AMSER Case of the Month November 2020

2 year old male with abdominal mass

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Patient Presentation

- HPI: 2 year old male with no PMH presented to outside ED with abdominal mass found on routine checkup by pediatrician. A mass was noted in the left upper quadrant on exam. The outside ED completed an ultrasound which showed a 13.5 cm x 12 cm x 9 cm mass, and transferred the patient for further workup.
- Medical and Surgical History: None
- Family History: Mesothelioma in paternal grandmother. Melanoma on both sides of family. History of HTN and TIA on father's side of family.

Physical Exam
Vitals: T: 36.9 BP: 112/68 HR: Unable to Record RR: 24 SpO2: 100%
General: Alert, NAD
Skin: Warm, intact
Cardiac: Regular rate and rhythm, no murmurs
Respiratory: Clear to auscultation bilaterally
GI: Soft, nondistended, non tender, **mass in LUQ that does not cross midline**
MSK: Normal ROM, no swelling

Labs were remarkable for a BUN of 22 and an LDH of 444.

What Imaging Should We Order?

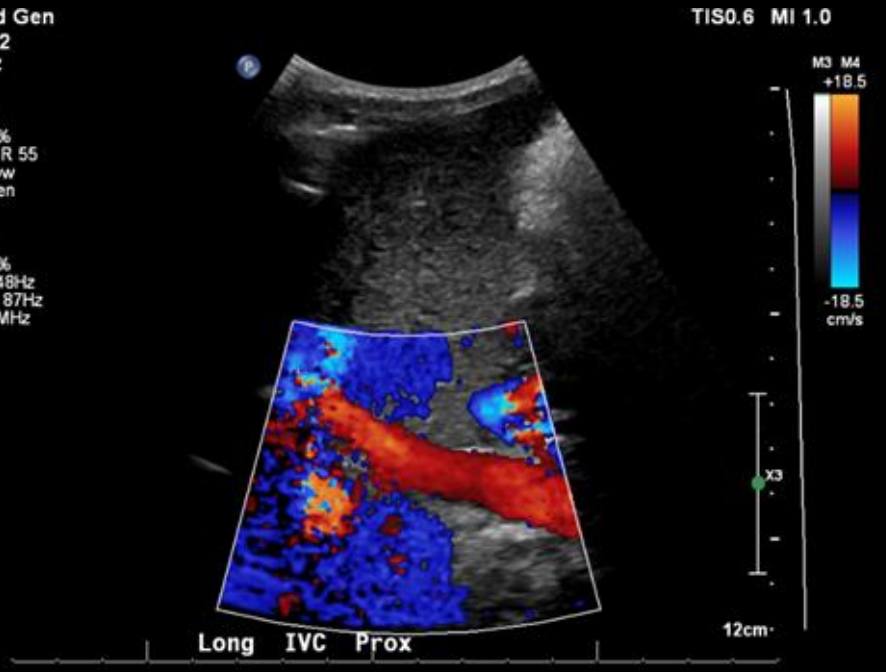
ACR Appropriateness Criteria

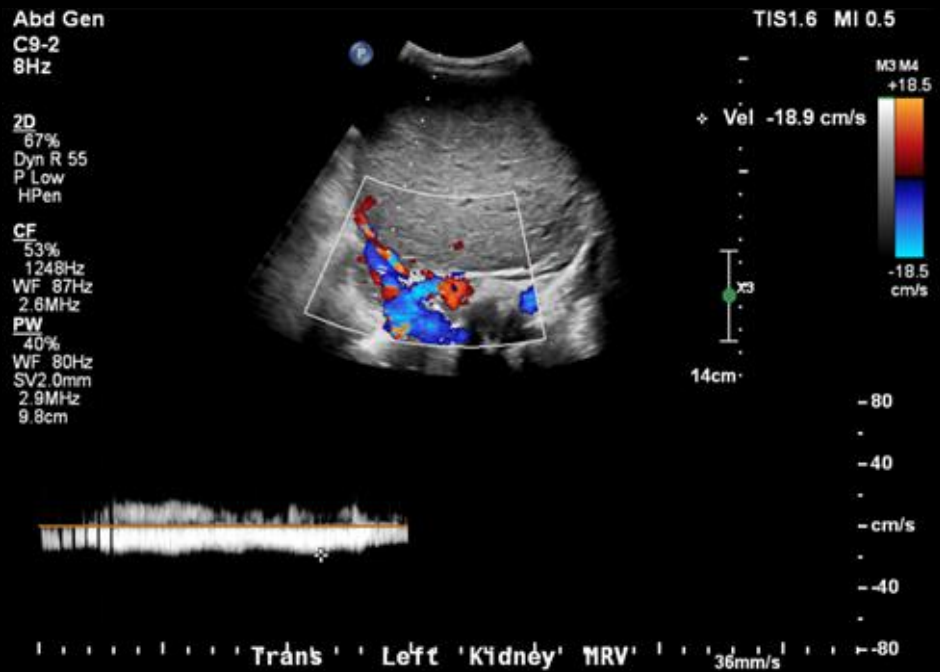
Variant 1: **Palpable abdominal mass. Suspected intra-abdominal neoplasm. Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
CT abdomen with IV contrast	Usually Appropriate	☢☢☢
US abdomen	Usually Appropriate	O
MRI abdomen without and with IV contrast	May Be Appropriate	O
CT abdomen without IV contrast	May Be Appropriate	☢☢☢
MRI abdomen without IV contrast	May Be Appropriate	O
CT abdomen without and with IV contrast	Usually Not Appropriate	☢☢☢☢
FDG-PET/CT skull base to mid-thigh	Usually Not Appropriate	☢☢☢☢
Radiography abdomen	Usually Not Appropriate	☢☢
Fluoroscopy contrast enema	Usually Not Appropriate	☢☢☢
Fluoroscopy upper GI series	Usually Not Appropriate	☢☢☢
Fluoroscopy upper GI series with small bowel follow-through	Usually Not Appropriate	☢☢☢

