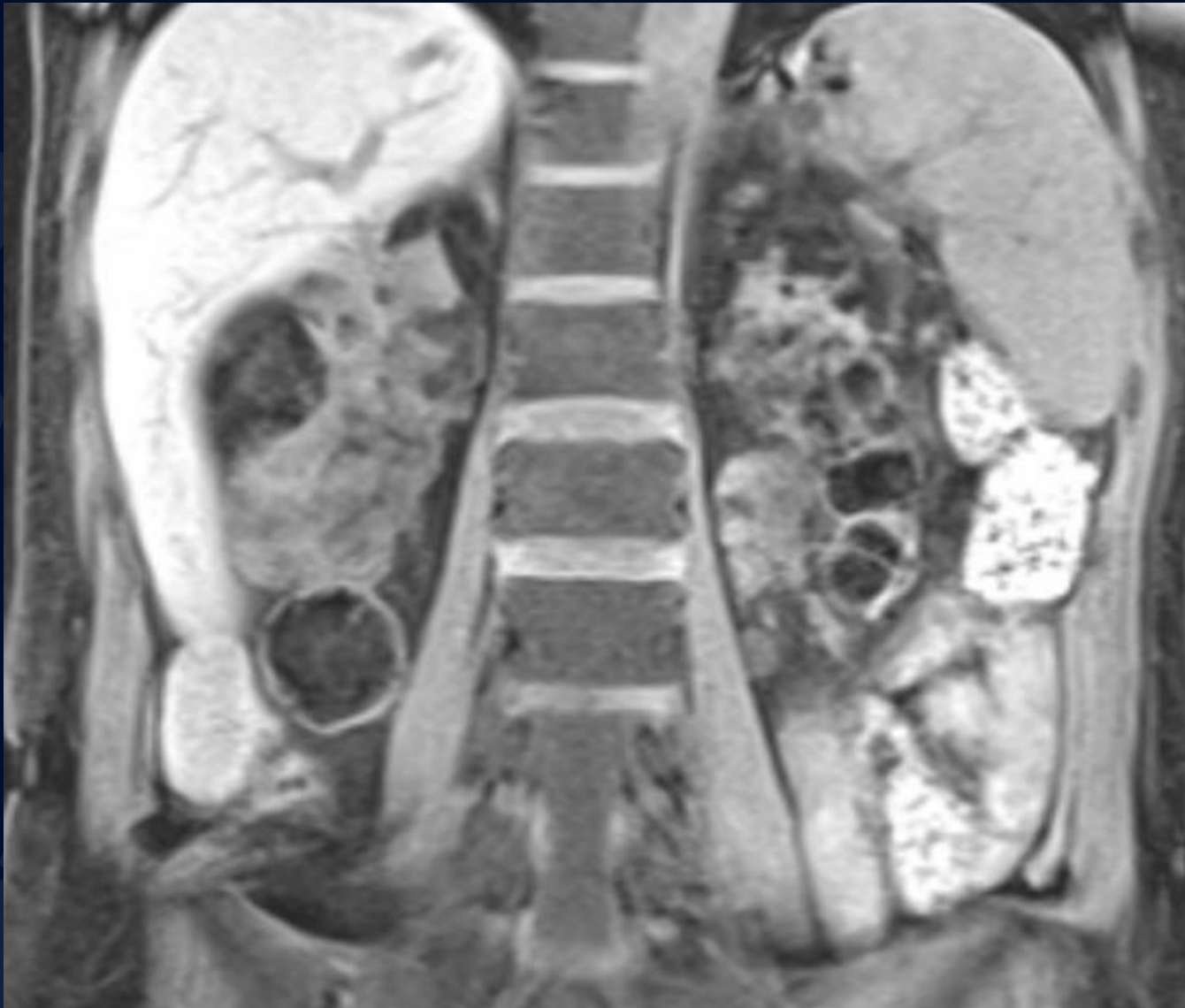
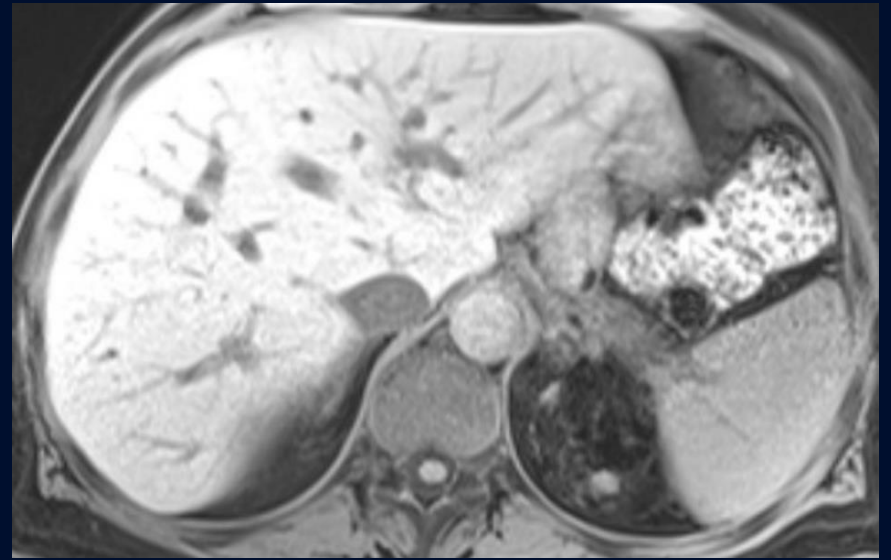
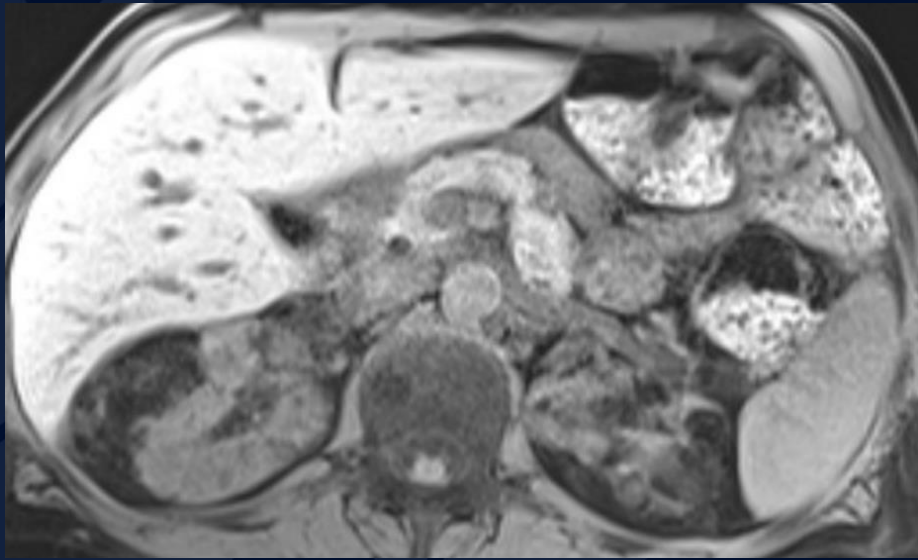


