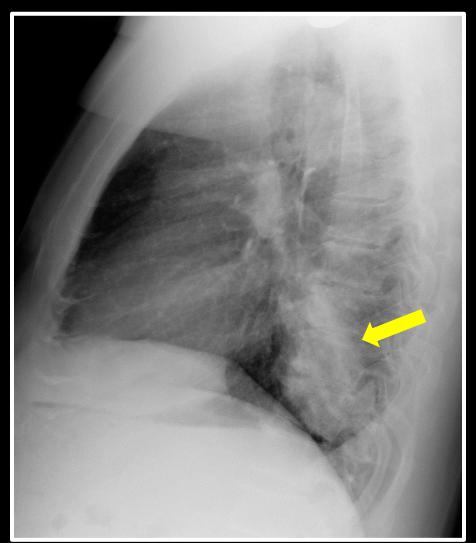
T2WI

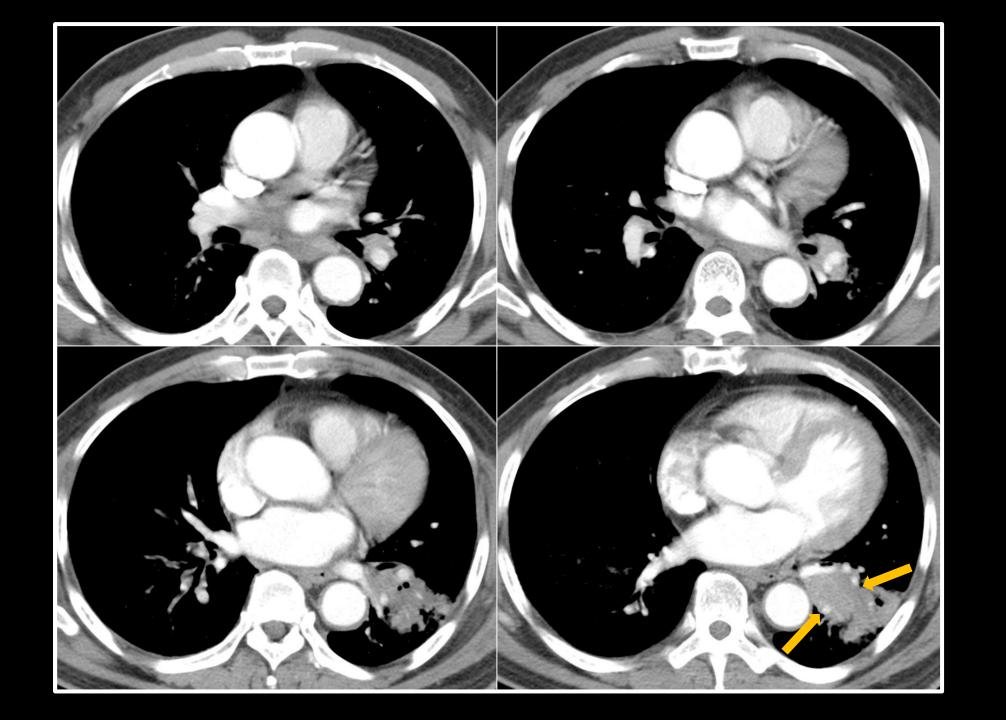
# 2018 RSNA Image Interpretation Session

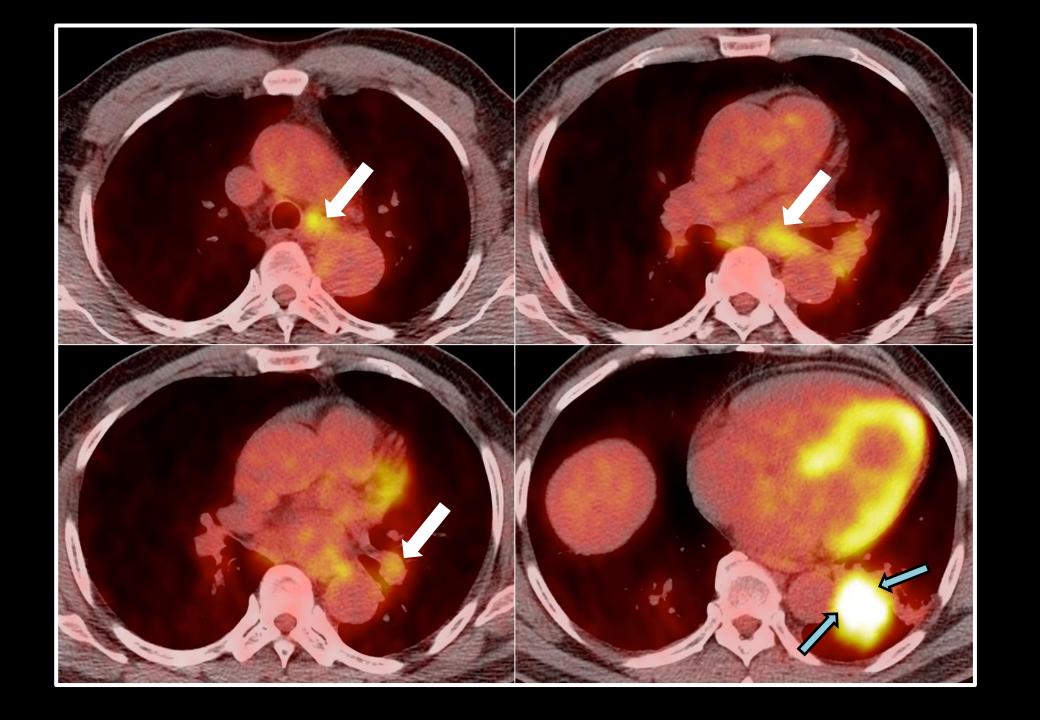
# Thoracic

# Middle age man; former smoker, hemoptysis









# **Imaging Findings**

- CXR: Ill-defined mass in the LLL extending from the left hilum.
- Contrast enhanced CT: Homogeneous non-cavitary softtissue mass with left hilar adenopathy, displacement of broncho-vascular structures and areas of peripheral collapse
- PET-CT: Intense homogenous increased glucose uptake of the mass and increased abnormal glucose uptake in left lower paratracheal, subcarinal nodal station and left hilum (> than reactive uptake)

