AMSER Case of the Month December 2021

24-year-old woman with diffuse abdominal pain

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Patient Presentation

- HPI: 24-year-old woman with no significant PMH or PSH presented to the emergency department with a five-day history of diffuse abdominal, suprapubic, and back pain that was sharp in nature and gradually worsened from 7/10 to 10/10. The pain worsened with sudden movements and was associated with nausea and decreased oral intake. Urine pregnancy test was negative.

- Vitals: T 97.7 F, HR 111, RR 18, BP 107/67, SpO2 100%

- PE: All abdominal quadrants tender to palpation; bowel sounds present. Pelvic: purulence in vaginal vault with putrid smell and cervical motion tenderness.
- Pertinent Labs: WBC 11.5

What Imaging Should We Order?

Select the applicable ACR Appropriateness Criteria

Variant 4: **Acute nonlocalized abdominal pain. Not otherwise specified. Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
CT abdomen and pelvis with IV contrast	Usually Appropriate	☢☢☢
CT abdomen and pelvis without IV contrast	Usually Appropriate	☢☢☢
MRI abdomen and pelvis without and with IV contrast	Usually Appropriate	O
US abdomen	May Be Appropriate	O
MRI abdomen and pelvis without IV contrast	May Be Appropriate	O
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 upper GI series with small bowel follow-through	Usually Not Appropriate	☢☢☢
Fluoroscopy contrast enema	Usually Not Appropriate	☢☢☢

This imaging modality was ordered by the ER physician

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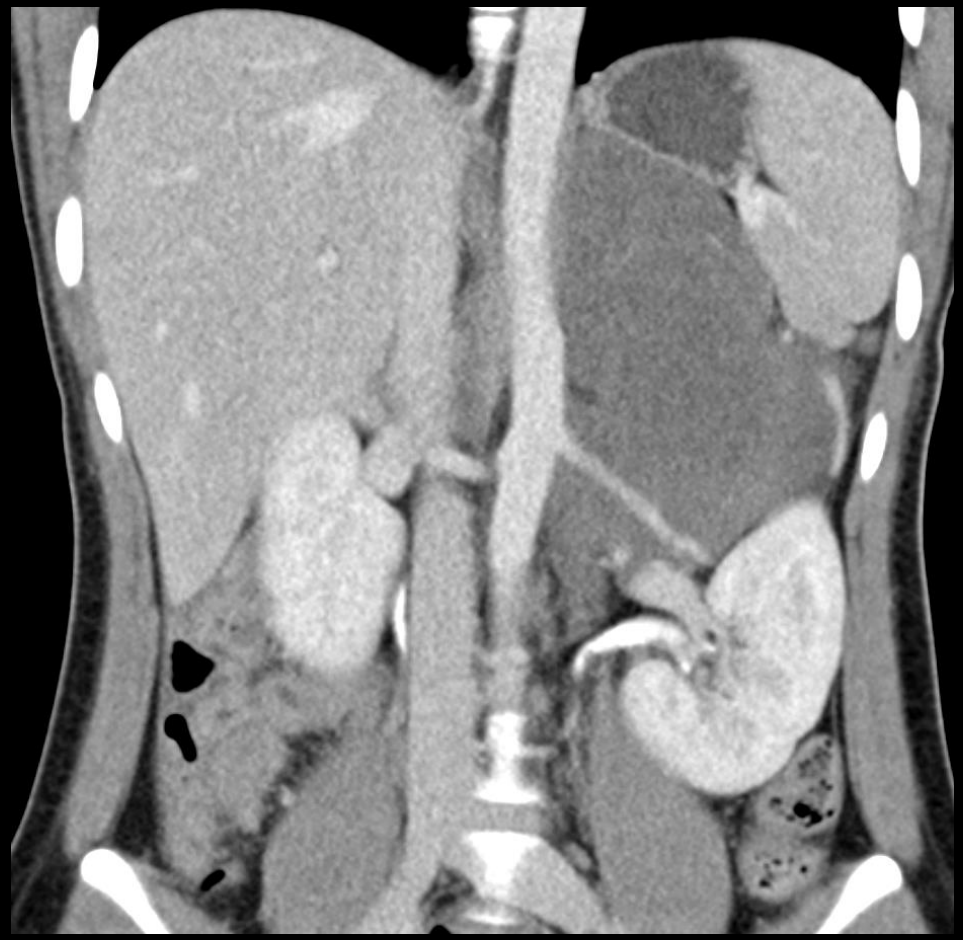
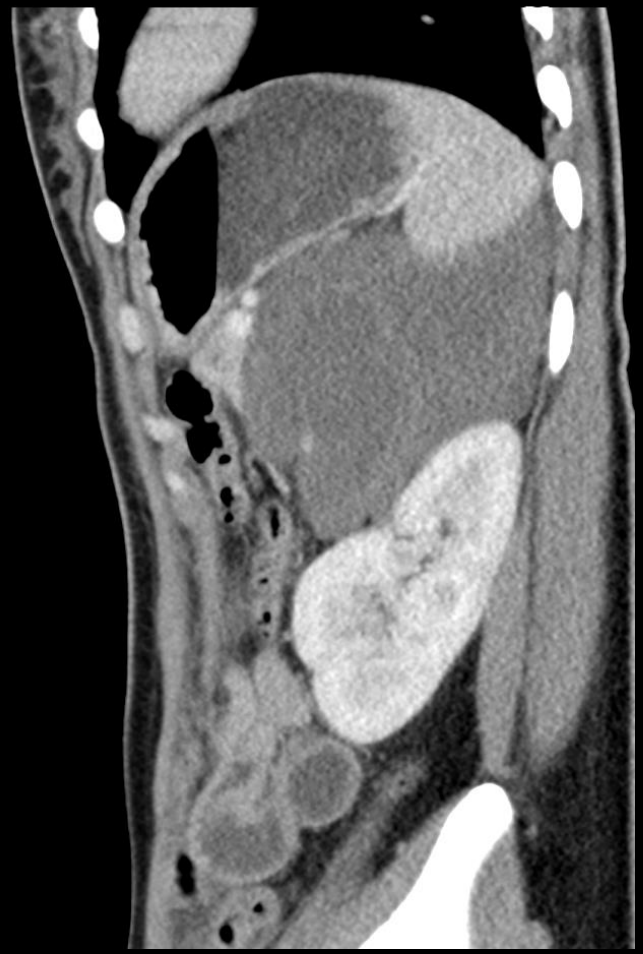
Findings (unlabeled): Coronal (left) and sagittal (right)





