AMSER Case of the Month:

32 Year Old Male With Back Pain

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Allegheny Health Network

Patient Presentation

- 32 year old male with 6 week history of back pain
- Past medical history
  - Migraine
  - Anxiety
- Family history
  - Melanoma in mother, brother, and half-brother
- No prior abdominal imaging
  - Previous XR lumbar spine and MRI lumbar spine with and without contrast
  - Indeterminate right renal mass noted on previous imaging
- No dysuria, urgency, frequency, hematuria, bothersome nocturia, or weak urine stream

Pertinent Labs

- Pre-op BMP
  - BUN 12
  - Creatinine 1.42 (elevated)

What Imaging Should We Order?

ACR Appropriateness Criteria

This imaging was ordered →

Variant 1: **Indeterminate renal mass. No contraindication to either iodinated CT contrast or gadolinium-based MR intravenous contrast. Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
US abdomen with IV contrast	Usually Appropriate	O
MRI abdomen without and with IV contrast	Usually Appropriate	O
CT abdomen without and with IV contrast	Usually Appropriate	☢☢☢☢
US kidneys retroperitoneal	May Be Appropriate	O
MRI abdomen without IV contrast	May Be Appropriate	O
CT abdomen with IV contrast	May Be Appropriate	☢☢☢
CT abdomen without IV contrast	May Be Appropriate	☢☢☢
CTU without and with IV contrast	May Be Appropriate	☢☢☢☢
Arteriography kidney	Usually Not Appropriate	☢☢☢
Radiography intravenous urography	Usually Not Appropriate	☢☢☢
Image-guided biopsy adrenal gland	Usually Not Appropriate	Varies
MRU without and with IV contrast	Usually Not Appropriate	O