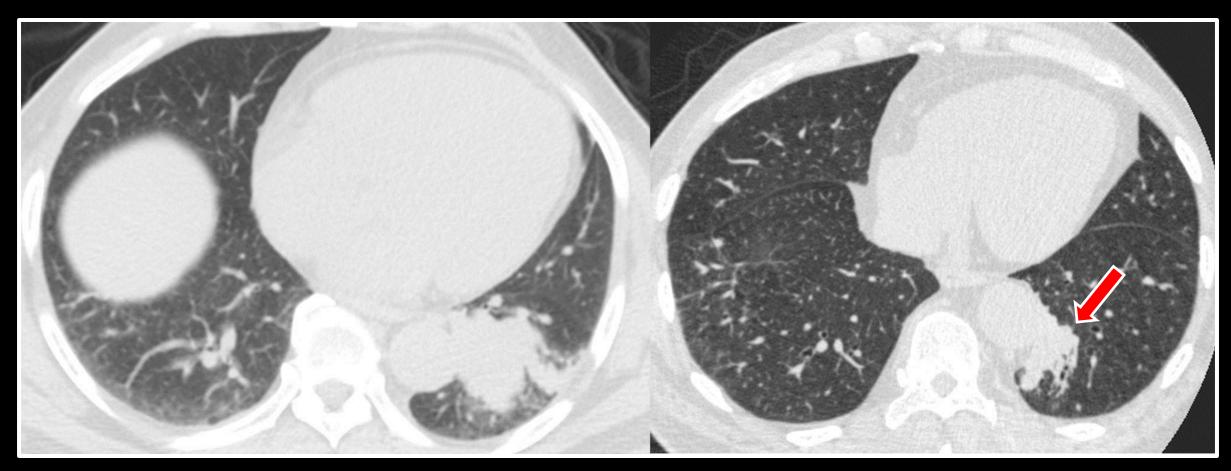
# **Differential Diagnosis**

Solitary lung mass with lymphadenopathies: many possibilities



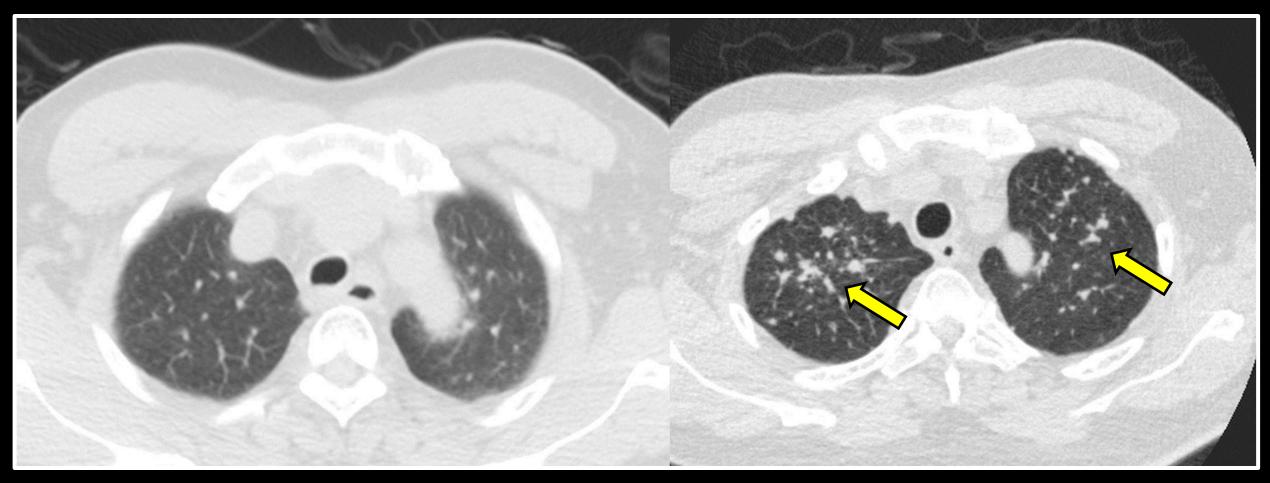
- 1. Neoplastic: Lung cancer (invasive ADK, undifferentiated neuroendocrine tumor (carcinoid), Maltoma,...
- 2. Inflammatory / Granulomatous: Sarcoidosis, IgG4 disease, GPA, Bronchocentric granulomatosis...
- 3. Infectious: Mycobacteria, Fungi

