

# AMSER Rad Path

## Case of the Month:

70-year-old man with left lower quadrant fullness and bright red blood in his stool

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# Patient Presentation

- The patient is a 70 yo M with a history of sialoadenitis (s/p biopsy 1999, sclerosing sialoadenitis) presents with bright red blood in his stool and subjective fullness in this left lower quadrant.
- **Past Medical History:** Type II IDDM, Facioscapulohumeral muscular dystrophy (FSHD), 10 years dry eyes, fatigue, 8 lb weight loss in past year (128 to 120)
- **Past Surgical History:** CCY in Hong Kong, 2015
- **Social History:** Denies tobacco, alcohol, or recreational drug use
- **Physical Exam:** Remarkable for LLQ palpable mass

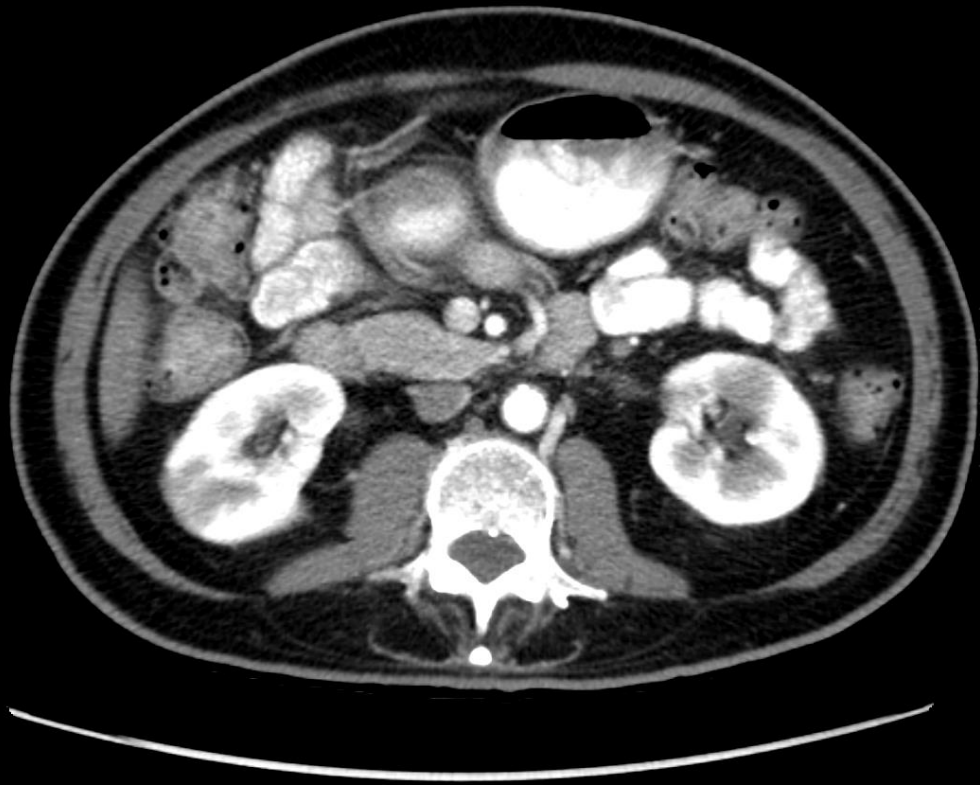
# ACR Imaging Guidelines

**American College of Radiology  
ACR Appropriateness Criteria®  
Palpable Abdominal Mass-Suspected Neoplasm**

**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      | ○                        |
| MRI abdomen without and with IV contrast                    | May Be Appropriate       | ○                        |
| CT abdomen without IV contrast                              | May Be Appropriate       | ⊕⊕⊕                      |
| MRI abdomen without IV contrast                             | May Be Appropriate       | ○                        |
| 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  | ⊕⊕⊕                      |