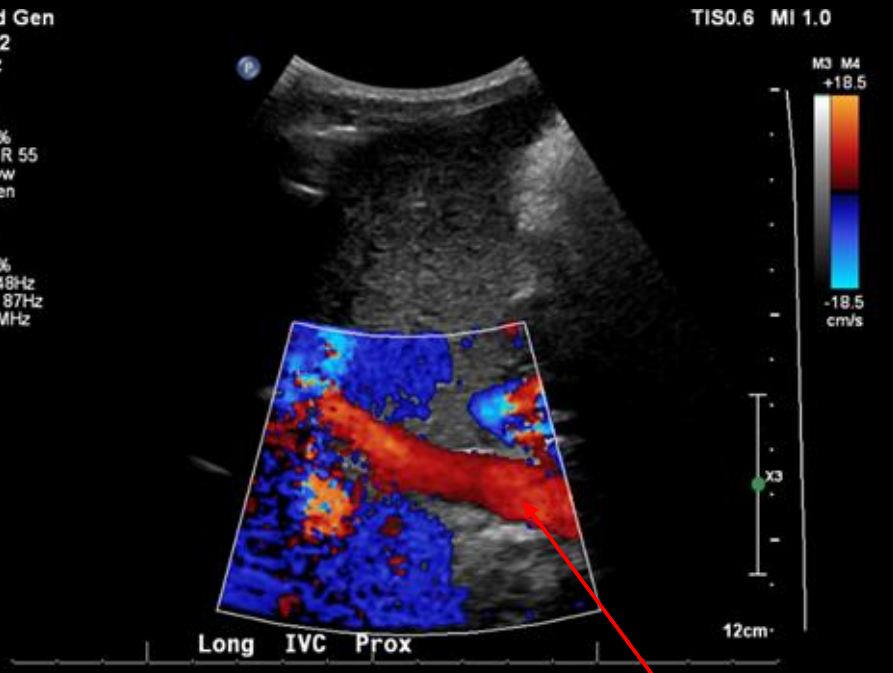
This was ordered by the outside hospital emergency room physician- dopplers were repeated by the ED at our facility

Findings (Unlabeled US)

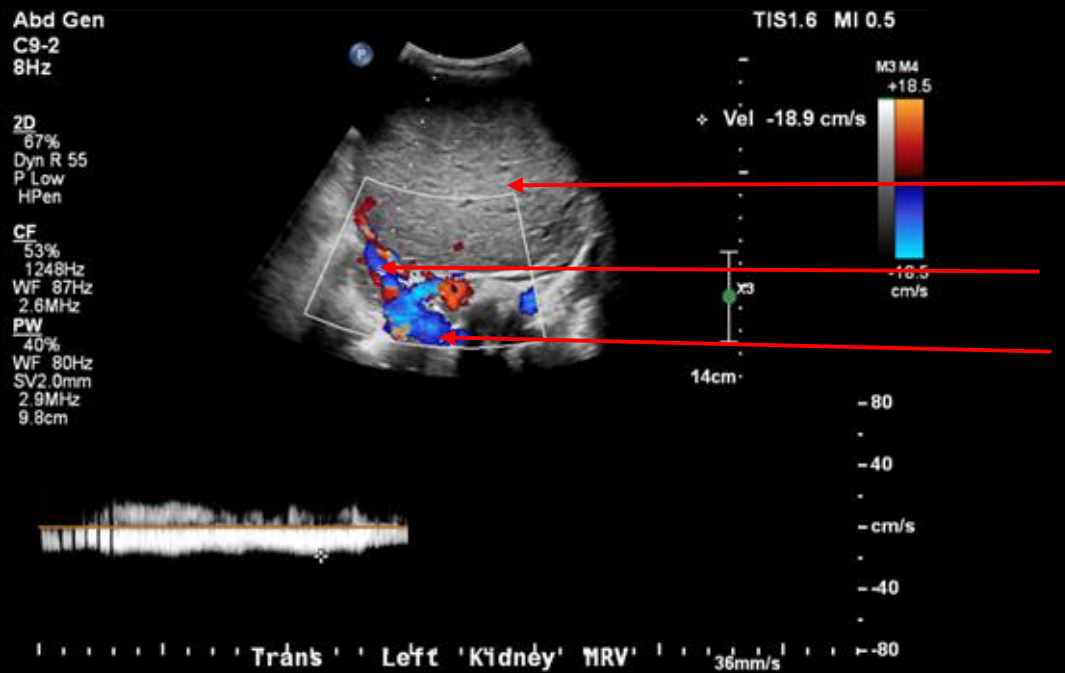




Findings (Labeled US)



