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ANGIOMYOLIPOMAS (AML)

Angiomyiolipoma (AML) is a benign mesenchymal tumour, belongs to a family of so-called PEComas (perivascular epithelioid cell tumours), characterised by the proliferation of perivascular epithelioid cells. Some PEComas can behave aggressively and even metastasize, while classic AMLs **are completely benign**

Overall prevalence **0.44%**. Only 5% present with multiple AMLs. These lesions are often discovered incidentally during imaging for other reasons or as part of screening in patients with **tuberous sclerosis** and pulmonary **lymphangioleiomyomatosis**.

CLINICAL MANIFESTATIONS:

- Asynthomatic (75%)
- Spontaneous **retroperitoneal hemorrhage** (1.55%) including Wunderlinch Syndrome due to microaneurysm rupture within the vascular component
- Palpable mass
- Flank pain (15%)
- Urinary tract infections
- Hematuria, Renal failure, Hypertension

HISTOLOGICAL VARIANTS:

- Classic: <10% epithellial cells. **Benign**
- Epitheloid: >80% epithellial cells. **Malignant**.
- Cystic: posivity to melanocytic markers. **Benign**

THE RISK OS BLEEDING IS PROPORTIONAL TO:
- The size of the lesion **(>4 cm** diameter).
- Aneurysm >5mm.
- Grade of angiogenic component
- Presence of **tuberous sclerosis**

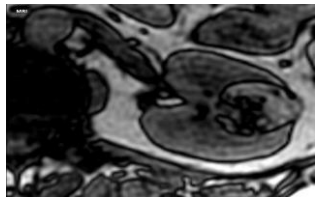
ULTRASOUND:

Hyperechoic lesions in the cortex and with beam attenuation posteriorly.

CT:

Most lesions involve the cortex and demonstrate macroscopic fat (**less than -20 HU**). Absence of ossification/calcification on imaging is in favor of AML

MRI:



Fat-saturated techniques: high signal intensity in non-fat-saturated sequences and loss of signal following fat saturation.
In-phase and Opposed-phase Imaging: Chinese ink artifact at the interface between fat (macroscopic) and non-fat, which will separate the AML from the rest of the kidney. The signal drop in the opposed-phase may indicate a lipid-poor AML (with microscopic fat).

Rarely renal cell carcinomas (RCC) may have macroscopic **fat components** and as such the presence of fat is strongly indicative of AML, but **not pathognomonic.**

Percutaneous biopsy is rarely useful

FOLLOW UP AND TREATMENT:

- **<2cm**: Re- evaluated every 3-4 years.
- **2-4 cm**: Re-evaluated every year with Ultrasound.
- **>4cm** (without previous treatment): Re-evaluated every 6 months with Ultrasound → every year.

Selective arterial embolisation (SAE) | **Thermal ablation** | **Nephron-sparing surgery (NSS)**

Active surveillance is the most appropriate option for most AMLs (48%)
When surgery is indicated, **nephron-sparing surgery (NSS) is the preferred option**, if technically feasible

The association between AML **size** and the **risk of bleeding remains unclear** and the traditionally used 4-cm cut-off **should not per se trigger active treatment**

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Treat angiomyolipoma (AML) with selective arterial embolisation or nephron-sparing surgery, in: • large tumours (a recommended threshold of intervention does not exist); • females of childbearing age; • patients in whom follow-up or access to emergency care may be inadequate; • persistent pain or acute or repeated bleeding episodes.	Weak
Offer systemic therapy (everolimus) to patients at need for therapy with surgically unresectable AMLs not amendable to embolisation.	Weak