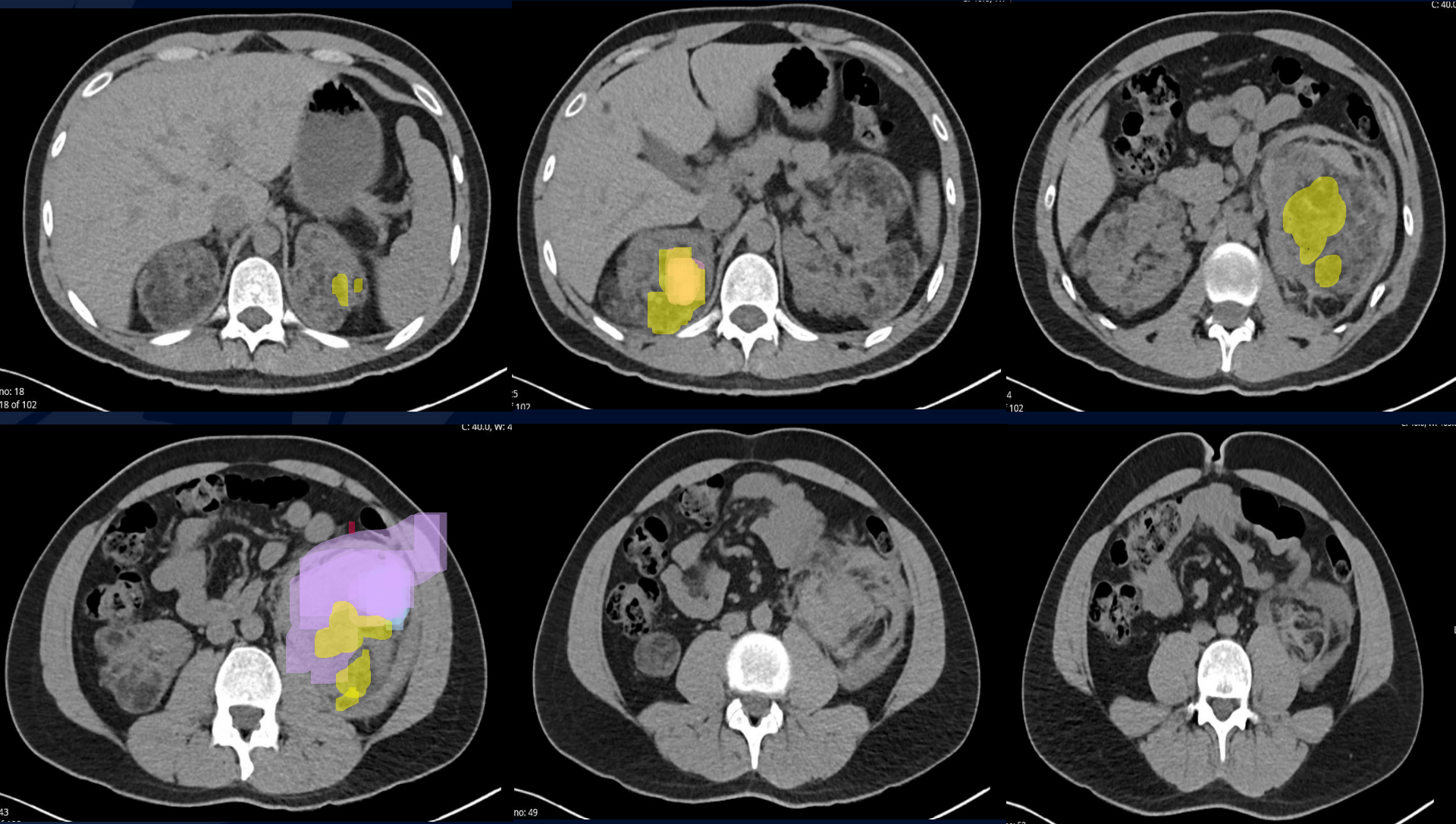


36-year-old male with tuberous sclerosis presenting with spontaneous, nontraumatic abdominal pain