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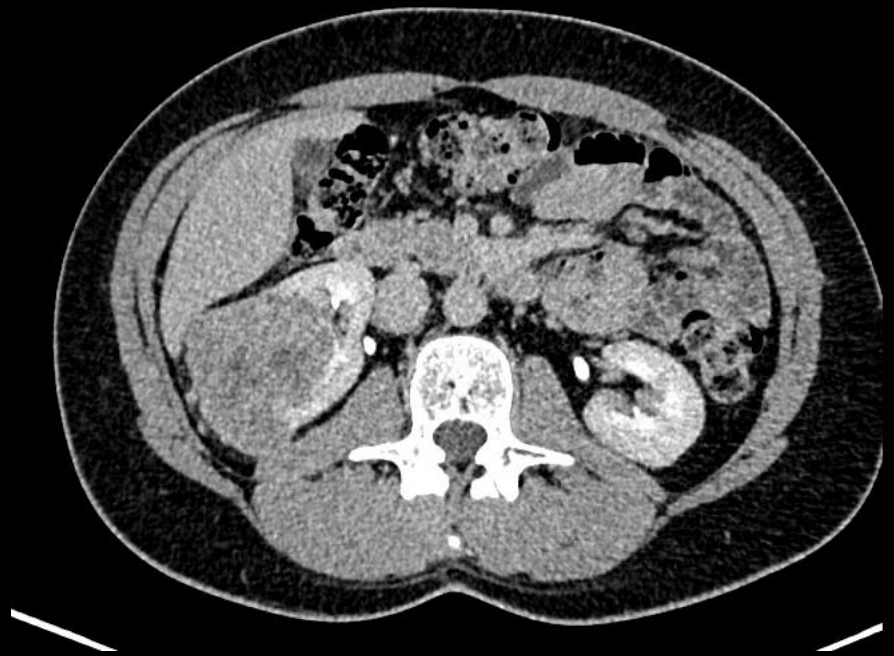
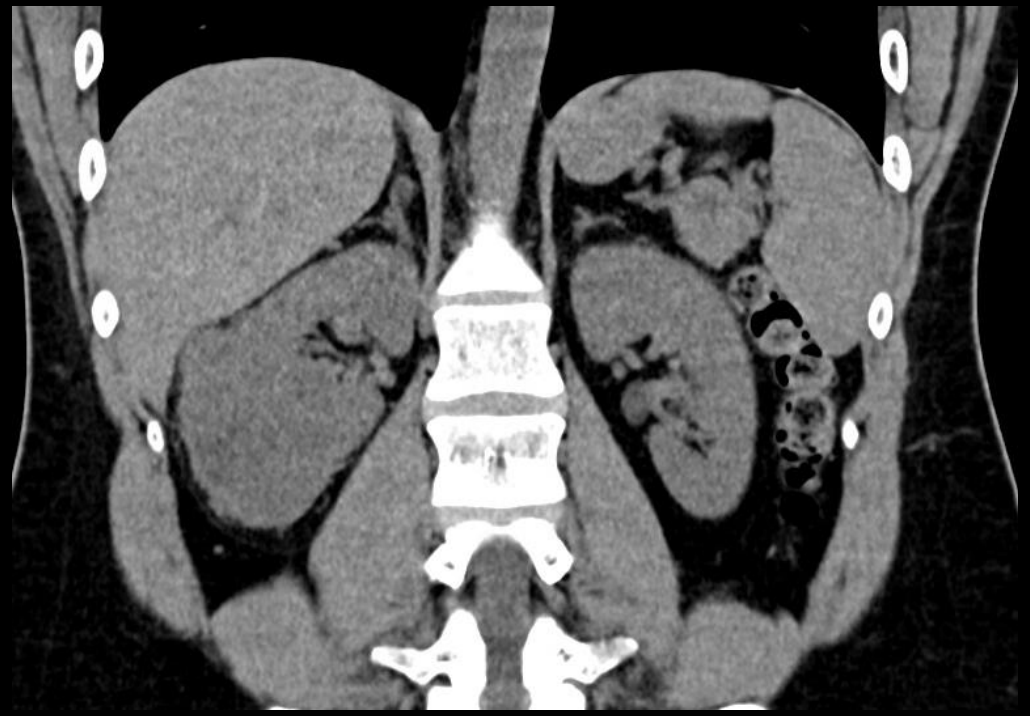
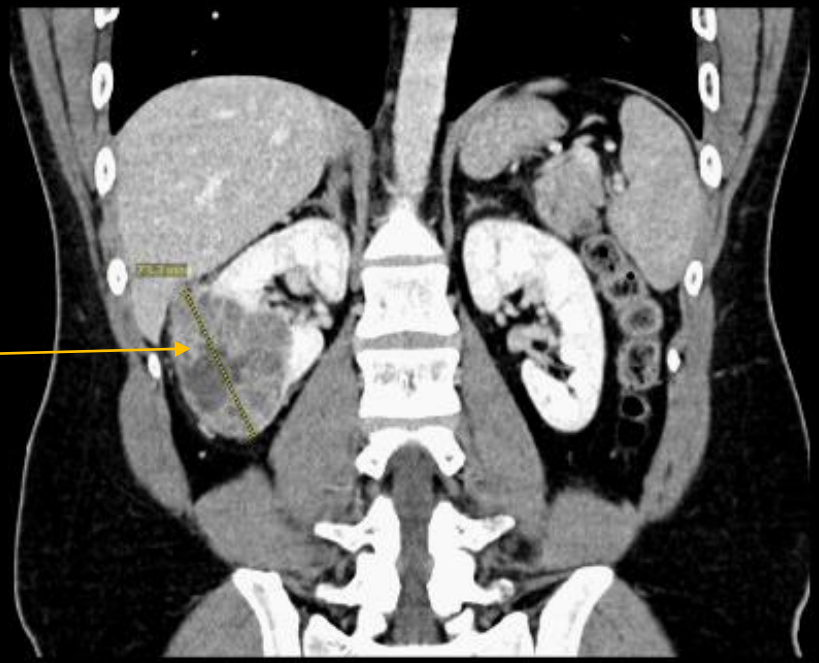
Findings (unlabeled)





Findings: (labeled)

- Non-contrast abdominal CT
- Findings: Right kidney, lobulated hyper-vascular exophytic mass
  - Measuring 7x7x6 cm
  - Assumed RCC
  - Abuts the medial inferior hepatic tip

