

# AMSER Case of the Month

## November 2021

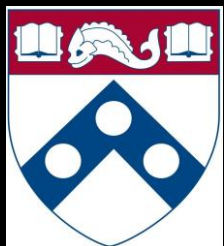
### 30-year-old Female with Nausea, Vomiting, and Abdominal Pain

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# Patient Presentation

- **HPI:** 30 y.o. F presenting with intermittent nausea, vomiting, and diffuse abdominal pain for the past week. She has also been experiencing paroxysmal chills and cough with SOB for the past few days.
- **PMH:** asthma
- **PSH:** none
- **FH:** stroke in uncle
- **SH:** no recent travel
- **Allergies:** no known allergies
- **PE:** 100.1 F, HR 117 , BP 134/72 General-no acute distress; HEENT- mucous membranes moist; Neck-supple; Respiratory- lungs clear to auscultation bilaterally; Cardiovascular- tachycardic with no murmurs, rubs, or gallops; Abdominal - soft with diffuse tenderness to palpation; Skin- no rashes or skin lesions; Musculoskeletal- no tenderness or swelling; Neurologic- sensation to touch intact and CNII-XII intact
- **Pertinent Labs:** WBC wnl, Covid +, Mono-, Echinococcus +

# What Imaging Should We Order?

American College of Radiology  
ACR Appropriateness Criteria®  
Acute Nonlocalized Abdominal Pain

**Variant 1:** Acute nonlocalized abdominal pain and fever. No recent surgery. Initial imaging.

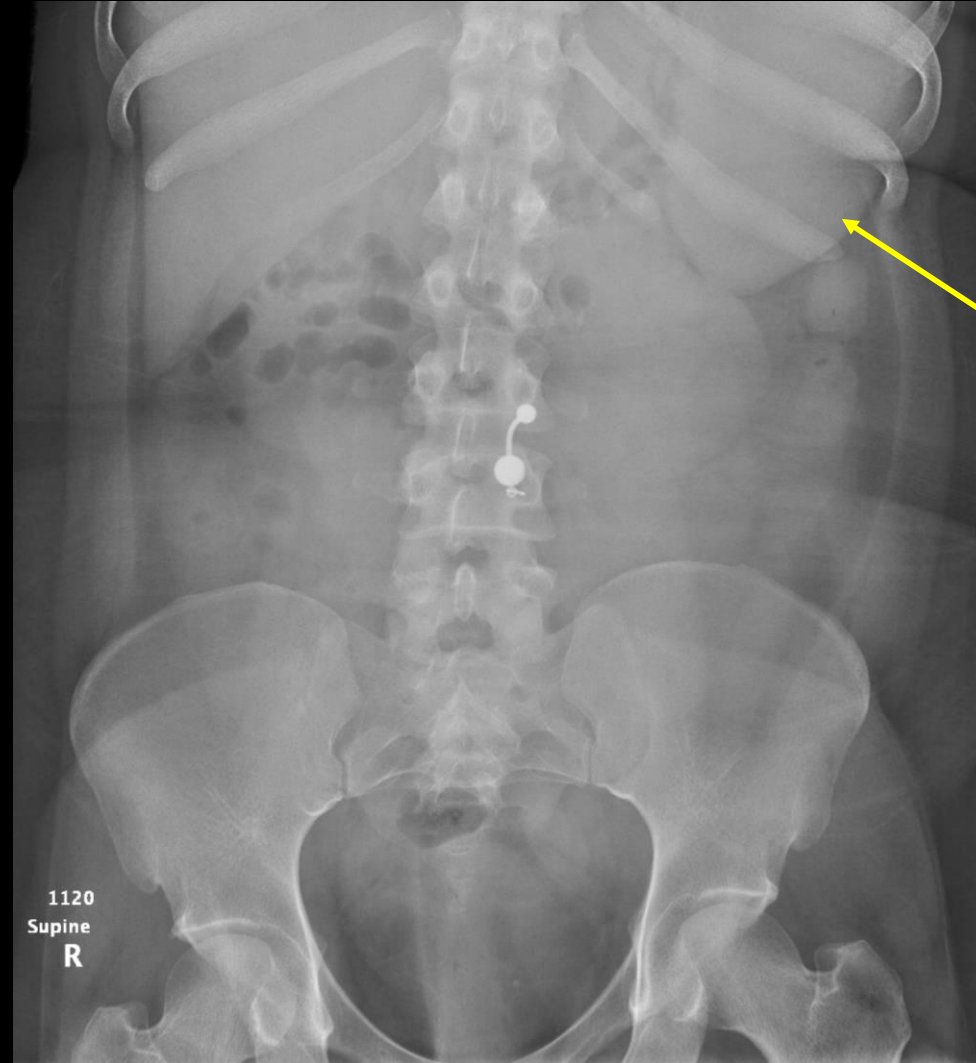
Procedure	Appropriateness Category	Relative Radiation Level
CT abdomen and pelvis with IV contrast	Usually Appropriate	⊕⊕⊕
MRI abdomen and pelvis without and with IV contrast	May Be Appropriate	○
US abdomen	May Be Appropriate	○
CT abdomen and pelvis without IV contrast	May Be Appropriate	⊕⊕⊕
MRI abdomen and pelvis without IV contrast	May Be Appropriate	○
CT abdomen and pelvis without and with IV contrast	May Be Appropriate	⊕⊕⊕⊕
Radiography abdomen	May Be Appropriate	⊕⊕
FDG-PET/CT skull base to mid-thigh	Usually Not Appropriate	⊕⊕⊕⊕
WBC scan abdomen and pelvis	Usually Not Appropriate	⊕⊕⊕⊕
Nuclear medicine scan gallbladder	Usually Not Appropriate	⊕⊕
Fluoroscopy contrast enema	Usually Not Appropriate	⊕⊕⊕
Fluoroscopy upper GI series with small bowel follow-through	Usually Not Appropriate	⊕⊕⊕