IVC without tumor thrombus



Solid left renal mass

Patent left renal vein

IVC without tumor thrombus

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This was ordered by the pediatric heme/onc team upon admission

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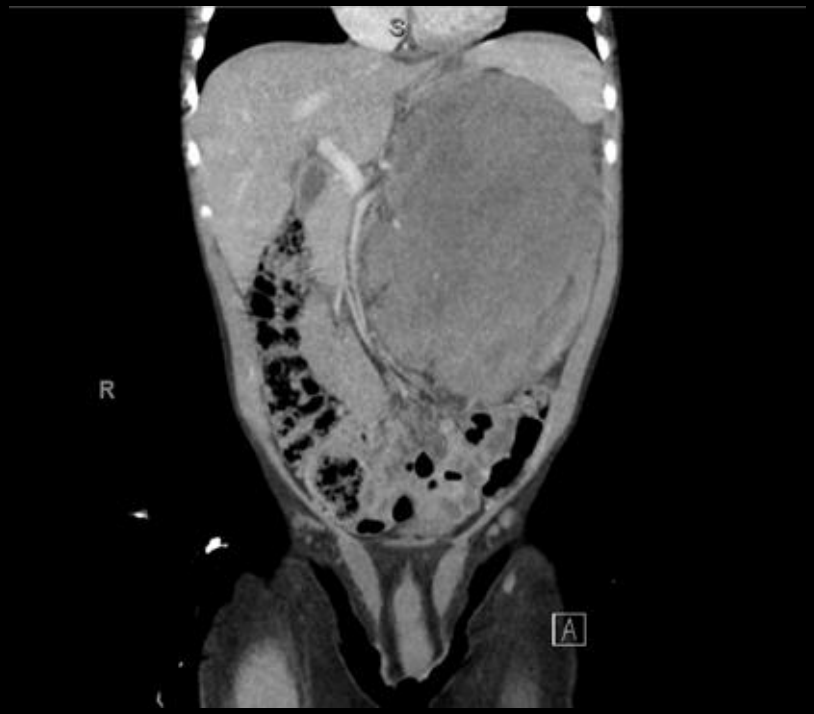
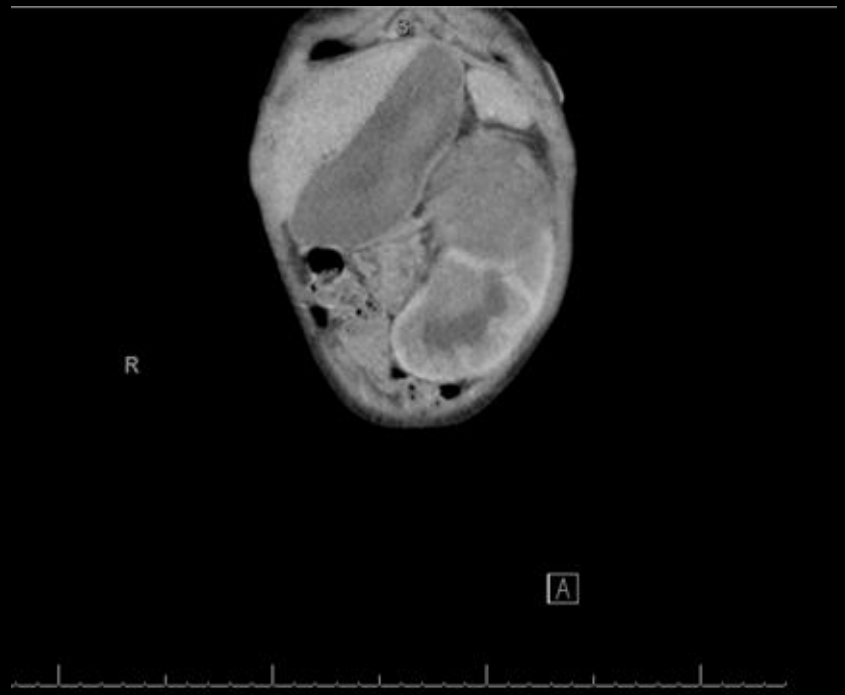
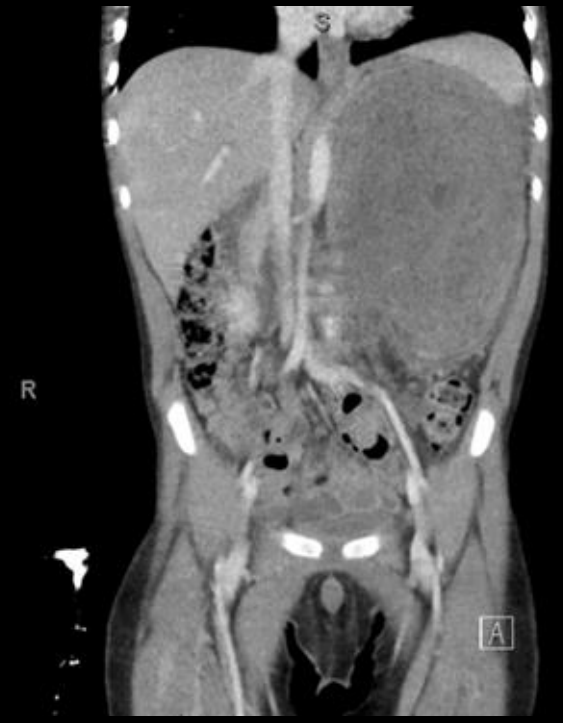
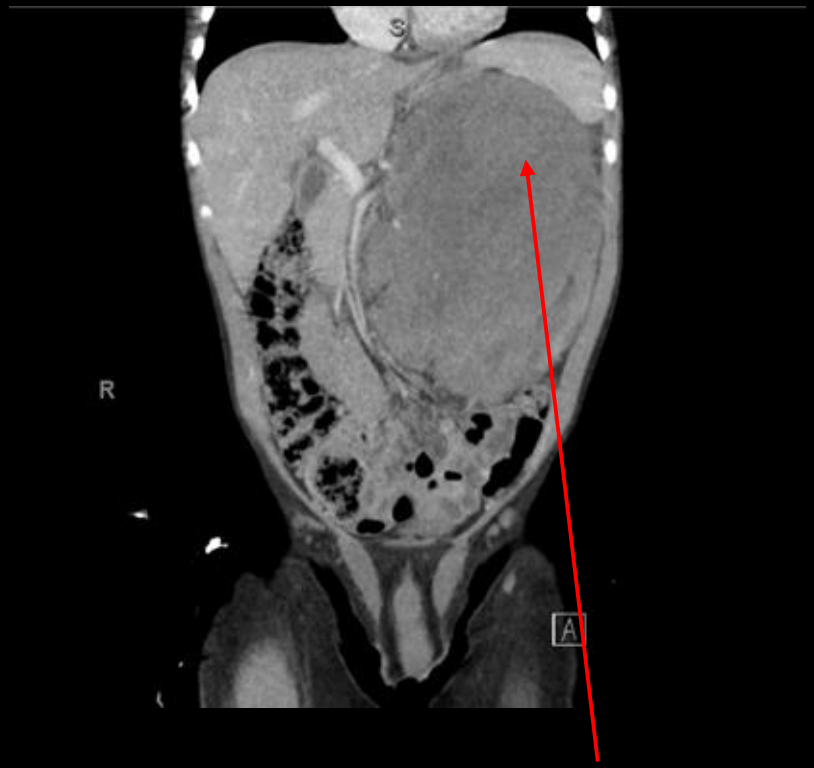
Findings - Unlabeled CT (Coronal)