# A diagnostic procedure is performed and treatment is started



Baseline

4 months later



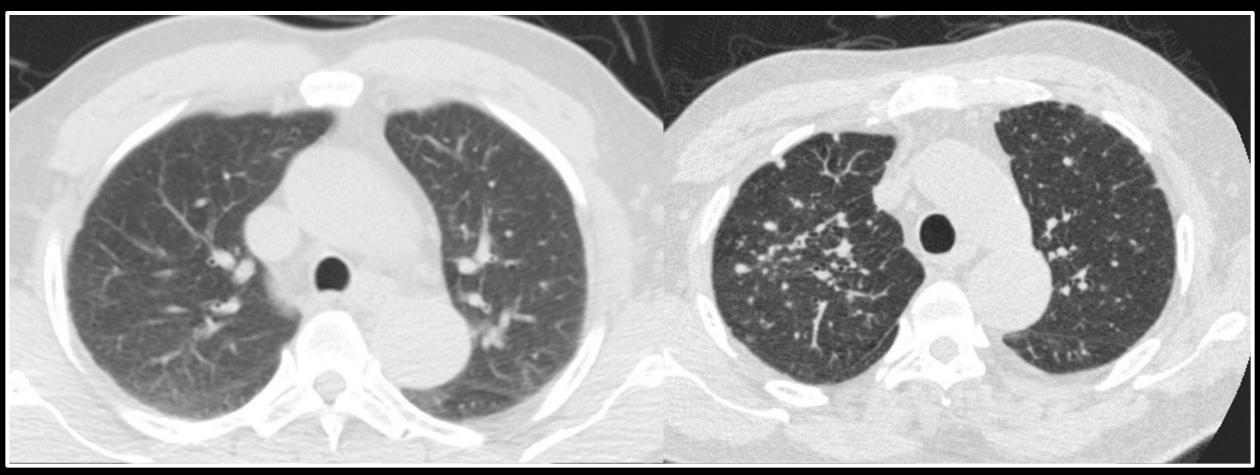
Baseline

4 months later



**Baseline** 

4 months later



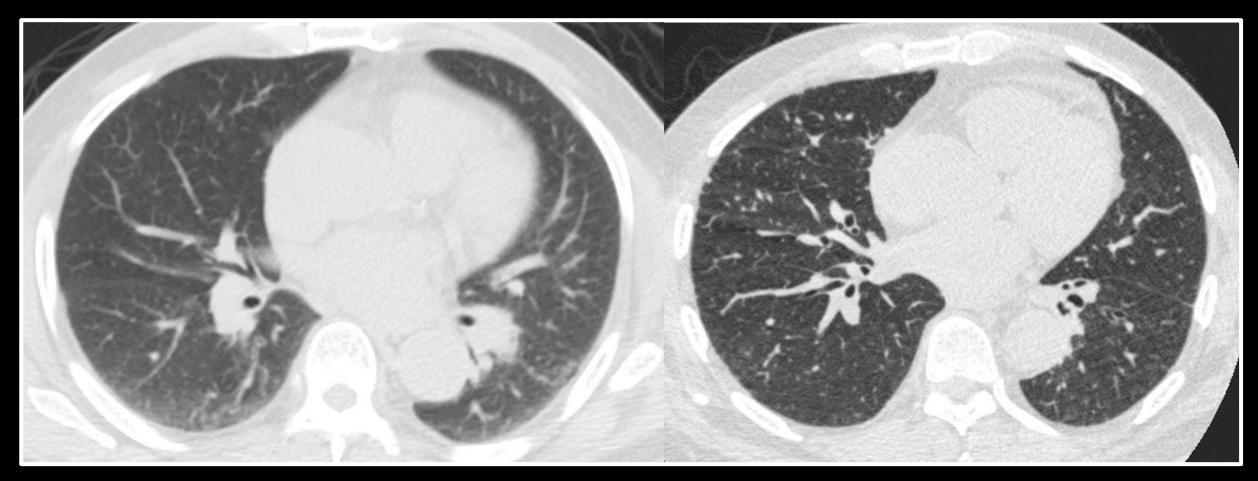
Baseline

4 months later



Baseline

4 months later

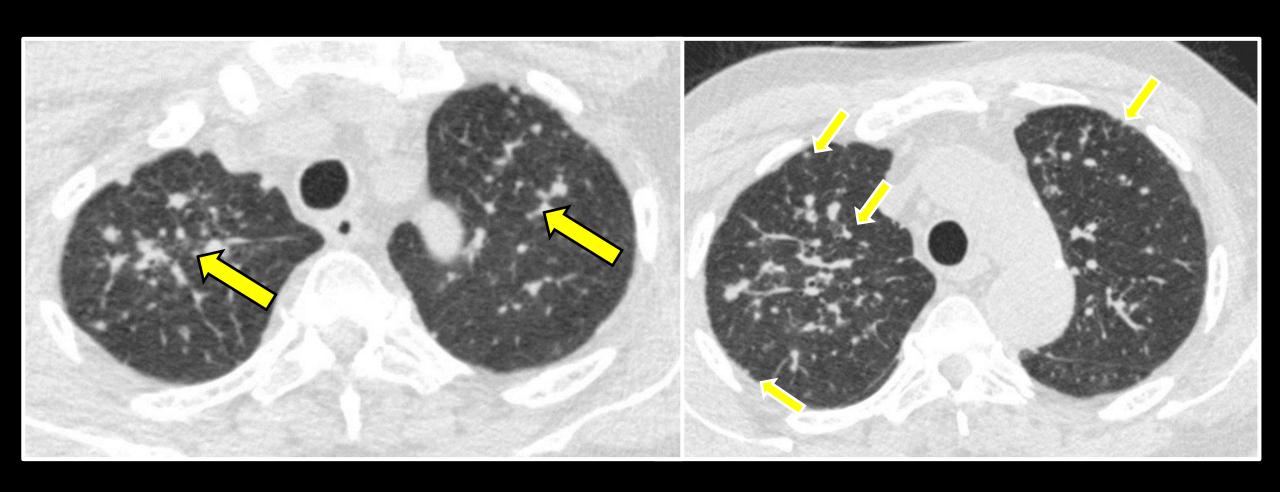


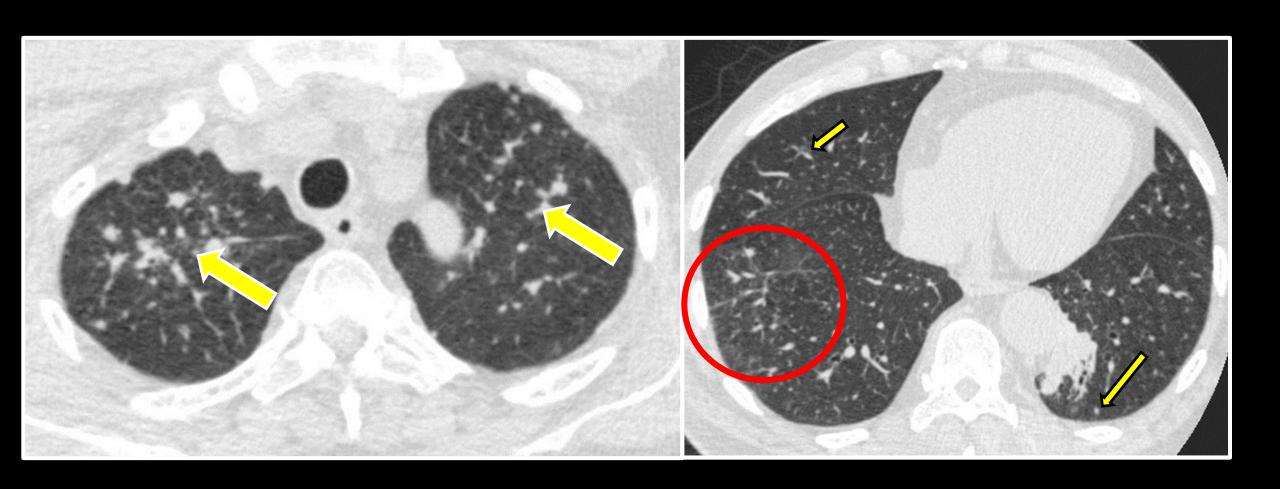
Baseline

4 months later

# **Imaging Findings**

CT: Multiple small nodules in peri-lymphatic distribution, subpleural, along the bronchovascular bundles, interlobular septum and in a symmetric and upper zone predominant distribution





# **Differential Diagnosis**

#### **Neoplastic:**

- 1. Lymphangitis carcinomatosa: ADK (breast, lung, stomach)
- 2. Lymphoproliferative disease
- 3. Pulmonary tumor thrombotic microangiopathy (PTTM): Gastric cancer is the most commonly associated malignancy

#### **Infection:**

1. Mycobacteria / Fungi

#### **Differential Diagnosis**

#### **Inflammatory / Granulomatous:**

- 1. **IgG4 related lung disease:** features are often excellent mimickers of malignancies, infections, and other immune-mediated disorders (vasculitis)
- 2. Sarcoidosis / Sarcoid reaction

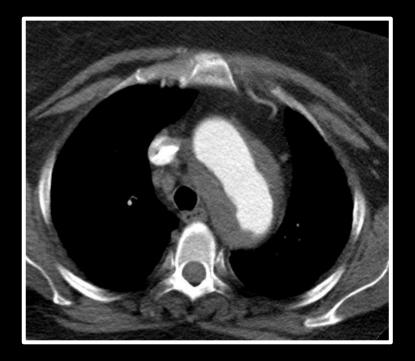
# 2018 RSNA Image Interpretation Session

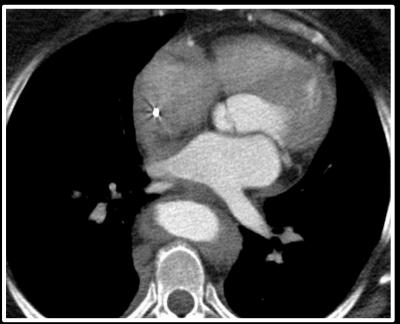
Cardiovascular

### History

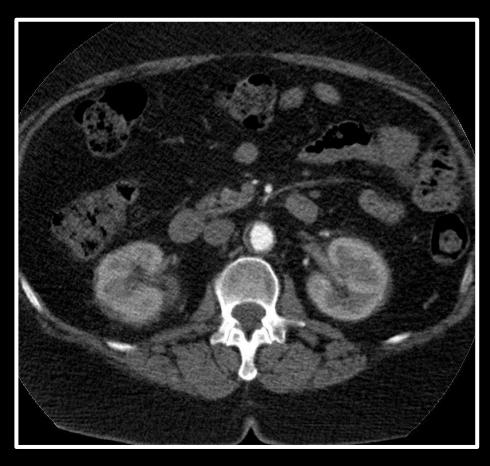
#### 57 year-old female presenting with:

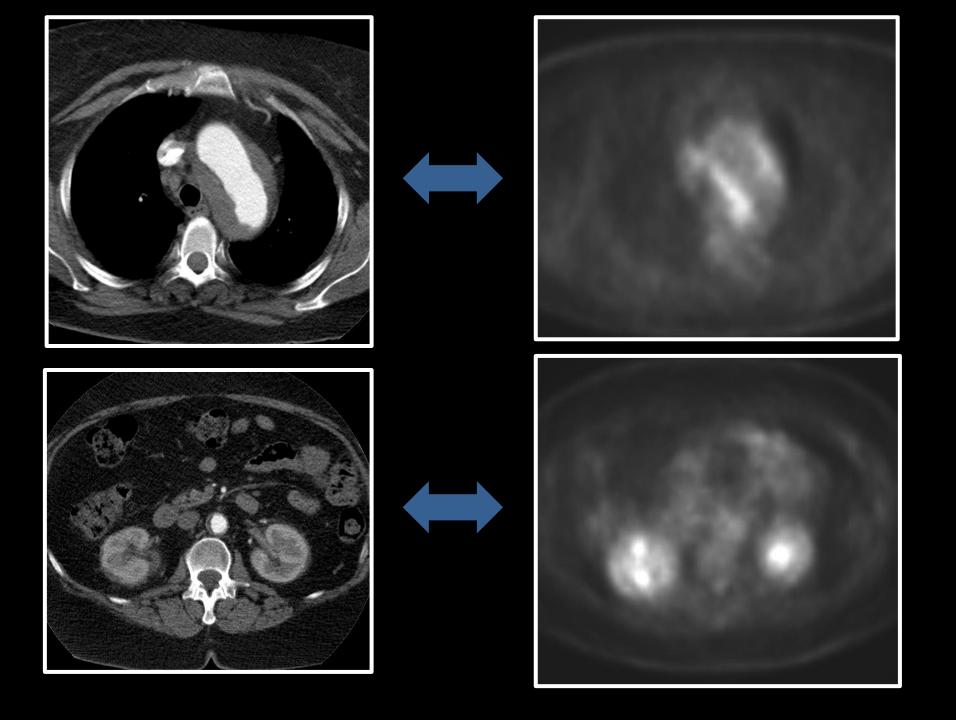
- 3-month history of daily exertional shortness of breath
- fatigue, and
- non-localized lower back pain













# Aortitis – diffuse inflammation of the aorta

- giant cell arteritis (GCA)
- Takayasu arteritis
- Cogan's syndrome
- systemic lupus erythematosus/rheumatoid arthritis
- HLA-B27 associated spondyloarthropathies (Reiter's and ankylosing spondylosis)
- ANCA-associated vasculitides (Wegener's, microscopic polyangiitis (MPA), and eosinophilic granulomatosis with polyangiitis (EGPA), previously known as Churg-Strauss)
- Behçet's disease
- sarcoidosis
- infectious (tuberculosis, syphilis, salmonella and other bacteria)
- idiopathic retroperitoneal fibrosis (Ormond's disease)/inflamed abdominal aortic aneurysm
- Erdheim-Chester
- idiopathic isolated