Maria Antony, MS3

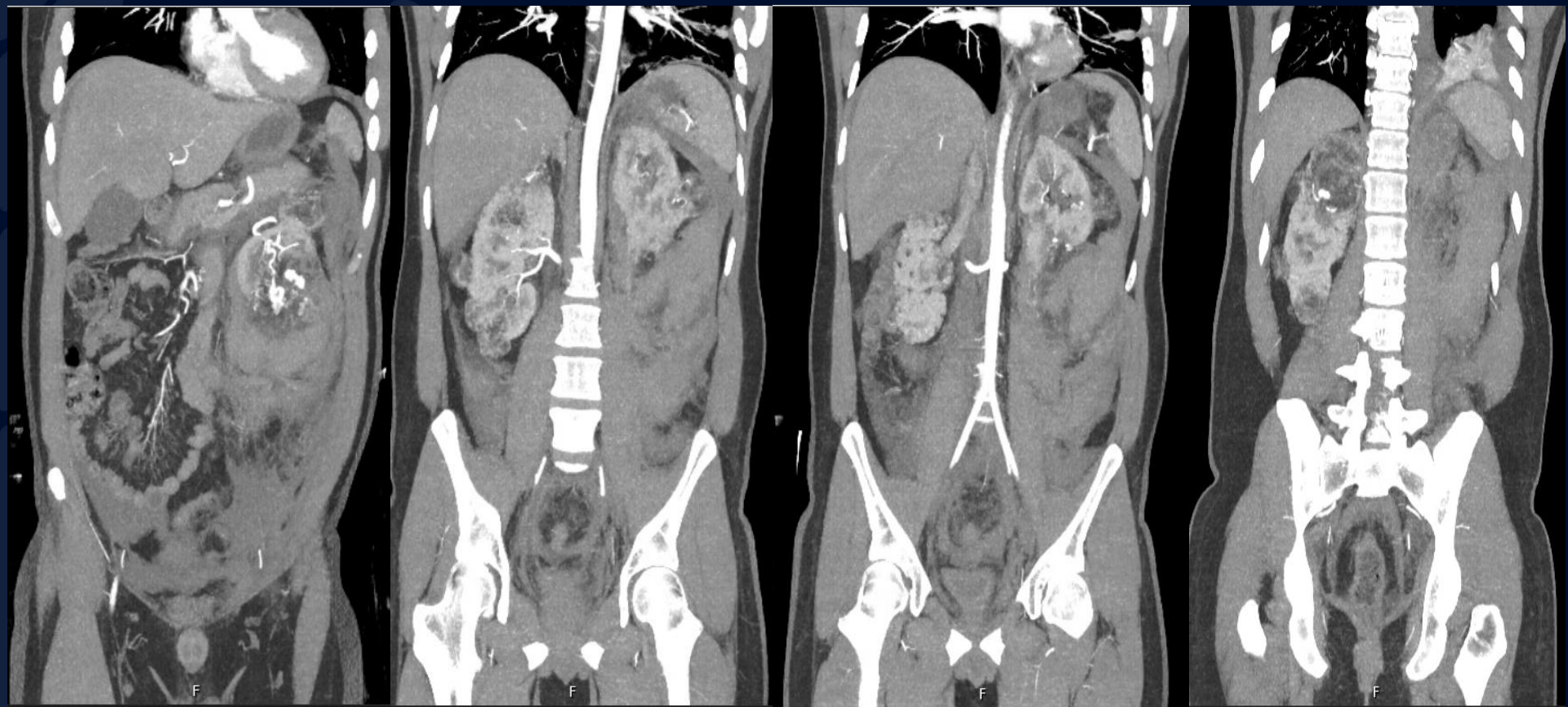
Non-contrast CT



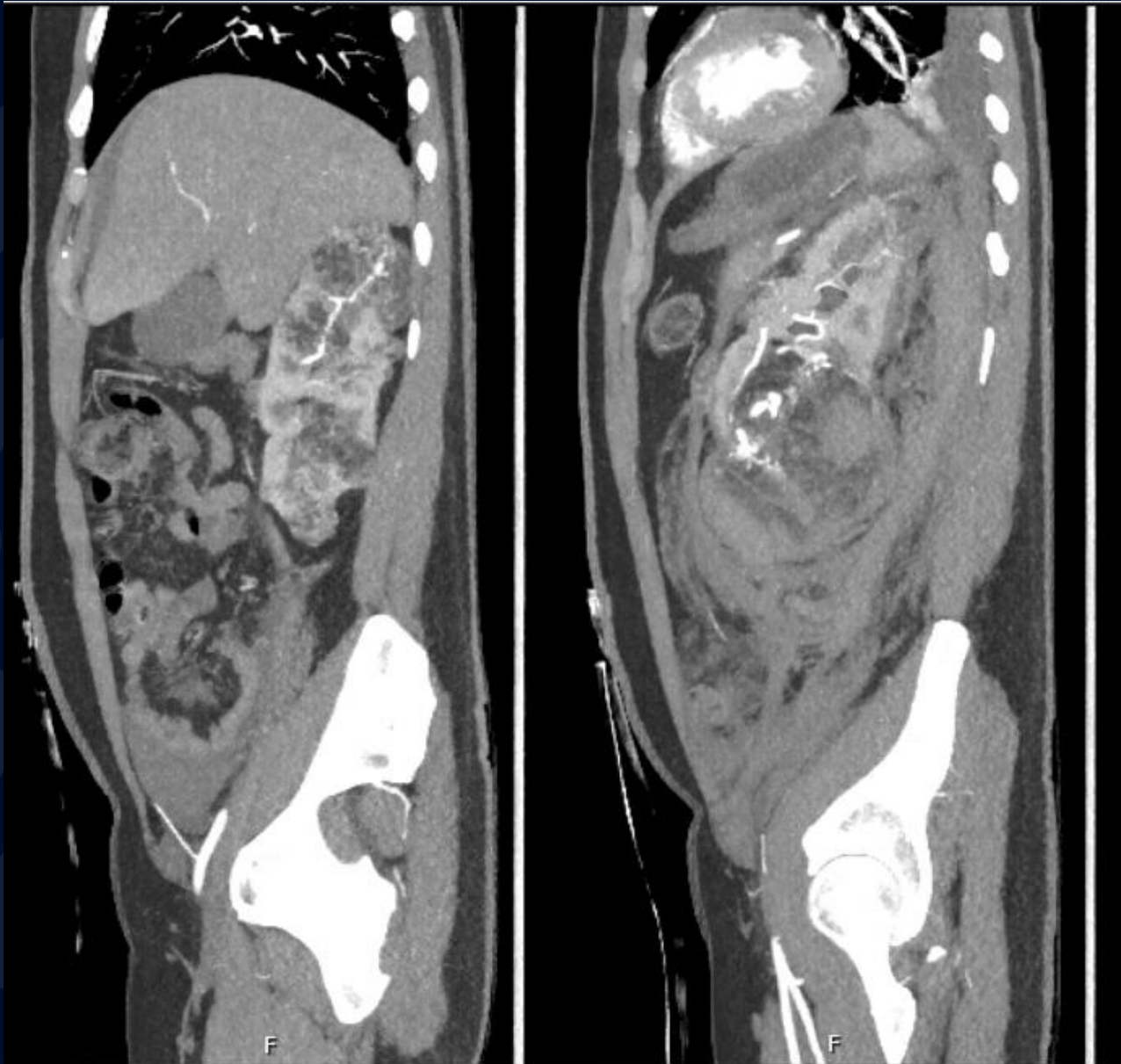
Non-contrast CT



Contrast enhanced CT, Nephrogenic Phase