# CT Abdomen/Pelvis With Contrast

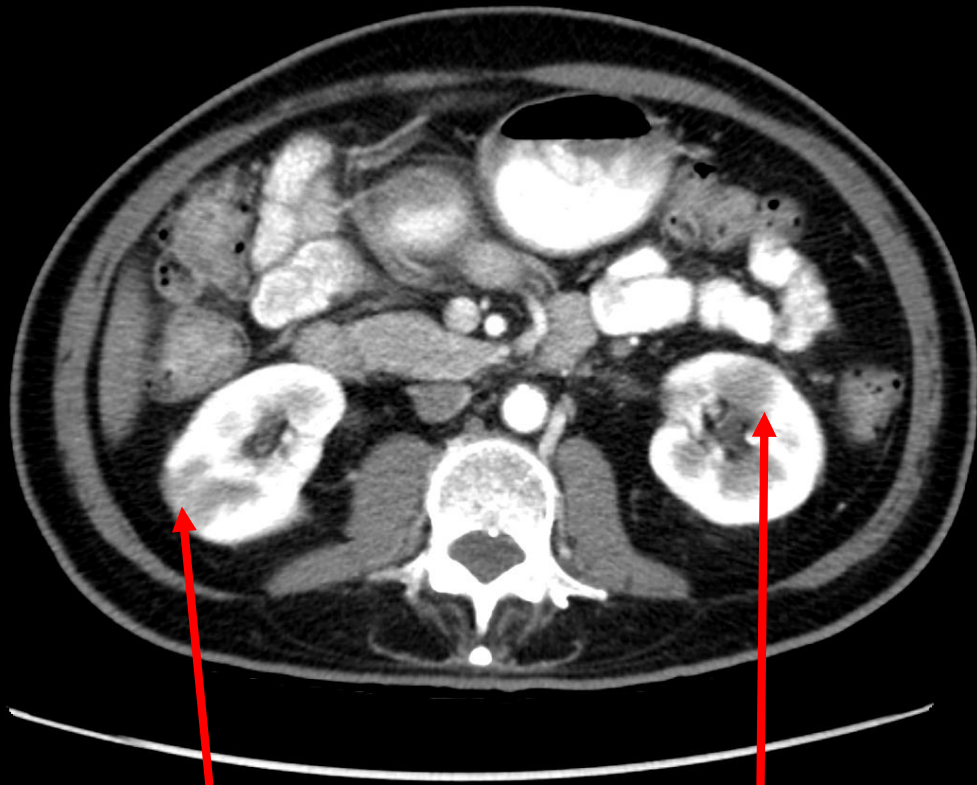


Axial Slice



Axial Slice

# CT Abdomen/Pelvis With Contrast (Labeled)



**Axial Slice**

Bilateral low-attenuation peripheral and cortical lesions



**Axial Slice**

Circumferential soft-tissue surrounding infrarenal abdominal aorta

# Differential Diagnosis

Multiple Wedge Shaped or Round Cortical Renal Lesions

1. Pyelonephritis
2. Vascular insult (i.e. infarct)
3. Metastases
4. Lymphoma

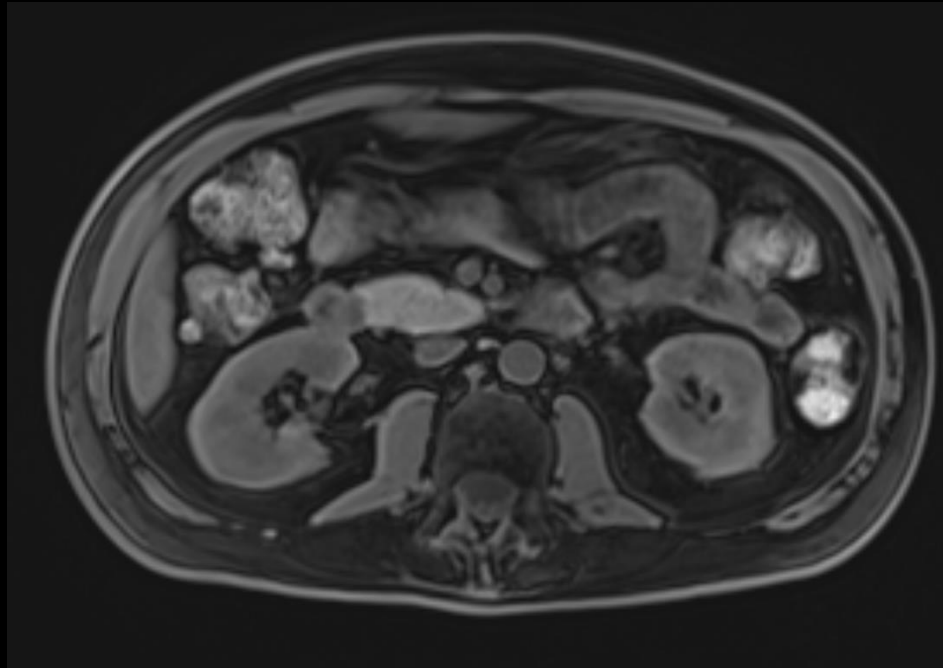
Periaortic Soft Tissue

1. Retroperitoneal fibrosis
2. Aortitis/vasculitis
3. Lymphoma
4. Metastases
5. Other

**Additional evaluation with MRI was recommended**

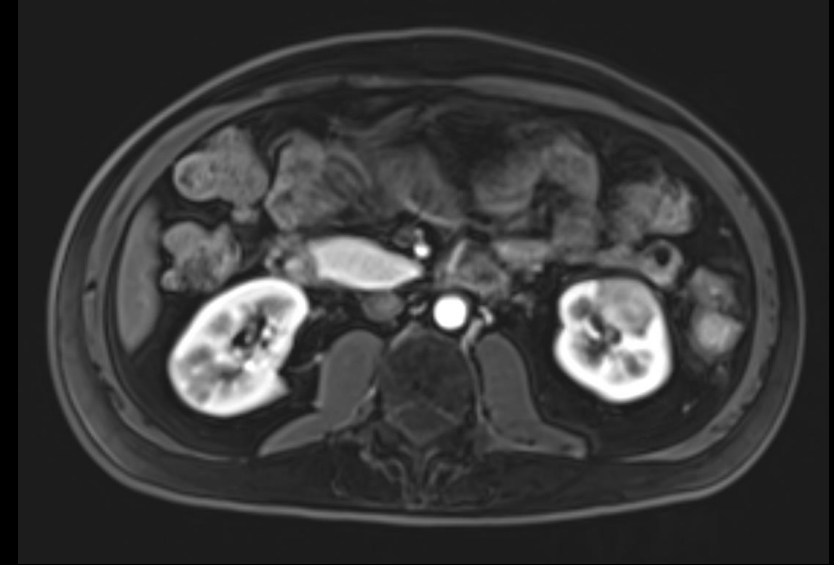


# MRI Abdomen With and Without Contrast

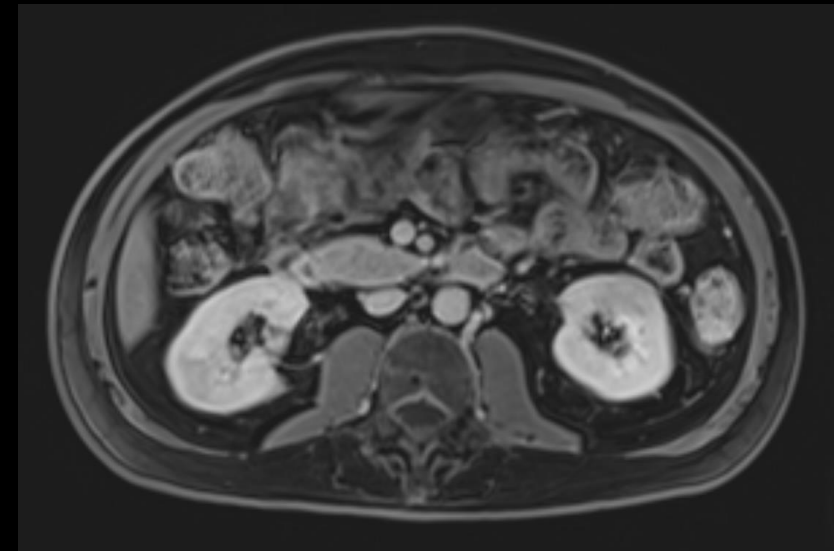


