Minimal or invisible fat angiomyolipoma: imaging findings in 86 cases

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<td>M. J. Díaz Candamio¹, J. Martel Villagrán², M. Sebastiá³, S. Martín⁴, M. Gil⁵, S. Comellas⁶, F. Matute⁷, P. Domínguez⁷; ¹Ferrol/ES, ²Alcorcón/ES, ³Barcelona/ES, ⁴Palma/ES, ⁵Lleida/ES, ⁶Badajoz/ES, ⁷Madrid/ES</td>
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Aims and objectives

Background

Angiomyolipoma (AML), a member of the perivascular epithelioid cells tumour group (PEComas), is the most common benign solid renal lesion. Triphasic, classic, AML, has three components: blood vessels (angio), spindle cells (myo) and fatty tissue (lipo) (Fig. 1). AMLs are usually found incidentally by ultrasound or CT, and when small require no treatment, although follow-up is recommended.

AML imaging diagnosis is usually easy, achieved by detecting a fatty component - a not pathognomonic finding- in a solid kidney lesion (Fig. 2).

In a little and unknow percentage of AML the fat component is hard to detect on imaging, being described as minimal or invisible fat AML (MIFAML). This is a radiological diagnosis without pathological correlate, since fat is always present microscopically in every AML.

MIFAML are still sometimes surgically resected due to a malignancy suspicion, since imaging findings overlap those of renal cell carcinoma, with the subsequent expenses and patient discomfort, that can even result to the loss of an entire kidney.

Previous studies described the MIFAML imaging findings useful for differentiation with malignant kidney lesions. These were small series, since it is a quite unusual AML presentation. The aim of the present study was to apply these know findings to analyse a larger MIFAML series.
Images for this section:

Fig. 1: Sagittal section of a human kidney containing multiple AML, seen as variably-sized yellow tumor nodules. Angiomyolipomas are multifocal in about 30% of cases and bilateral in about 15% of cases. Multifocality and bilaterality is frequently associated with tuberous sclerosis.

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Methods and materials

We retrospectively analysed the CT and MRI findings of pathologically proven AML cases retrieved during the last ten years from the electronic health records of seven public hospitals in Spain:

Complejo Hospitalario Universitario de Ferrol, A Coruña.
Hospital Universitario Fundación Alcorcón, Madrid.
Hospital Clínic i Provincial de Barcelona.
Hospital Universitari Son Espases, Palma de Mallorca.
Hospital Universitari Arnau de Vilanova, Lleida.
Hospital Perpetuo Socorro, Badajoz.
Hospital Universitario Clínico San Carlos, Madrid.

Only those cases with presurgical cross-sectional imaging studies reporting a solid kidney lesion with a renal cell carcinoma suspicion were included, and they were a priori considered to be MIFAML.

Two cases of epithelioid AML have not been included in this MIFAML series, since they have histologically no fat and they have malignant potential.

Imaging MIFAML findings were reviewed at the respective hospital PACS, looking for the data previously considered to be useful to the differential diagnosis with renal cell carcinoma.

The whole sample in our study consisted of 86 patients with MIFAML, including 62 women and 24 men (age range, 22-84 years; mean age, 61 years).

Most of the lesions were resected by means of tumorectomy / partial nephrectomy (n=55) or radical nephrectomy (n=28). The other three pathology samples were obtained by percutaneous biopsy, and the lesion was not resected.
Results

CT findings (n=86):

All patients underwent a 64 or 16-slice multidetector CT study. A baseline, non-contrast study was performed only to 40 patients. All patients underwent a post intravenous contrast administration CT study, at varying timing phases (arterial, corticomedullary, portal, nephrographic, delayed) (Fig. 5 - Fig. 10)

One of the patients had a tuberous sclerosis diagnosis.

Six patients had another simultaneous AML and two a coincidental renal cell carcinoma.

In three cases a coincidental ipsilateral renal vein thrombosis was found.

The mean size of MIFAML was 18 mm (range 7-41mm).

All but one lesion had well-defined margins.

Calcifications were present only in three lesions.

Haemorrhage was detected in four of the lesions.

82.5 % of the lesions were cortical based (n=71)

At unenhanced CT (n=40), 65% of NIFAML were hyperattenuating (high tumour-to-cortex attenuation ratio) (n=26), 90% homogeneous (n=75), and 35 % showed negative pixels indicating the presence of fat in any phase (n=30).

At enhanced CT, 75% of the lesions showed homogeneous enhancement (n=64), and 35% an angular interface (n=31)

MIFAML usually presented as a well-defined homogeneous solid lesion on CT, without haemorrhage or calcifications. Important ancillary findings such as the high attenuation of the lesion in the baseline study and an angle interface can add further clues to the MIFAML diagnosis.