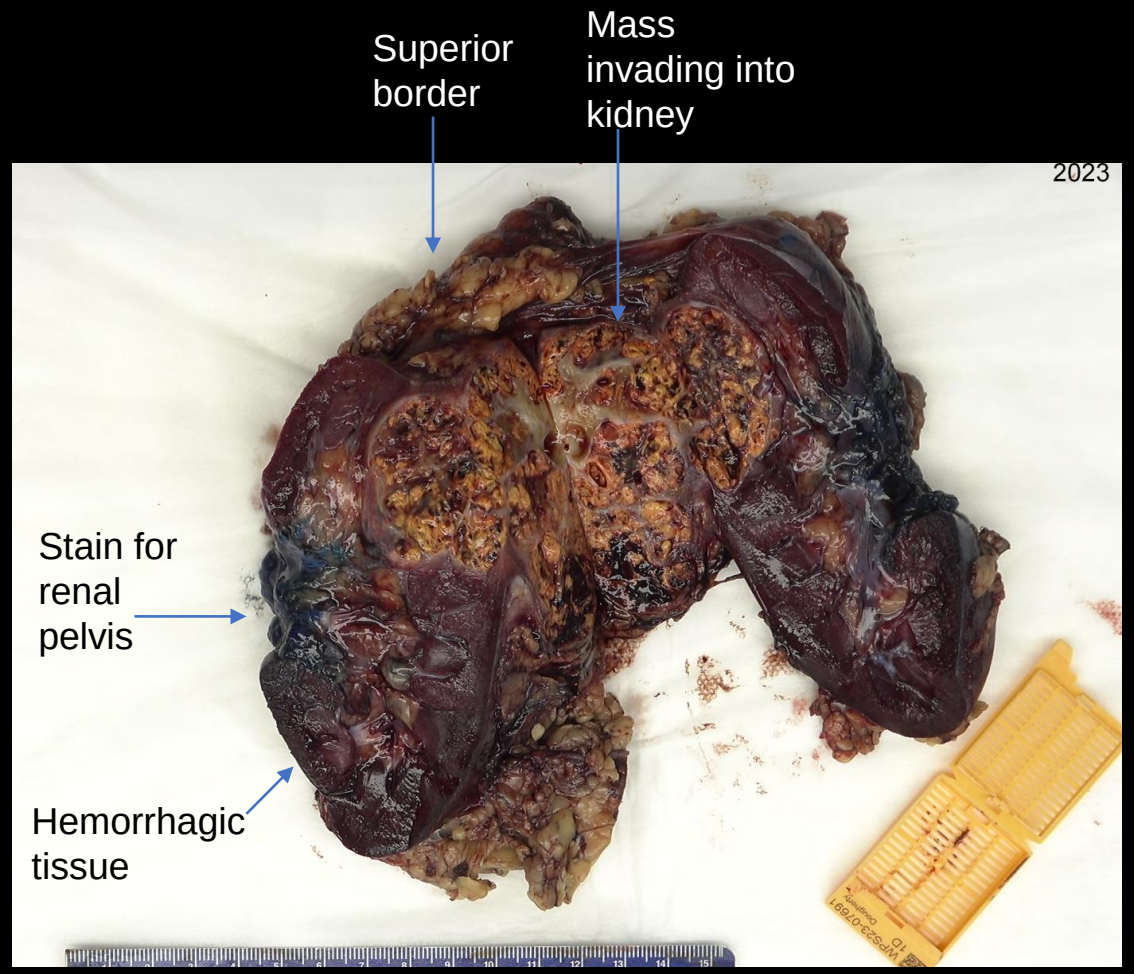


Differential Diagnosis

- Renal cell carcinoma
  - Clear cell type is focally positive for CD10
- Oncocytoma
  - Negative for CD10
- Leiomyoma
- Metastases

