AMSER Case of the Month
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Patient Presentation

- HPI: 14 y.o. female presenting with 2 weeks of sudden onset left lower abdominal pain with no noted trauma. The pain has been gradually worsening and it radiates to her left flank.
- ROS: Negative for hematuria, dysuria, SOB, fatigue, fever, weight loss, easy bruising/bleeding, or rash
- PMH: Seasonal Allergies
- PSH: None
- SHx: Denies any past or current sexual activity; denies any drug/tobacco/alcohol use.
- Physical Exam: ”Splenomegaly” noted on abdominal exam. No other pertinent physical exam findings.
- Labs: BUN 16, Cr 0.9, UA Negative, Plt 660 K

Pediatric Flank Pain DDx

- Nephrolithiasis/Ureteral Obstruction
- Urinary Tract Infection/Pyelonephritis
- Vesicoureteral Reflux
- Renal Tumor
  - Wilms Tumor
  - Renal Cell Carcinoma (RCC)
  - Renal Adenoma
  - Renal Oncocytoma
  - Renal Angiomyoplipoma
- Adrenal Tumor

What Imaging Should We Order?

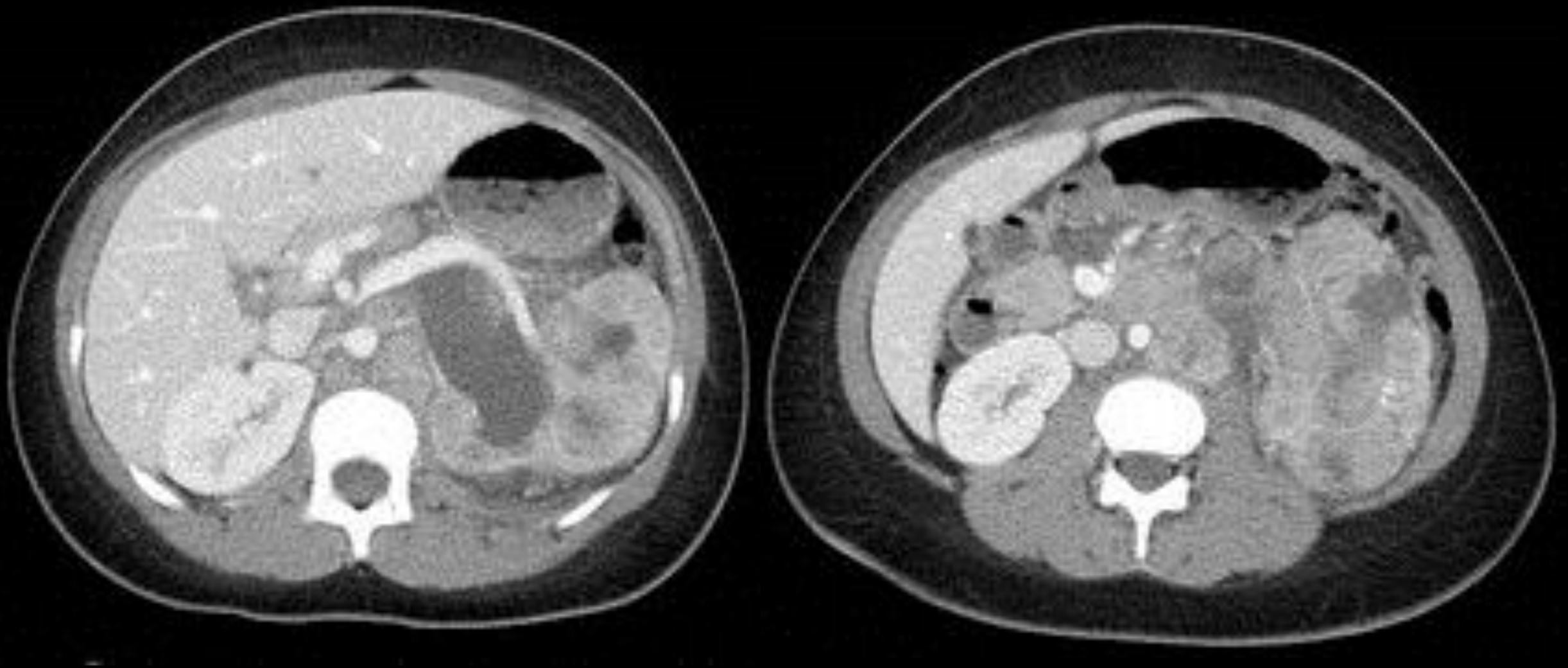
Select the applicable ACR Appropriateness Criteria

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 imaging modality was ordered by the ER physician

CT Abdomen w/ Contrast (unlabeled)