Contrast enhanced CT, Nephrogenic Phase

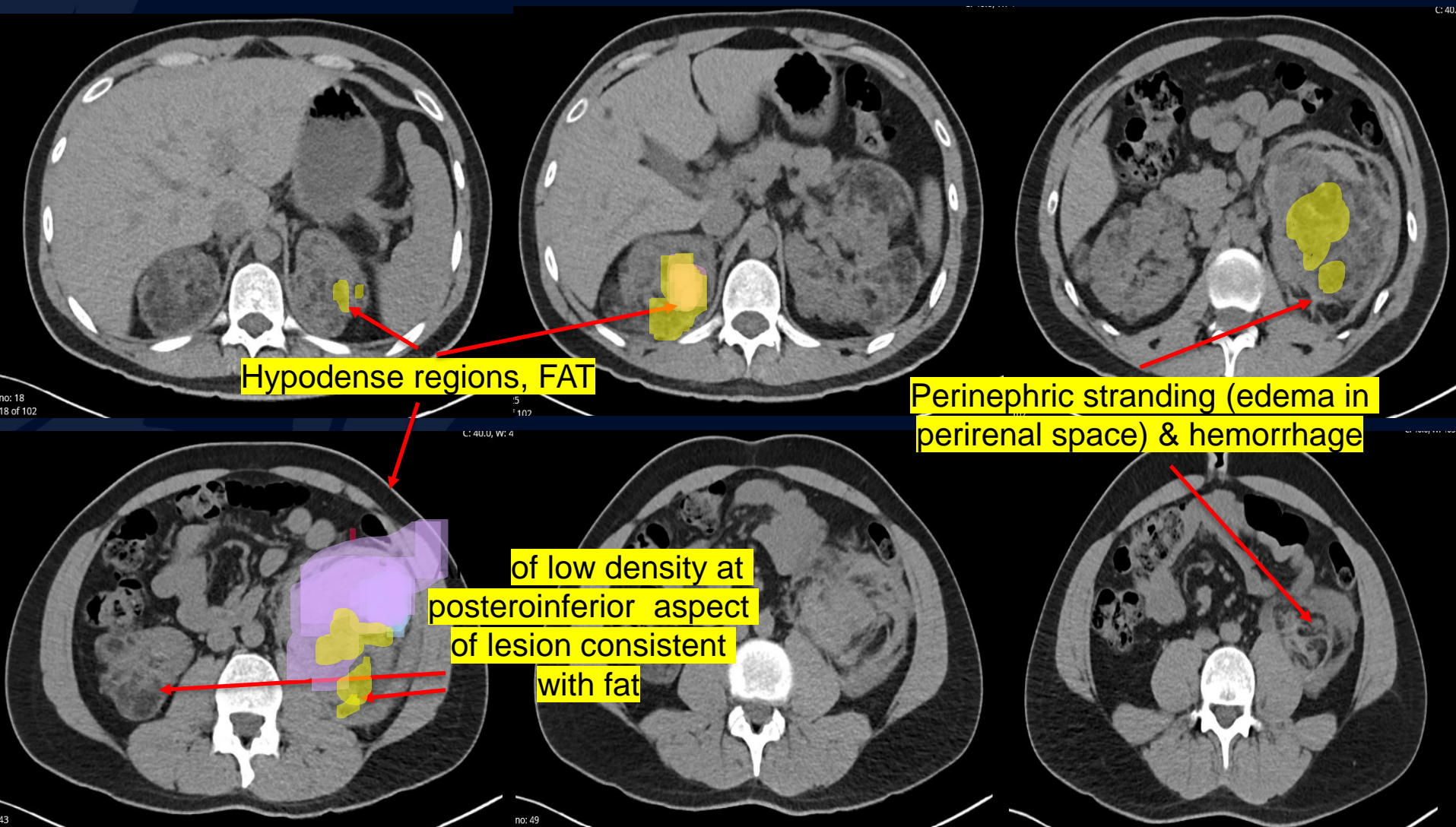


A large, stylized oak leaf graphic in a dark blue color, positioned on the left side of the slide.

?