T1 Fat Sat Pre-Contrast

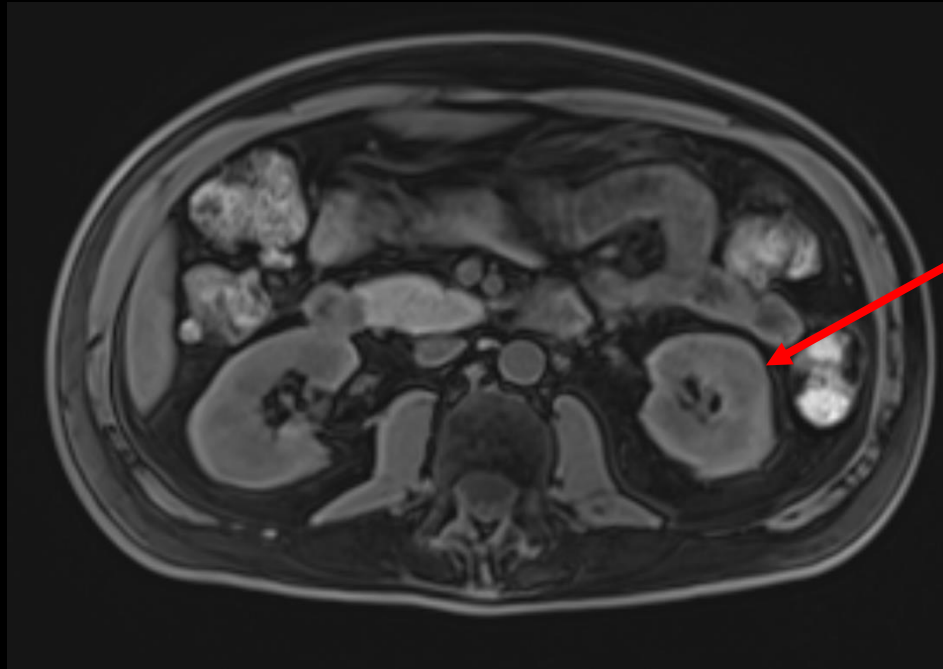
T1 Fat Sat  
Post-Contrast  
Early



T1 Fat Sat  
Post-Contrast  
Late

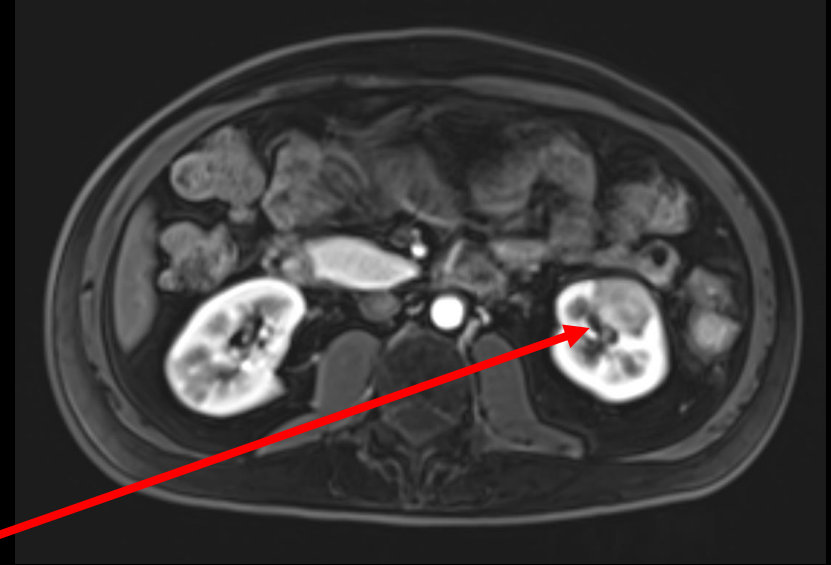


# MRI Abdomen With and Without Contrast (Labeled)



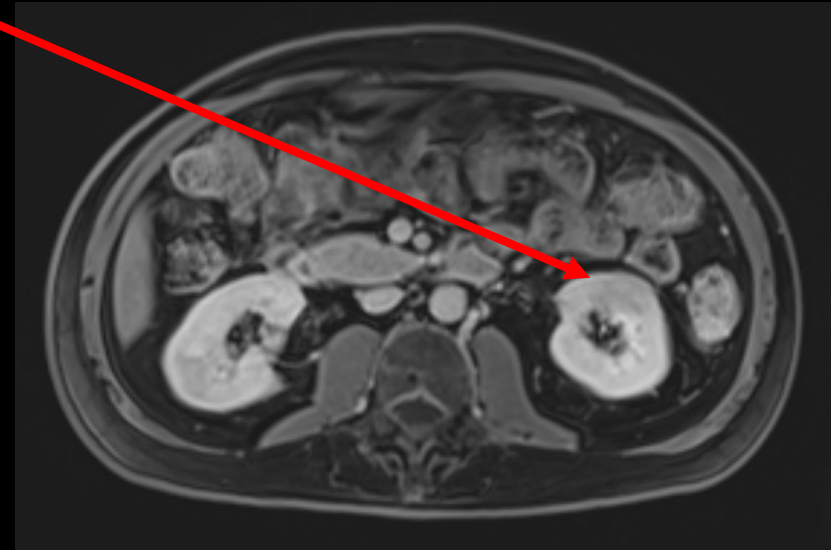
T1 Fat Sat Pre-Contrast

T1 Fat Sat  
Post-Contrast  
Early



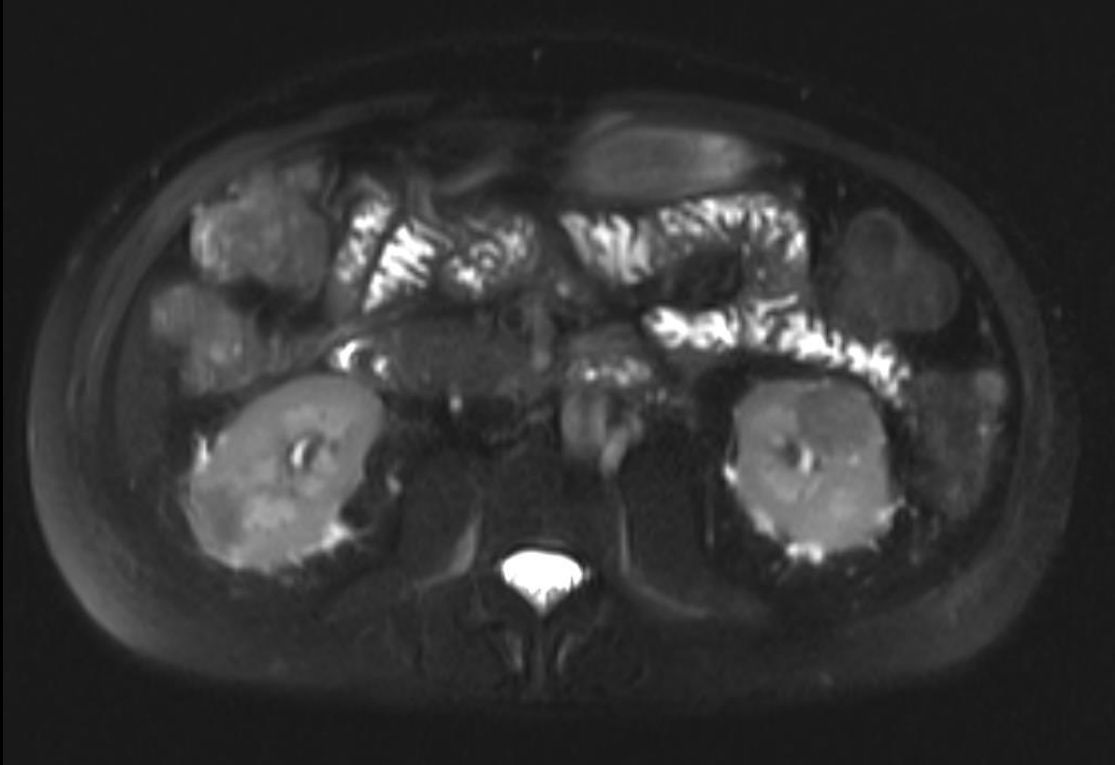
Isointense  
Hypoenhancement

T1 Fat Sat  
Post-Contrast  
Late



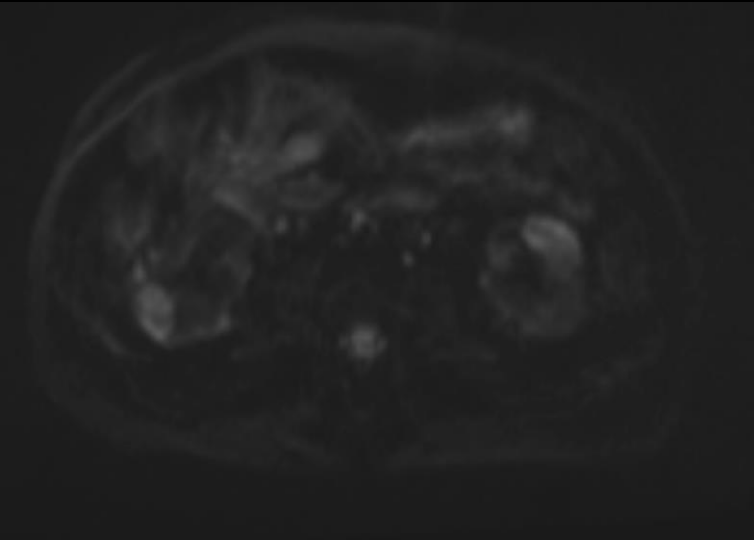


# MRI Abdomen With and Without Contrast

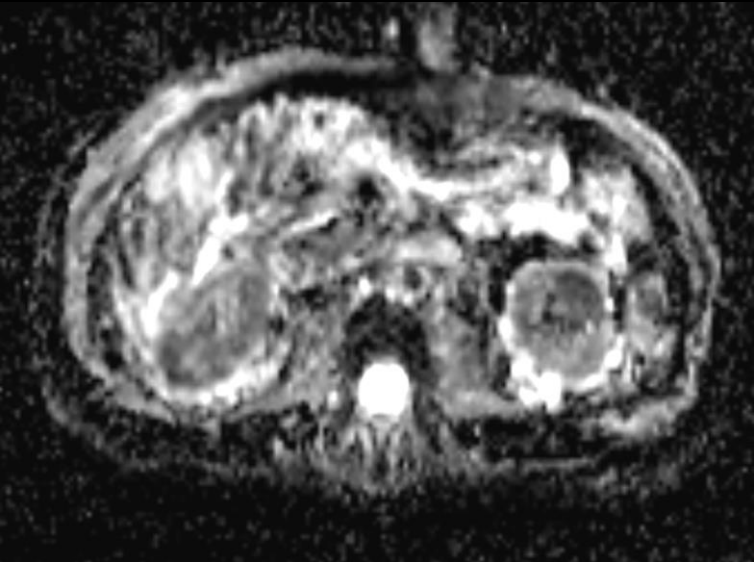


T2 Fat Sat

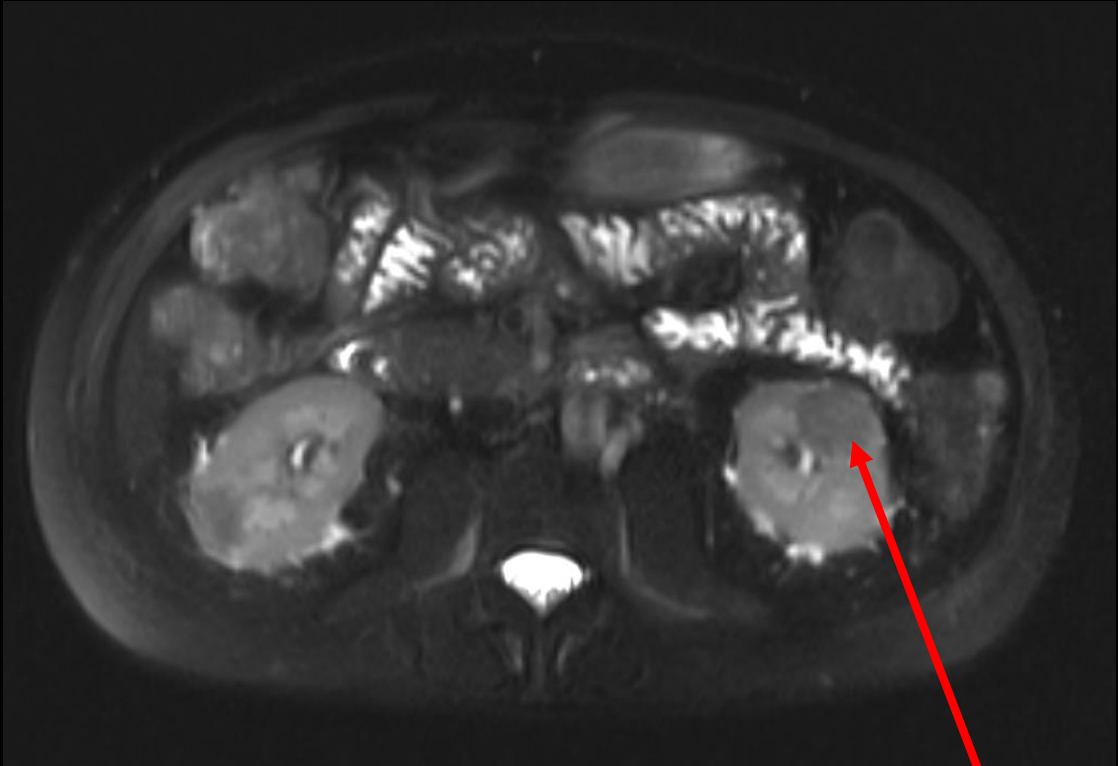
DWI



ADC



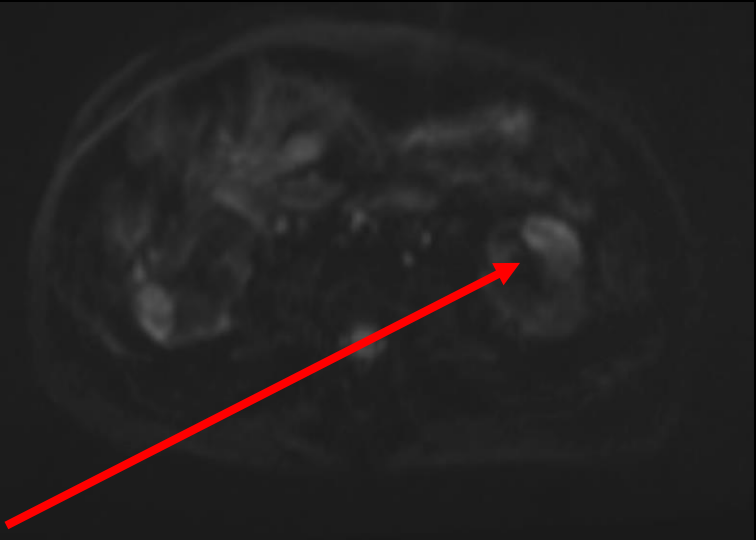
# MRI Abdomen With and Without Contrast (Labeled)