These imaging modalities were ordered by the provider

# Radiography Abdomen (Findings Unlabeled)

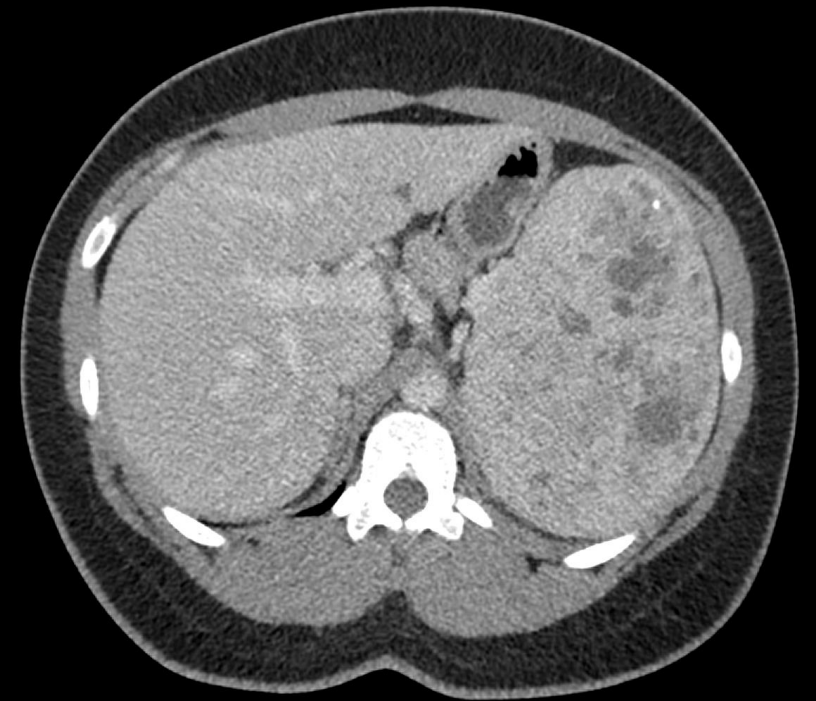
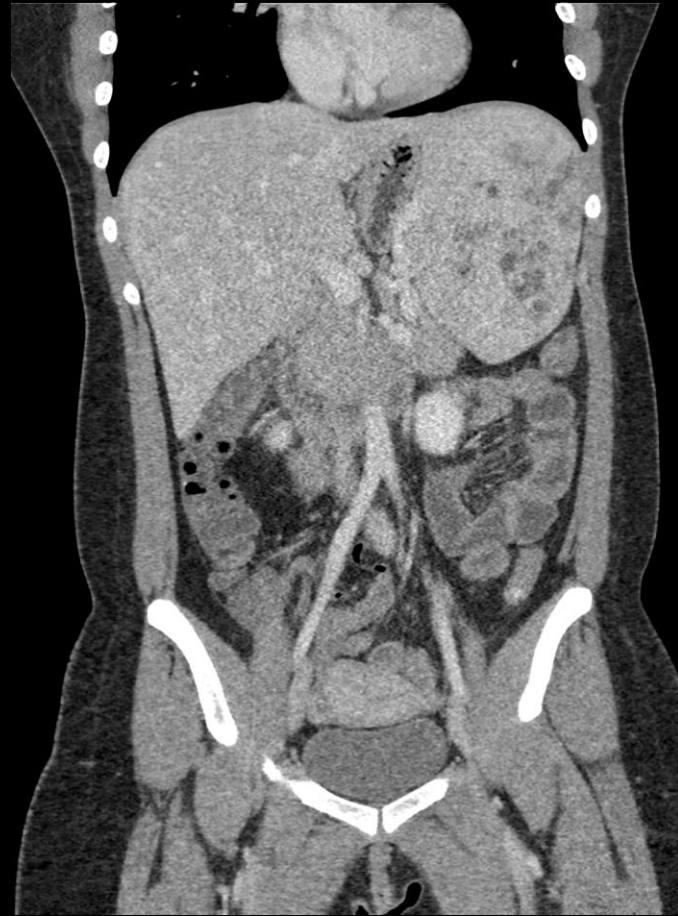
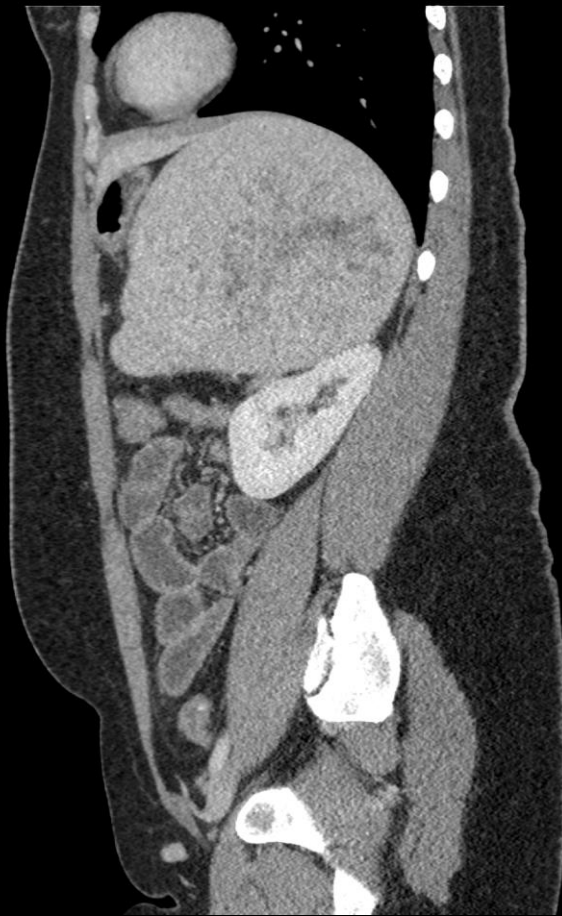