54-year-old female with a history  
of seizures presenting with  
elevated creatinine and  
proteinuria

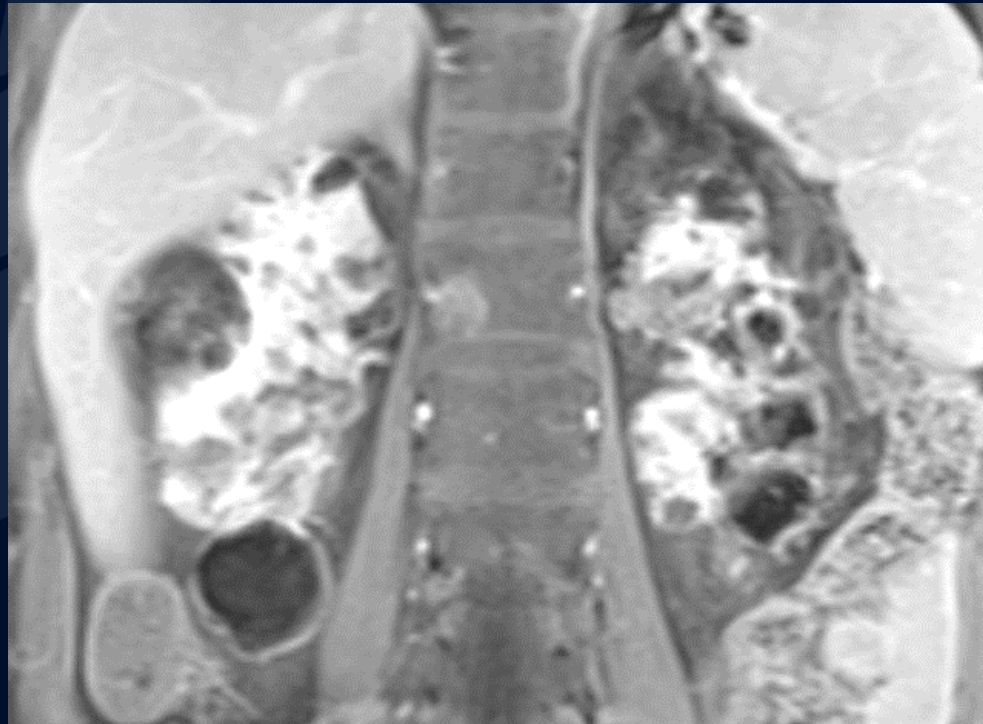
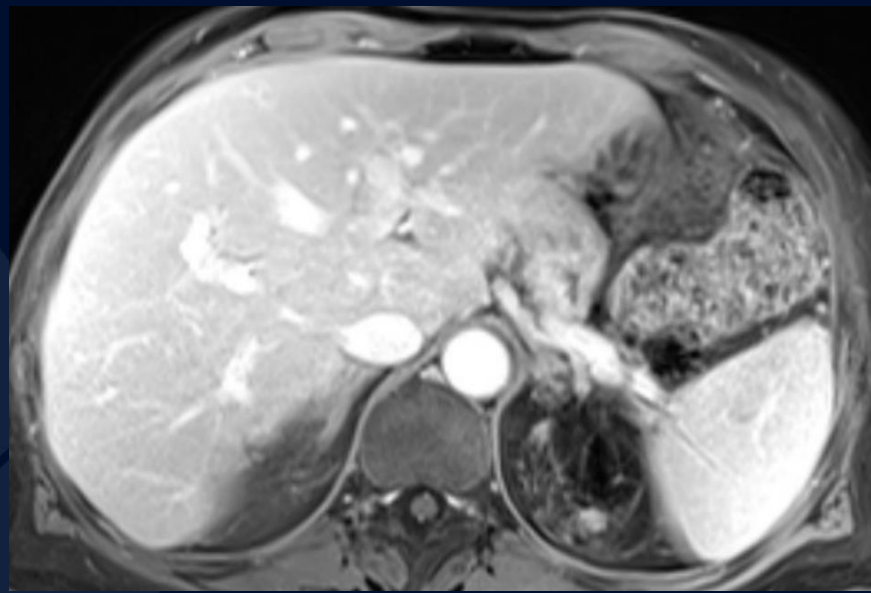
Kiran Singh-Smith, MS3