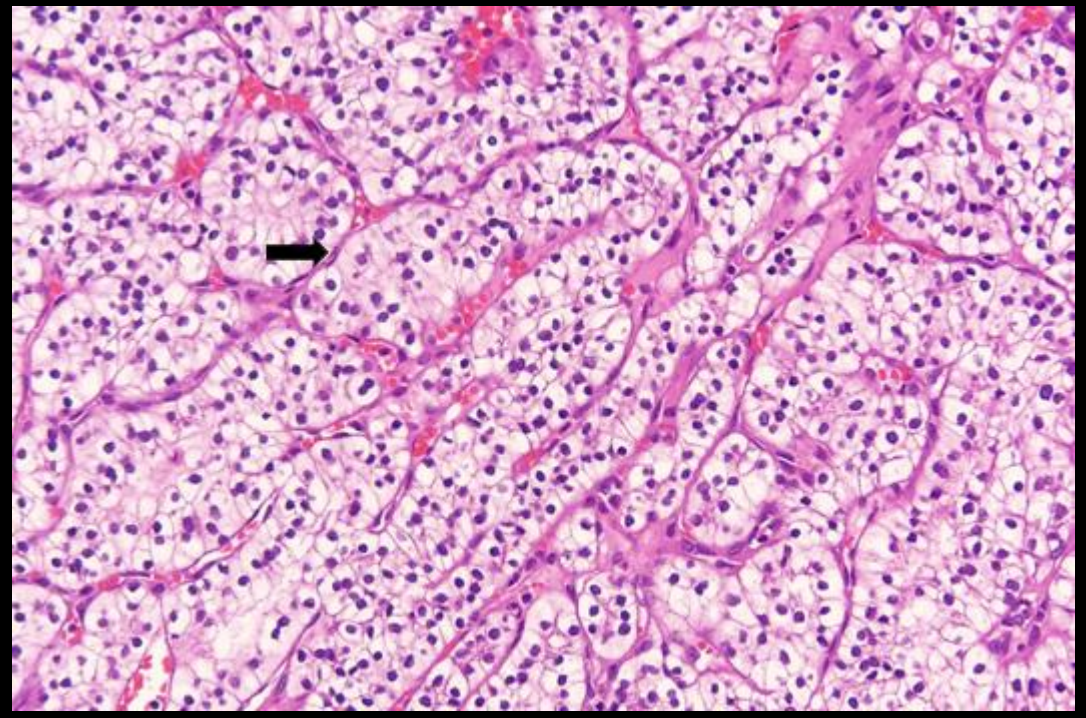
Gross Pathology

- Bivalve of the right kidney with an exophytic mass
- Tissue is grossly hemorrhagic
- Gross pathology consistent with renal cell carcinoma



Pathology

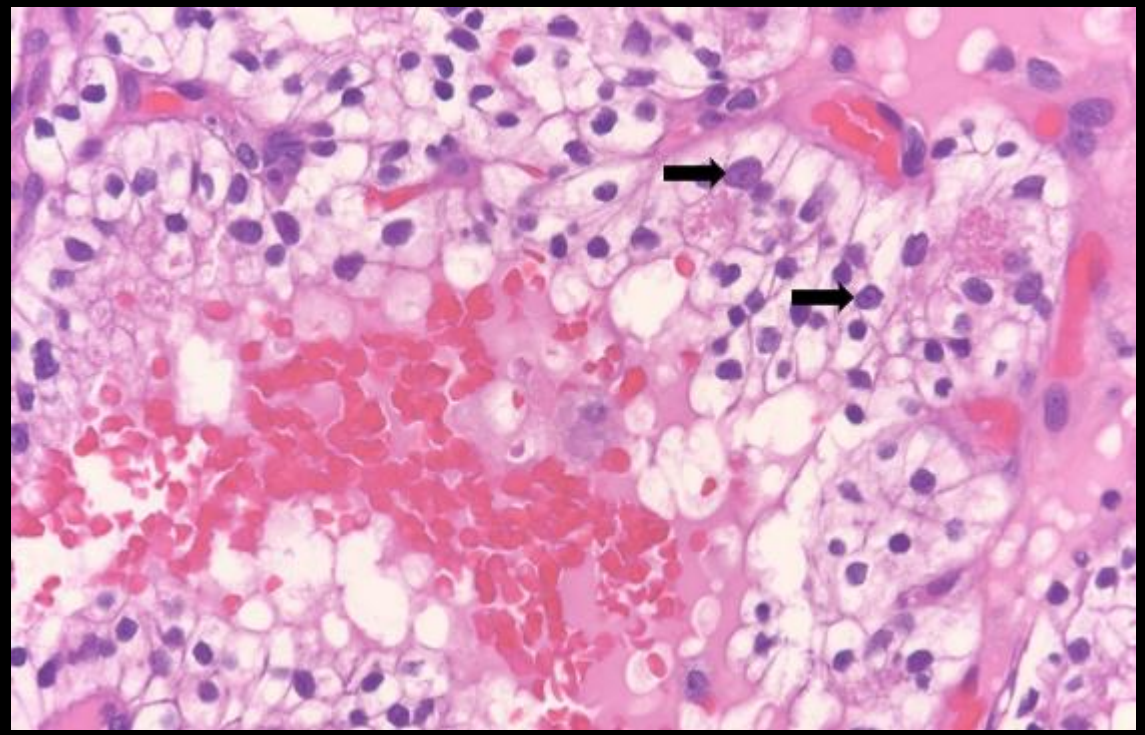
- Right renal cell carcinoma
  - Clear cell type
    - CD10+
    - CK7-
  - Invasion into peri-nephritic tissue
    - Beyond the renal capsule