# Radiography Abdomen (Findings Labeled)

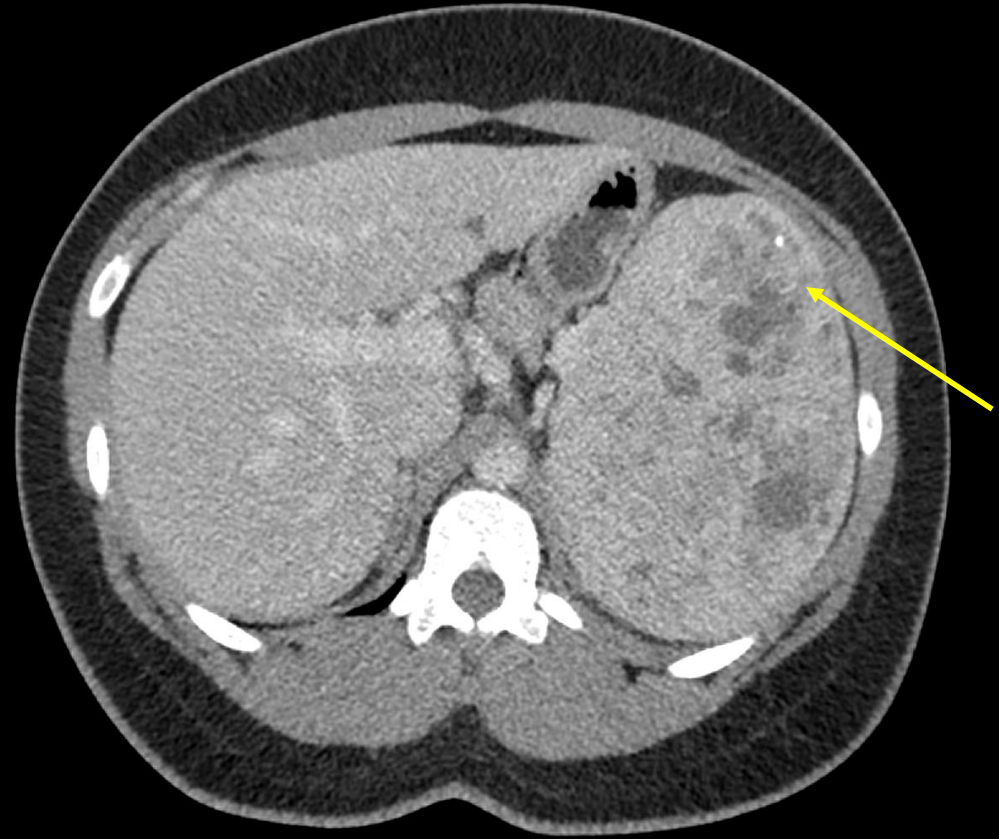