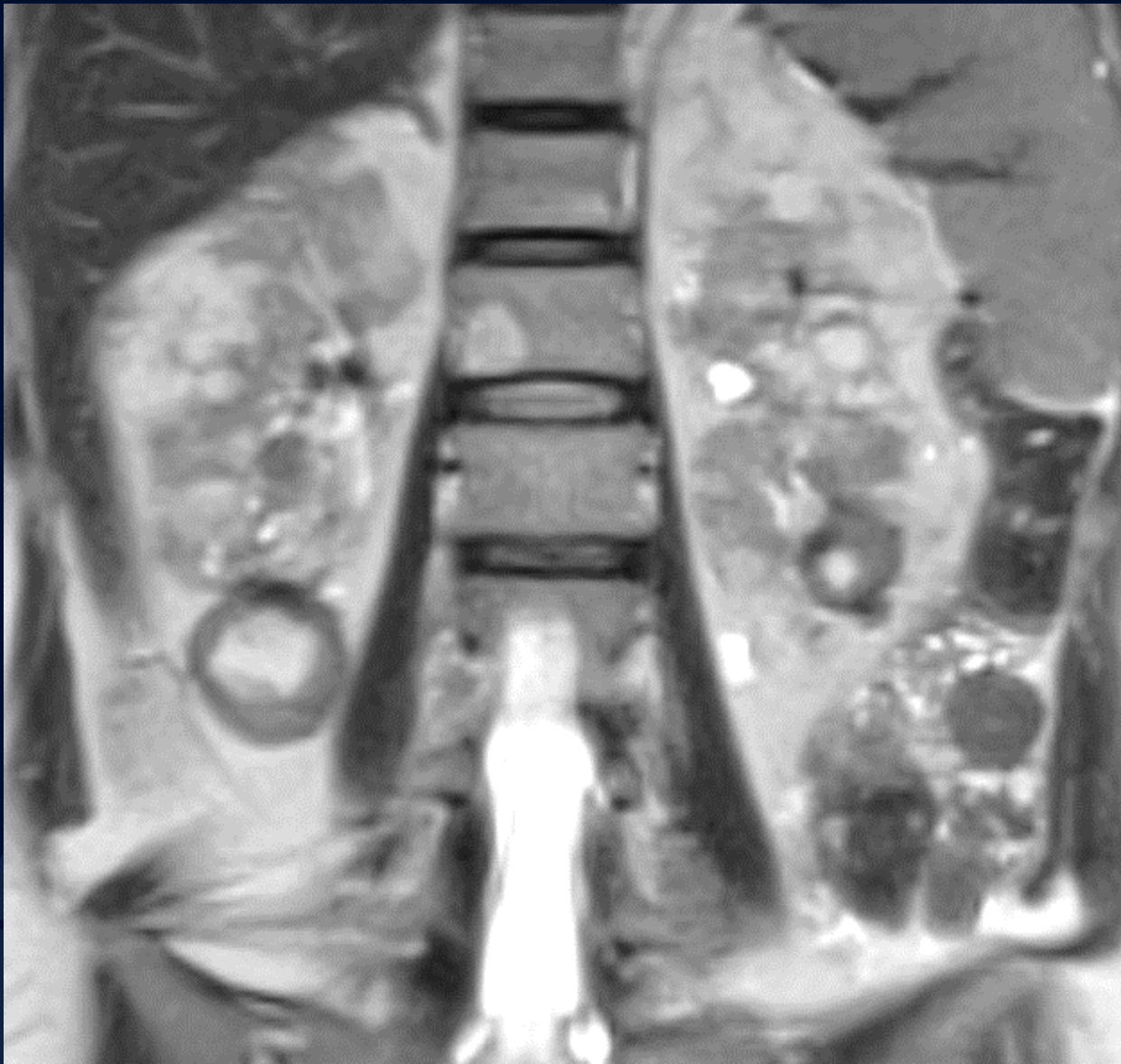
MR T1 Fat-Saturated



MR T1 Fat-Saturated



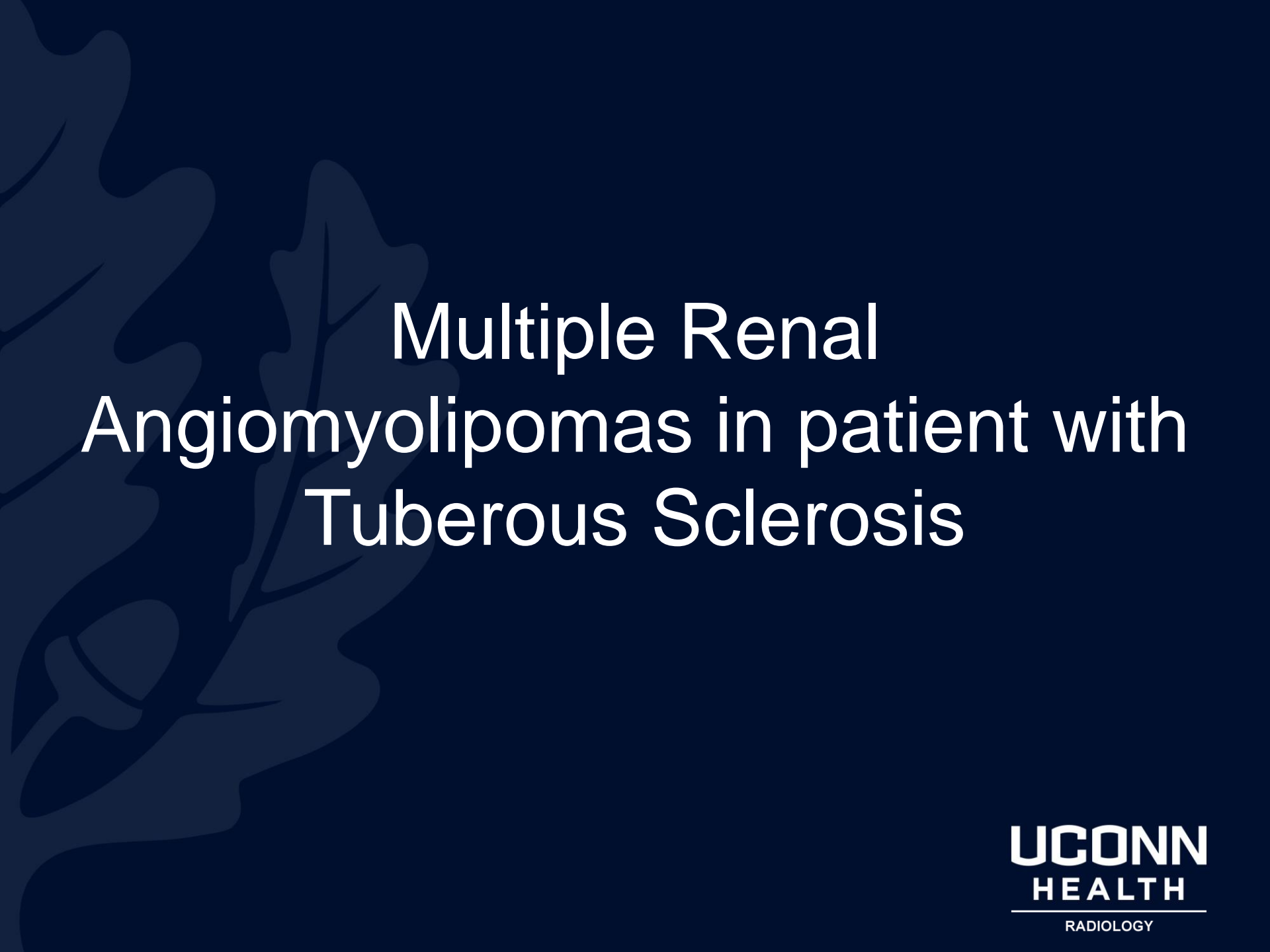
