AMSER Case of the Month
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52-year-old-woman with fatigue and altered mental status

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Patient Presentation

- A 52-year-old woman presents to emergency department for a lab test showing elevated WBC at 29, fatigue and dizziness. Her current risk factors include VP shunts. She was seen by the neurosurgical service one day prior for shunt flow rate adjustment. She currently reports dizziness, headache, and not feeling right for the last week, but remains confused as to why she is in the ED.

Patient Presentation

- Pertinent past medical history includes MAC CNS infection, seizures, recurrent urinary tract infections, and problems with her left kidney
- Surgical history includes IVC Filter placement and VP shunt with multiple revisions
- Physical Exam:
  - Vitals: BP(104/44), Temp(98.4 F), Pulse: 90 bpm, Resp: 18
  - Head: Atraumatic, occiput with shunts in place, mild tenderness to palpation
  - Neurological: A&O x2, unaware of date/president or events
    - Strength: 3/5 LLE(baseline) and 5/5 RLE
  - No other relevant physical exam findings

Pertinent Labs

- BMP
  - BUN: 26
  - Cr: 0.8
  - Na, K, Cl, bicarb, anion gap, glucose, Ca, Mg, PO4 within normal limits
- CBC:
  - Hemoglobin: 9.5
  - WBC: 23.2
  - PLT: 492
- Urinalysis:
  - Bacteria/HPF: Few
  - Leukocytes: small
  - WBC/HPF: 26 (nl 0-5)
  - Other values wnl
- Urine culture
  - Final report: No growth at 1000 CFU/mL

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What Imaging Should We Order?

Select the applicable ACR Appropriateness Criteria

Variant 2: **Suspected acute pyelonephritis. Complicated patient (eg, recurrent pyelonephritis, diabetes, immune compromise, advanced age, vesicoureteral reflux, or lack of response to initial therapy). Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
CT abdomen and pelvis with IV contrast	Usually Appropriate	☢☢☢
US abdomen	May Be Appropriate	O
US color Doppler kidneys and bladder retroperitoneal	May Be Appropriate	O
MRI abdomen and pelvis without and with IV contrast	May Be Appropriate	O
MRI abdomen and pelvis without IV contrast	May Be Appropriate	O
CT abdomen and pelvis without IV contrast	May Be Appropriate	☢☢☢
CT abdomen with IV contrast	May Be Appropriate (Disagreement)	☢☢☢
CT abdomen and pelvis without and with IV contrast	May Be Appropriate (Disagreement)	☢☢☢☢
Fluoroscopy voiding cystourethrography	Usually Not Appropriate	☢☢
Radiography abdomen and pelvis	Usually Not Appropriate	☢☢
Fluoroscopy antegrade pyelography	Usually Not Appropriate	☢☢☢
Radiography intravenous urography	Usually Not Appropriate	☢☢☢
MRI abdomen without and with IV contrast	Usually Not Appropriate	O
MRI abdomen without IV contrast	Usually Not Appropriate	O
MRU without and with IV contrast	Usually Not Appropriate	O
MRU without IV contrast	Usually Not Appropriate	O
CT abdomen without IV contrast	Usually Not Appropriate	☢☢☢
DMSA renal scan	Usually Not Appropriate	☢☢☢
CT abdomen without and with IV contrast	Usually Not Appropriate	☢☢☢☢
CTU without and with IV contrast	Usually Not Appropriate	☢☢☢☢

This was ordered by the hospitalist to rule out acute pyelonephritis

Findings (unlabeled)