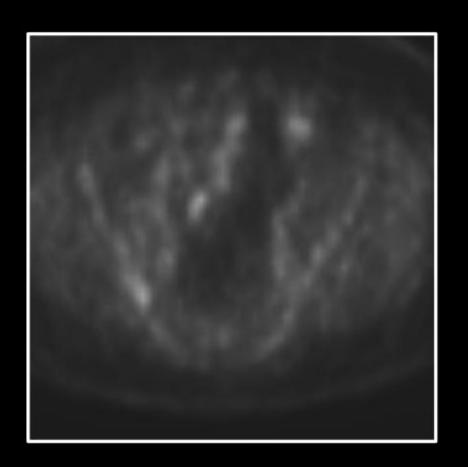
#### **Exertional SOB**

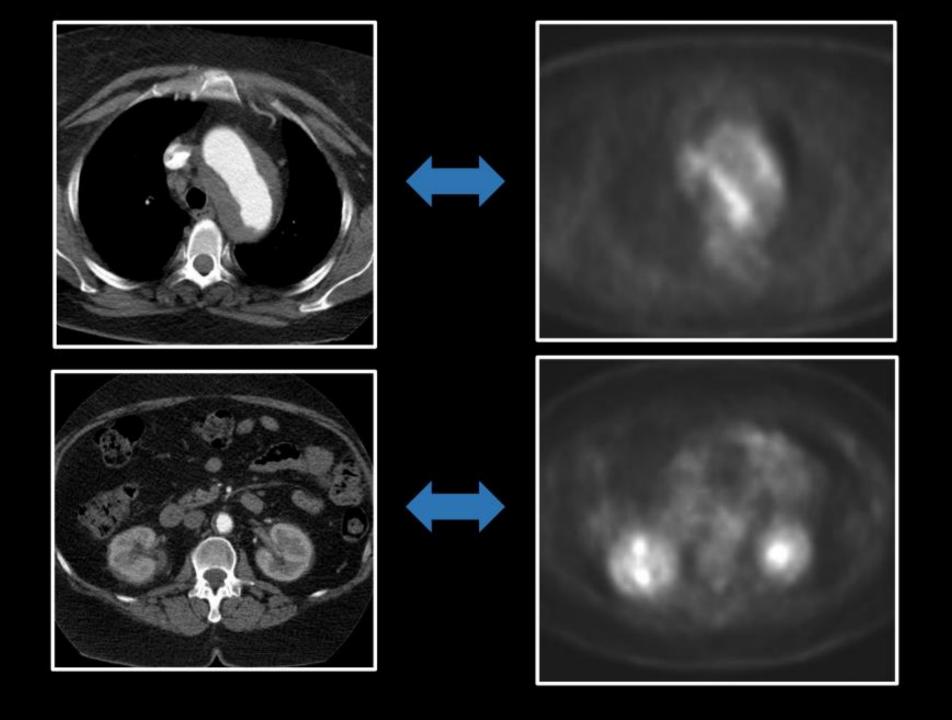
- Lung SLE, RA, sarcoid, HLA-B27
   associated, ANCA-associated vasculitides,
   Behçet's, infectious, giant cell arteritis
- Cardiac giant cell, Takaysu, Cogan's, SLE, sarcoid, HLA-B27 associated, ANCAassociated, Behçet's, infectious



#### Back pain? Sacroiliac joint, other bone involvement?



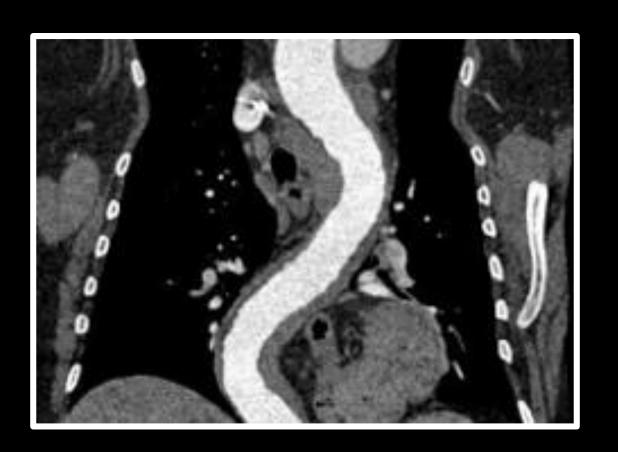


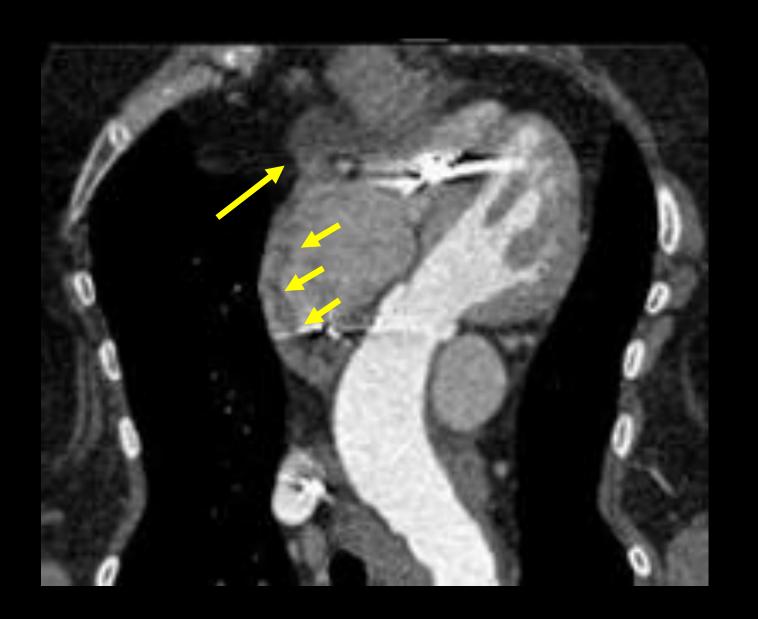




### "Coated aorta"







# 2018 RSNA Image Interpretation Session

Abdomen: GI



### History

- 66 year-old male with chronic abdominal pain
- Acute exacerbation
- Chronic medical history, includes
  - Gout
  - Hypertension
  - "Congenital emphysema": Rx inhalers and prn steroids
    - Recurrent pneumothoraces
    - Recurrent infections
    - Chronically short of breath

### History

- Relevant Surgical history
  - Bilateral carpal tunnel releases; right side 1970's
  - Bilateral finger amputations: etiology?
  - Hx of intussusception: small bowel resection

- Relevant Surgical history
  - Bilateral carpal tunnel releases; right side 1970's
  - Bilateral finger amputations: etiology?







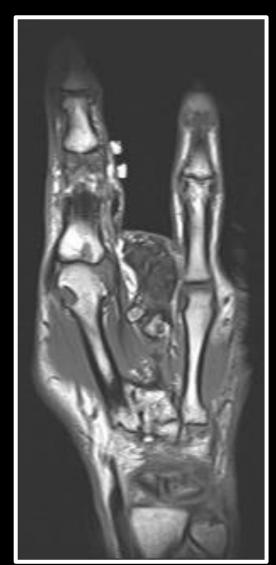
### Findings: X-ray



- Third finger amputation
- Macrodactyly (disproportionate overgrowth)
- Course trabeculation of enlarged phalanxes
- Soft tissue hypertrophy (metacarpus and around second digit)
- Abnormally calcified connective tissue

### Findings: MR

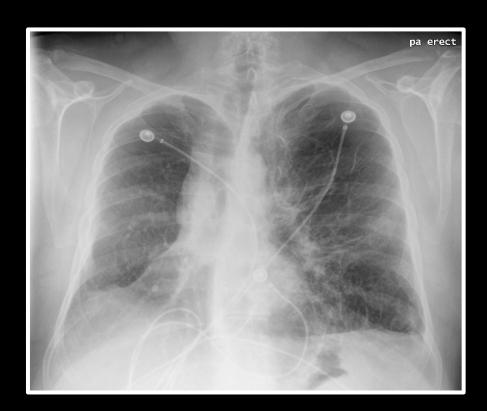
- Soft-tissue fibrotic lesion (low signal on T1/T2), i.e. fibrous hamartoma
- Bone cystic lesions



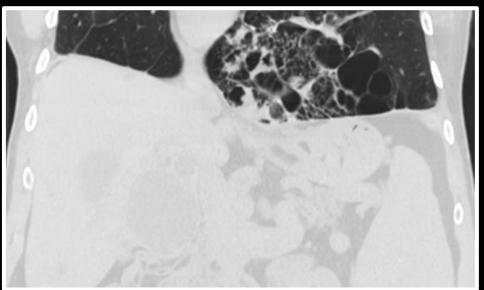


- Overgrowth of connective tissues (bone, fat)
- Progressive disease
- Onset at young age

- "Congenital emphysema": Rx inhalers and prn steroids
  - Recurrent pneumothoraces
  - Recurrent infections
  - Chronically short of breath



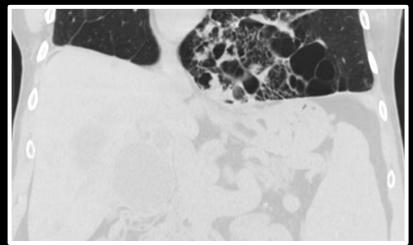






- Chest X-ray
  - Asymmetric hemithoraces; hyperinflation of left lung
  - Redistribution of blood flow to the upper lung zones with abnormal enlargement of the upper lobe vessels
  - Prominent hila with reticular and streaky densities in both perihilar areas

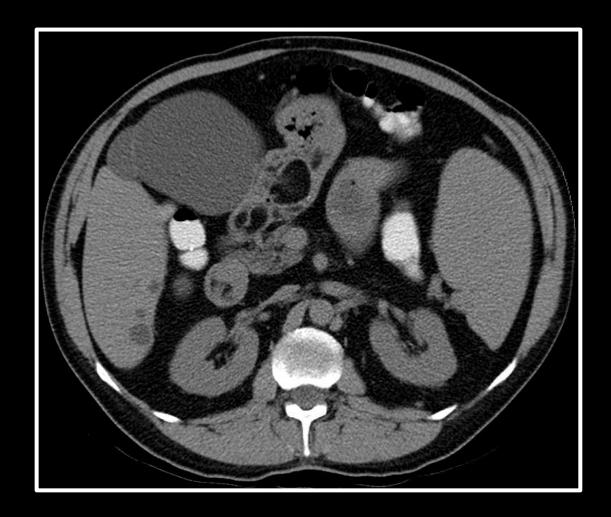




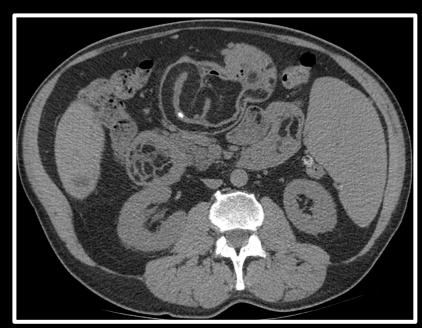
- Lung CT
  - Hyperinflation of the left lung
  - Emphysematous changes
  - Areas of scarring and cystic changes in the left lower lobe