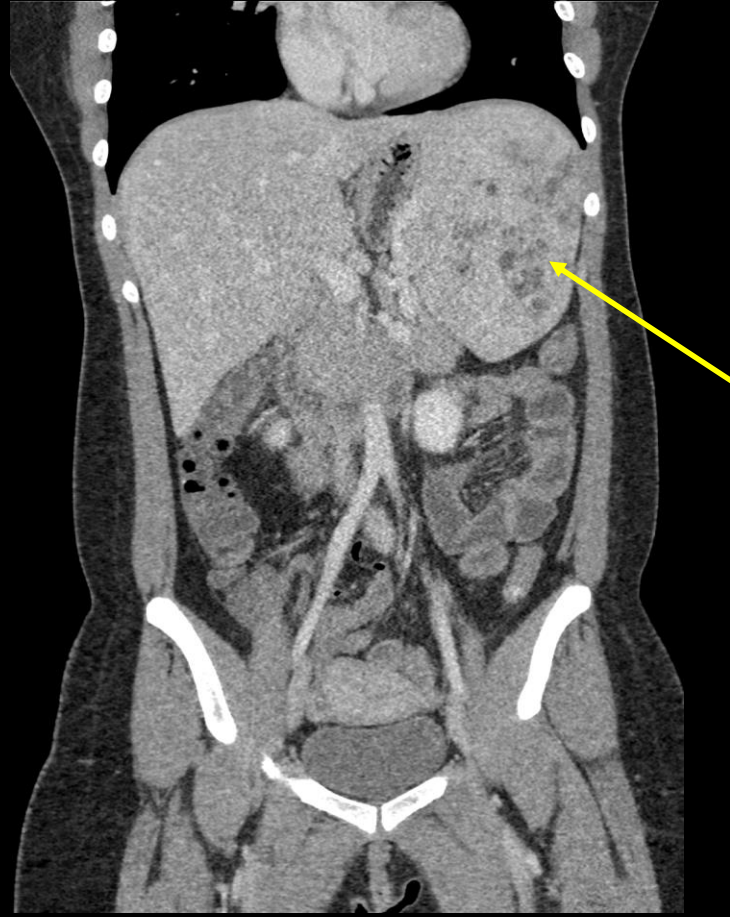
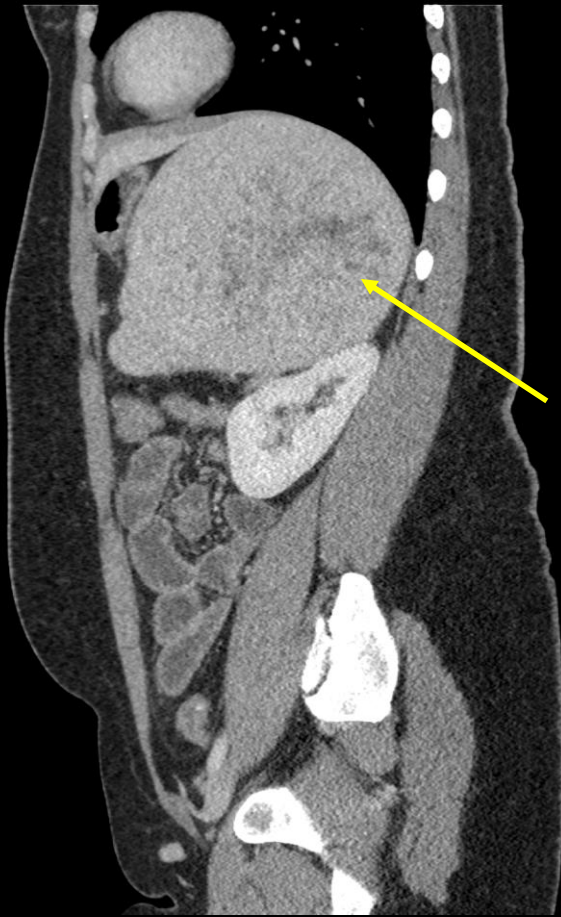