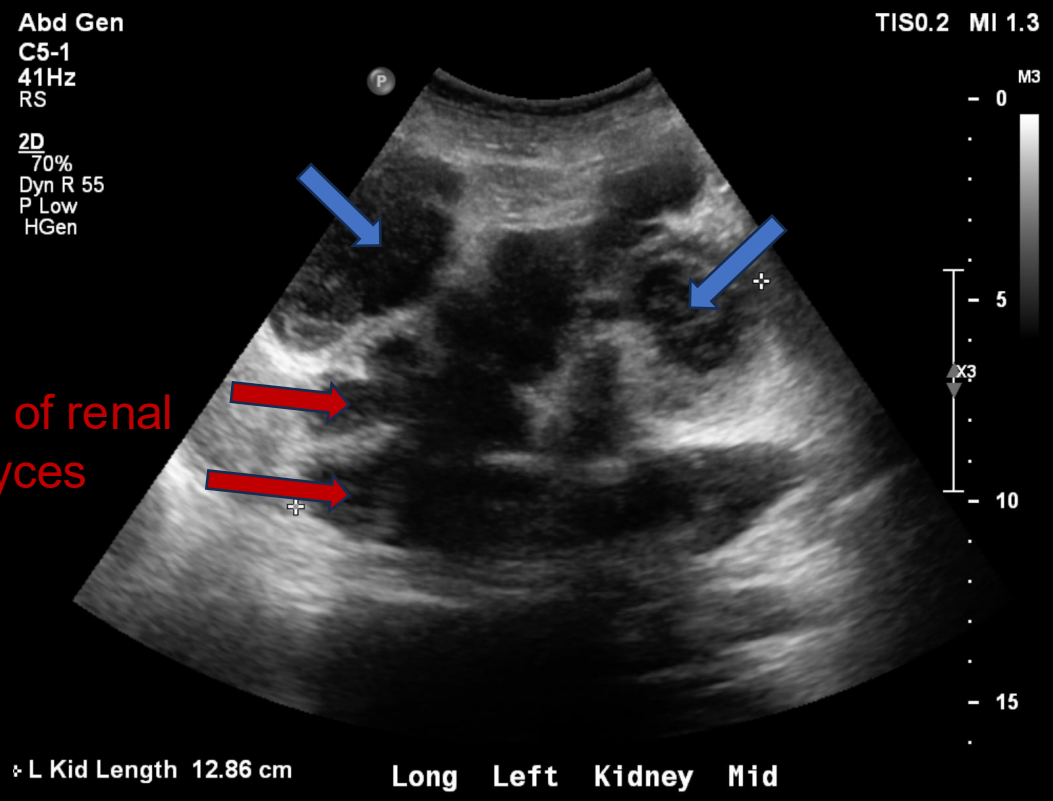
Findings (unlabeled)

Findings (labeled)

6.0 x 3.4 x 2.0 cm mass

Findings (labeled)

Heterogenous hypoechoic/anechoic lesions



Dilation of renal calyces

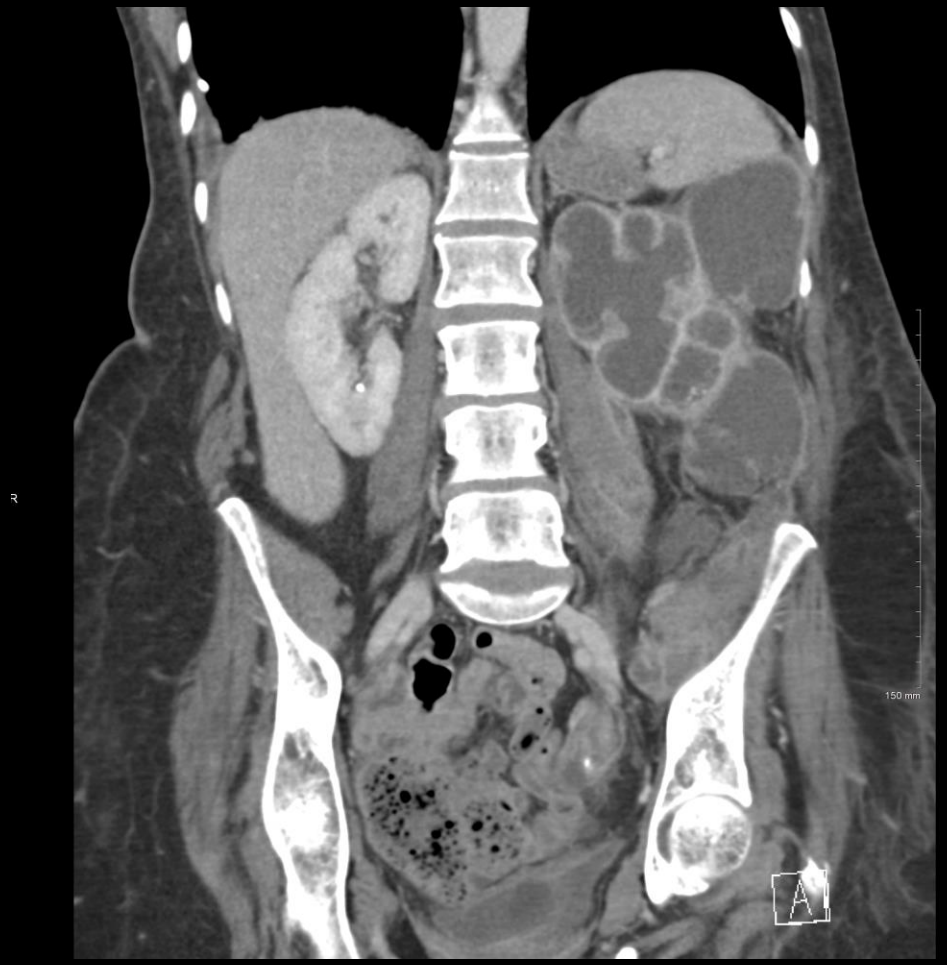
Select the applicable ACR Appropriateness Criteria