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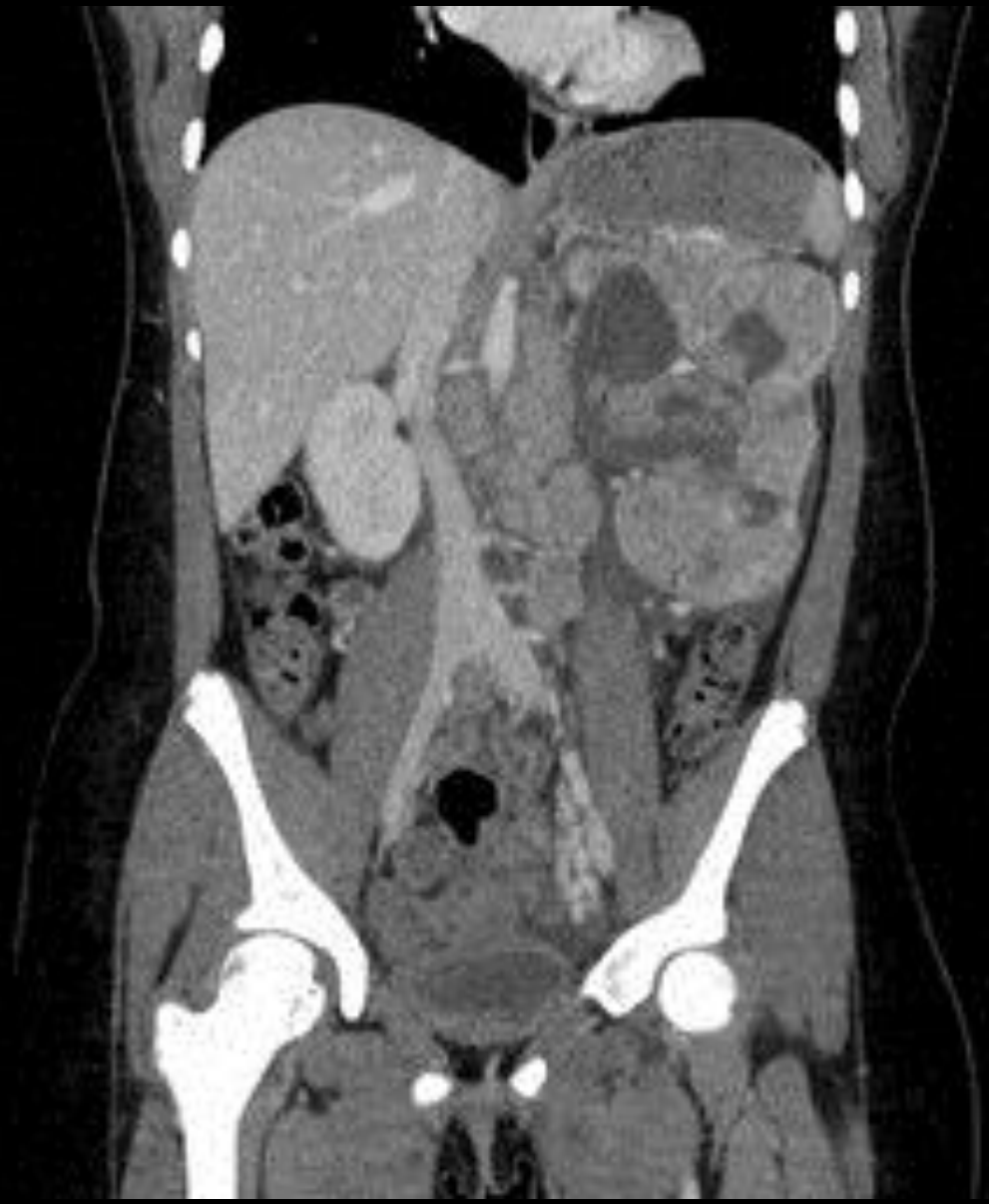
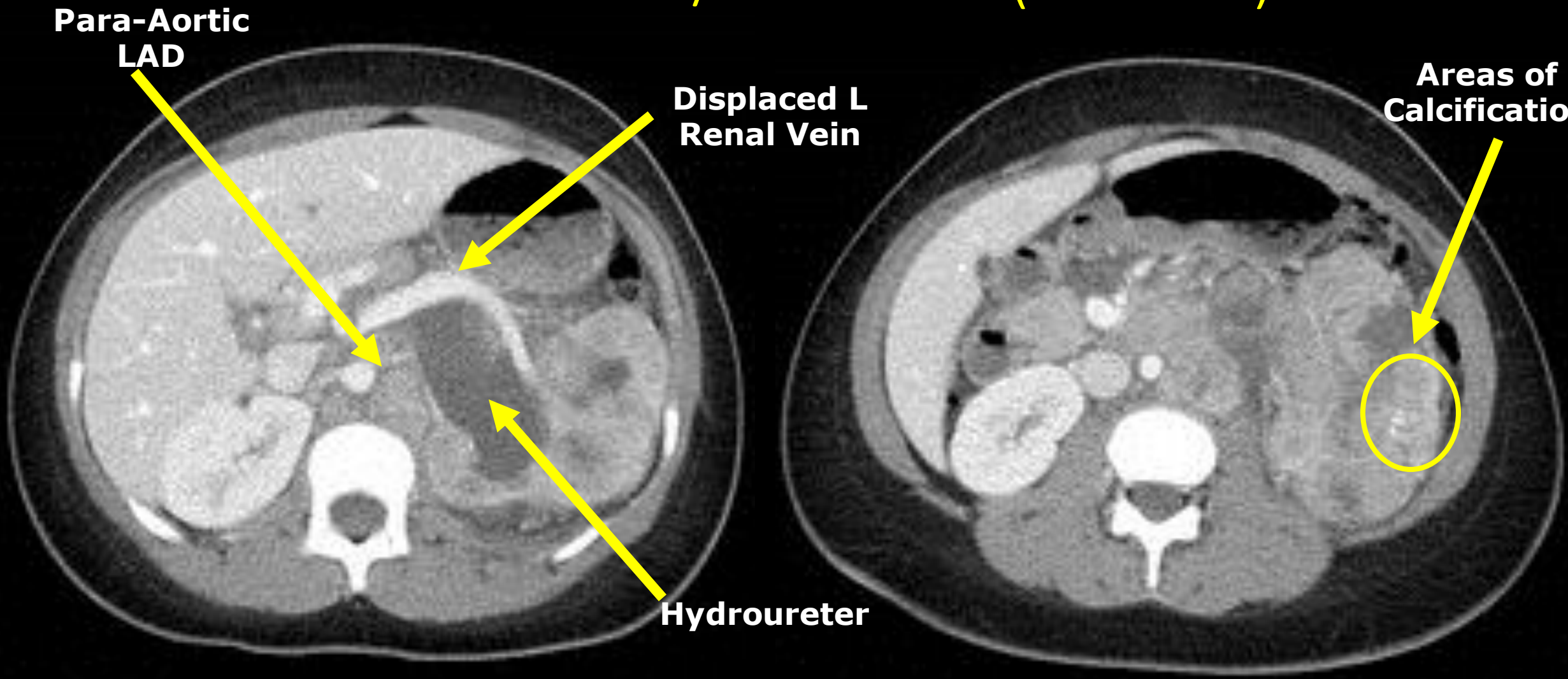