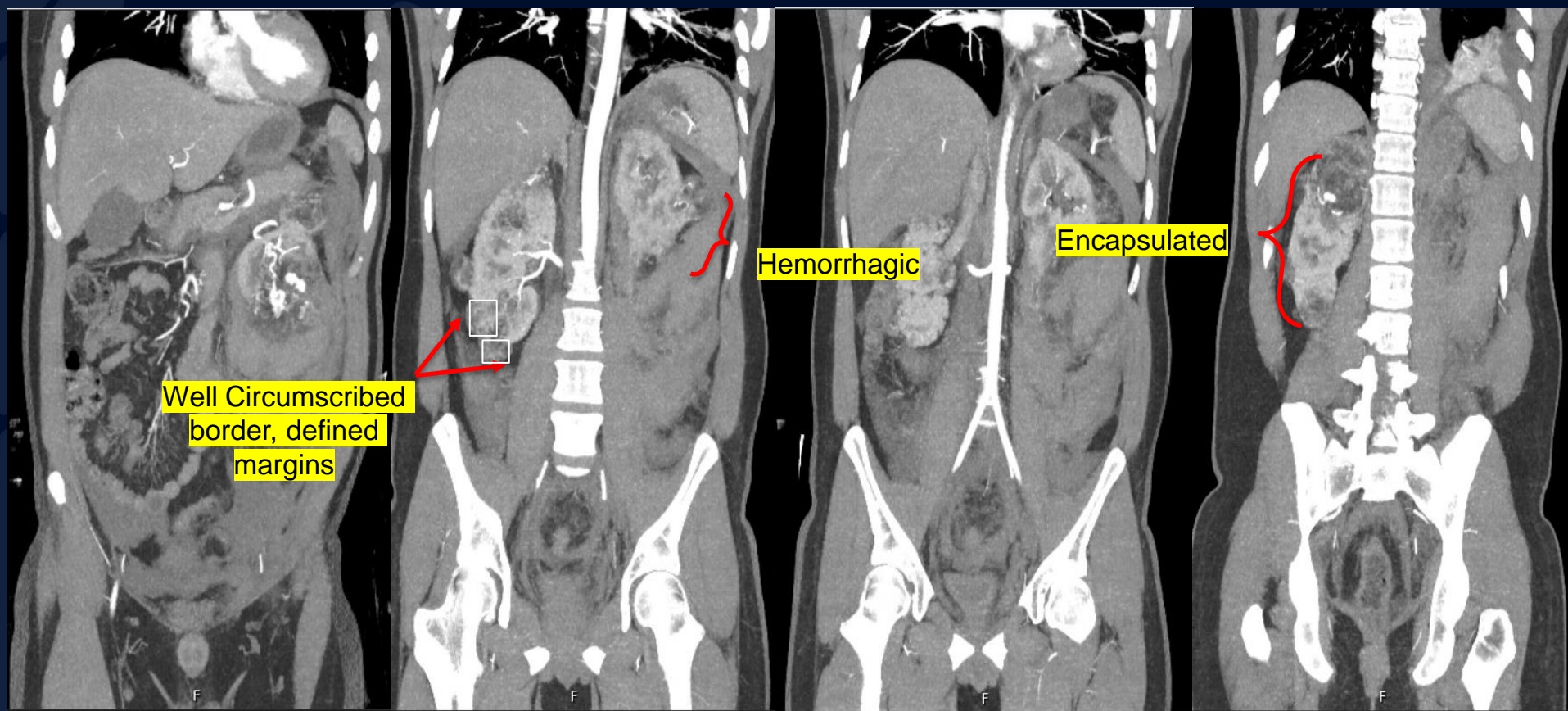
Angiomyolipoma

Non-contrast CT