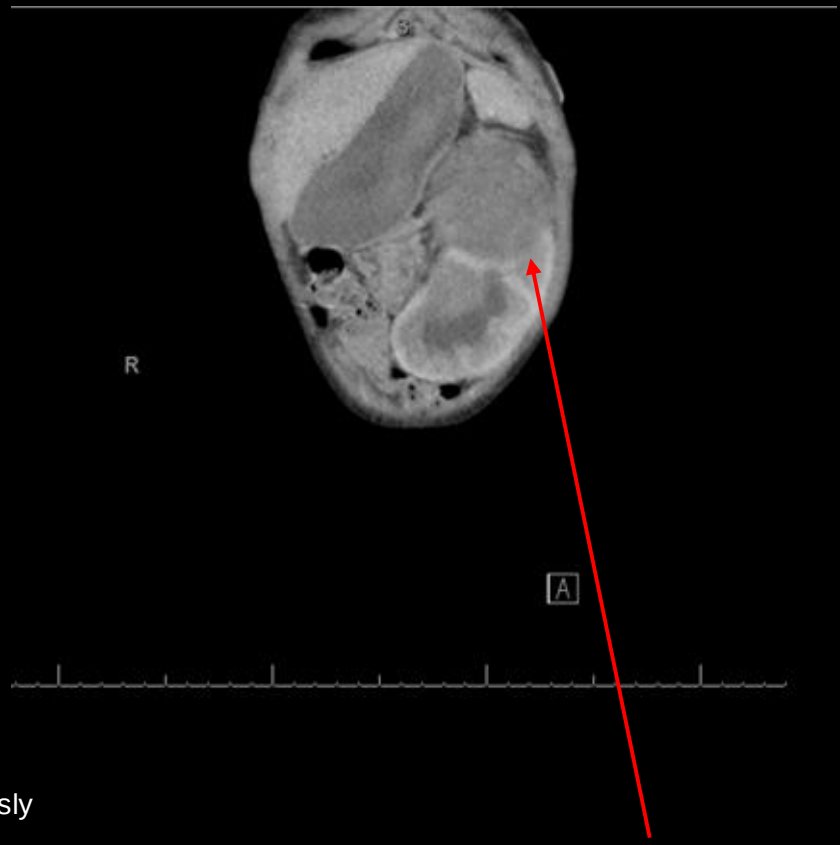




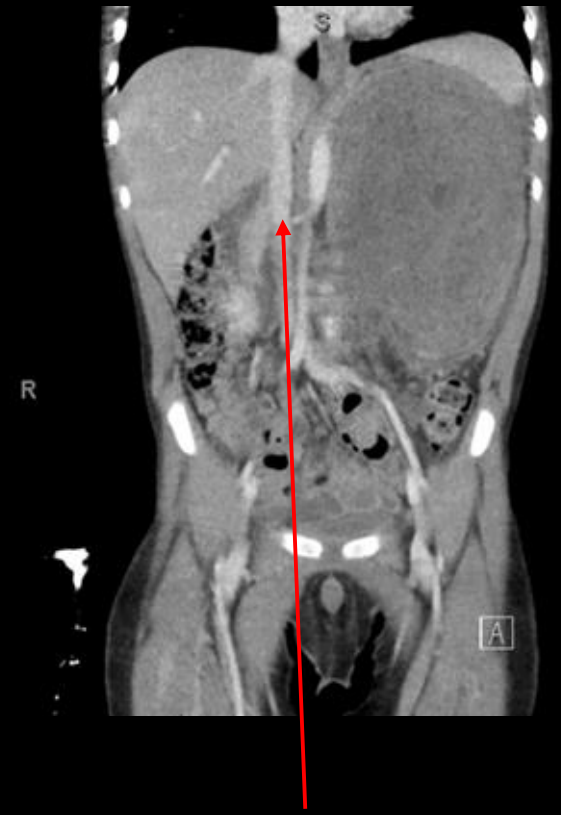
Findings - Labeled CT (Coronal)



Left renal heterogeneously enhancing soft tissue mass

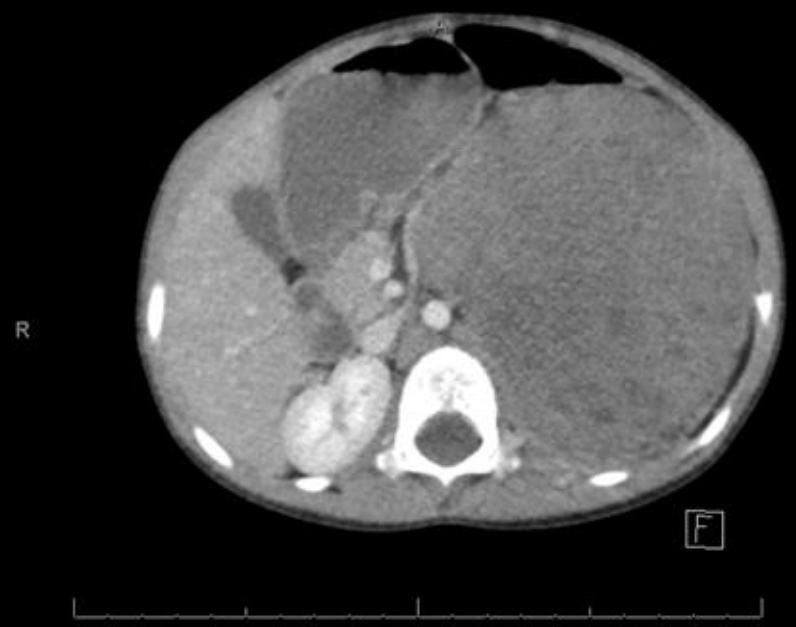


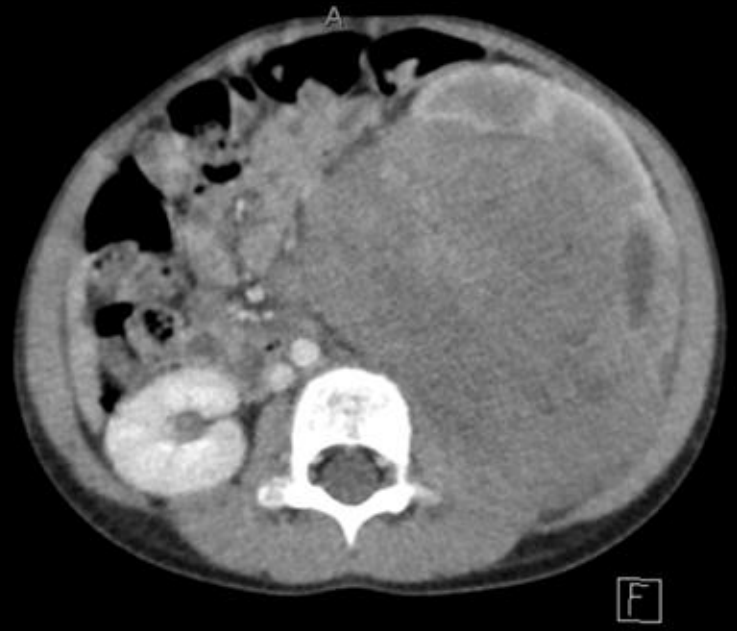
Claw sign indicating mass originates from left kidney