#### History

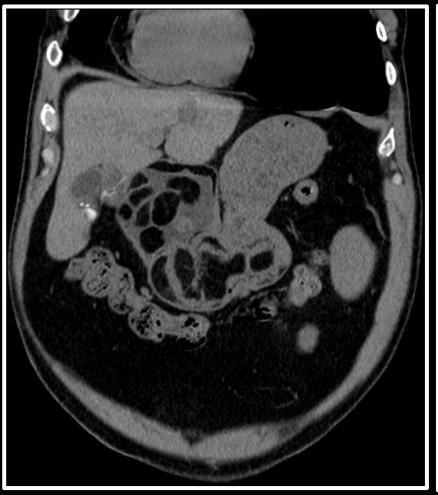
• Hx of intussusception: small bowel resection





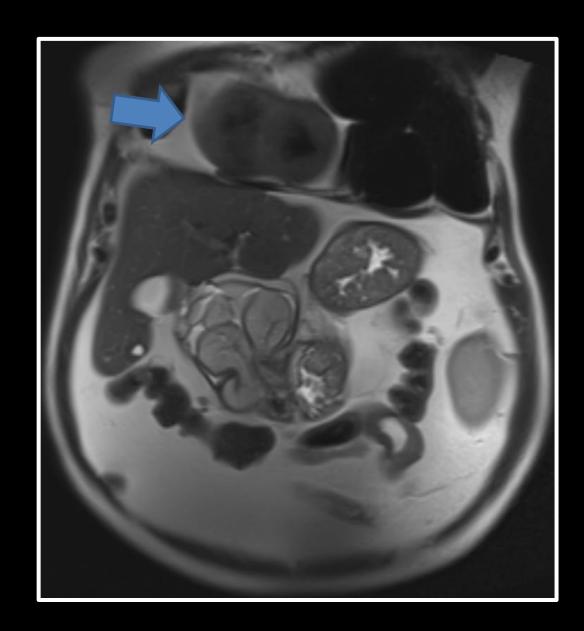


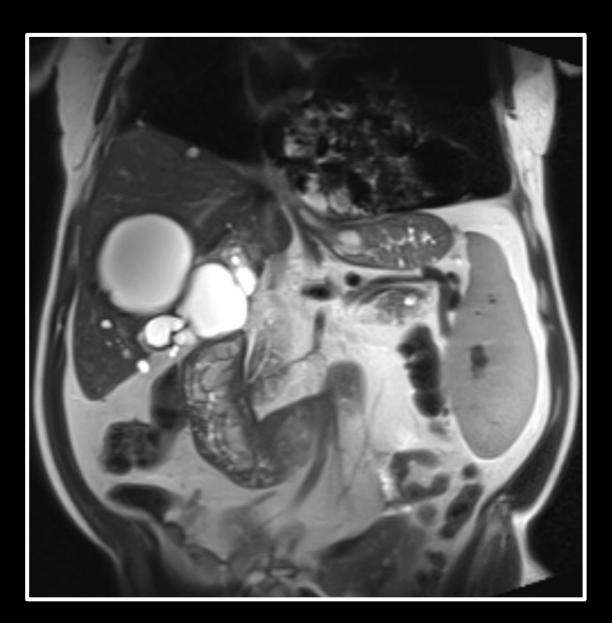


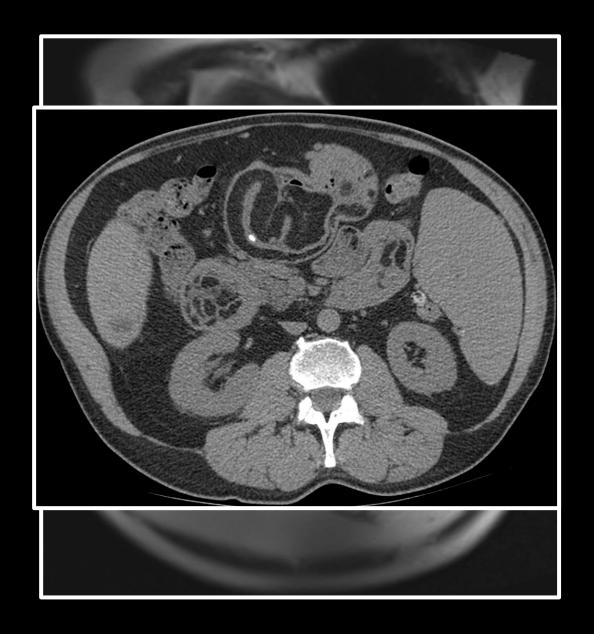












- Multiple encapsulated lipomas in the stomach, duodenum and jejunum
- Fatty overgrowth in the omentum
- Lipohypoplasia of subcutaneous fat
- Dysregulated adipose tissue
- Splenomegaly
- ?Thick wall of the right ventricle

#### DIFFERENTIAL DIAGNOSIS

Macrodystrophia lipomatosa

Multiple symmetric lipomatosis (Madelung disease)

 CLOVE (Congenital Lipomatous Overgrowth, Vascular malformations, and Epidermal nevi, Skeletal) syndrome

Proteus syndrome

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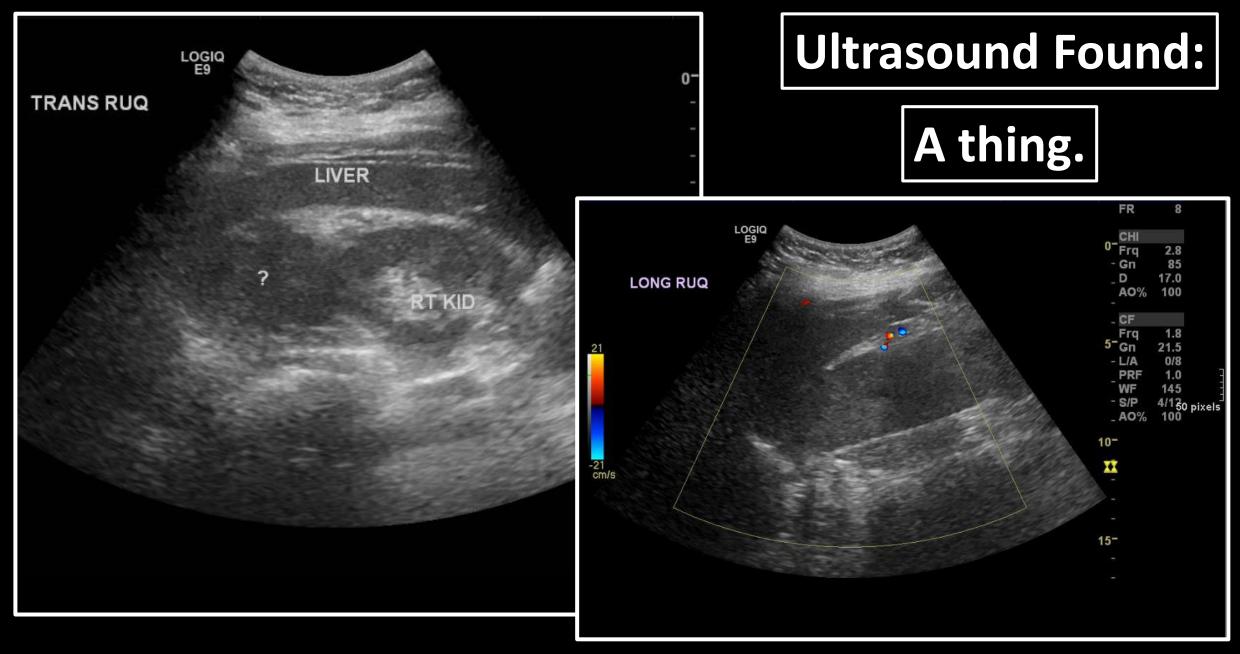
Abdomen: GU