CT Abdomen w/ Contrast (labeled)



LEFT: Axial CT image shows enlarged left kidney with infiltrative neoplasm that extends into the collecting system with resulting hydronephrosis. The left renal vein is patent but displaced anteriorly by the mass. RIGHT: Axial CT image displaying areas of small calcification within the renal mass.

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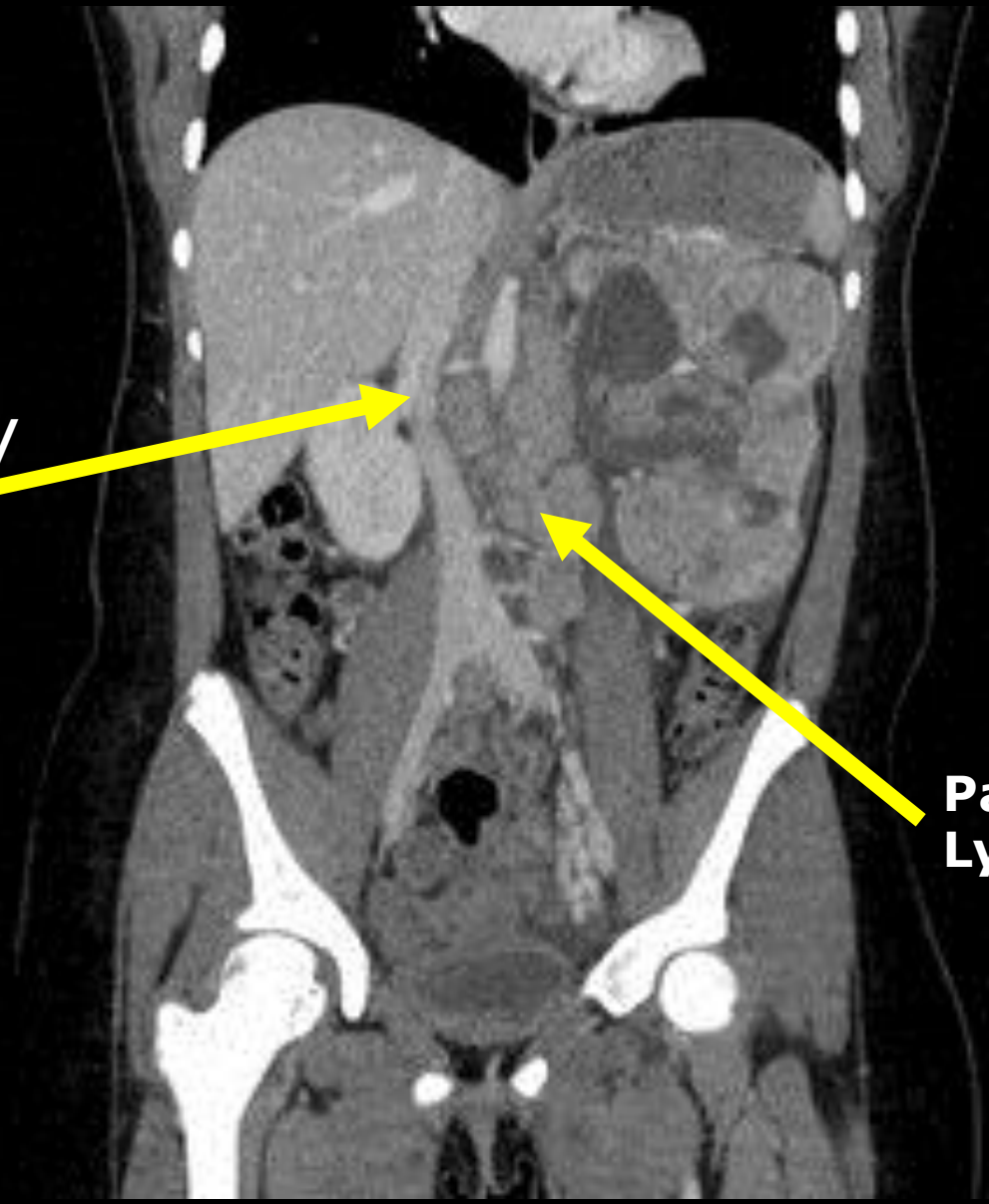
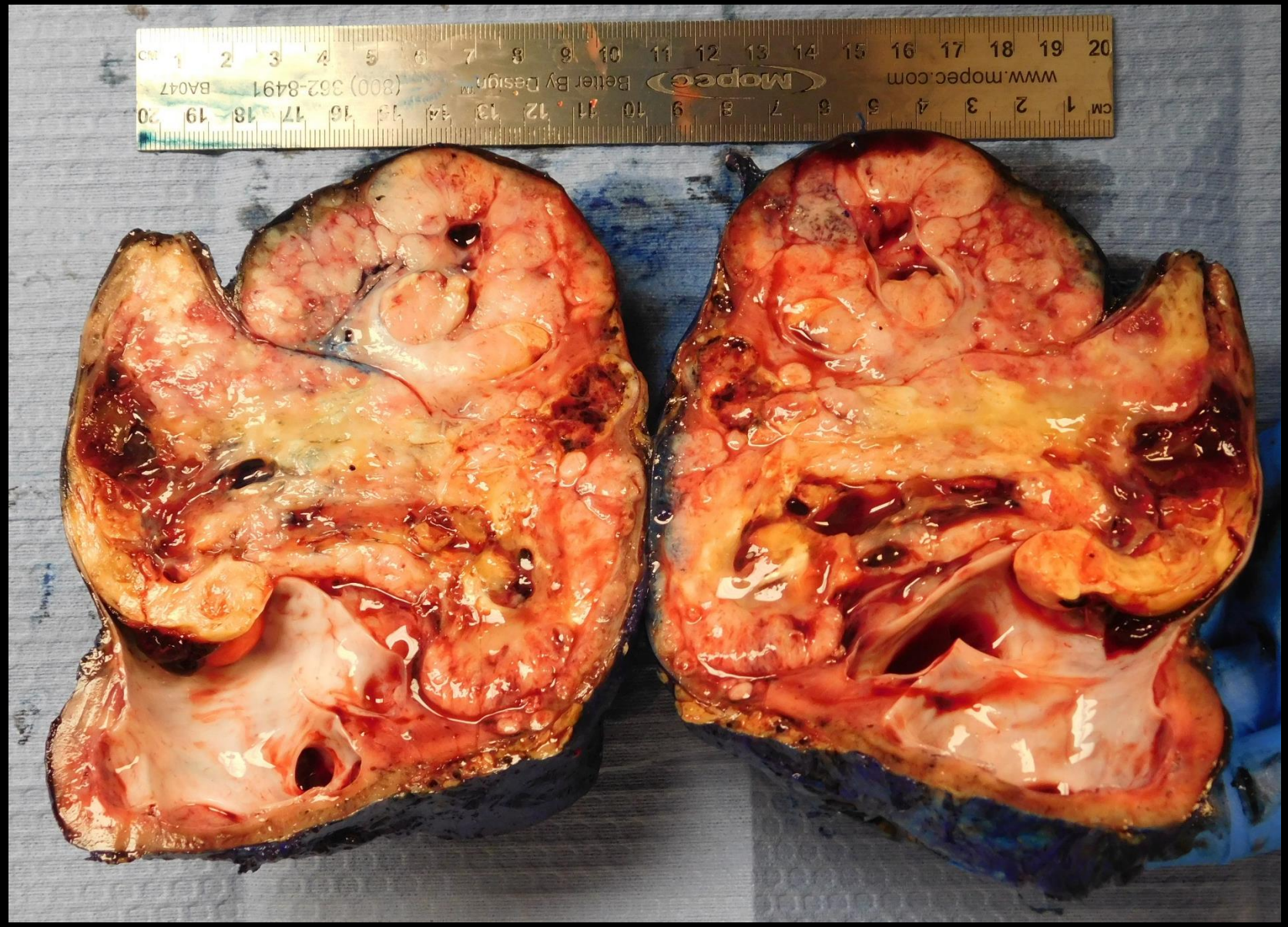
CT Abdomen w/ Contrast (labeled)