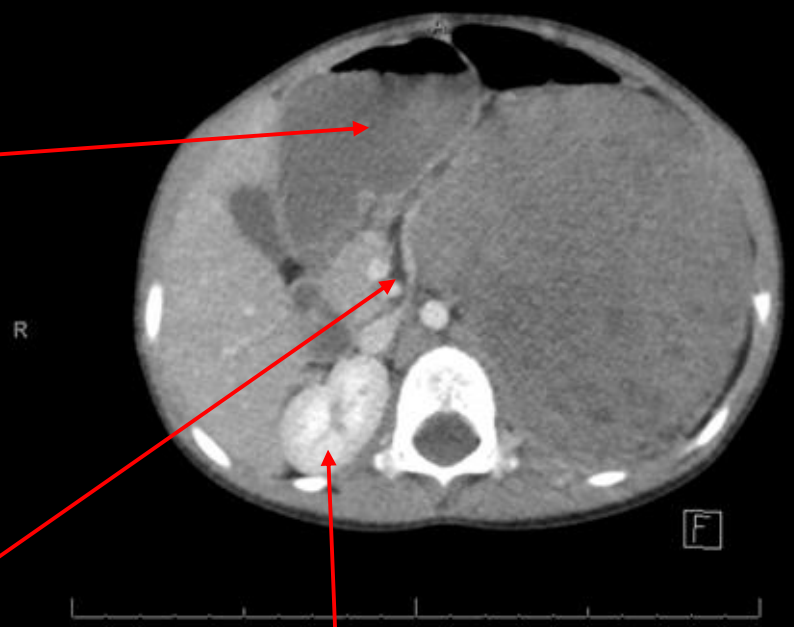
Patent IVC without thrombus

Findings - Unlabeled CT (Axial)





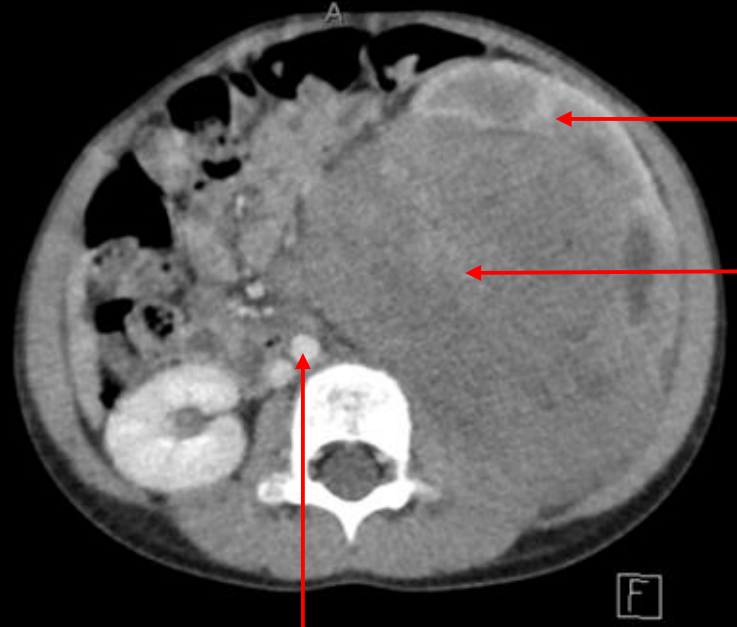
Findings - Labeled CT (Axial)