T2 Fat Sat

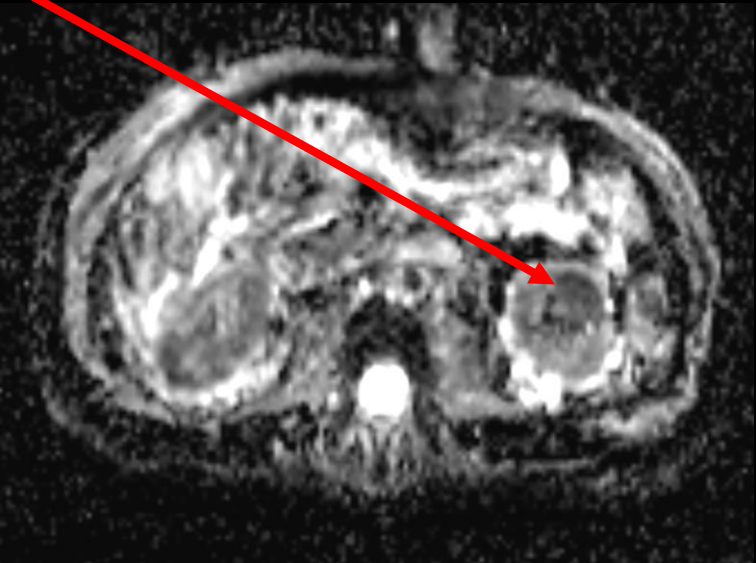
Decreased  
T2 intensity

DWI

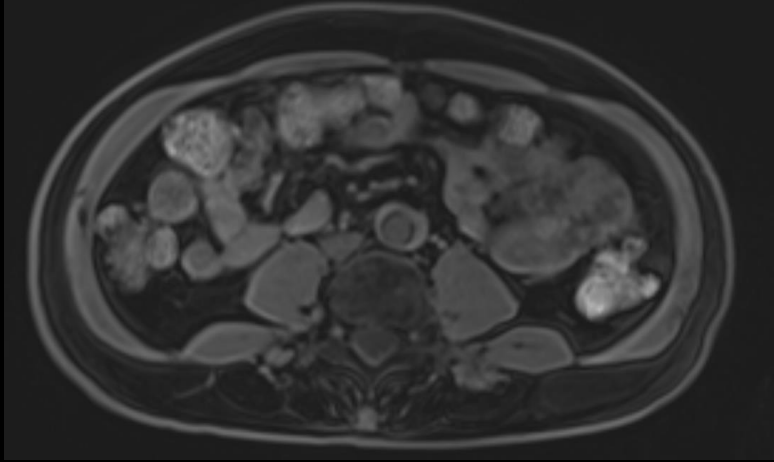


Restricted  
Diffusion

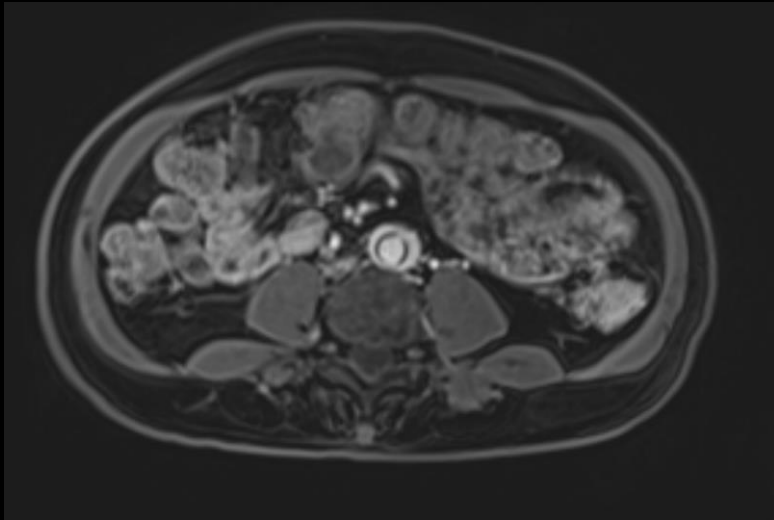
ADC



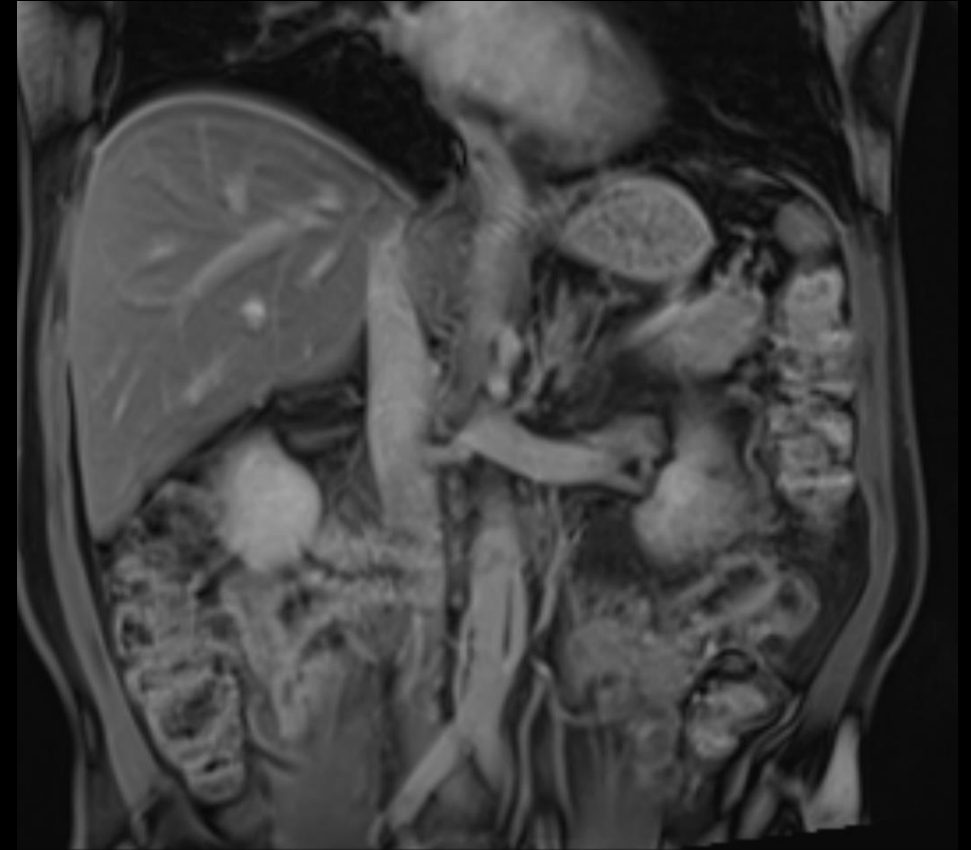
# MRI Abdomen With and Without Contrast



T1 Fat Sat  
Pre-Contrast

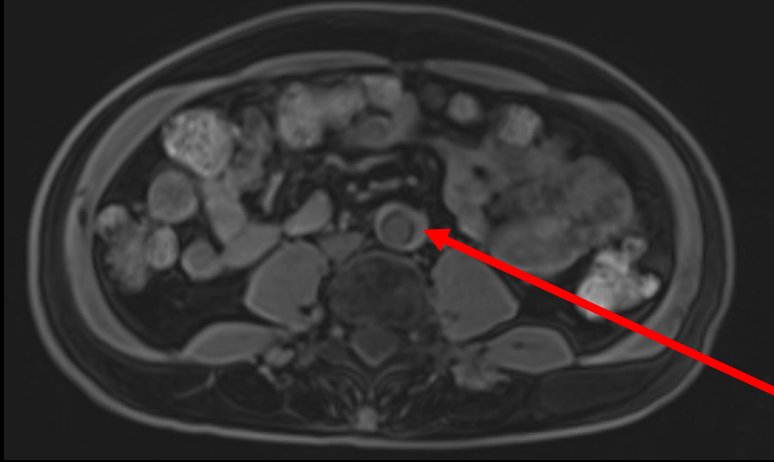