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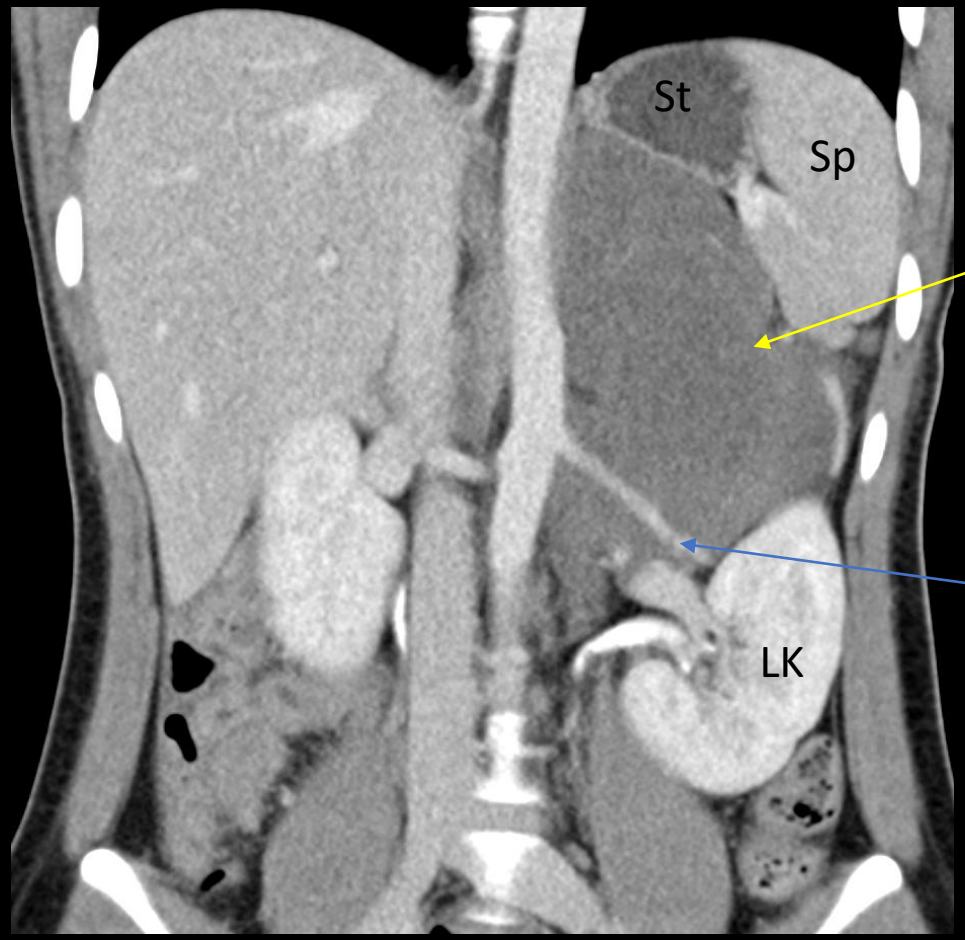
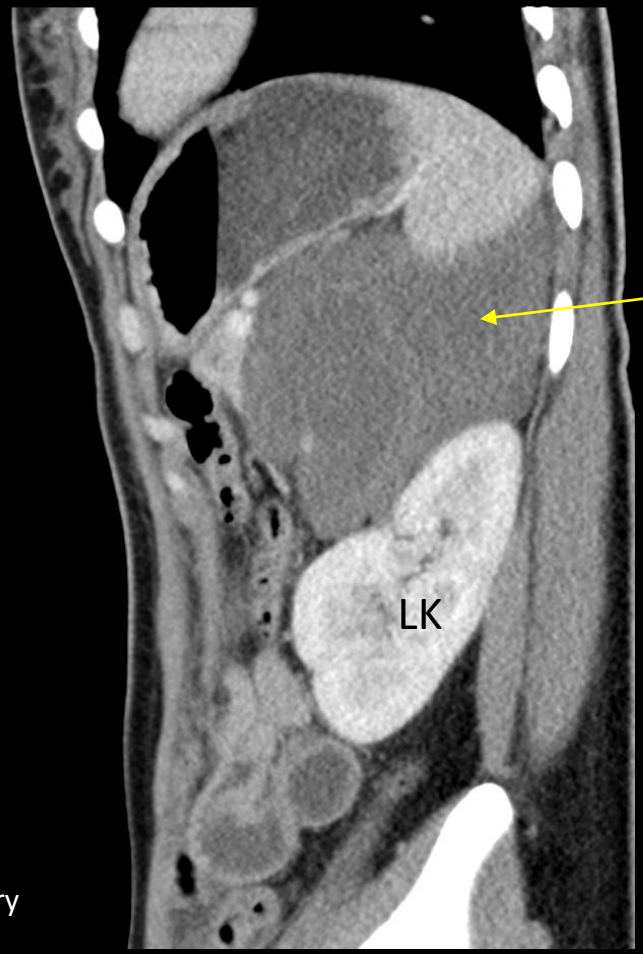


