Renal Angiomyolipoma

* Benign tumor composed of fat, smooth muscle cells and thick-walled vessels; believe to originate from **perivascular epithelioid cells** (PEC) (aka, **PEComa**).
* Related to other PEComas such as lymphangioleiomyomatosis, "sugar" tumor of lung, cardiac rhabdomyomas, etc.
* May arise in association with **tuberous sclerosis:**
  + >50% of patients with TS develop AMLs; <50% of AML has TS.
  + Tend to be asymptomatic, bilateral, small, and multiple.
  + Associated with genetic alteration in TSC1 (chr 9q34) and TSC2 (chr 16p13.3).
* Abundant fat containing tumor recognizable radiologically.
* Gross:
  + Range from 1-20 cm (average 9 cm).
  + Golden-yellow appearance is common, but depends on the ratio (>fat looks like lipoma; > smooth muscle looks like leiomyoma) [**(image A)**](https://www.auanet.org/images/education/pathology/kidney-mesenchymal/renal-figureA_Big.jpg).
  + Usually well-demarcated but not encapsulated.
  + May show local "invasion" or extension into perinephric or sinus fat.
* Histology:
  + Consists of varying amounts of smooth muscle cells, mature fat and thick-walled vessels   
    [**(image B)**](https://www.auanet.org/images/education/pathology/kidney-mesenchymal/renal-figureB_Big.jpg) and [**(image C)**](https://www.auanet.org/images/education/pathology/kidney-mesenchymal/renal-figureC_Big.jpg).
  + Vessels abnormal, often hyalinized and thick with eccentric lumen.
  + Smooth muscle cells appear to originate and "radiate" off the vessels [**(image D)**](https://www.auanet.org/images/education/pathology/kidney-mesenchymal/renal-figureD_Big.jpg).
  + AML with benign epithelial cysts (AMLEC) is a rare variant.
* IHC: spindle cells reactive with melanoma marker (HMB-45+, Mart1+) and actin+ thereby distinguishing it from other spindle cell lesions of the kidney.
* DDX: lipoma (for fat predominant AML), leiomyoma (for smooth muscle predominant AML).
* Benign course.
* Surgical intervention recommended for tumors >4 cm (retroperitoneal hemorrhage is common complication)