T1 Fat Sat  
Post-Contrast



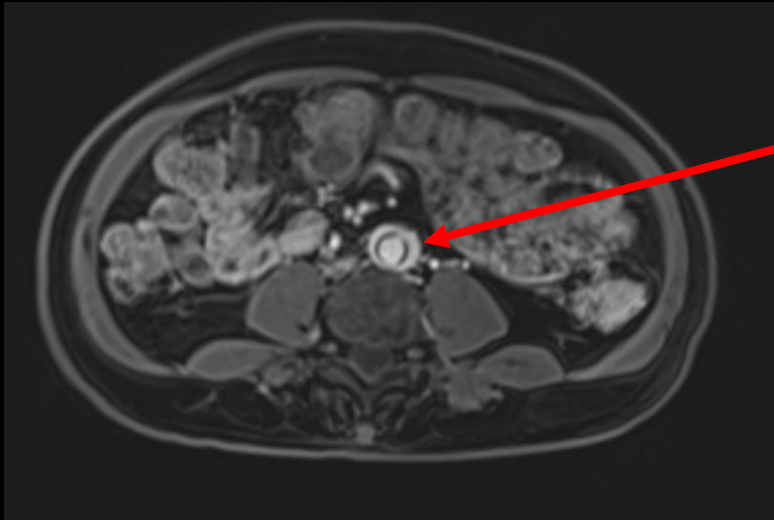
T1 Coronal Fat Sat Post

# MRI Abdomen With and Without Contrast (Labeled)



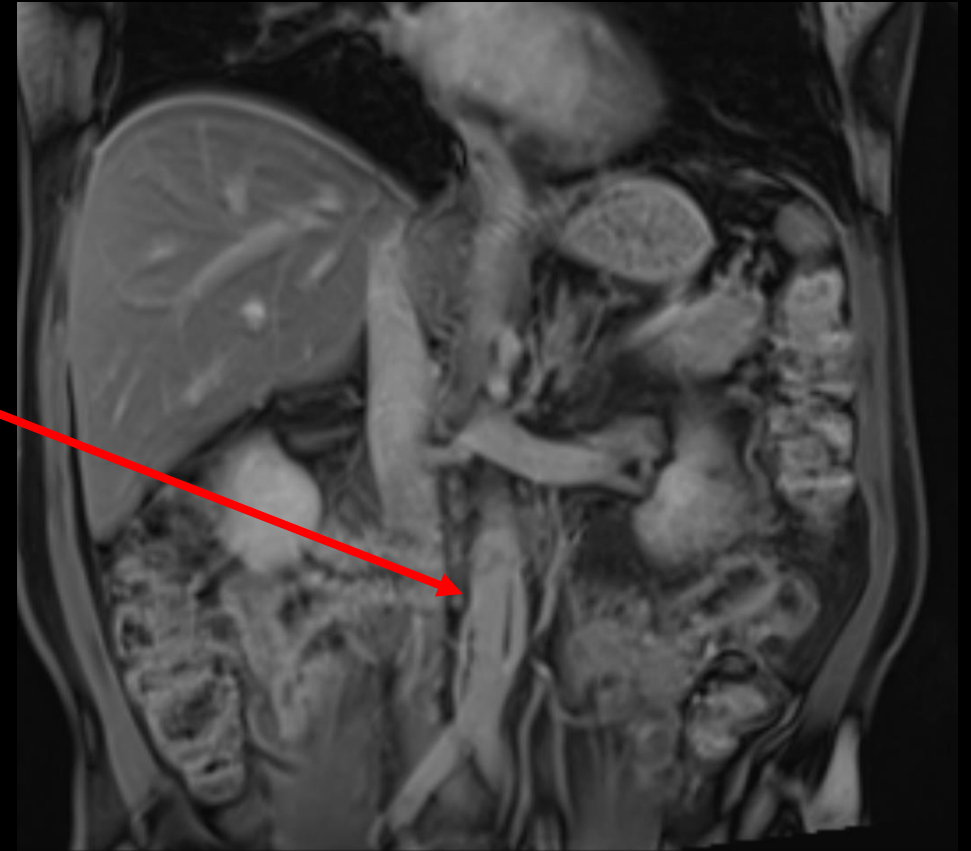
T1 Fat Sat  
Pre-Contrast

Isointense on T1



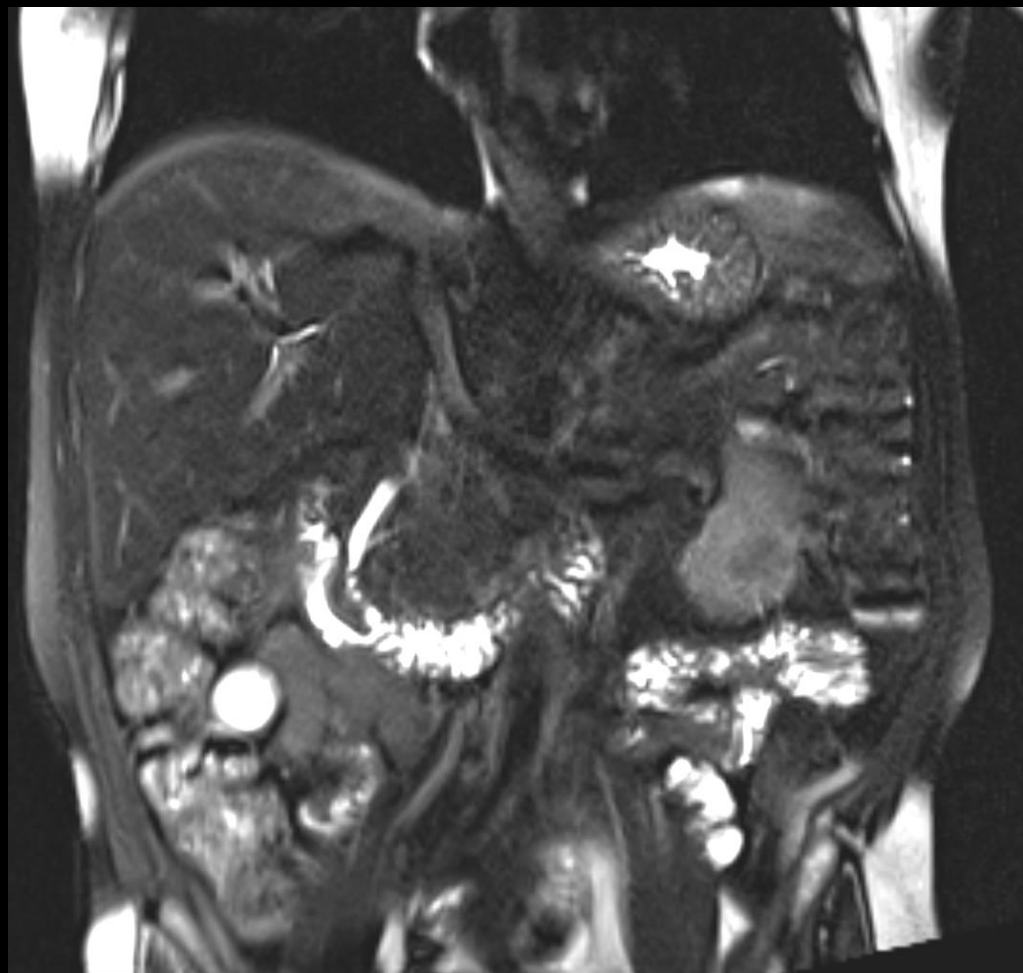
Soft tissue  
thickening around  
the aorta