### History

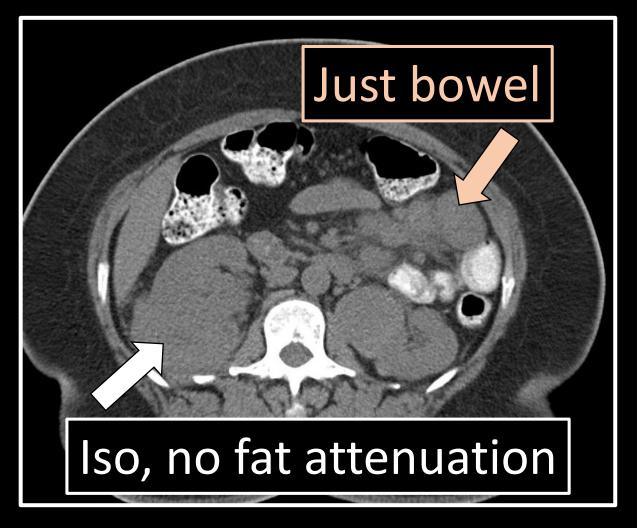
#### 63 year-old female with:

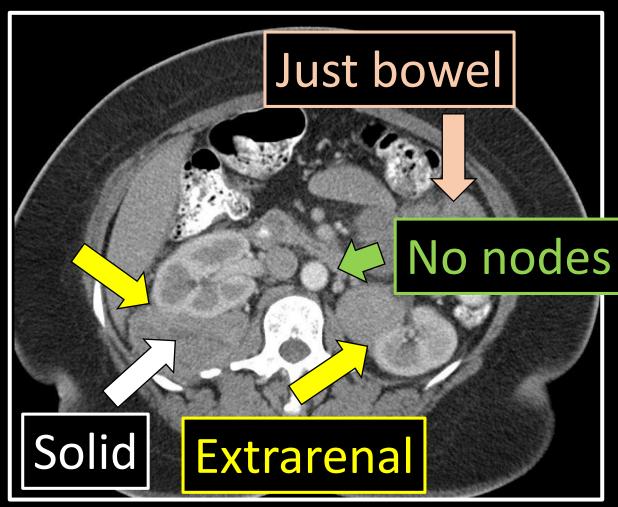
- right abdomen/flank pain
- long standing history of seizures
- 5.0 kg unintentional weight loss

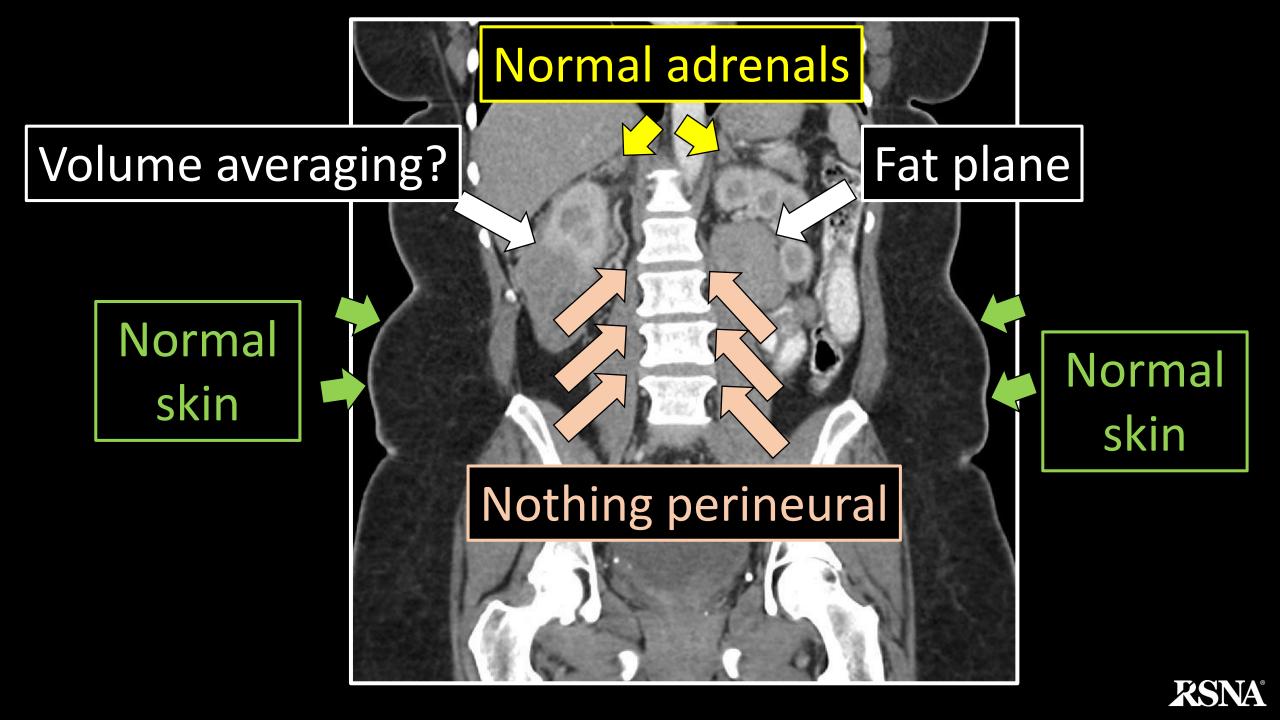


CT shows:

#### Large bilateral solid perinephric masses

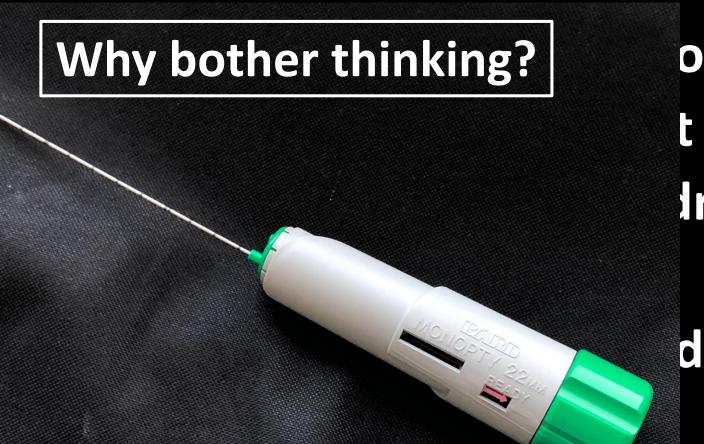






### Thought Process

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- 2. Rena
- 3. Seizi
- 4. Mult
- 5. "Gue



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### Perirenal Differential Diagnosis

#### **Perirenal**

Lymphoma or mets

Histiocytosis conditions

**Plasmacytomas** 

Paragangliomas

Extramedullary

Hematopoesis

**Erdheim Chester** 

Mass-forming IgG4



### Neuro-Oncologic Differential Diagnosis

#### **Perirenal**

NF-1

**vHL** 

Many others

**Tuberous Sclerosis** 

Some AMLs nasty: Epithelioid AMLs (aggressive)

#### Final Differential and Plan

#### Reasonable possibilities

- -- Fat-poor epithelioid AMLs in setting of TS
- --Histiocytosis (Rosai-Dorfman [non-Langerhans cell])
- --Lymphoma
- --Paragangliomas
- -- Mass-forming IgG4

#### Plan:

Look at chart (do they have TS?)
Comparisons (esp. CNS)
Metanephrines – if above (-)
Set up for biopsy

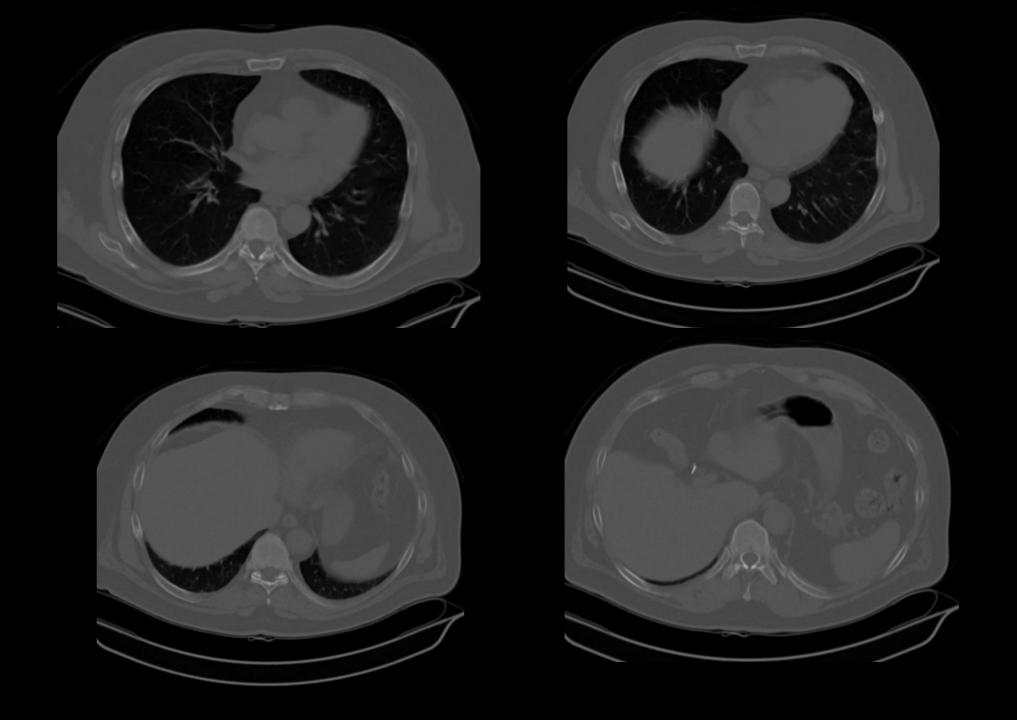
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**MSK** 