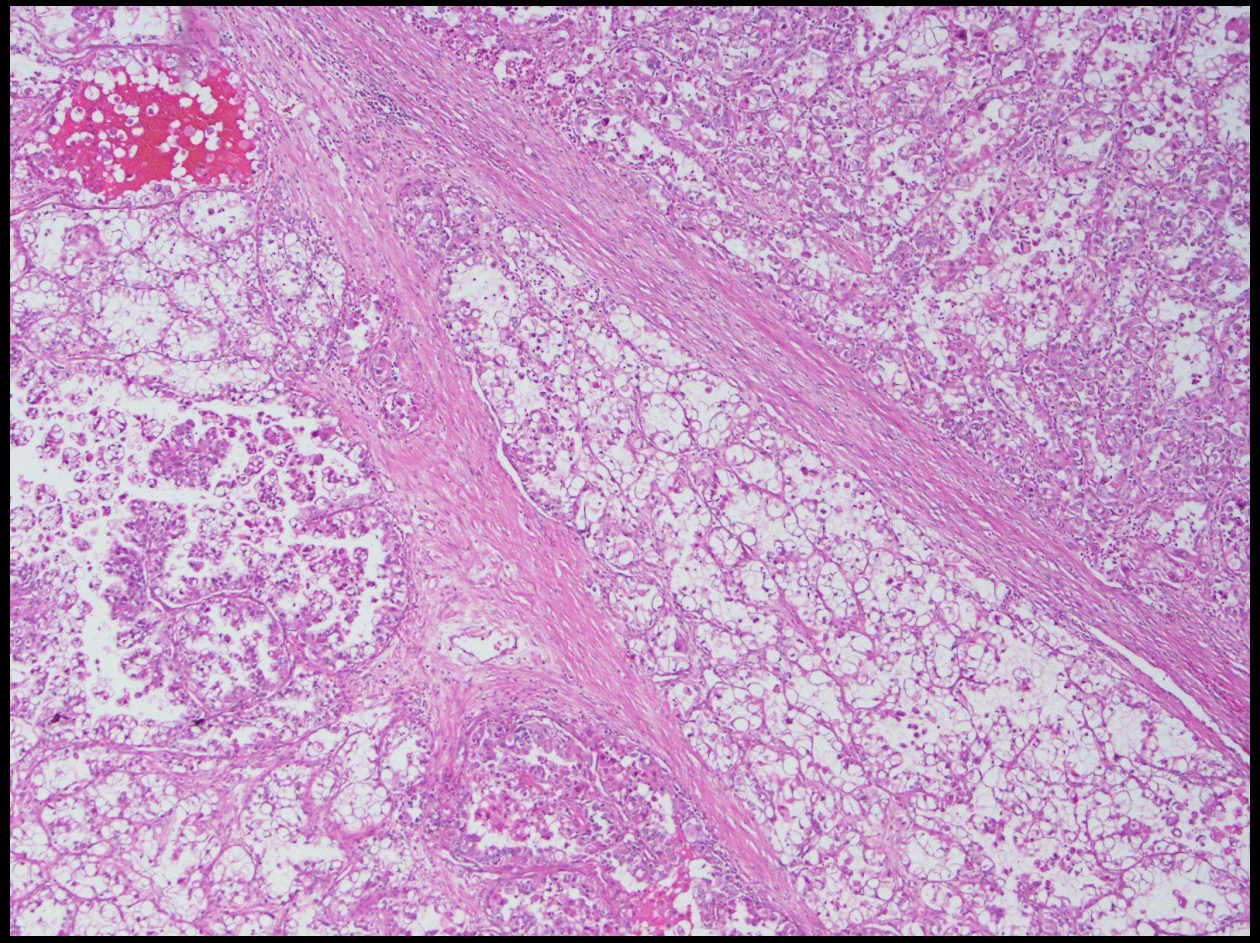


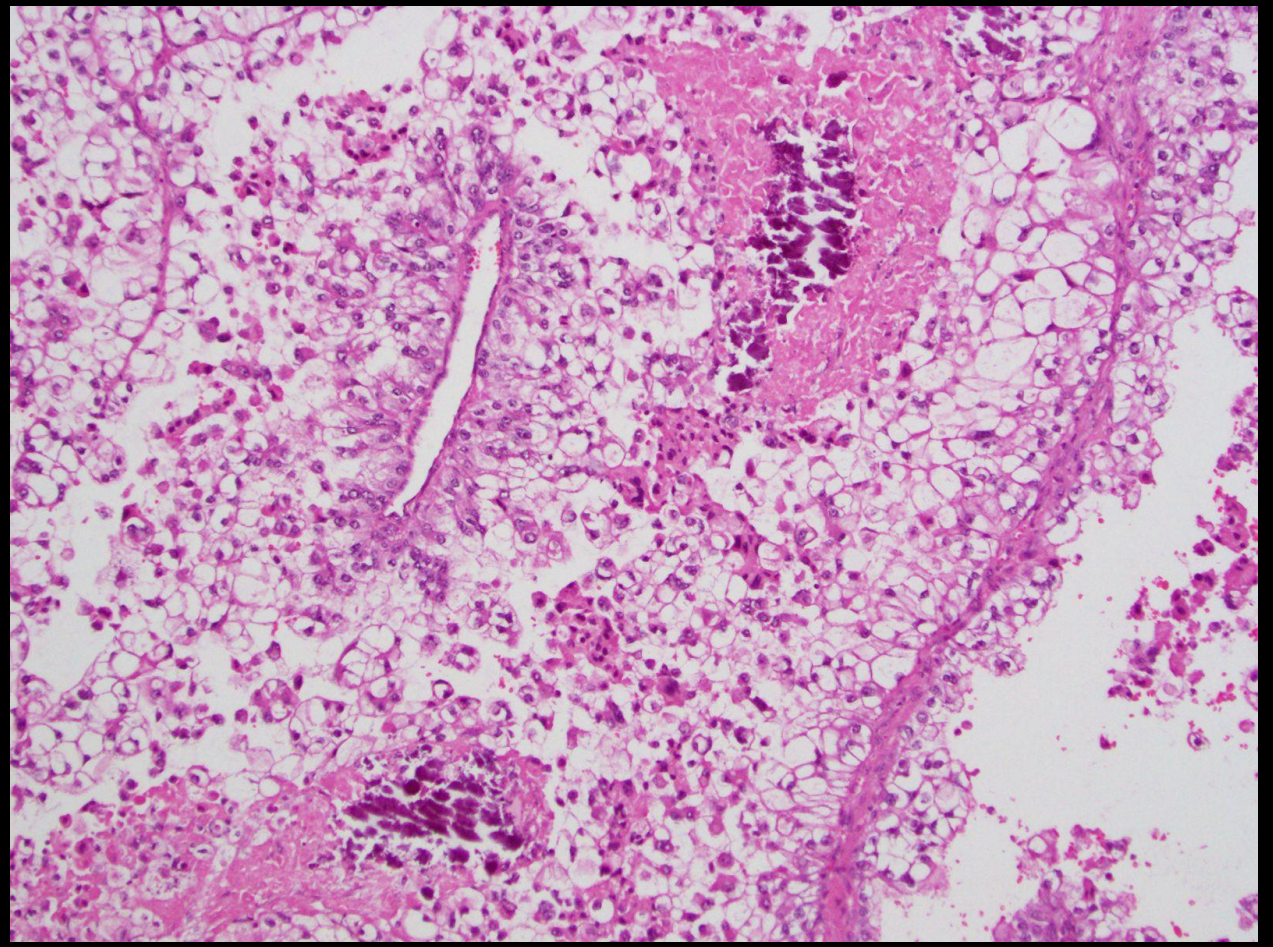
Coronal CT further demonstrating the extent and of size the neoplasm and subsequent mass effect on intraabdominal structures. Extensive retroperitoneal/para-aortic lymphadenopathy is also noted.

Pathology (unlabeled)



Pathology (unlabeled)