Nests separated by delicate chicken-wire-like fibrovascular structures (black arrow)

Pathology

- ISUP nucleolar grade 2 of 4
  - Based on nucleoli appearance at 400x
- Background renal parenchyma shows no significant pathological findings

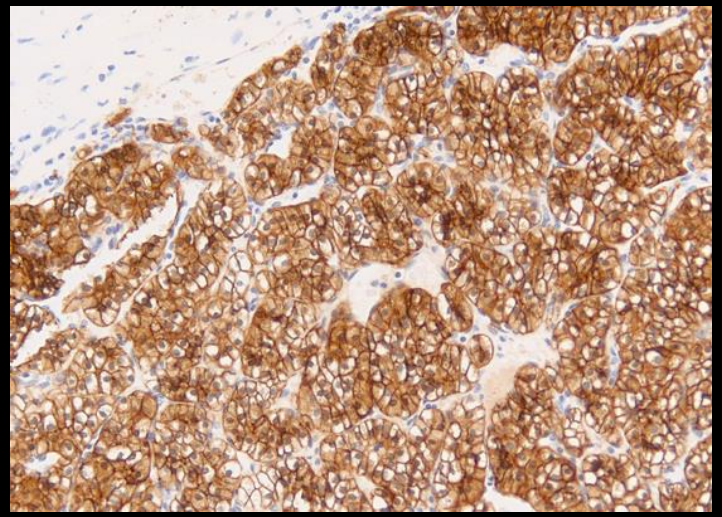


Conspicuous eosinophilic nucleoli visible at 400x (black arrows).