MR T1 Fat-Saturated + Gad



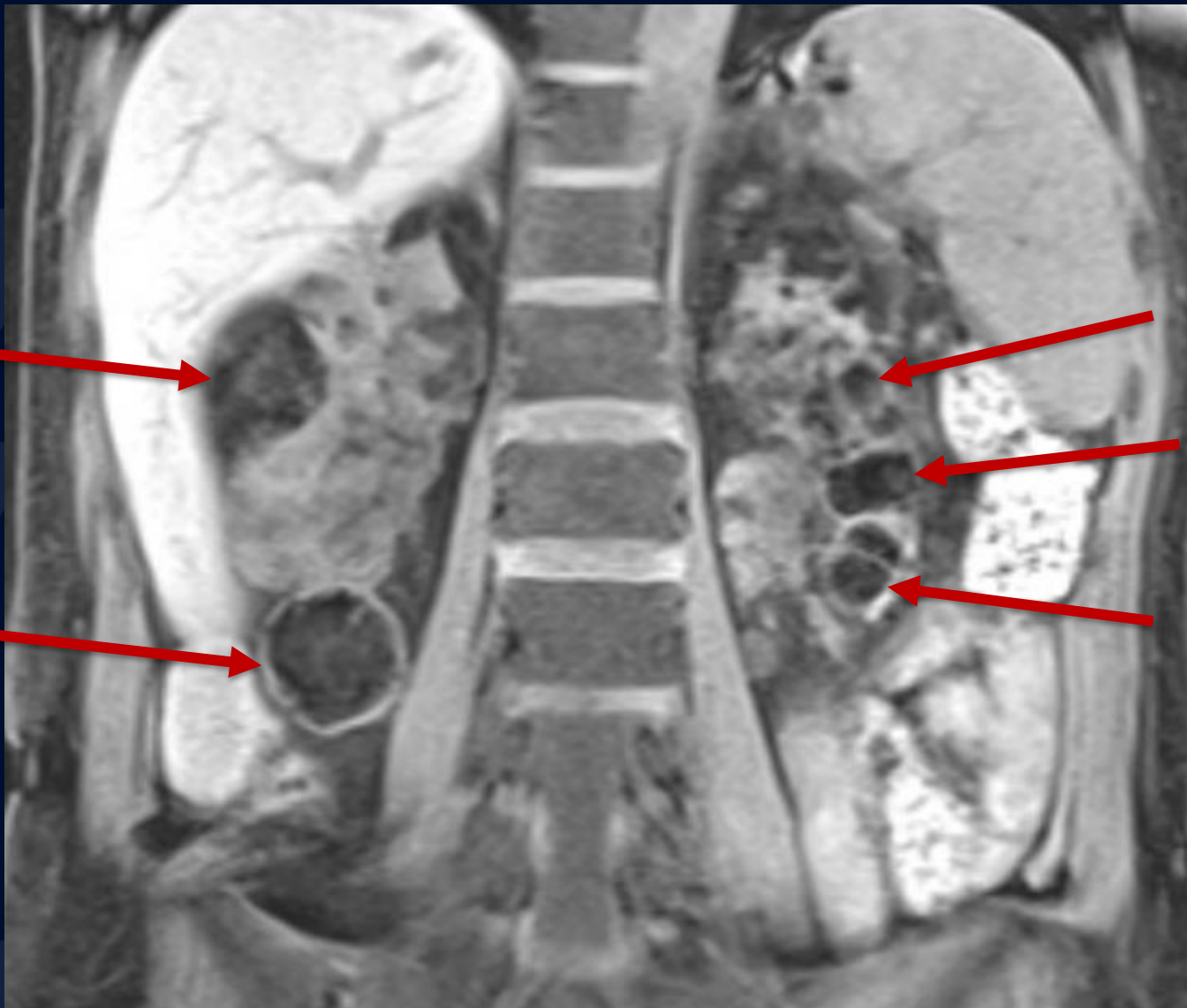
MR T2

A large, stylized oak leaf graphic in a dark blue color, positioned on the left side of the slide. It features detailed vein patterns and a lobed edge.