Mass effect on stomach

Patent left renal vein

Normal right kidney

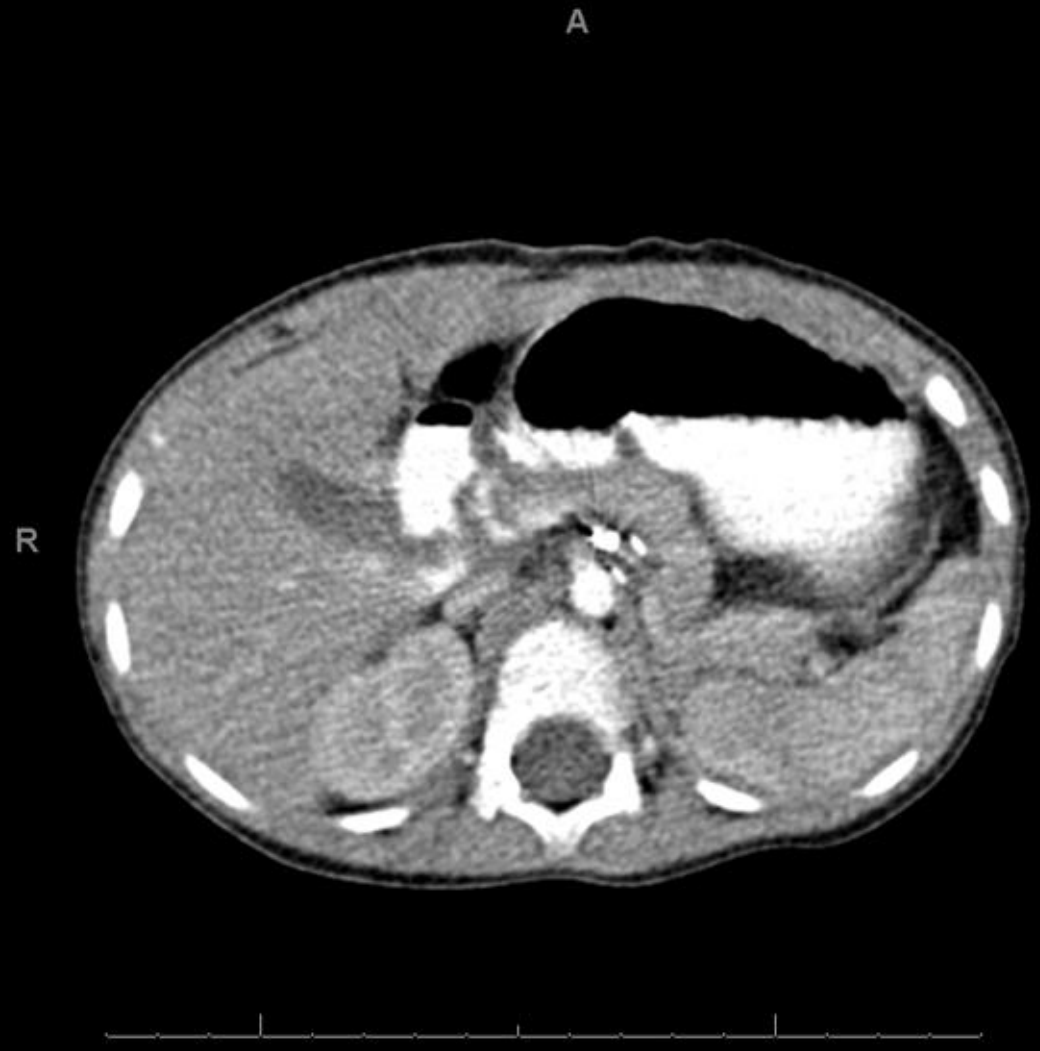