T1 Fat Sat  
Post-Contrast

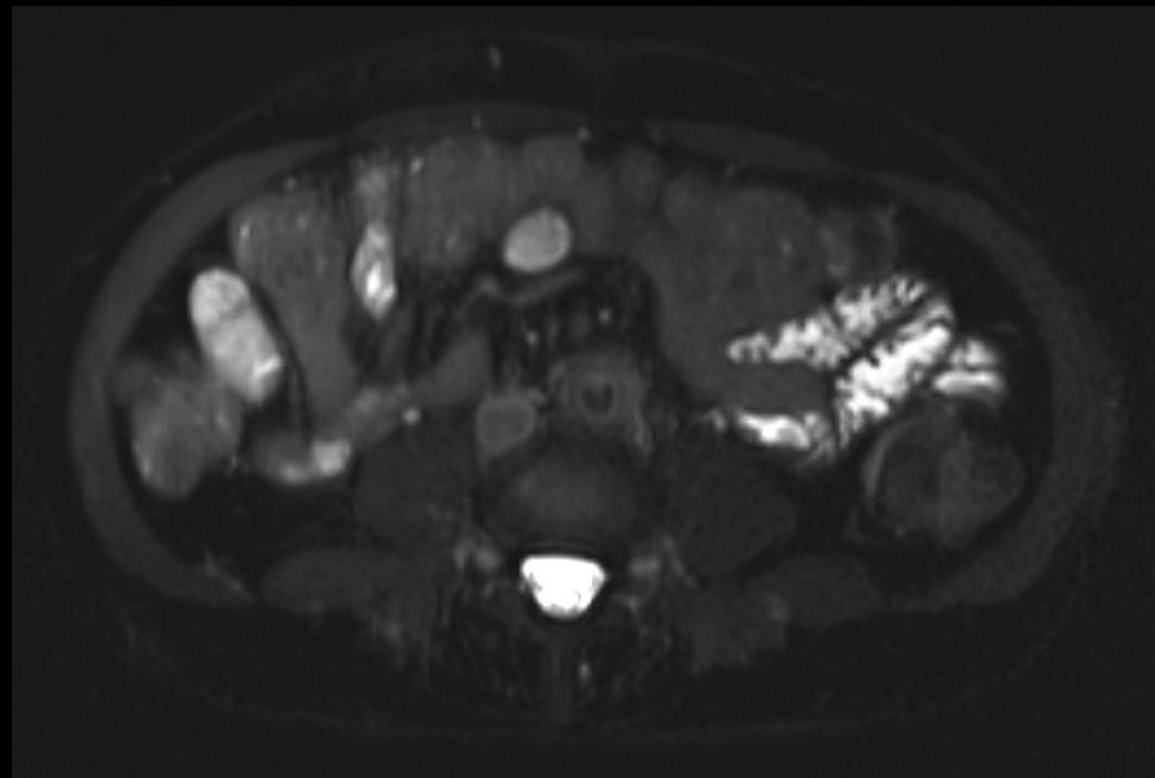


T1 Coronal Fat Sat Post

# MRI Abdomen With and Without Contrast



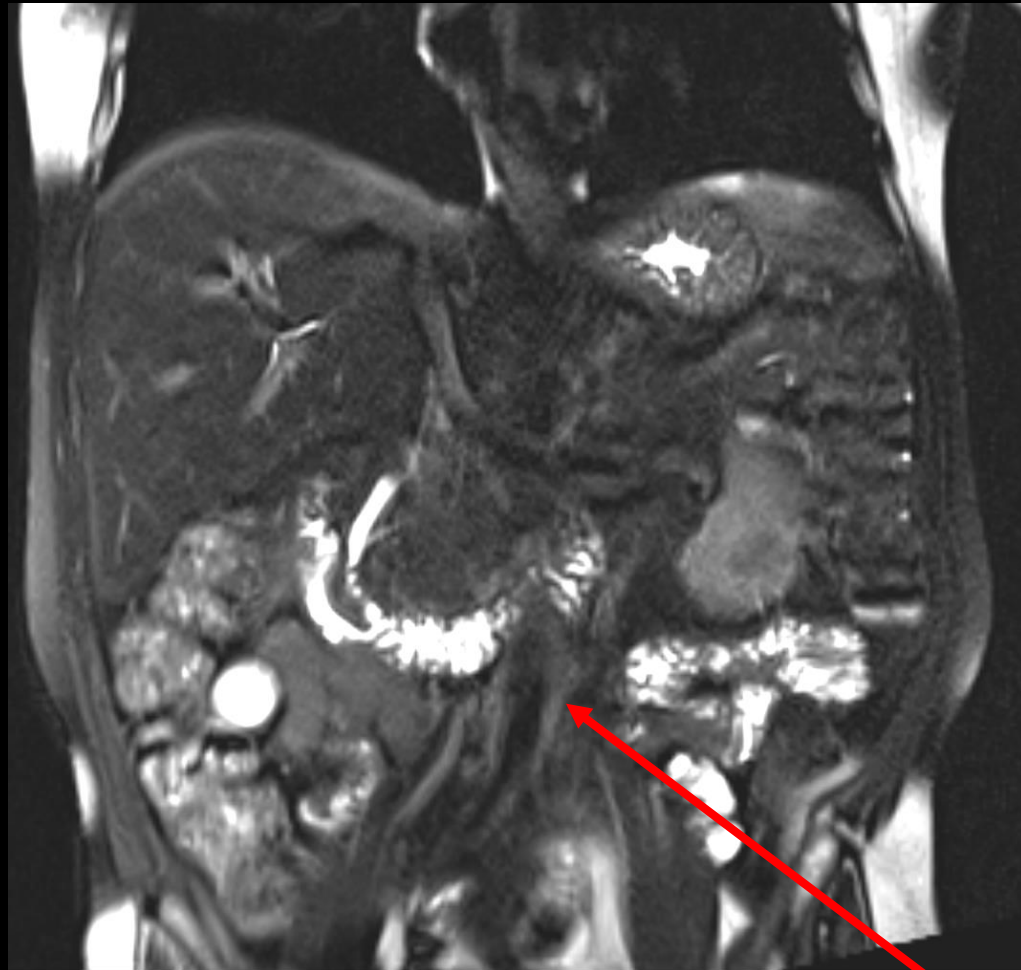
T2 Coronal



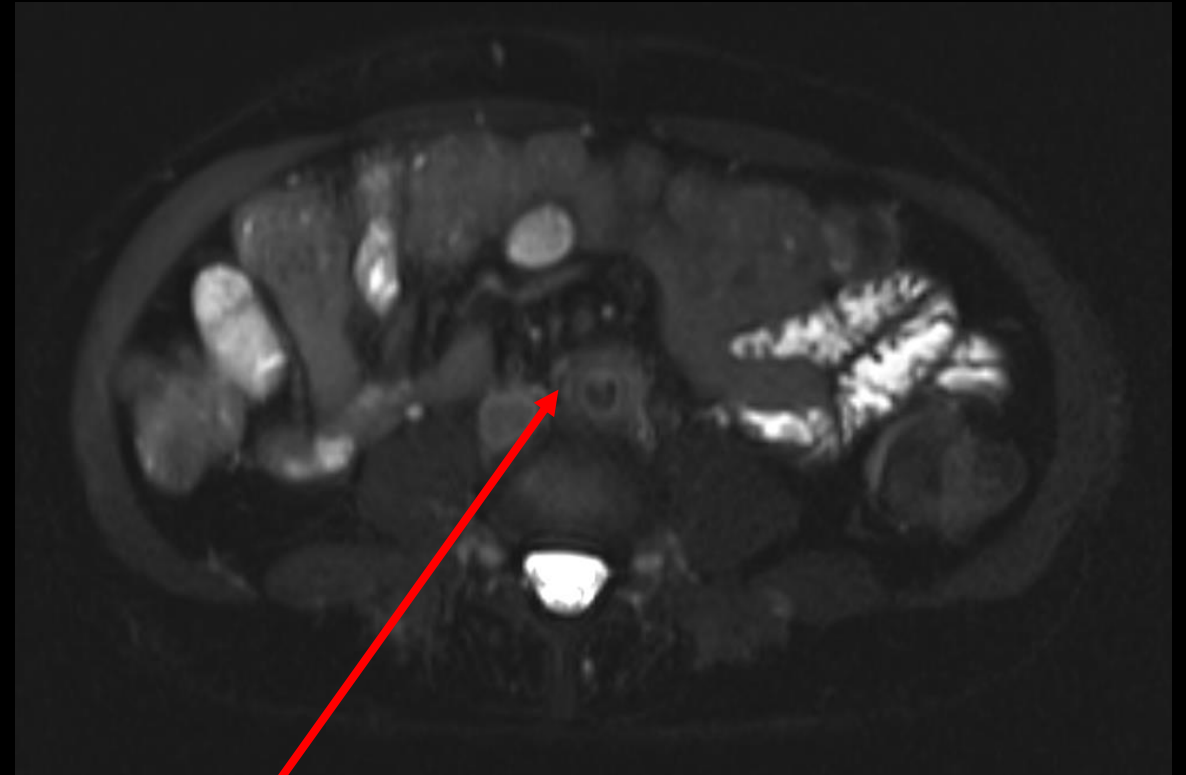
T2 Axial



# MRI Abdomen With and Without Contrast (Labeled)



T2 Fat Sat Coronal



T2 Fat Sat Axial

Increased T2 signal  
around the aorta



# DDX (based on imaging)

- Vasculitis
- Aortitis
- Inflammatory vs Infectious
  - IgG – related disease
- Lymphoma
- Metastases