?



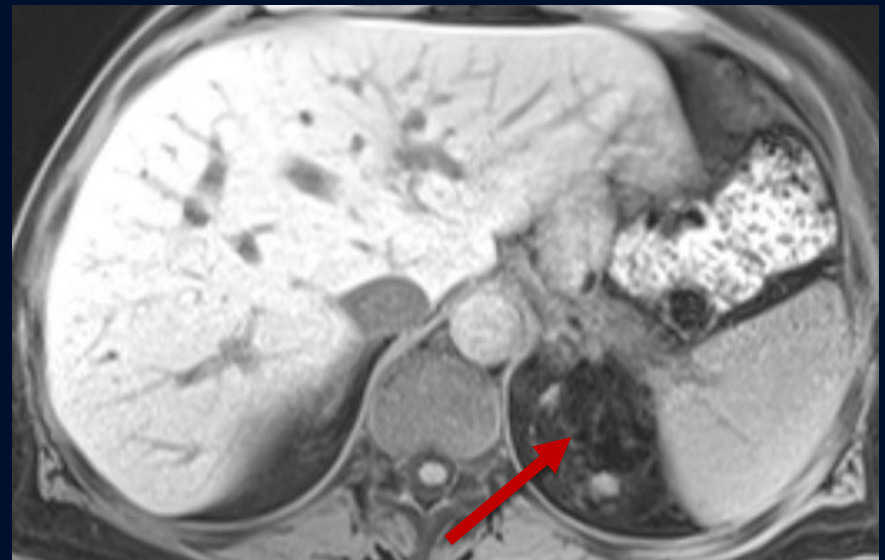
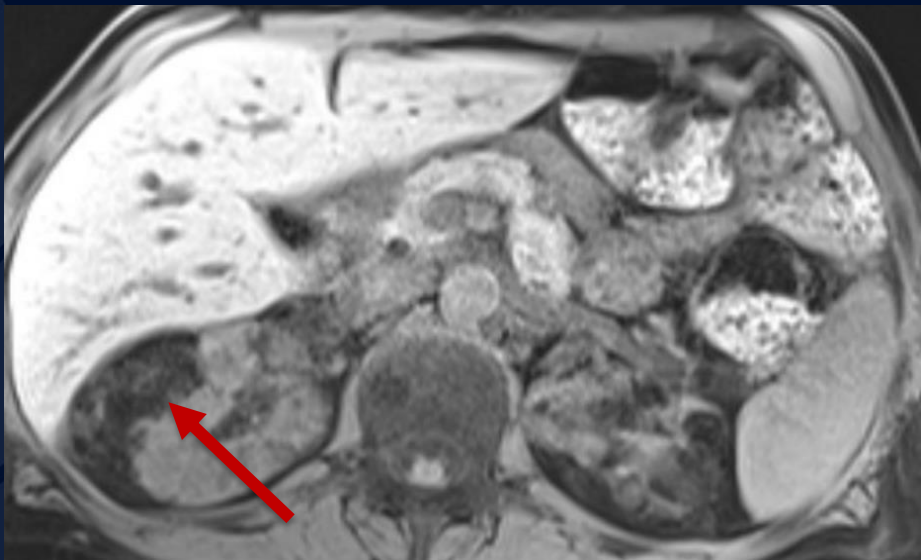
# Multiple Renal Angiomyolipomas in patient with Tuberous Sclerosis