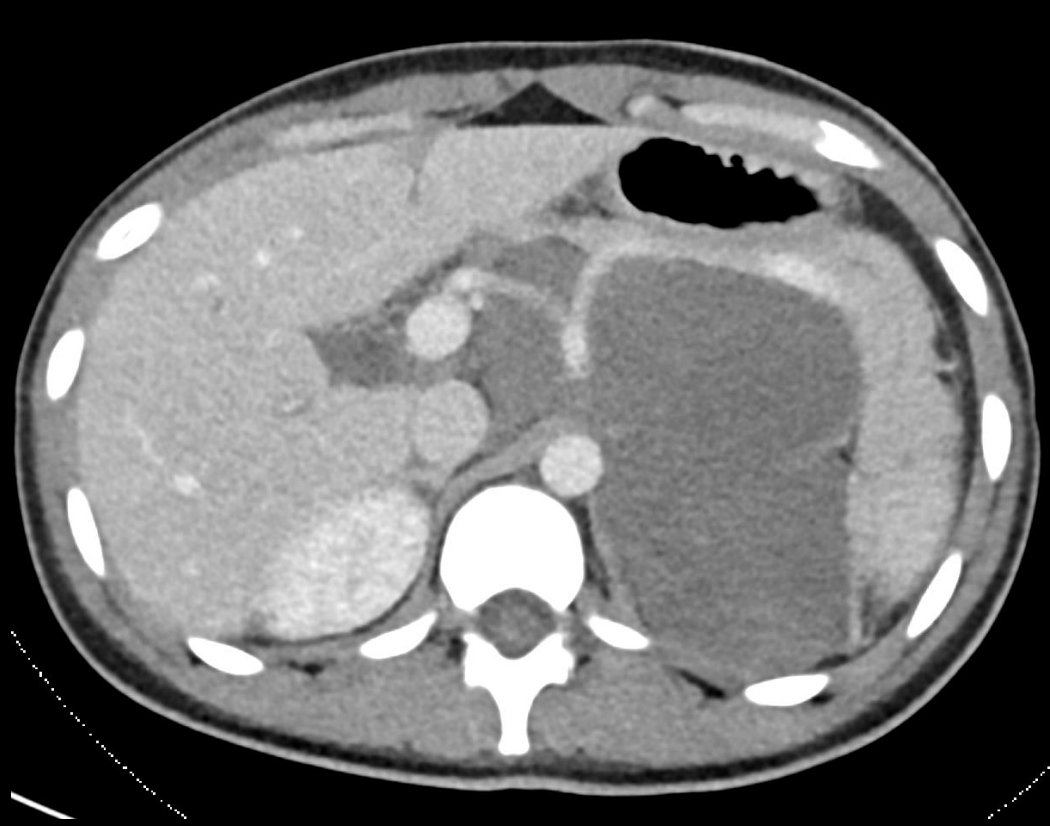
L retroperitoneal mass encases the aorta, left renal artery and exerts mass effect on L kidney displacing it inferiorly