KUB  
demonstrates  
enlarged spleen

# CT w/Contrast (Findings Unlabeled)

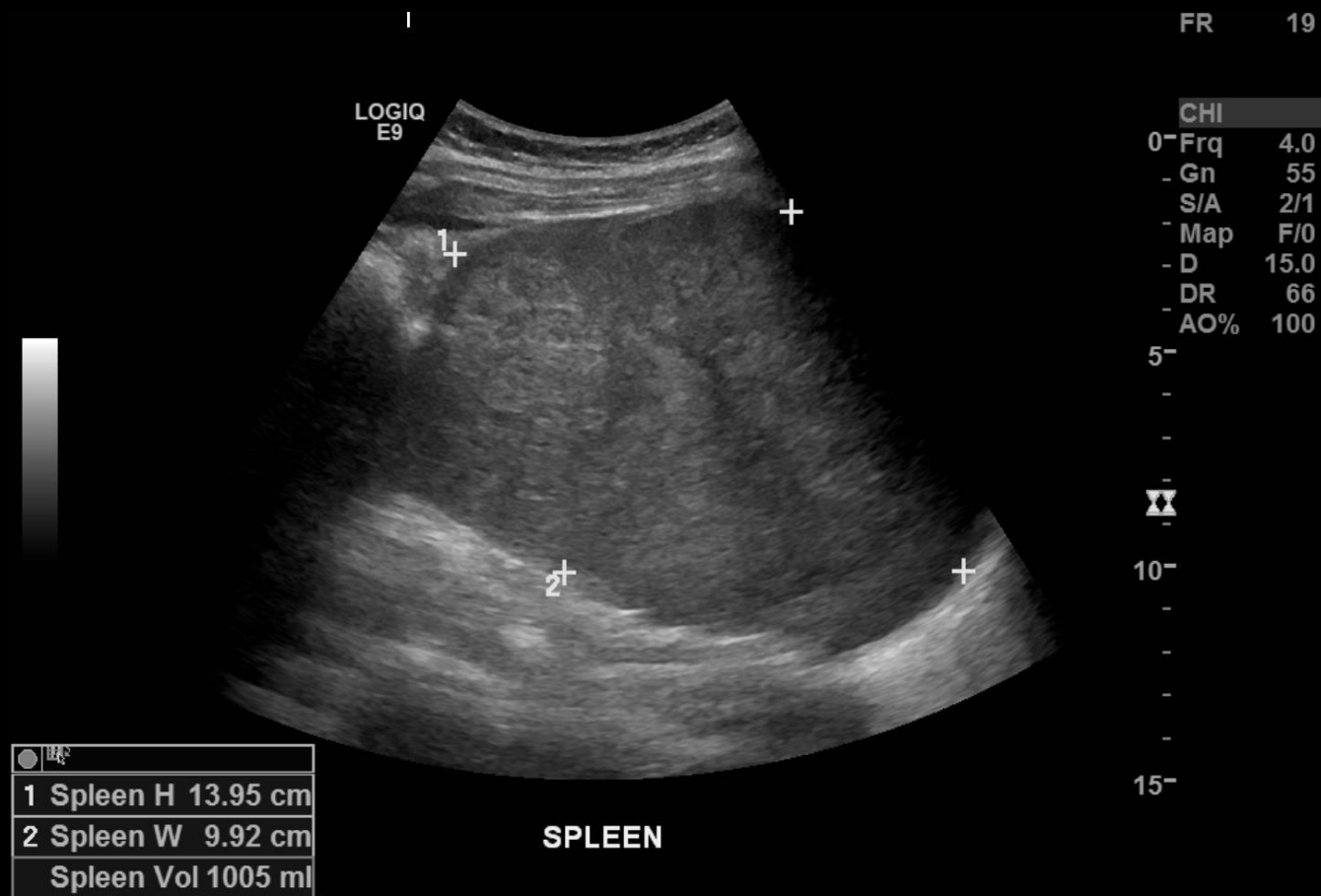


# CT w/Contrast (sagittal, coronal and axial images) (Findings Labeled)