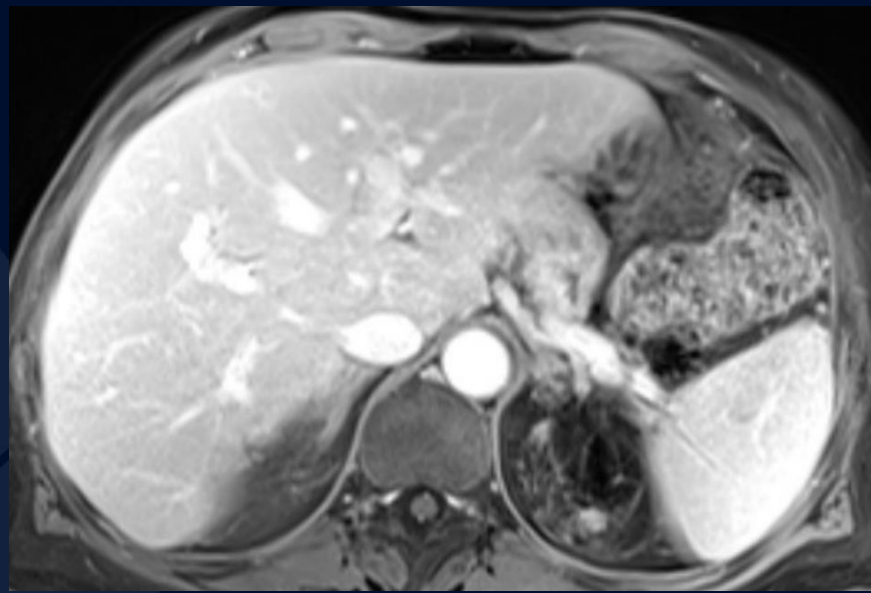
MR T1 Fat-Saturated

Multiple bilateral, well-defined,  
hypointense, round renal masses

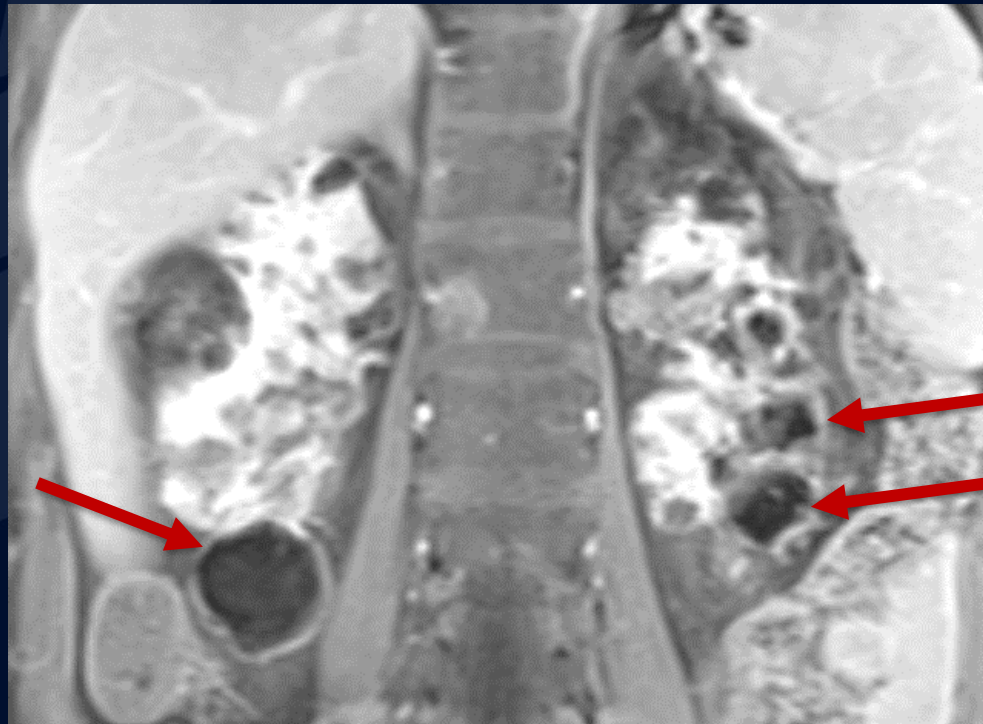




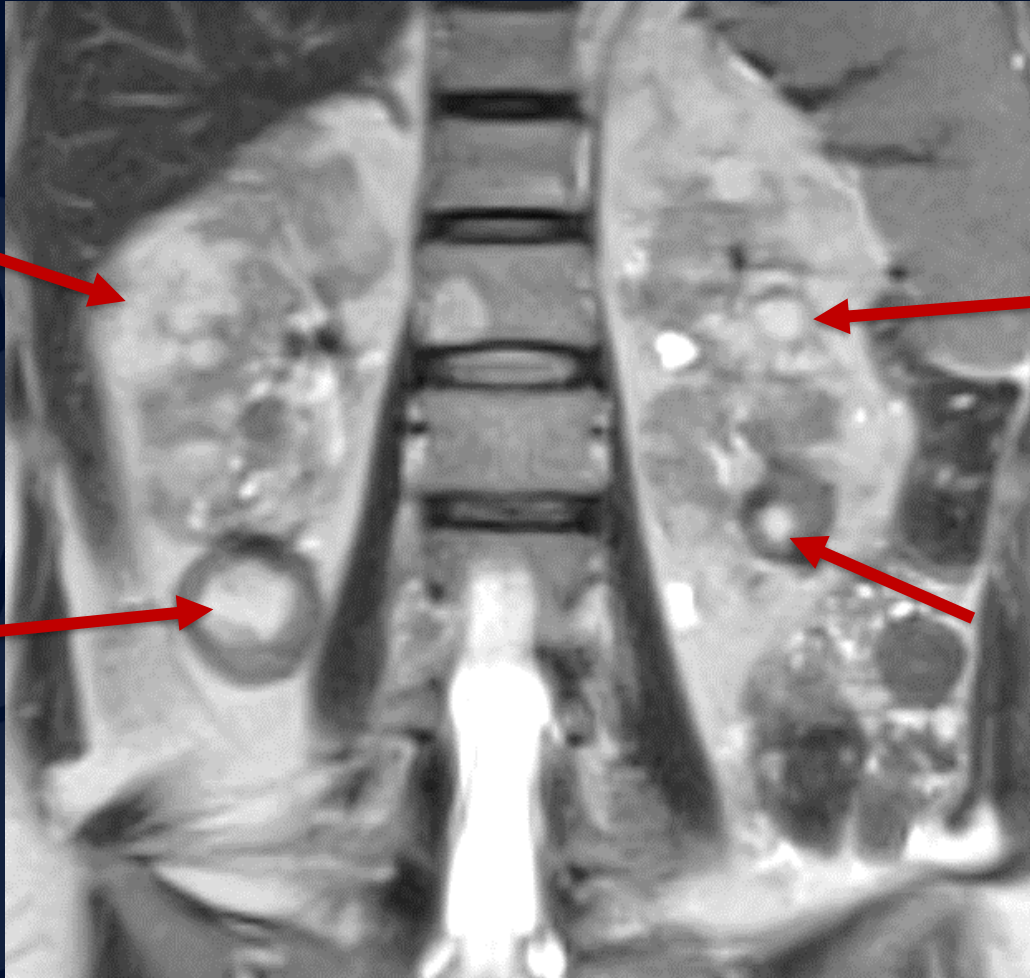
MR T1 Fat-Saturated  
Multiple bilateral, well-defined,  
hypointense, round renal masses



Multiple bilateral,  
well-defined, non-  
enhancing renal  
masses



MR T1 Fat-Saturated + Gad



MR T2

Multiple bilateral, predominantly  
hyperintense renal masses

# Tuberous Sclerosis and Renal AMLs

## Pathophysiology & Genetics

- Neurocutaneous disorder characterized by development of widespread hamartomas and predominantly benign neoplasms
- Caused by sporadic or autosomal dominant mutations in TSC1 (hamartin) or TSC2 (tuberin) tumor suppressor genes leading to overactivation of mTOR pathway → tumor formation

## Clinical Features

- Renal – angiomyolipomas
- Caused by clonal proliferation of epithelioid cells distributed around blood vessels
  - Possible presentation:
    - Asymptomatic, incidentally found on imaging
    - Hemorrhage, causing hematuria and/or anemia if >4 cm
    - Abdominal/flank masses and/or pain
    - Hypertension, renal insufficiency
    - Rare malignant transformation

# Tuberous Sclerosis and Renal AMLs

- Brain – cortical tubers, subependymal nodules, white matter lesions, arachnoid cysts
  - Possible presentation:
    - Seizures/epilepsy