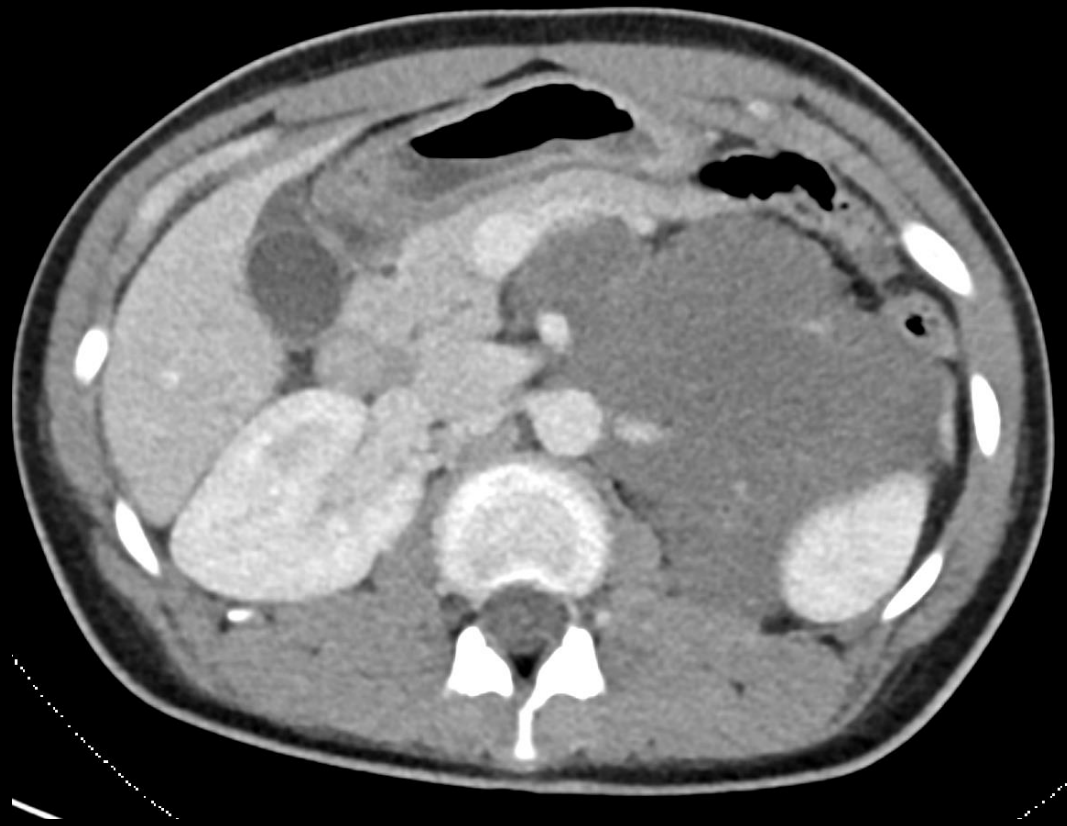
LK: left kidney
Sp: spleen
St: stomach
LRA: left renal artery



Hypodense L retroperitoneal mass is separate and distinct from L kidney. Lies superior to kidney, abuts the spleen.