Left kidney

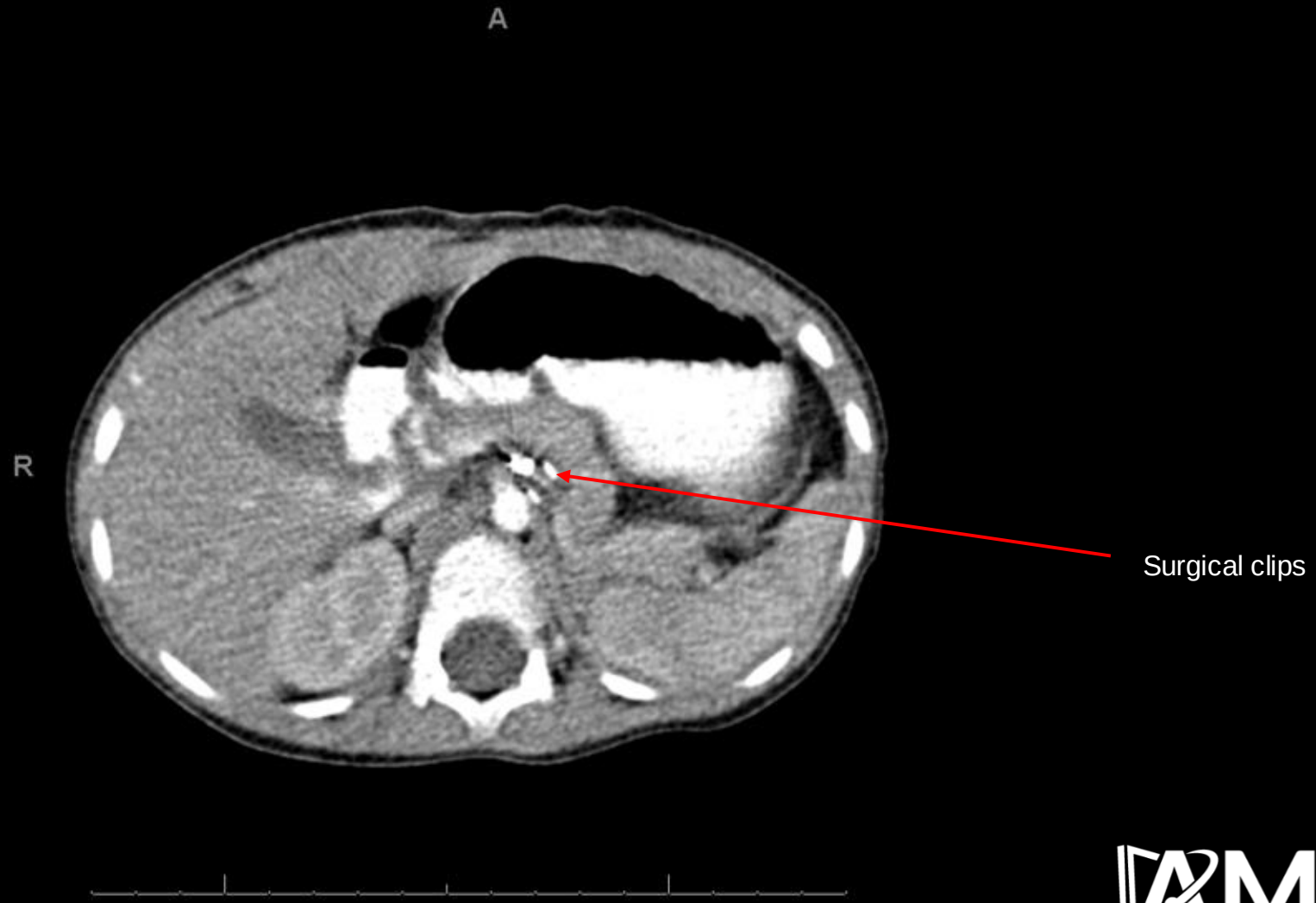
Heterogeneously enhancing soft tissue mass