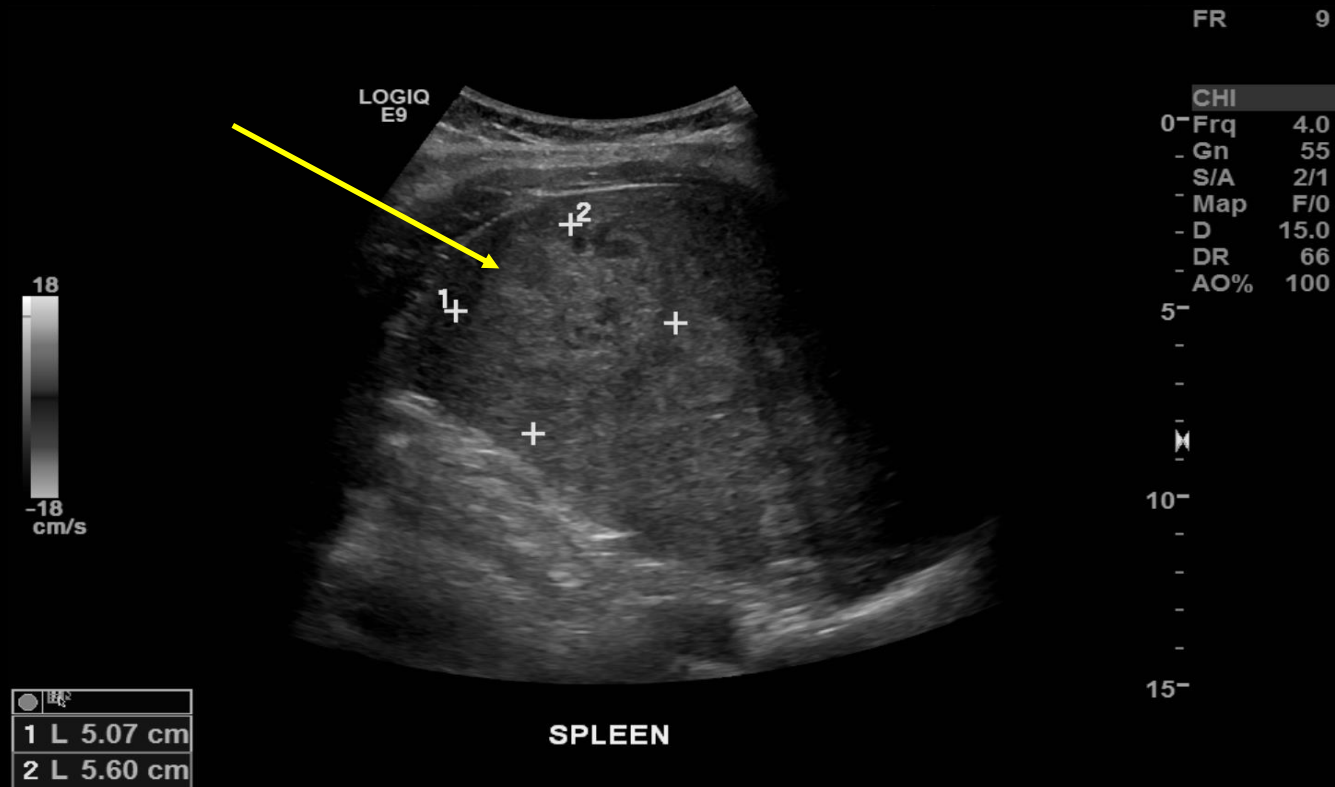
Markedly enlarged **spleen** with a multiloculated solid and cystic mass spanning the entirety of the spleen and demonstrating scattered punctate calcifications.