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

This imaging modality was ordered to further evaluate the renal findings identified on ultrasound

Findings (unlabeled)



Coronal CT Scan

Findings (unlabeled)

Axial CT

Coronal CT

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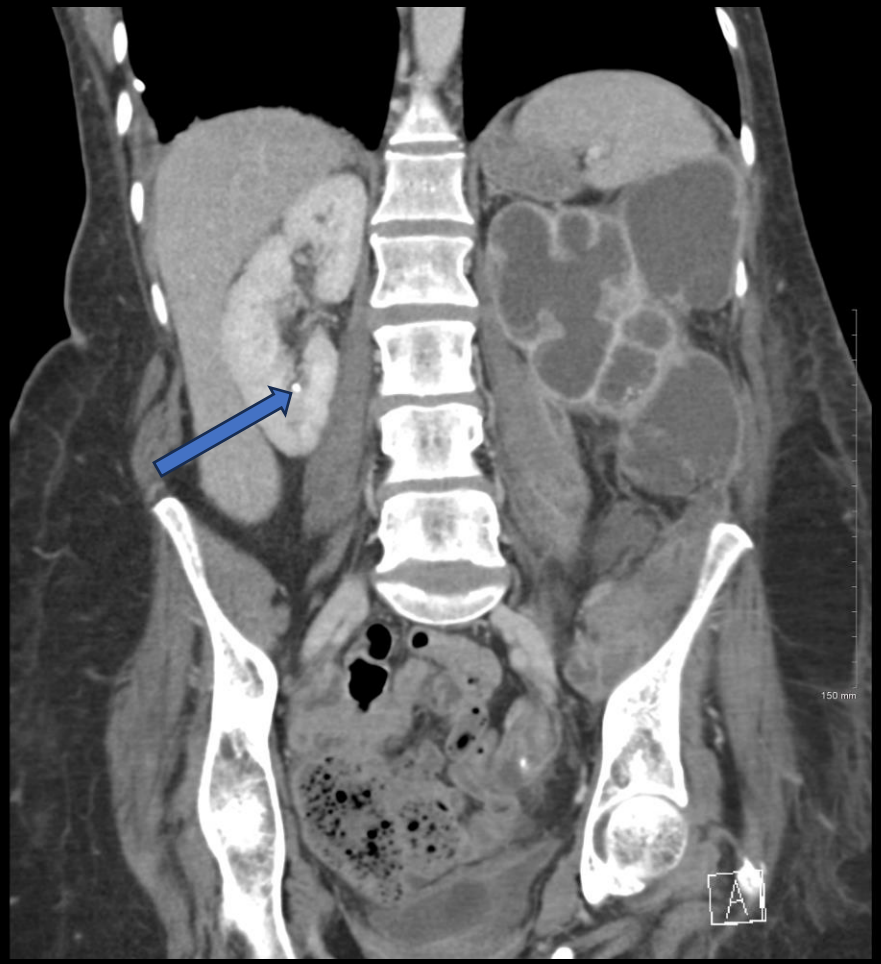
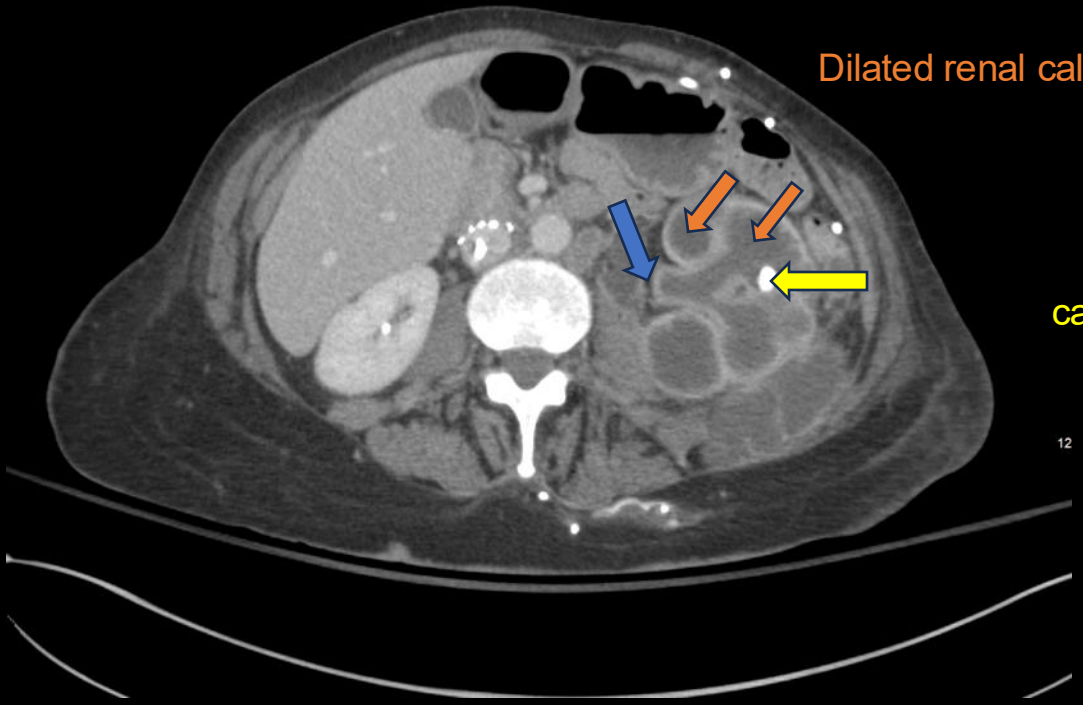
Findings (labeled)