Aorta displaced to right

Post-Op Axial CT (unlabeled)



Post Left Radical Nephrectomy Axial CT (Labeled)



Abdominal Mass Differential Diagnosis

- Wilms tumor
- Neuroblastoma
- Mesoblastic nephroma
- Renal cell carcinoma
- Renal Rhabdoid Tumor
- Clear cell sarcoma
- Renal medullary carcinoma

Final Dx:

Wilms Tumor

Case Timeline

- Abdominal US was performed at outside hospital with concern for neuroblastoma versus Wilms tumor. Patient was transferred for further workup
- Abdominal US with dopplers was performed at VCU ED, which showed no tumor extension into the renal vein or IVC. Chest, abdomen and pelvis CT with contrast was ordered. Chest CT was unremarkable. Abdominal CT showed mass arising from left kidney
- Left radical nephrectomy was performed
- Follow up CT showed post-surgical changes

Case Discussion

Epidemiology and Presentation

- Accounts for 87% of pediatric renal masses
- Peak incidence of 3-4 years of age with 80% of patients presenting before 5 years of age
- Can be bilateral in 4-13% of children
- May be associated with other congenital anomalies such as cryptorchidism, hemihypertrophy, hypospadias, and sporadic aniridia
- Discovery of tumor most commonly follows detection of palpable mass
- Hematuria and pain are infrequent clinical findings, and hypertension can be noted in 25% of cases

Features

- Tumor arises from mesodermal precursors of renal parenchyma
- Solid intrarenal mass with pseudocapsule and distortion of renal parenchyma and collecting system
- Tumor typically spreads by extension and displaces adjacent structures but does not elevate or encase the aorta (which would be a distinguishing feature of a neuroblastoma)
- May demonstrate vascular invasion of renal vein and inferior vena cava with occasional extension into right atrium
- Metastases are most commonly found in the lungs, liver and regional lymph nodes

Case Discussion

Radiographic Features

- Often very large on presentation and displaces adjacent structures without insinuating between them
- Initial evaluation on ultrasound the tumor has heterogeneous echogenicity representing hemorrhage, fat, necrosis or calcification
- Examination of IVC using dopplers on ultrasound is crucial to detect tumor extension
- CT demonstrates heterogeneous soft tissue mass with infrequent areas of calcification and fat density regions
- Claw sign on CT can help differentiate Wilms tumor from neuroblastoma
- Important to assess contralateral kidney for presence of disease
- 10-20% of cases have lung metastases found on CT at the time of diagnosis, so it is important to complete a chest, abdomen and pelvis CT with contrast in order to evaluate for metastases
- While MRI is the most sensitive modality for evaluating caval patency, it may require sedation in many children and so surgical planning can be done using US and CT
- If MRI is completed, T1 imaging shows heterogeneous hypointensity and T2 imaging shows heterogeneous hyperintensity

Case Discussion

Treatment

- Unilateral Wilms tumors are treated by a combination of nephrectomy and chemotherapy
- Occasionally, especially in cases where tumors are bilateral, chemotherapy can precede surgery
- Radiotherapy has a limited role in treatment
- Cure is possible in 90% of cases
- Recurrence can be seen in the tumor bed or with metastases in the lungs or liver

References

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Chung EM, Graeber AR, Conran RM. Renal Tumors of Childhood: Radiologic-Pathologic Correlation Part 1. The 1st Decade: From the Radiologic Pathology Archives. Radiographics. 2016 Mar-Apr;36(2):499-522. doi: 10.1148/rg.2016150230. PMID: 26963460

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Servaes SE, Hoffer FA, Smith EA, Khanna G. Imaging of Wilms tumor: an update. Pediatr Radiol. 2019;49(11):1441-1452. doi:10.1007/s00247-019-04423-3