    - Intellectual disability
    - Autism spectrum disorder
    - Neuropsychiatric disorders like ADHD, insomnia, behavioral issues
- Skin – malar angiofibromas, unguinal fibromas, Shagreen patches
- Ophthalmic – retinal hamartomas, chorioretinal depigmentation
- Cardiac – rhabdomyomas
- Pulmonary – lymphangiomyomatosis (LAM)

## Management

- Depends on individual manifestations and severity
- Renal AMLs – mTOR inhibitors, elective embolization, ablative therapies, surgery
- Surveillance with imaging (every 1-3 years)

# AML Imaging Findings

## Ultrasound

- Well-defined, hypoechoic mass relative to normal renal parenchyma with acoustic shadowing
  - Subset of minimal fat containing AMLs are isoechoic to slightly hypoechoic and lack shadowing

## CT

- Well-marginated, hypodense (<10 HU) mass with macroscopic fat arising from renal cortex
  - Notch sign: AML originates from triangular or rectangular, notch-like defect in cortex
  - 5% contain minimal fat and cannot reliably be diagnosed by CT
  - Hemorrhage more likely in large AMLs (> 4 cm)

## MRI

- Lipid-rich AMLs (95%):
  - T1 and T2 hyperintense compared to renal parenchyma
  - T1 FS will demonstrate loss of signal on fat saturated images
- Lipid poor AMLs (5%):
  - T1 and T2 hypointense relative to renal parenchyma
  - Poor to no fat suppression

# References

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