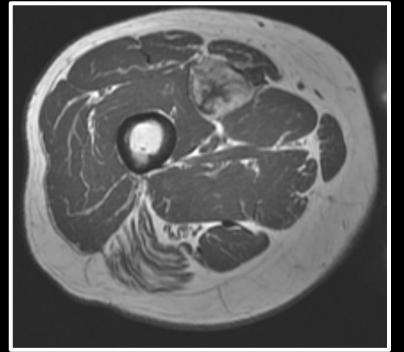


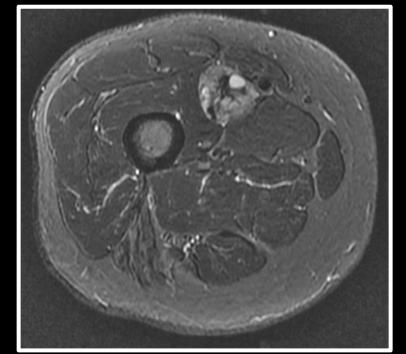
# History

- Male in his 60's
- Several years of generalized pain, fatigue, and muscle weakness with abnormal gait
- History of pubic ramus fractures and rib fractures
- Recent diagnosis of prostate cancer
  - Watchful waiting
- 2-3 year history of pain and discomfort in right thigh









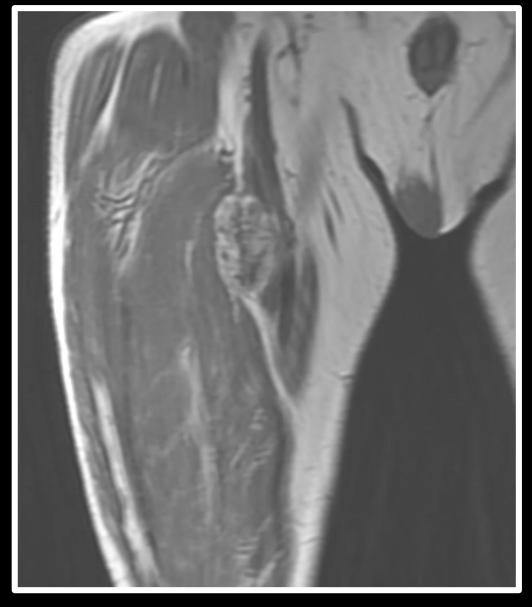
T1

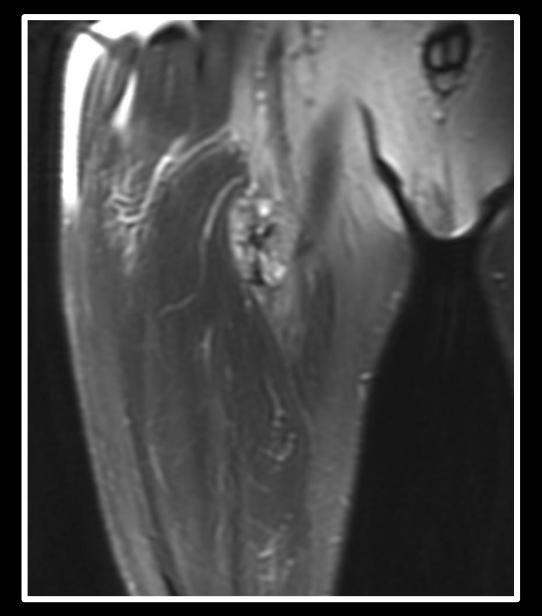


STIR

T1 with contrast





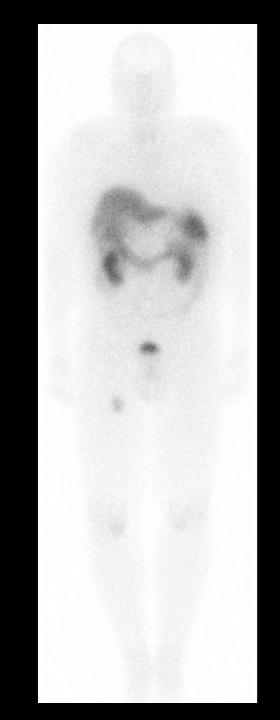


T1 STIR



## **Additional Data**

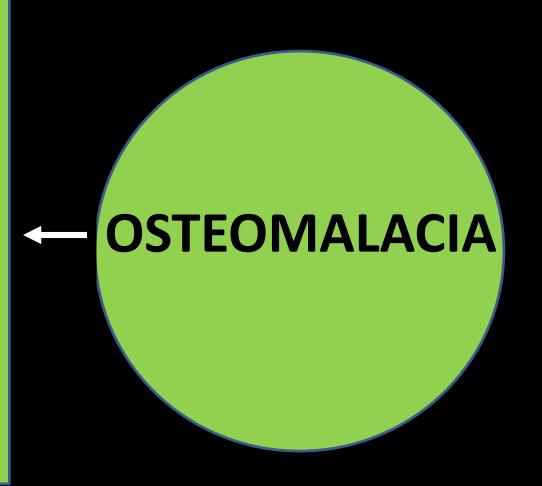
Bone density evaluation → osteopenia



KSNA®

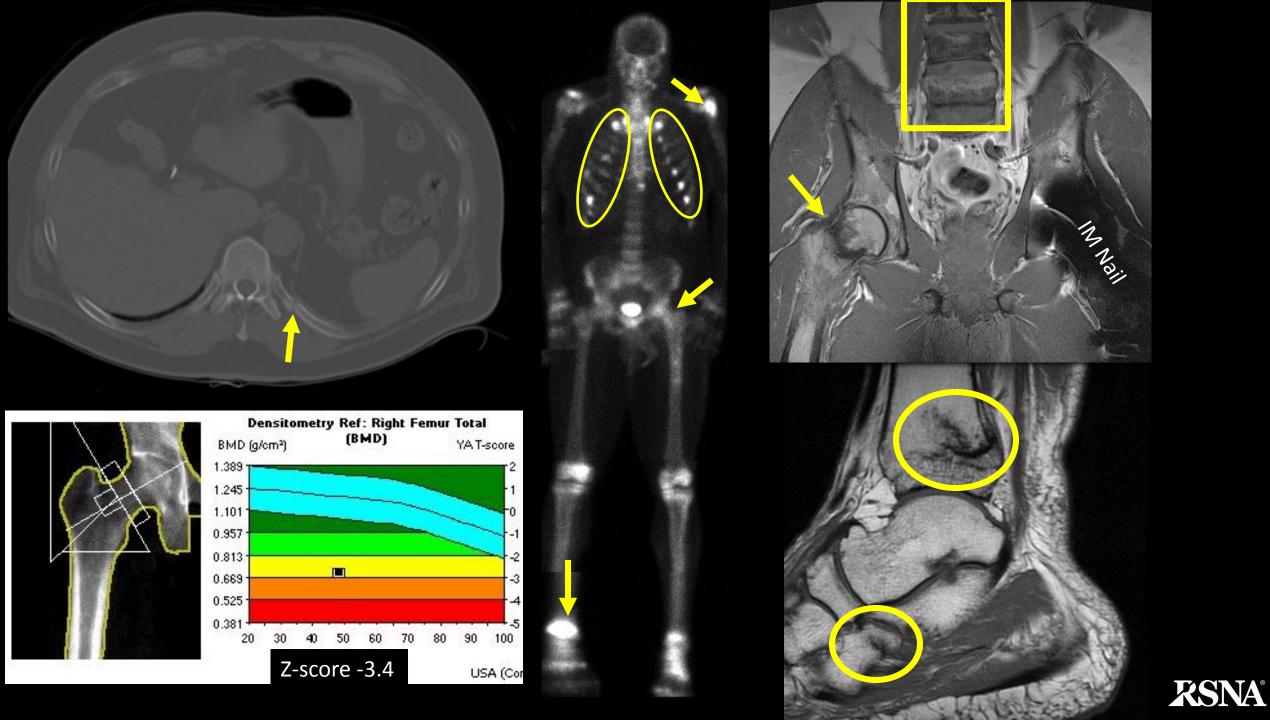
# **Clinical History**

- Vitamin D
   deficiency/liver dz
- X-linked hypophosphatemia
- Drug toxicity
- Tumor-induced