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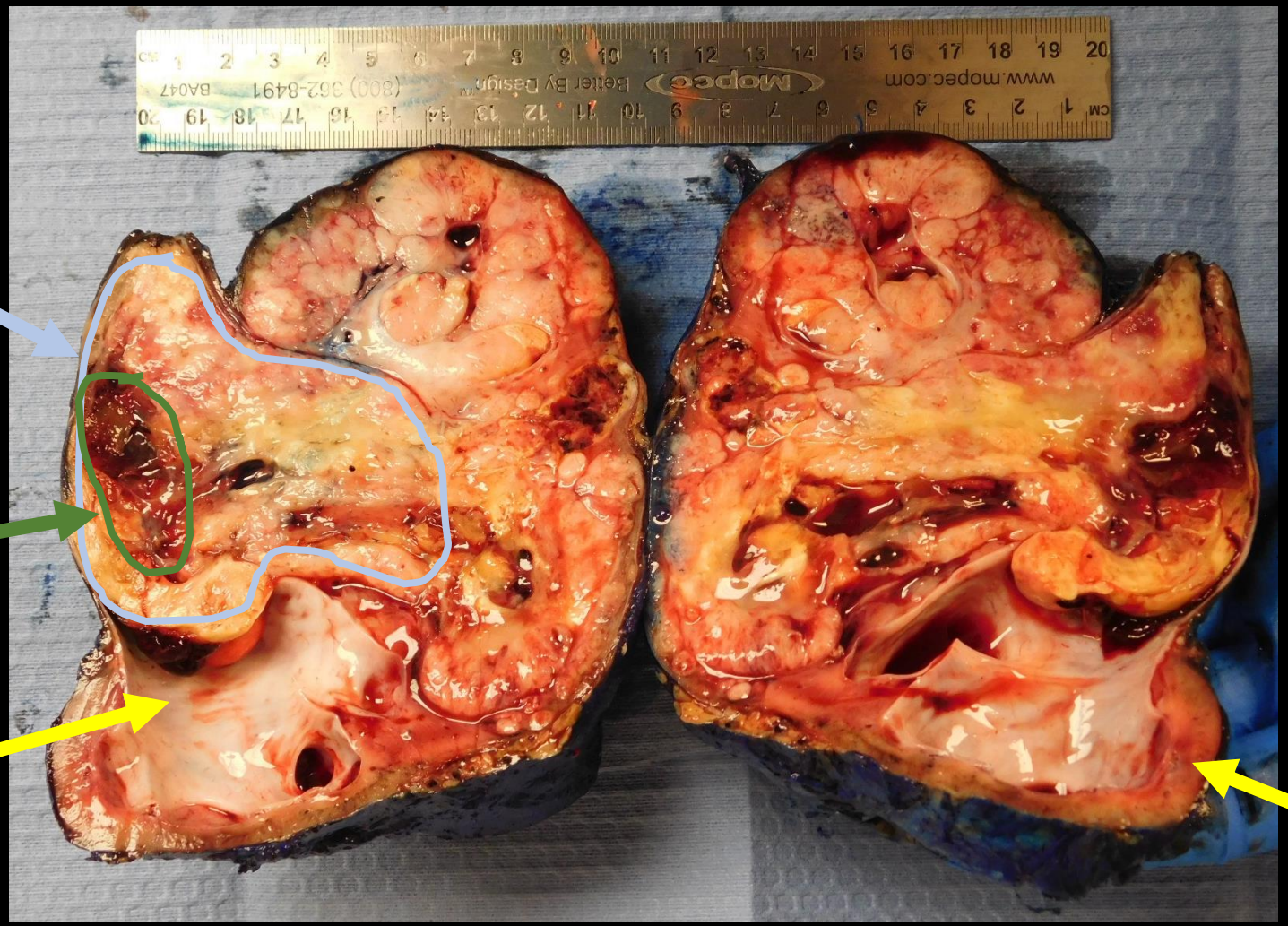
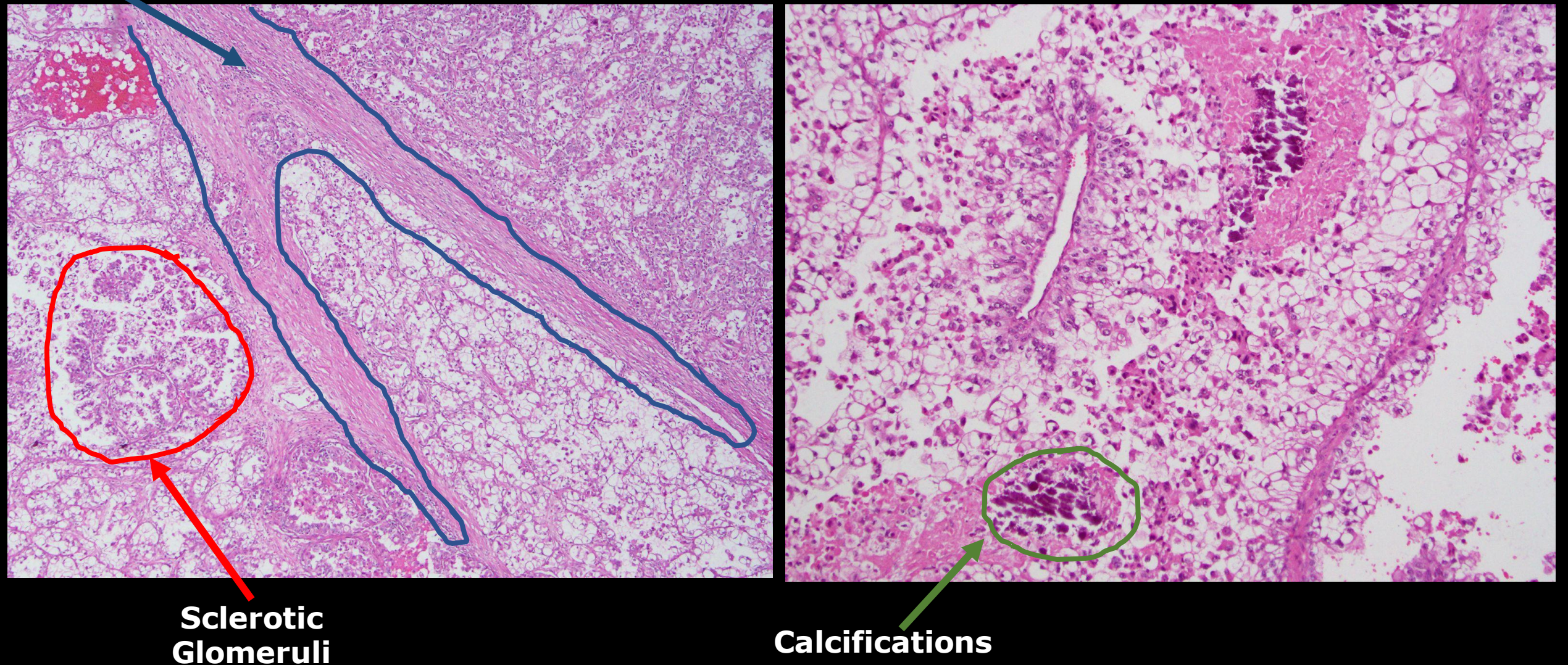