Findings (unlabeled): Axial





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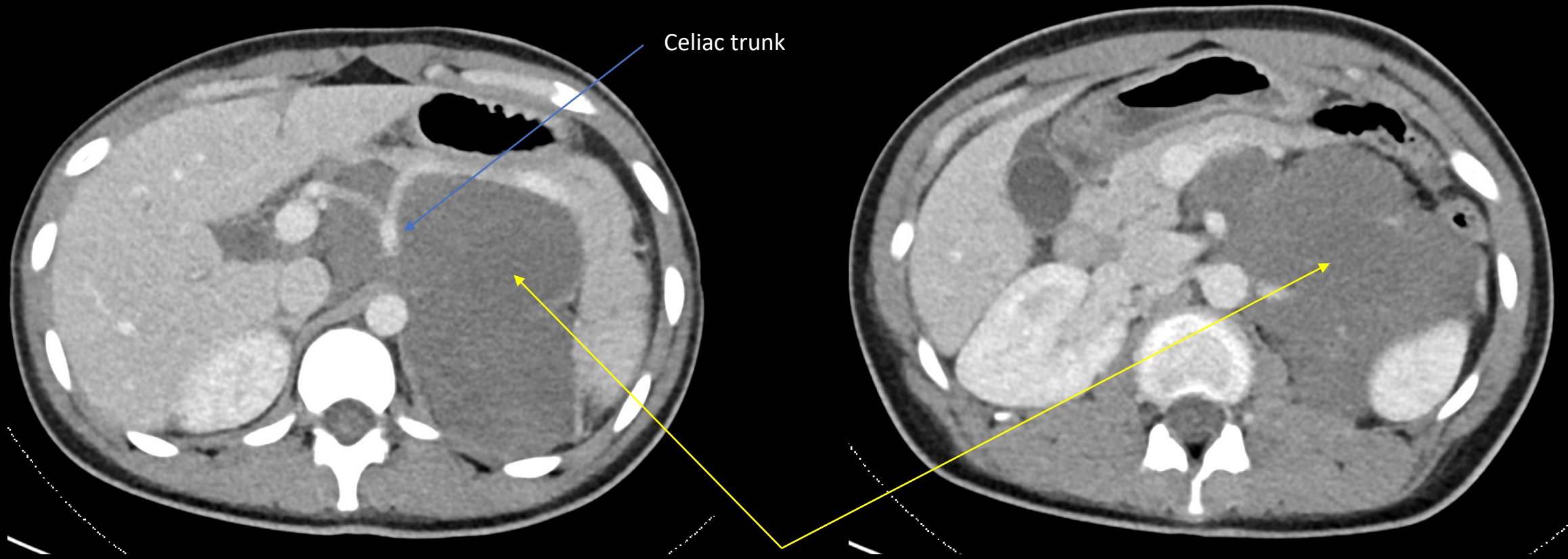
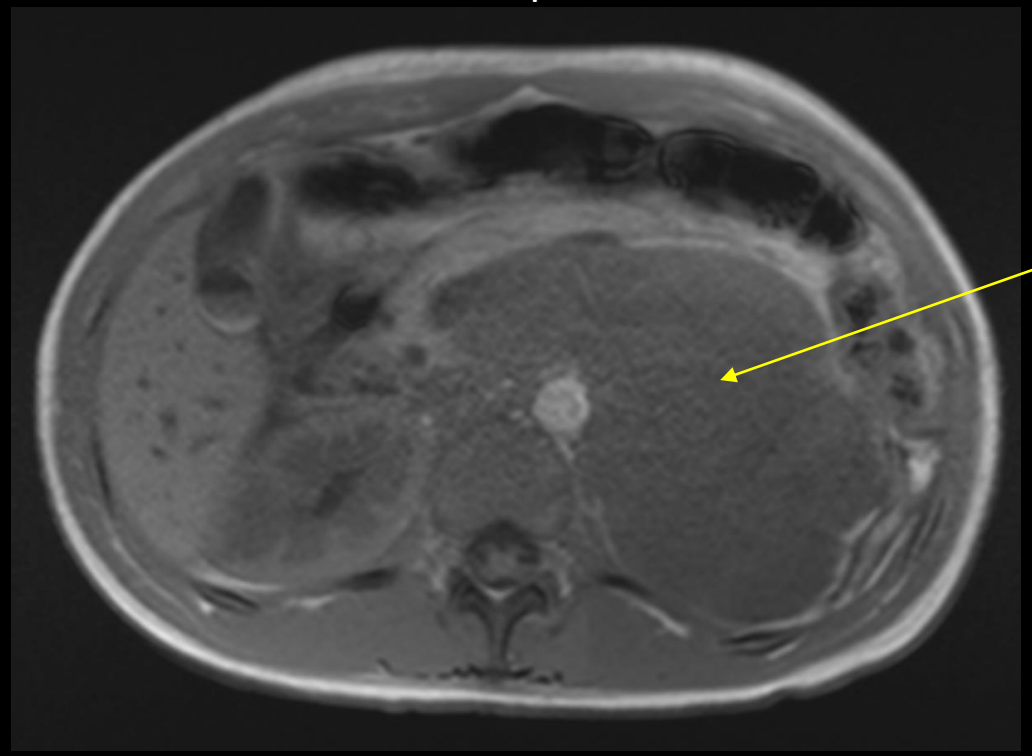