Not prominent at 100x

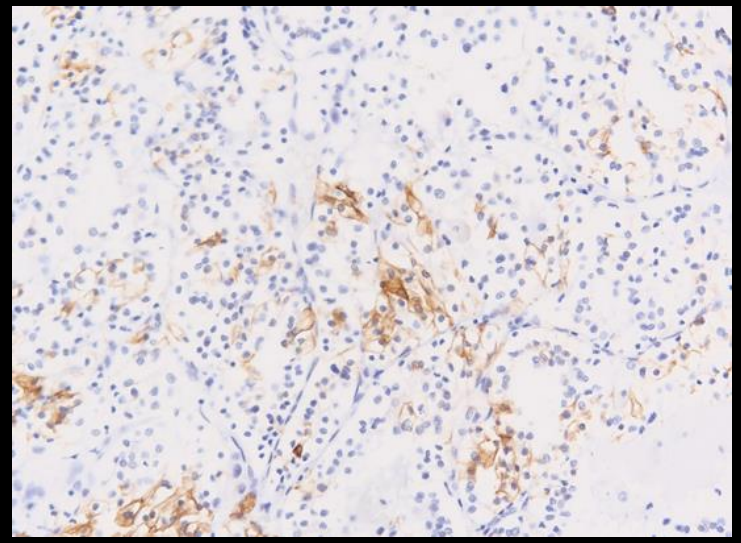
Pathology

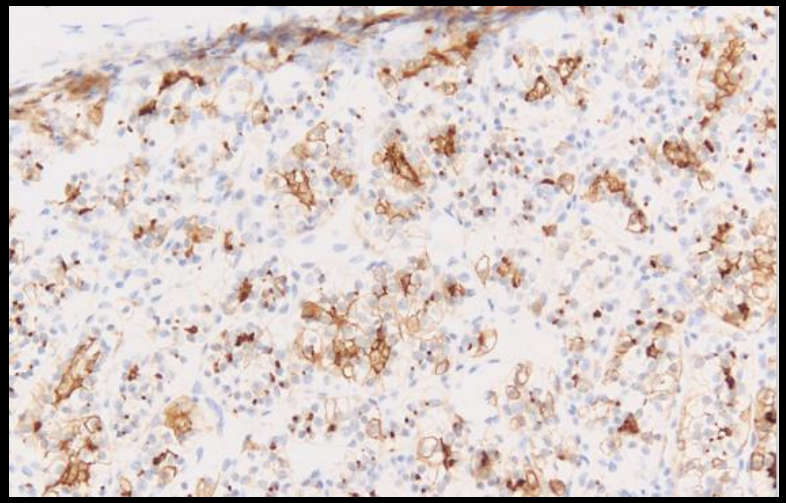
Positive for carbonic anhydrase IX and CD10. CK7 positive in cystic areas.
Consistent with clear cell RCC