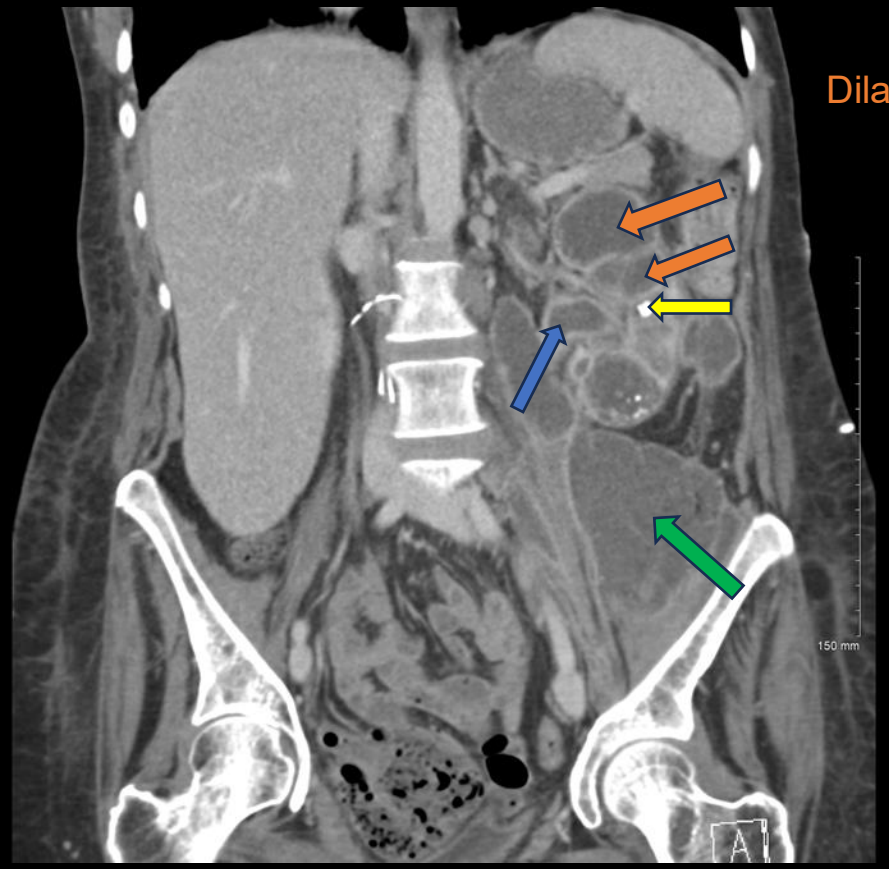
Non-obstructing 5 mm renal calculus



CT shows no mass involving right kidney. The right renal mass on ultrasound turned out to be a pseudomass.

Findings (labeled)





Differential Diagnosis

- Renal abscess
- Renal cell carcinoma
- Xanthogranulomatous pyelonephritis
- Renal tuberculosis

Though these conditions can have clinically similar presentations, differences can be seen on CT and ultrasound imaging. The presence of multiple low attenuation rounded spaces (dilated renal calyces and pelvis) surrounded by a rim of enhancing tissue, also known as the bear paw sign, indicates…

Final Dx:
Xanthogranulomatous Pyelonephritis

Case Discussion: Pathophysiology

- Xanthogranulomatous pyelonephritis(XGP) is a rare, aggressive variant of pyelonephritis[2]
- Chronic urinary obstruction and UTI are thought to play a role in development[2]
  - MCC organisms include E. Coli, Proteus Mirabilus, Pseudomonas, E. Faecalis, and Klebsiella
  - Primary cause of urinary obstruction is nephrolithiasis
    - Staghorn calculus is found in 80% of patients[2]
- Risk factors include immunocompromise, recurrent UTI, ureteropelvic junction obstruction, and vesicoureteral reflux[2]
- Symptoms include flank pain, malaise, and fever[2]
  - Urinary symptoms may include dysuria, hematuria, increased urinary frequency
  - Anorexia, chills, weight loss, and malaise

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Case Discussion: Workup

- Lab findings associated with XGP may include:
  - CBC: anemia and leukocytosis[2]
  - Elevated ESR and CRP[2]
  - Renal function tests may show elevated BUN and creatinine[2]

Case Discussion: Workup

- Imaging Findings:
  - Ultrasound:
    - Enlarged with distortion of renal outline; central shadowing calculus may be seen[3]
    - Multiple fluid collections corresponding to dilated calyces and parenchymal destruction[3]
  - CT
    - Bear paw sign[4]