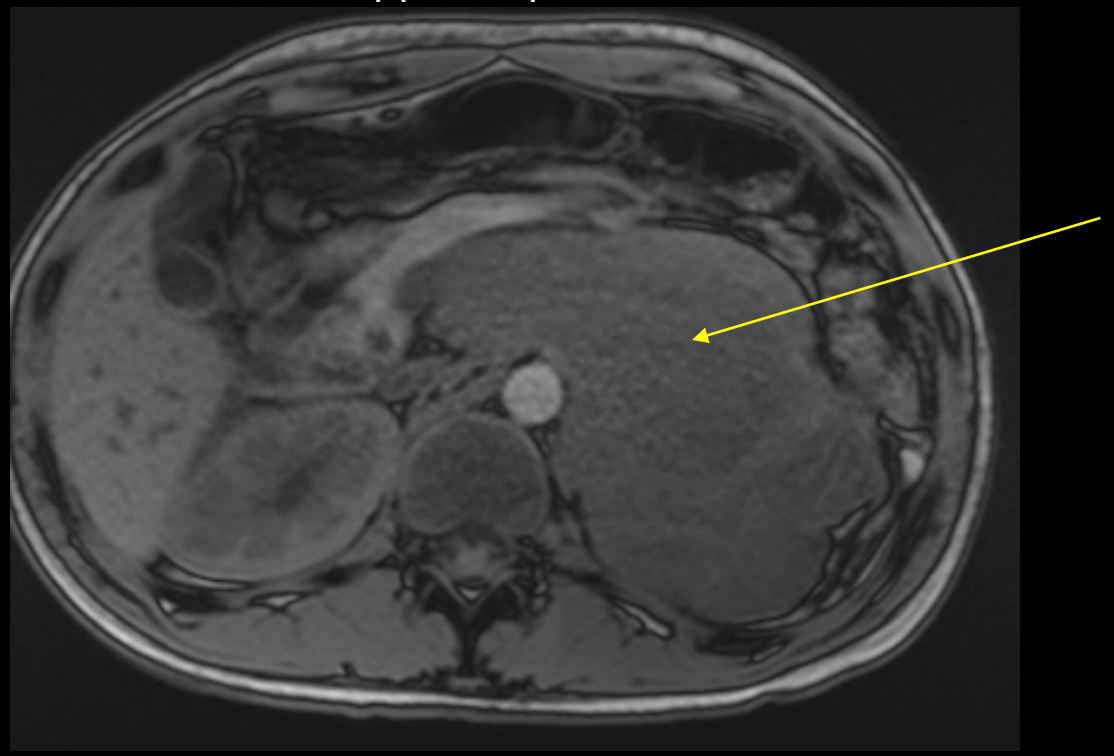
Hypodense L retroperitoneal mass encases celiac trunk and abuts pancreas. **Abdominal MRI recommended for further evaluation**.

Abdominal MRI: In phase and opposed phase

In phase



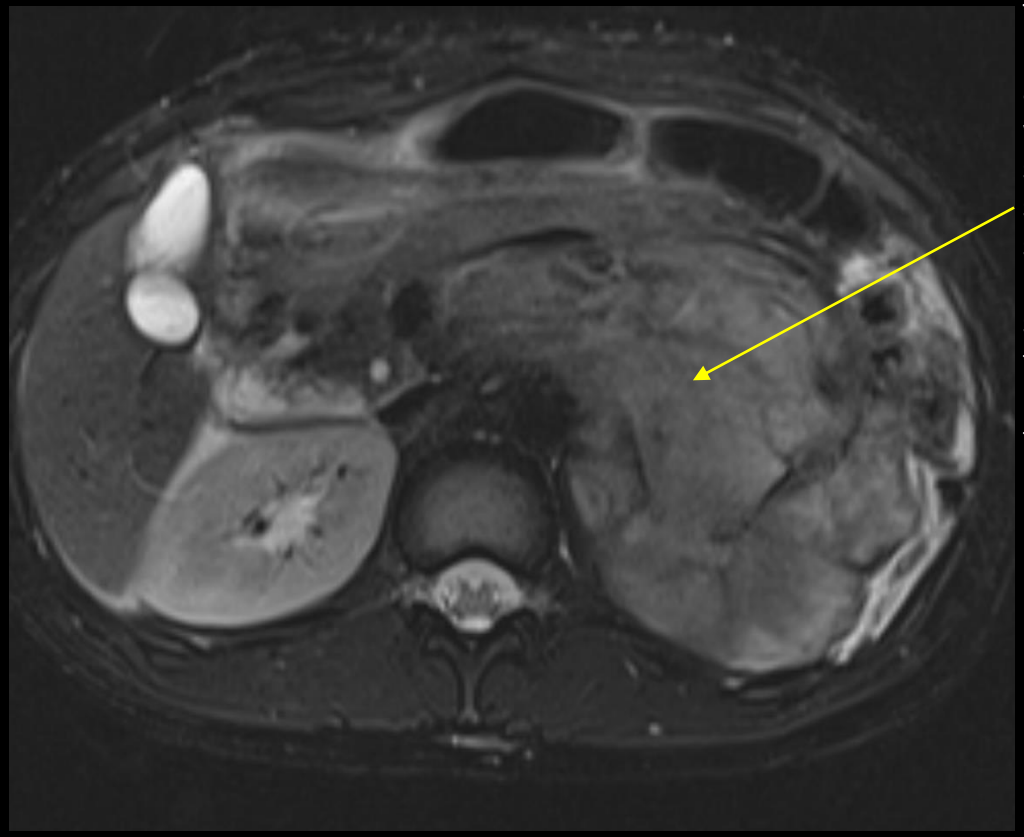
Opposed phase



In phase and opposed phase images of the mass show no change in signal intensity between the two, indicating no microscopic fat, which can be present in adrenal adenomas. Thus, imaging findings are not consistent with a lipid-rich adrenal adenoma.