Diffusely and strongly positive for CA-IX

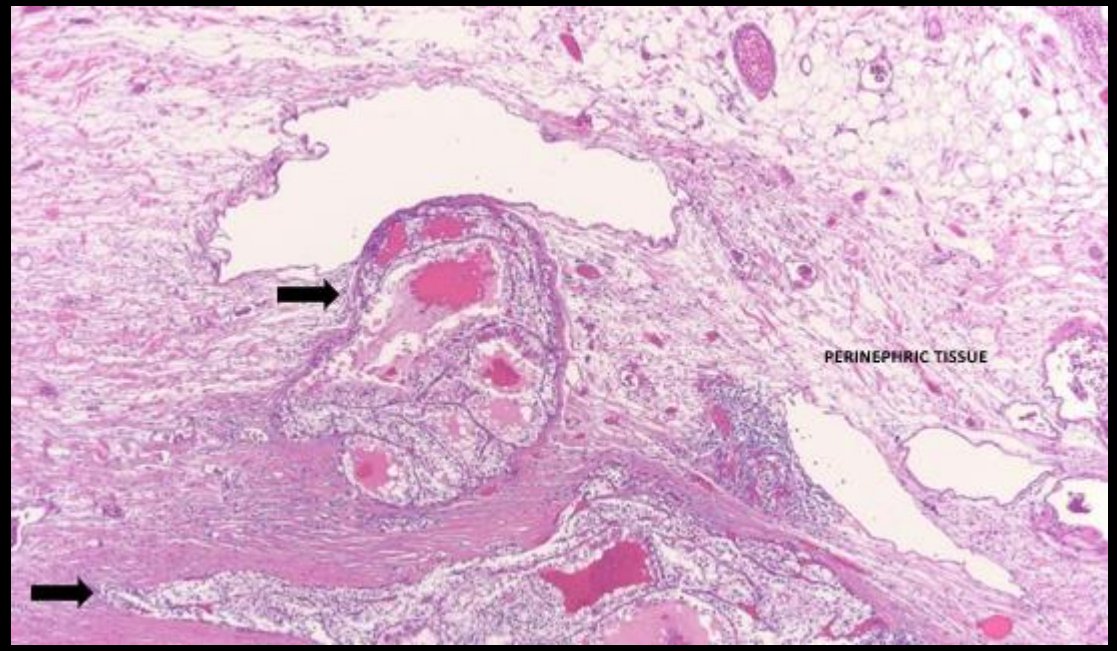
Focally positive for CD10



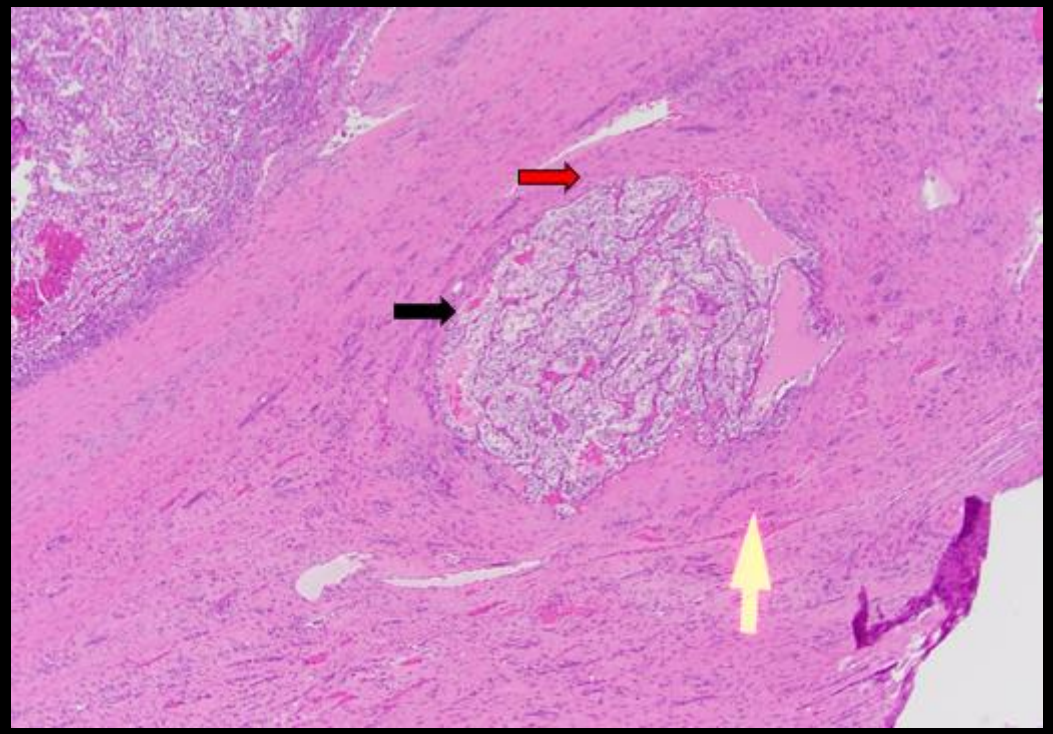


Negative for CK7

Pathology- Staging and Invasion



Invasion into and beyond the renal capsule (black arrows) and into the perinephric tissue
Stage pT3a (AJCC 8th Edition)



Small foci of tumor (black arrow) which is concerning for possible lymphovascular invasion. CD31+ which is also consistent with lymphovascular invasion.

Red arrow- ring-like possible vessel with has been invaded by tumor

Final Dx:

Renal Cell Carcinoma, Clear Cell Type

Case Discussion

- Imaging was consistent with renal cell carcinoma
  - Stage 3 RCC is generally treated with radical nephrectomy
    - Treatment is often curative
- Radical right nephrectomy was performed
  - Pathology report: “All surgical margins of resection are negative for carcinoma”
- Stage
  - pT3aN0
  - Treatment consistent with recommendation based on post-operative staging
  - Role of imaging for staging of RCC
    - CT with and without contrast can add valuable staging information