      - Replacement of renal parenchyma by the infectious process leading to hypoattenuating masses arranged in a “hydronephrotic” pattern[4]
      - This appearance resembles a bear's paw
      - Loss of normal renal outline, enlarged with paradoxical contracted renal pelvis[5]
    - Focal forms may show a low attenuation mass with associated calculus adjacent to a calyx[5]
  - MRI
    - Heterogenous signal on all sequences[5]

Case Discussion: Treatment and Prognosis

- Staging: Classified into focal, segmental, and diffuse forms[2]
  - Focal is further split into Stages
    - Stage 1(Nephritic) – Disease limited to the kidney
    - Stage 2(Perinephric) – Disease involves renal pelvis or perinephric fat within Gerota's fascia
    - Stage3(Paranephric) – Involves a wider area, including the retroperitoneum
- XGP may present with extrarenal extension[3]
  - Extension may be into the perirenal space, ipsilateral psoas muscle, diaphragm, posterior abdominal wall, bowel
- XGP is generally managed through nephrectomy[1]
  - Due to inflammation which affects renal function and inflammation which obscures surgical planes
  - It is unclear whether partial vs total nephrectomy and robotic/laparoscopic vs open is superior
  - Treatment with antibiotics and drainage alone is rare[1]
    - Likely due to major destruction of the affected kidney
    - NM renal scans typically show little renal function in the affected kidney in 80% of cases
- With prompt identification and treatment, prognosis is improved[1]
  - Unilateral XGP is associated with better outcomes than bilateral XGP which is often fatal

References:

1. Jang TL, McKoy T, Hakim J, Polenakovik HM. Xanthogranulomatous pyelonephritis – A diagnostic and therapeutic dilemma. *The American Journal of the Medical Sciences*. 2023;365(3):294-301. doi:https://doi.org/10.1016/j.amjms.2022.11.004
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