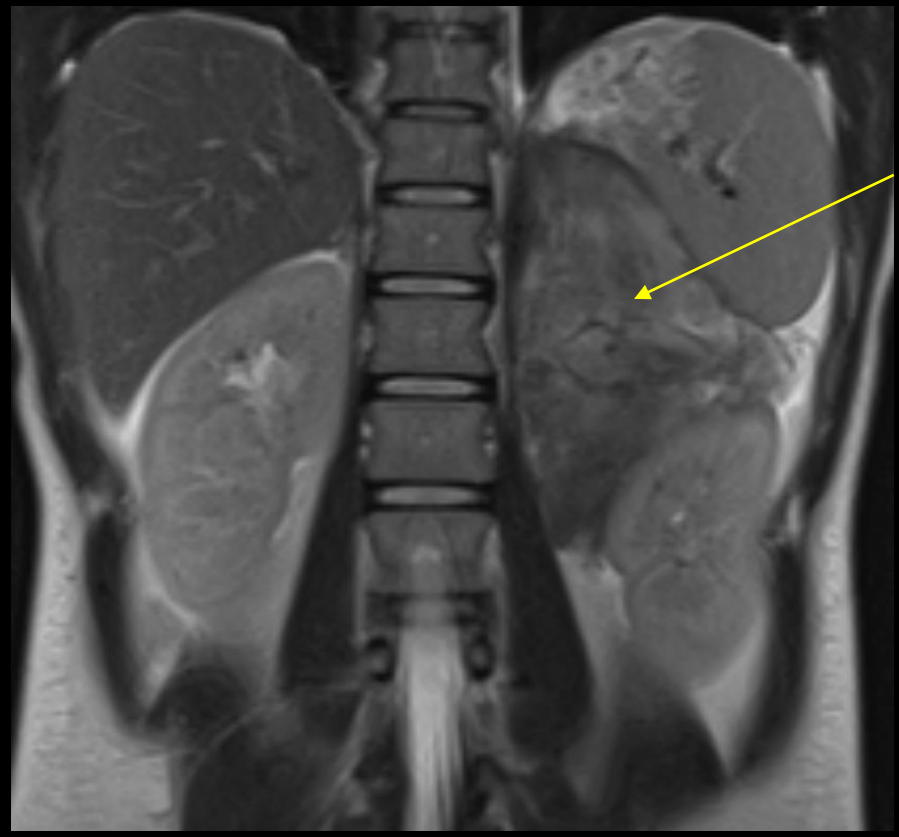
Abdominal MRI: T2

T2 fat saturation



T2 fat saturated images demonstrate a heterogenous mass with intermediate T2 signal. There was no signal loss on T2 fat saturated images, relative to non-fat saturated T2 images, to suggest macroscopic fat in the mass.

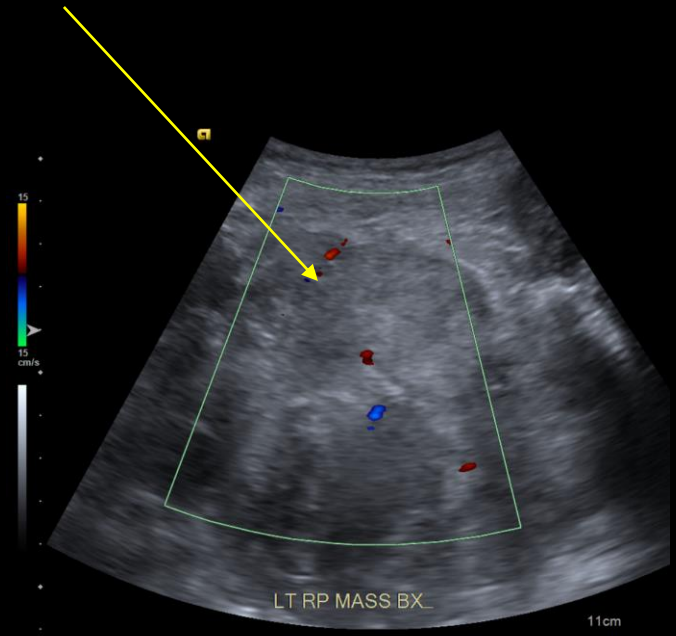
T2-weighted coronal (no fat saturation)