Case Discussion

Table 1: Reporting Elements for Indeterminate Renal Masses

Reporting Elements	Guidance	Importance
Basic Characteristics		
Size	Provide three orthogonal dimensions	Essential
Mass type	Describe solid vs cystic	Essential
Macroscopic fat	Describe present vs absent	Essential
Enhancement	Provide quantitative numbers	Essential
Laterality	Describe right vs left	Important
Change in size over time	Describe change in size from most recent and oldest examinations	Important
Bosniak classification	Provide specific category	Important
Margins	Describe >90% well-marginated vs infiltrative	Important
Necrosis	Provide estimated percentage	Optional
MRI microscopic fat	Describe present vs absent	Optional
MRI signal characteristics	Describe signal characteristics	Optional
Location		
Capsular location	Describe endophytic, <50% exophytic, or >50% exophytic	Important
Polar location	Describe upper vs lower	Important
Polar extent	Describe 100% polar, >50% polar, or <50% polar	Important
Axial location	Describe anterior vs posterior	Important
Distance to sinus fat	Provide distance measurement	Important
Detailed axial location	Provide more detail for interventional planning	Optional
Invasiveness		
Collecting system	Describe no invasion or invasion	Important
Perirenal fat	Describe no invasion or invasion	Important
Perirenal fascia	Describe no contact, contact, or invasion	Important
Adjacent organs	Describe no contact, contact, or invasion	Important
Vascular		
Venous thrombus	Describe present vs absent; give detail on anatomic extent	Important
Venous anatomy	Describe major ipsilateral veins	Optional
Arterial anatomy	Describe major ipsilateral arteries and identify early branches	Optional
Other		
RENAL Nephrometry Score (5)	Provide details and final score	Optional
Clear cell likelihood score	Provide T2-weighted imaging and enhancement characteristics	Optional
Favored histology	Provide likelihood of favored histology	Optional
Follow-up recommendations	Use AUA guidelines (6)	Optional

Source.—Reference 7. Definitions of terms are described in more detail in reference 7.
Note.—AUA = American Urological Association, RENAL = radius, exophytic or endophytic, nearness to collecting system or sinus, anterior or posterior, and location relative to polar lines.

Table 2: AJCC TNM Staging System

Category	Definition
Tumor	
Tx	Primary tumor cannot be assessed
T0	No evidence of primary tumor
T1	Primary tumor is ≤7 cm in greatest dimension and confined within the renal capsule
T1a	Primary tumor is ≤4 cm in greatest dimension and confined within the renal capsule
T1b	Primary tumor is >4 but ≤7 cm in greatest dimension and confined within the renal capsule
T2	Primary tumor is >7 cm in greatest dimension and confined within the renal capsule
T2a	Primary tumor is >7 cm but ≤10 cm in greatest dimension and confined within the renal capsule
T2b	Primary tumor is >10 cm in greatest dimension and confined within the renal capsule
T3	Primary tumor extends into major veins or perinephric tissues but not into the ipsilateral adrenal gland and not beyond the perirenal (Gerota) fascia
T3a	Primary tumor extends into the renal vein, renal sinus fat, and renal capsule but not beyond the perirenal (Gerota) fascia
T3b	Primary tumor invades the IVC below the diaphragm
T3c	Primary tumor invades the IVC above the diaphragm
T4	Primary tumor invades beyond the perirenal (Gerota) fascia or invades the ipsilateral adrenal gland
Lymph nodes	
Nx	Lymph nodes cannot be assessed
N0	No regional (retroperitoneal) lymph node metastasis
N1	Regional (retroperitoneal) lymph node metastasis
Distant metastasis	
M0	No distant metastasis
M1	Distant lymph node or other metastasis, including noncontinuous adrenal involvement

Source.—Reference 15.
Note.—AJCC = American Joint Committee on Cancer, IVC = inferior vena cava.

T3a staging of the tumor

Case Discussion

- Pathology was consistent with VHL gene mutation
  - Clear cell renal carcinomas show loss of the short arm of chromosome 3
    - This site includes the VHL gene which predisposes to cancer due to loss of heterozygosity
  - “lesional cells are diffusely and strongly positive for CA-IX”
    - Seen with sporadic or inherited VHL RCC
  - Von Hippel-Lindau Syndrome
    - Autosomal dominant inheritance
      - Germline VHL mutation
    - Hemangioblastomas, RCC, pheochromocytomas, pancreatic cysts, neuroendocrine tumors, endolymphatic sac tumors, and epididymal and broad ligament cysts.
    - Early detection important
      - Removal of tumors can prevent disease complications

Case Discussion

Other genetic disorder associated with renal cell carcinoma

- Tuberous Sclerosis Complex
  - Autosomal dominant inheritance
    - Mutations in TSC1 and TSC2
  - RCC presentation at a younger age

References:

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