# US Abdomen (Findings Unlabeled)





# US Abdomen (Findings Labeled)



Spleen is heterogenous and enlarged, measuring 13.9 x 14.0 x 9.9 cm, with volume of 1005mL. There are complex, mostly solid, partially cystic splenic masses with the **largest discrete lesion** delineated (+) and measuring 5.6 x 5.1 x 4.9 cm.

## Final Diagnosis:

Littoral Cell Angioma of the Spleen

# Case Discussion

- Littoral cell angioma of the spleen is a rare, benign primary vascular tumor arising from the littoral cells lining the splenic red pulp sinuses
- **Epidemiology**
  - Rare with few cases documented in literature
  - Occurs in all ages but most cases are reported in middle-aged adults
  - Not associated with gender
- **Clinical Findings**
  - Often found incidentally but may present with abdominal pain or splenomegaly on physical exam
  - Laboratory results may show signs of hypersplenism such as anemia or thrombocytopenia

# Case Discussion

- **Imaging Differential Diagnosis**

- metastases
- lymphoma
- sarcoidosis
- abscesses
- other primary splenic vascular tumors
  - Benign - hemangioma, hamartoma, hemangioendothelioma
  - Malignant - angiosarcoma

- **Imaging Findings**

- CT: hypoattenuating masses with progressive homogeneous enhancement, later than normal splenic parenchyma
- MR: T1 and T2 – hypointense masses (due to hemosiderin)
- US: heterogeneous echotexture; variable vascularity

# Case Discussion

## Comparative Imaging Findings:

### Littoral Cell Angioma

#### CT

- Hypoattenuating masses
- Progressive homogenous contrast enhancement

#### MRI

- T1 and T2- Hypointense

#### US

- Heterogeneous echotexture
- Variable vascularity

### Splenic Hemangioma

#### CT

- Hypoattenuating masses
- Centripetal enhancement

#### MRI

- T1- Iso to hypointense to splenic parenchyma
- T2- Hyperintense to splenic parenchyma

#### US

- Homogeneous echotexture
- Predominantly hyperechoic

### Angiosarcoma

#### CT

- Multiple nodular masses of heterogenous hypoattenuation
- Heterogenous enhancement

#### MRI

- T1 and T2- Nodular, hypointense to splenic parenchyma

#### US

- Heterogeneous echotexture (cystic and solid components)
- Increased vascularity on doppler

### Lymphoma

#### CT

- Iso attenuating masses
- Hypoenhancement (best appreciated in late venous phase)

#### MRI

- T1 and T2- Iso to hypointense compared to splenic parenchyma

#### US

- Homogeneous echotexture
- Hypoechoic

### Abscess

#### CT

- Centrally hypoattenuating lesions
- Peripheral enhancement

#### MRI

- T1- Hypointense (increased signal with proteinaceous content)
- T2-Hyperintense

#### US

- Heterogeneous echotexture
- Ranges from predominantly hypoechoic with some internal echoes to hyperechoic

# Case Discussion

- **Diagnosis and Treatment:**
  - Splenectomy performed for:
    - symptom relief
    - histological diagnosis to differentiate benign, littoral cell angioma from malignant, littoral cell angiosarcoma and from littoral cell hemangioendothelioma
  - Pathology: multiple masses composed of numerous anastomosing vascular channels filled with blood and lined with tall, hemophagocytic, endothelial cells
  - Immunophenotype: Vascular endothelial markers (CD31, CD34, and factor VIII) and Histiocytic markers (CD68)
  - Littoral cell angioma has been associated with extra-splenic, visceral organ malignancies and Crohn's Disease, so patients should be assessed accordingly
  - Massive splenomegaly (weight  $\geq$  1500gm) has been associated with malignancy requiring post-splenectomy patients to undergo surveillance for recurrence
- **Our case:** Patient experienced gastric outlet obstruction requiring splenectomy. Pathology of the lesion showed co-expression of CD31, CD68, and CD163

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