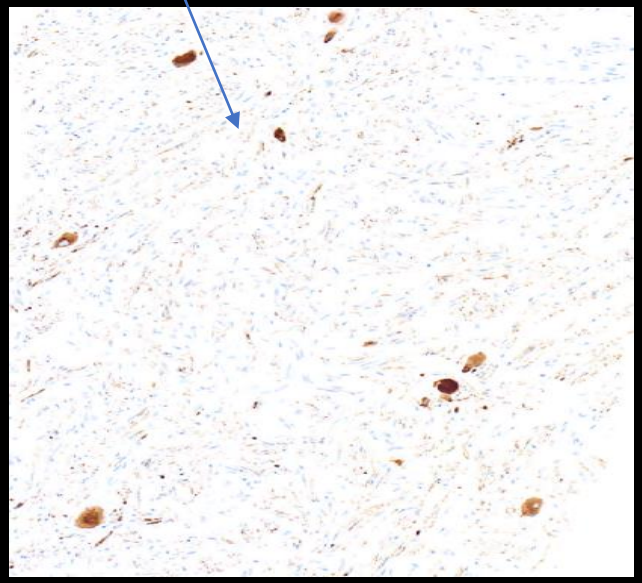
Left retroperitoneal mass exerting mass effect on the left kidney, abuts the spleen and stomach, and with heterogeneous signal on T2-weighted images, including areas of T2 hyperintense signal.

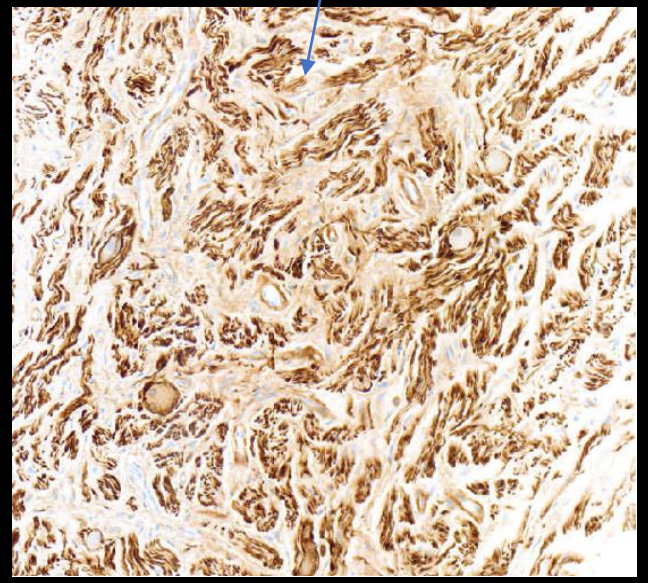
Discussion: Follow-up

- IR consulted for biopsy of L retroperitoneal mass; ultrasound-guided biopsy performed.



- Biopsy results: Schwann cell bundles positive for S100 and synaptophysin





- Final diagnosis: Left retroperitoneal ganglioneuroma, patient referred for outpatient follow-up with surgical oncology.

Discussion: Ganglioneuromas

- **Epidemiology**: Benign tumors which occur in the young adult and children population, more frequently in females. Most often develops in abdomen/pelvis region. Data such as incidence and prevalence are not available due to this tumor being exceedingly rare.

- **Clinical presentation;** Nonspecific, diagnosis is often made incidentally. Usually asymptomatic, but patients may present with abdominal pain, distention, and various sequelae because of hormonal secretion from the tumor, such as hypertension or Cushing's syndrome.

- **Pathophysiology:** Develop from neural crest tissue of the sympathetic nervous system ganglia. They grow slowly and may secrete catecholamines or steroid hormones.

Discussion: Ganglioneuromas

- **Diagnosis and Treatment**: Distinction from other sympathetic nervous system tumors (neuroblastoma, pheochromocytoma, schwannoma) based on imaging is difficult, so complete resection with comprehensive pathological confirmation is the ideal treatment to avoid overlooking a malignancy.

- **Prognosis:** Outcomes are generally very good, even if surgical resection is incomplete. Head/neck ganglioneuromas are associated with the most post-surgical complications due to complexity of the operation. Recurrence or metastases from ganglioneuromas are exceedingly rare.

References

- Scheirey, Fowler, Therrien, et al. ACR Appropriateness Criteria Acute Nonlocalized Abdominal Pain. Available at https://acsearch.acr.org/docs/69467/Narrative/. American College of Radiology. Accessed 10/21/2021.

- Fliedner SMJ, Winkelmann PER, Wesley R, Vonthein R, Lehnert H. Ganglioneuromas across age groups: Systematic review of individual patient data. Clin Endocrinol (Oxf). 2021 Jan;94(1):12-23. doi: 10.1111/cen.14297. Epub 2020 Aug 7. PMID: 32702779.

- Wu, Z., Zeng, Q., Zhang, X., Tu, S., & Zhang, F. (2018). Ganglioneuroma in unusual sites: clinical, radiologic and pathological features. *International journal of clinical and experimental pathology*, *11*(10), 4862–4866.

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