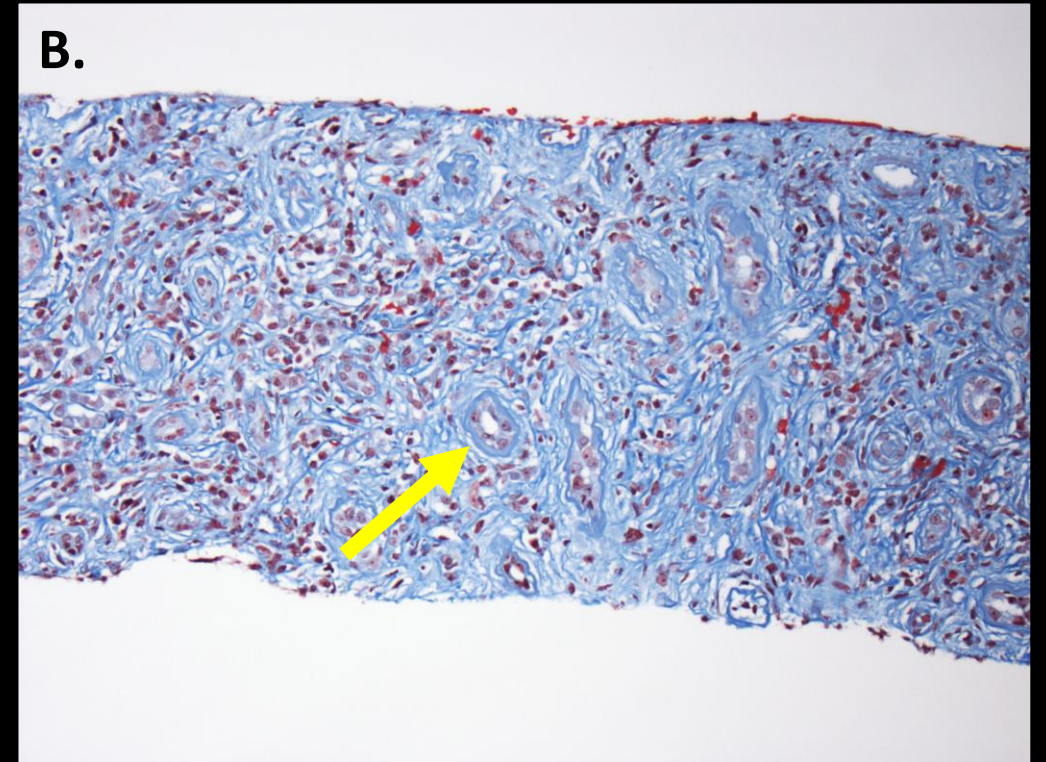
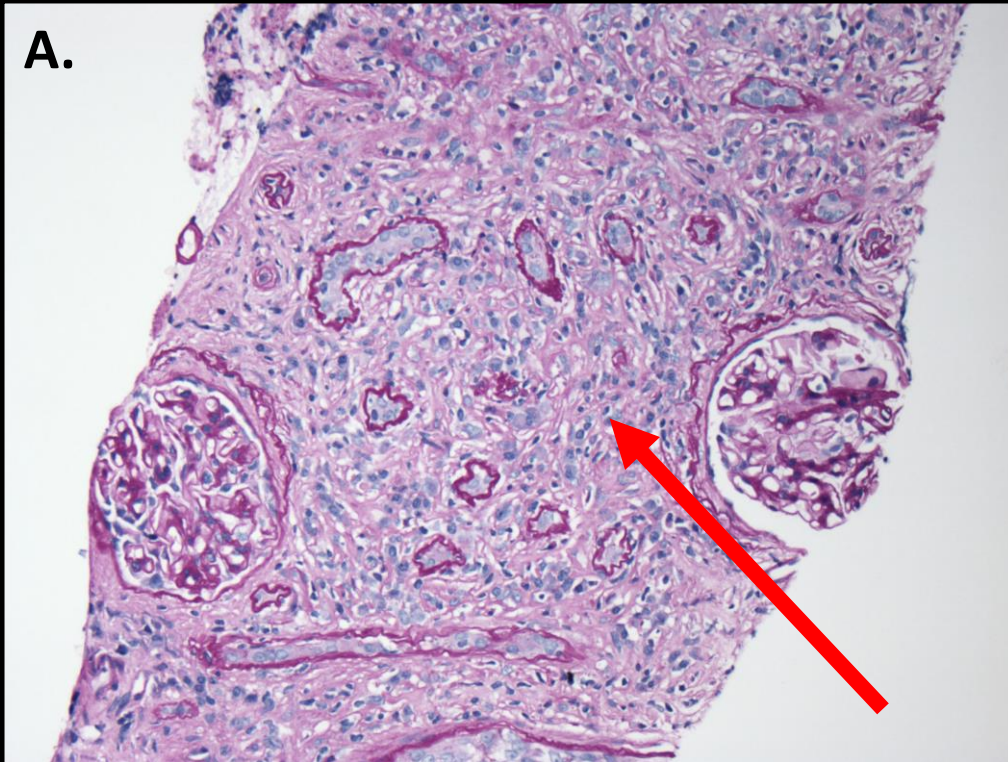
To further work up the renal masses, labs were ordered, and the patient was referred for a Nephrology consult and renal biopsy.

# Supporting Labs and CT Guided Biopsy

- Hct, Hgb, WBC all wnl
- **ESR elevated to 47**
- CRP, ANA, RF, C3, C4 all wnl
- ANCA negative
- **IgG4 = 975.9 mg/dL (ref 4 – 86 mg/dL)**



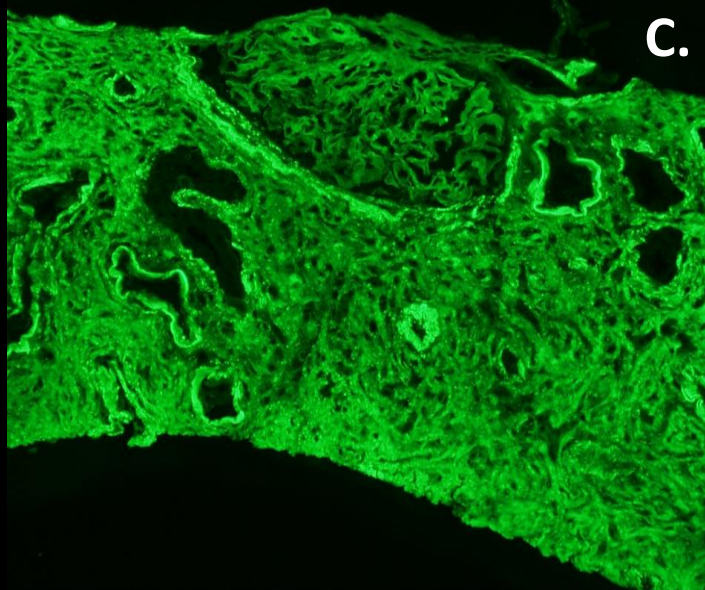
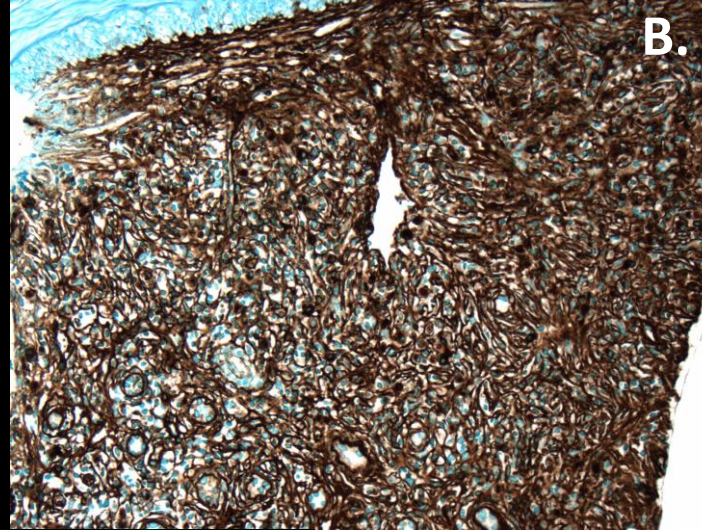
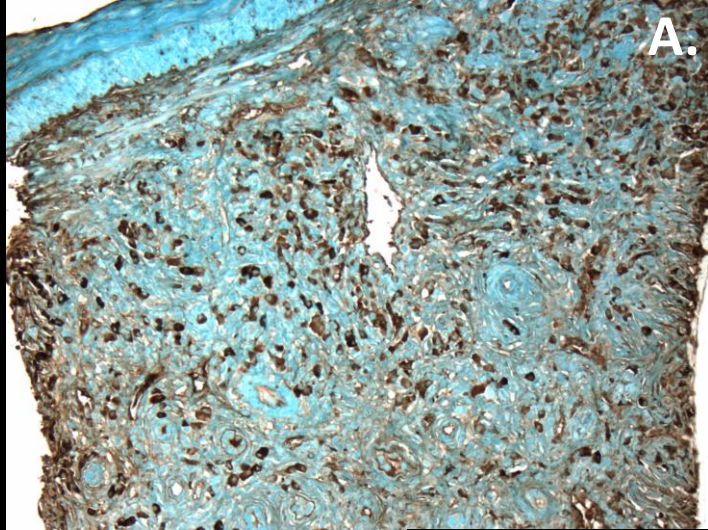
# Histopathology



PAS (A) and Trichrome (B) stains: **Diffuse interstitial fibrosis** in a **storiform pattern** expanding the interstitium with many infiltrating plasma cells. The **tubules** present are **atrophic**.