Cut gross-surgical specimen displaying hemorrhagic, yellow-tan mass invading renal parenchyma and portions of the collecting system.

Pathology (labeled)



LEFT: Medium-power histology of the mass shows sclerotic glomeruli, tubular atrophy and fibrovascular septa. RIGHT: Papillary architecture with clear cells and scattered psamommatous calcifications.

Final Dx:

TFE3-Positive Translocation Xp11 Renal Cell Carcinoma

Renal Cell Carcinoma

Clinical Features

- Classified by the histology and where tumor originates in nephron (i.e. Papillary, Chromophobe, Collecting Duct RCC)
- "Classic" triad of symptoms present in only 10-15% of cases:
  - macroscopic hematuria: 60%
  - flank pain: 40%
  - palpable flank mass: 30-40%
- May develop varicocele if tumor invades renal vein
- Mets to lung and bone most commonly

Prognosis

- Stage I-IV= 90-5% 5-year-survival
- Nephrectomy is the standard treatment; Clinical trials and/or immunotherapy trials may be considered adjuvantly
- Abdominal CT imaging surveillance should occur at 2 and 5 years for T2-T3 stage tumors

Paraneoplastic Syndromes (25% of RCC cases)

- **Hypercalcemia** from Parathyroid-Hormone Related Peptide Secretion
- **Hypertension** due to excess renin production
- **Polycythemia** due to erythropoietin secretion

Imaging of RCC

- Multiplanar reformat CT is the primary modality to detect and stage RCC
  - Non-contrast images plus post-contrast (arterial, late arterial, nephrographic, and excretory phases) should be acquired
  - Allows delineation of renal vasculature and evaluates for tumor thrombus in the renal vein/IVC
- Clear Cell RCC is typically heterogeneous and shows more enhancement
  - On average 84 HU's increase in attenuation in corticomedullary phase
- Papillary RCC is more commonly homogenous and has low enhancement
- Chromophobe RCC tends to have a peripheral pattern of enhancement but this is non-specific
- Oncocytomas cannot be reliably differentiated from RCC by imaging
- Further evaluation with Chest CT, Brain MRI, or Bone Scan if tumor is locally aggressive or there is clinical evidence of metastatic disease

Genetic Causes of Renal Tumors

Xp11 (TFE-3) Translocational RCC

- Translocation of the gene encoding Transcription Factor E3 with various partners causes overexpression of TFE3
- Most common subtype of pediatric renal neoplasms
- Increased risk after chemotherapy (Particularly DNA topoisomerase inhibitors)
- Radiographically, may have extensive psammomatous calcifications
- Commonly has a microscopic pattern of clear cells and papillary architecture

von Hippel-Lindau syndrome (Chromosome 3 Gene Deletion)

- Renal cysts and clear cell renal cell carcinoma (50% bilaterally)
- Also hemangioblastomas, visceral cysts, and pheochromocytomas

Birt-Hogg-Dubé syndrome (Mutation in Folliculin gene)

- Oncocytoma, clear cell carcinoma, or hybrid oncocytic / chromophobe tumors with areas of clear cells

Tuberous sclerosis (TSC1 and TSC2 gene mutations)

- Multiple, bilateral renal angiomyolipomas

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