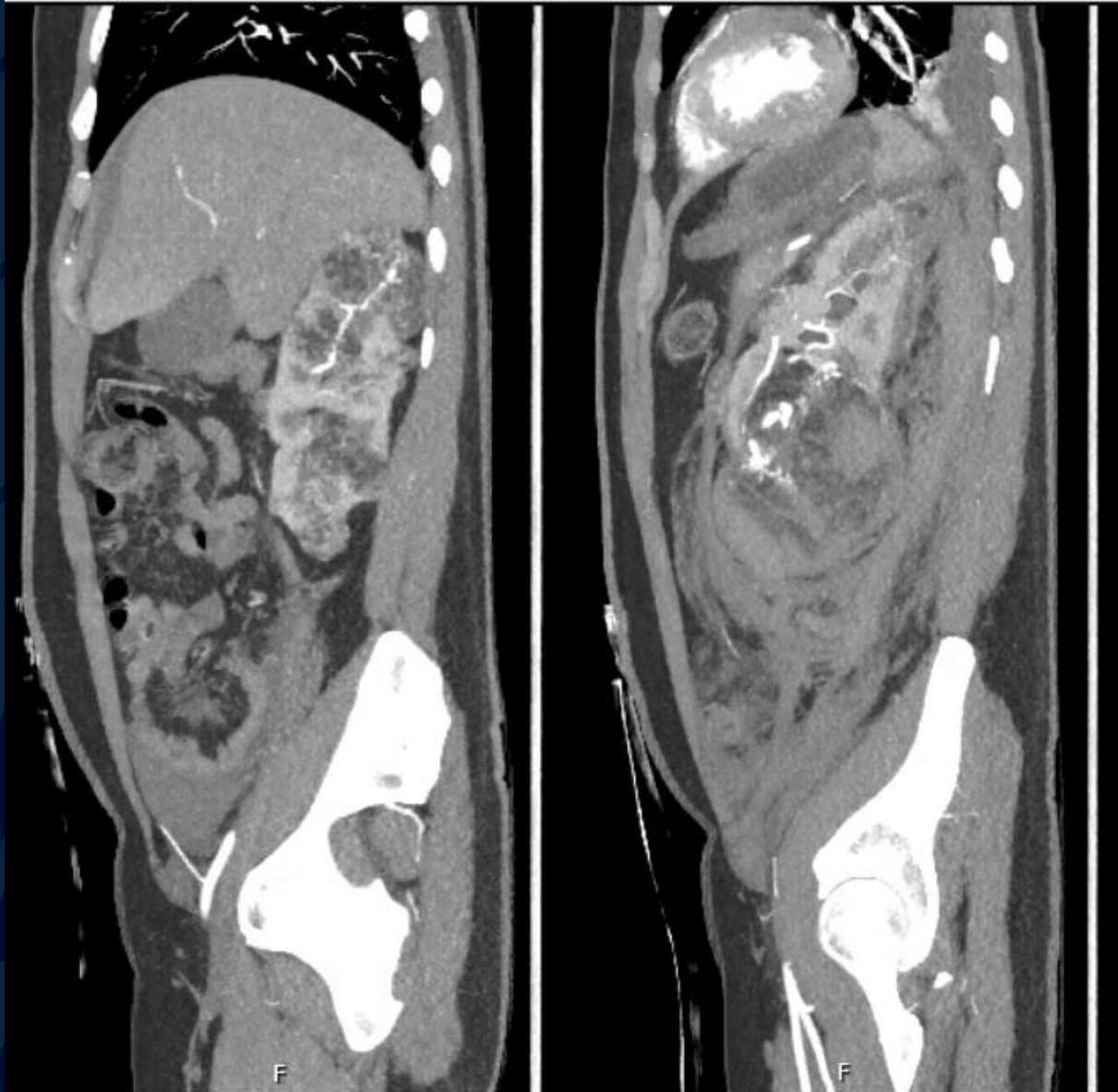
> 10 cm left exophytic, lower pole of heterogenous density