# Laboratory

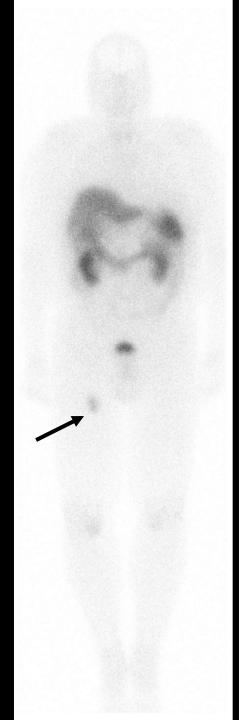
	Ca	PO <sup>4</sup>	<sup>25</sup> D	<sup>1,25</sup> D
<ul><li>Vitamin D deficiency/liver dz</li></ul>	<b>↓</b>	<b>↓</b>	<b>↓</b>	<b>↓</b>
<ul> <li>X-linked hypophosphatemia</li> </ul>	<b>+</b>	<b>↓</b>	N	N
<ul> <li>Drug toxicity</li> </ul>	N	N	N	N
• Tumor-induced	+ \		N	



#### T1 STIR T1+Gd

#### **Imaging**

- Size
- Location
- Density
- Enhancement
- \*Tumor matrix
  - Amorphous
  - Punctate



# Octreoscan (Octreotide, <sup>111</sup>In-pentetreotide)

- Tumors high expression somatostatin receptors
  - -Neuroendocrine tumors
  - -Adrenal medullary tumors
  - -Merkel cell tumor of skin
  - -Pituitary adenoma
  - -Small-cell lung carcinoma
  - -Phosphaturic mesenchymal tumors

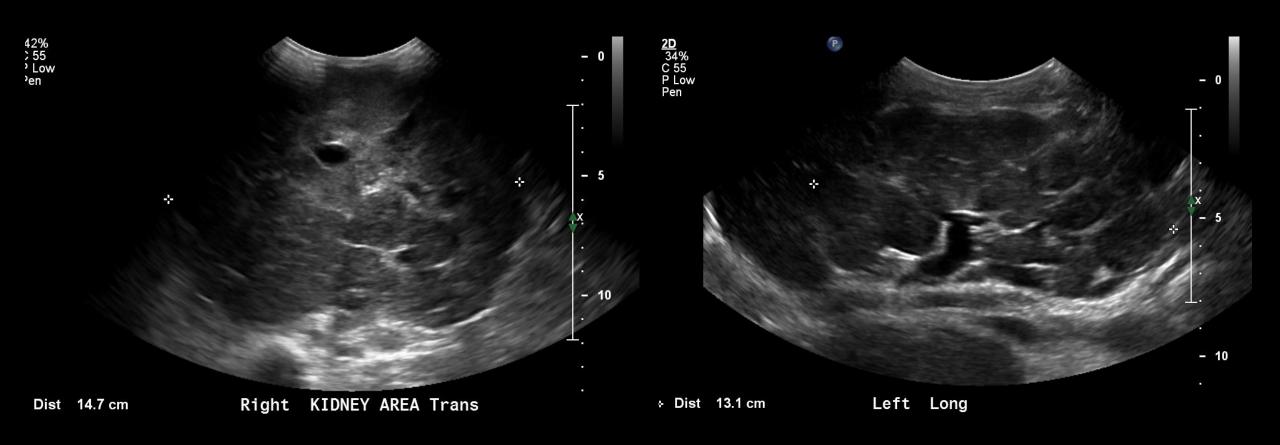
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Peds

# History

- 2 year old girl who originally presented at an outside practice
  - "renal abnormalities"
  - abdominal distension
- Followed by renal ultrasound

#### Renal Ultrasound at 2 Years of Age

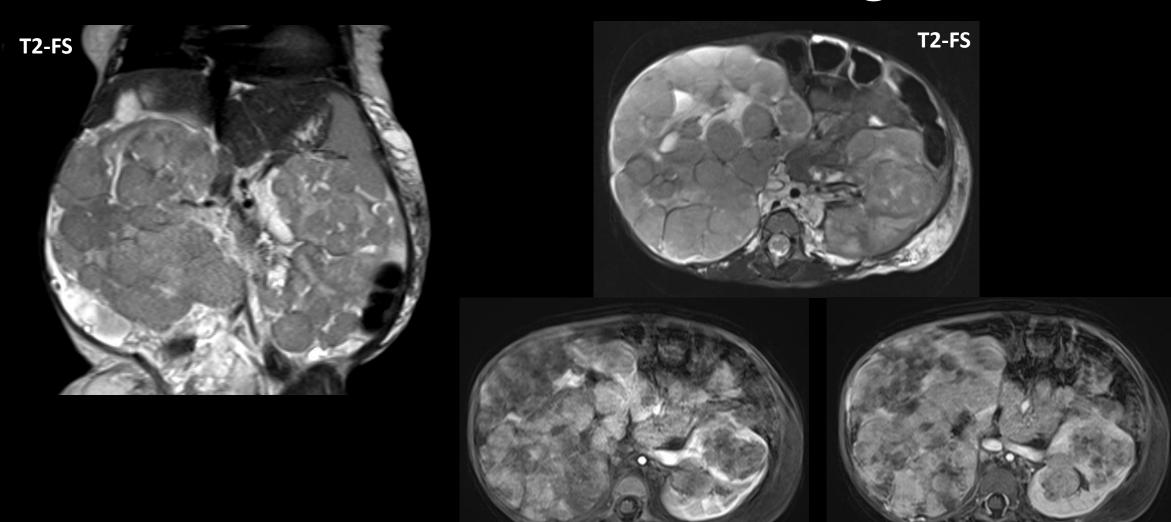




## History

- 2 year old girl who originally presented at an outside practice
  - "renal abnormalities"
  - abdominal distension
- Followed by renal ultrasound
- At 26 months of age:
  - enlarged right kidney; mass suspected
  - MRI performed

### MRI at 26 Months of Age

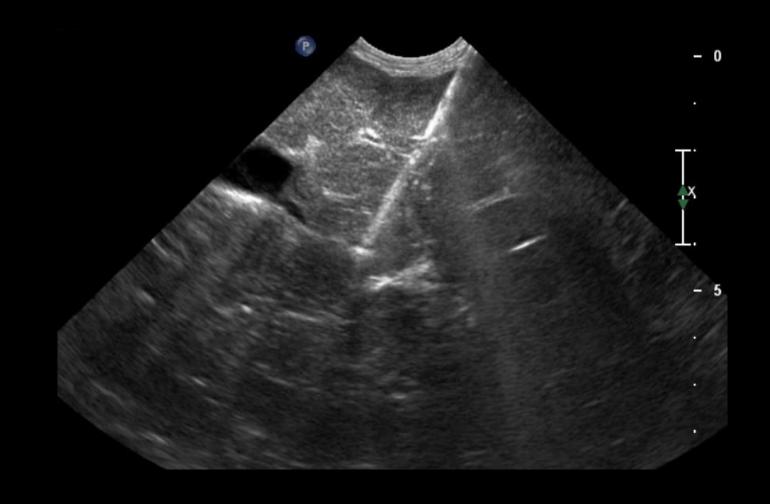


**T1+C Arterial Phase** 

T1+C PV Phase



# Subsequently, US-Guided Right Renal Biopsies: eventual path diagnosis was "Wilms"



#### **Findings**

 Young child with asymmetric nephromegaly and multiple solid masses replacing normal renal parenchyma with loss of cortico-medullary distinction



- Nephroblastomatosis +/- Wilms tumor >>>>>> lymphoma, metastases
- Very difficult to distinguish hyperplastic nephrogenic rests from Wilms tumor on biopsy
- It can't be that straightforward, can it?

#### **Additional Findings**

• Fluid-filled locules within left abdominal wall, para-aortic/para-renal retroperitoneum, and right pelvis; ? thoracic and right hip lipomatosis; otherwise, decreased adipose tissue



- Overgrowth syndrome with veno-lymphatic vascular malformations
- Isolated hemihypertrophy, Beckwith-Wiedemann, Perlman, Soto, and Simpson-Golabi-Behmel syndromes are associated with overgrowth, nephrogenic rests and Wilms tumor
- So, what could be the diagnosis in this case?



### Deepest Gratitude



Neuro:

Michael Malinzak, MD



**Breast:**Jay Baker, MD



Abd GI: Rendon Nelson, MD



**Thoracic:**Page McAdams MD



MSK: Lee Cothran, MD



CVI & Abd GU:
Daniele Marin, MD



Peds:
Gary Schooler, MD



# Thank You