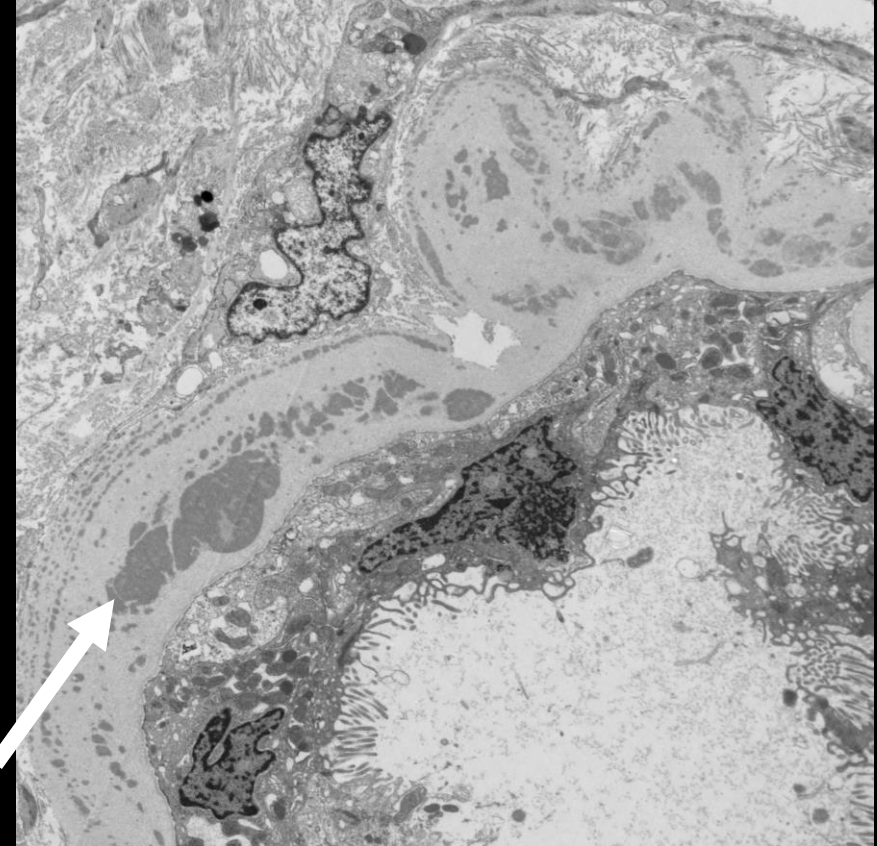
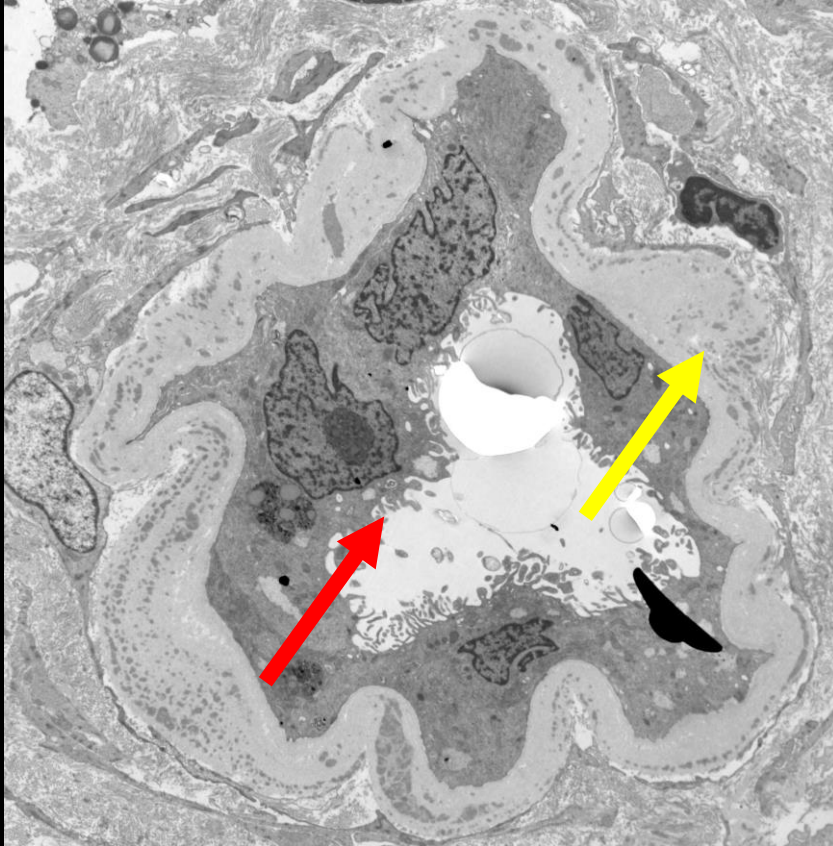
# Immunohistochemistry/Immunofluorescence



- IHC stains show increased **IgG positive plasma cells** (A) and increased **IgG4 positive plasma cells** (B)
- IgG4 is much darker, related to the increased deposits present throughout interstitium and along tubular basement membranes
- IgG4 subclass staining by immunofluorescence shows **fine granular staining** along tubular basement membrane, interstitium, and Bowman's capsule (C)



# Electron Microscopy



Electron microscopic images show damaged tubules with **degenerative epithelial changes** and **thickened basement membranes**. There are many **electron dense deposits** in the tubular basement membranes.

Final Dx:

IgG4 – Related Disease



# Case Discussion

- IgG4 is an immune mediated disease that results in fibrosis and has many systemic manifestations.<sup>1, 2</sup>
- Common clinical manifestations<sup>3</sup>
  - **Sclerosing Sialadenitis**
  - Orbital disease
  - Autoimmune Pancreatitis (AIP)
  - **Retroperitoneal fibrosis** – accompanied often by chronic **periaortitis**, and ureteral involvement with hydronephrosis
  - Sclerosing cholangitis
- Can mimic SLE, Sjogren's, GPA, RA, APLS, and others

# Case Discussion

## Epidemiology

- Exact numbers are difficult because IgG4-RD has no ICD-10 code
- In Japan, 0.28-1.08/100,000<sup>4</sup>
- Grouped into four categories<sup>3</sup>
  - Pancreato-hepato-biliary
  - RPF and/or aortitis
  - Head-and-neck limited
  - Mikulicz syndrome (lacrimal, salivary, parotid, submandibular) with systemic symptoms

## Pathogenesis

- Autoimmune, but not completely understood
- Consensus is IgG4 are down-regulatory response to other unknown processes<sup>5</sup>
  - Also high in allergic disorders, EGPA, sarcoidosis

# Treatment and Prognosis

- Remission induction
  - Heavily debated – prednisone or rituximab monotherapy vs prednisone + rituximab
- Resistance to initial therapy and steroid dependence
  - Azathioprine, mycophenolate mofetil, 6-mercaptopurine
- Maintenance
  - "Watchful waiting" – evaluate every 6 months or sooner for new symptoms

Our Patient – He is recovering well from rituximab injection and labs have normalized.

# References:

1. Stone JH, Zen Y, Deshpande V. IgG4-related disease. *N Engl J Med*. 2012 Feb;366(6):539-51.
2. Kamisawa T, Zen Y, Pillai S, Stone JH. IgG4-related disease. *Lancet*. 2015 Apr;385(9976):1460-71. Epub 2014 Dec 4.
3. Martinez-de-Alegria A, et al. IgG4-related Disease from Head to Toe. *RadioGraphics*. 2015;35:2007-2025. Epub 10.1148/rg.357150066.
4. Umehara H, et al. A novel clinical entity, IgG4-related disease (IgG4RD): general concept and details. *Mod Rheumatol*. 2012 Feb;22(1):1-14. Epub 2011 Sep 1.
5. Aalberse RC, Stapel SO, Schuurman J, Rispens T. Immunoglobulin G4: an odd antibody. *Clin Exp Allergy*. 2009;39(4):469.