Contrast enhanced CT, Nephrogenic Phase



Large heterogenous mass with macroscopic fat content, measuring about 17 cm in craniocaudal dimension

Contrast enhanced CT, Nephrogenic Phase



Suspicion for active bleed from AML on the left side

Multiple AML's > 4 cm

Abnormal aneurysmal dilations within AML's bilaterally

Angiomyolipoma (AML)

Common benign renal mass, often detected incidentally or following an acute bleed (particularly in masses > 6 cm)

Given the multifocality and bilaterality of this presentation, there is high suspicion for underlying hereditary etiology

- Differential Diagnosis for hereditary AML:
 - Tuberous Sclerosis Complex (most likely)
 - Up to 80% of patients with TSC will develop AML
 - Von Hippel Lindau
 - Neurofibromatosis

CT

- Both non-contrast and contrast CT Abdomen consistent with multiple non-enhancing hypodense, heterogenous renal masses suggestive of fat containing masses

Treatment for AML in Patients with TSC

- 1st line treatment of asymptomatic AML's > 3cm is low dose Everolimus
- 2nd line treatment is prophylactic embolization or nephron sparing surgery

References

- Oesterling JE, Fishman EK, Goldman SM et al. The management of renal angiomyolipoma. J Urol. 1986;135:1121.
- Nelson CP, Sanda MG. Contemporary diagnosis and management of renal angiomyolipoma. J Urol. 2002;168:1315.
- Fujii Y, Ajima J, Oka K et al. Benign renal tumors detected among healthy adults by abdominal ultrasonography. Eur Urol. 1995;27:124.
- Curatolo P, Bombardieri R, Jozwiak S. Tuberous sclerosis. Lancet. 2008;372:657.
- Franz DN. Everolimus: an mTOR inhibitor for the treatment of tuberous sclerosis. Expert Rev Anticancer Ther. 2011;11:1181.
- Meraj R, Wikenheiser-Brokamp KA, Young LR et al. Lymphangioliomyomatosis: new concepts in pathogenesis, diagnosis, and treatment. Semin Respir Crit Care Med. 2012;33:486.
- Seyam RM, Bissada NK, Kattan SA et al. Changing trends in presentation, diagnosis and management of renal angiomyolipoma: comparison of sporadic and tuberous sclerosis complex-associated forms. Urology. 2008;72:1077.
- Sooriakumaran P, Gibbs P, Coughlin G et al. Angiomyolipomata: challenges, solutions, and future prospects based on over 100 cases treated. BJU Int. 2010;105:101.
- Dyer R, DiSantis DJ, McClennan BL. Simplified imaging approach for evaluation of the solid renal mass in adults. Radiology. 2008;247:331.
- Bosniak MA. Angiomyolipoma (hamartoma) of the kidney: a preoperative diagnosis is possible in virtually every case. Urol Radiol. 1981;3:135.
- Simpson E, Patel U. Diagnosis of angiomyolipoma using computed tomography--region of interest ≤ 10 HU or 4 adjacent pixels ≤ 10 HU are recommended as the diagnostic thresholds. Clin Radiol. 2006;61:410.
- Steiner MS, Goldman SM, Fishman EK et al. The natural history of renal angiomyolipoma. J Urol. 1993;150:1782.
- Davenport MS, Neville AM, Ellis JH et al. Diagnosis of renal angiomyolipoma with Hounsfield unit thresholds: effect of size of region of interest and nephrographic phase imaging. Radiology. 2011;260:158.