Although MIFAML are defined as AML lesions where the fatty component cannot be detected at cross-sectional imaging, at 35% of the lesions studied here we retrospectively were able to detect fatty components, defined as the presence of negative pixels (under -10 H.U.). The inclusion of a basal phase and the close evaluation of these sometimes tiny fatty components could have avoided many of the performed surgeries.
Some MIFAML cases were particularly hard to diagnose, such as the one presenting as a pelvis renal soft tissue lesion mimicking urotelial carcinoma Fig. 9), or the three cases presenting with a concurrent renal vein thrombosis, that would make plausible a renal cell carcinoma diagnosis.

**MRI (1.5T) findings (n=10):**

70% of the lesions had low tumour-to-cortex SI ratios on T2-weighted MRI sequences (n=7).

60% of the MIFAML showed intravoxel fat on opposed-phase and in-phase gradient-echo MRI (n=6) (Fig. 11)

Our small sample of MIFAML patients to whom MRI was performed does not allow us to draw conclusions, although it confirms the previously reported findings that MIFAML usually have a low T2 SI and not always show the india ink artifact at the in and out of phase GRE study.
Images for this section:

**Fig. 1:** Sagittal section of a human kidney containing multiple AML, seen as variably-sized yellow tumor nodules. Angiomyolipomas are multifocal in about 30% of cases and bilateral in about 15% of cases. Multifocality and bilaterality is frequently associated with tuberous sclerosis.

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**Fig. 2:** Typical non contrast enhanced CT appearance of a renal AML: easy diagnosis based on the lesion fatty components detection. On the contrary, MIFML are characterized by minimum or absent fat attenuation at cross-sectional imaging, which makes it difficult to diagnose.

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Fig. 3: MIFAML contrast enhanced CT study. Well circumscribed exophytic homogeneous cortical soft tissue nodule. Attenuation (60 HU) is maintained at arterial (a) and portal (b) phases. No basal CT study was performed. The lack of a non contrast phase precludes the assessment of the most relevant MIFAML findings. Instead of percutaneous biopsy, a laparoscopic tumorectomy was performed. Final diagnosis: AML.
Fig. 4: MIFAML basal (a), arterial phase (b) and portal phase (c) CT study. A well defined soft tissue exofitic lesion showing hiperattenuation at basal CT and homogeneous and maintained enhancement after intravenous contrast administration. No fatty pixels were detected, but these findings should rise to a MIFAML suspicion and a percutaneous biopsy should have been carried out. Nevertheless, a laparoscopic tumorectomy was performed, with a AML histological diagnosis.
Fig. 5: MIFAML mimicking renal cell carcinoma. A basal non contrast-enhanced study (a) shows an homogeneous well-defined 2 cm nodule hyperattenuating in relation to the renal cortex in which negative attenuation pixels are detected. The nodule exhibits heterogeneous enhancement, stronger at the arterial (b) than at the portal (b) phase. Although the enhancement pattern mimics renal cell carcinoma, the finding of a tiny fatty component should have led to percutaneous biopsy. Laparoscopic tumorectomy was performed. Anatomopathological diagnosis: AML.

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Fig. 6: MIFAML basal (a), arterial (b) and portal phase (c) CT study. No fatty pixels were detected in this case, but the hiperattenuation of the lesion at basal CT and the homogeneous and maintained enhancement make the MIFAML diagnosis possible. Percutaneous biopsy should have been carried out. A laparoscopic tumorectomy was performed, with a AML pathology diagnosis.

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**Fig. 7:** A well defined hiperattenuating at basal CT (a) and homogeneously enhancing at portal phase CT (b) lesion. The nodule shows an angular interface with the cortical. All these findings suggest MIFAML, that should have been ruled out by percutaneous biopsy. A laparoscopic tumorectomy was performed, with AML as final diagnosis.

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Fig. 8: MIFAML corticomedullary phase (a), nephrographic phase (b) and delayed phase (c) coronal CT reformatting. No basal phase was done in this case. Nevertheless, tiny fatty pixels were detected even after intravenous contrast. A suspicion MIFAML diagnosis and a percutaneous biopsy should have been done. A laparoscopic tumorectomy was performed, with an AML diagnosis.

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Fig. 9: MIFAML mimicking an urotelial tumor. No basal study was performed. At a portal phase CT study (a) a pelvis renal soft tissue lesion was found, and a 10 min delayed phase (b) was performed. After nephrectomy, the lesion was anatomopathologically characterised as a AML. Retrospectively, low attenuation pixels were seen at the delayed phase.

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**Fig. 10:** Epithelioid AML presenting as pelvis renal lesion, mimicking urotelial tumour. No negative pixels or other findings suggesting MIFAML were found. Pathological diagnosis was epithelioid AML, a rare AML variant. In fact, two cases of epithelioid AML have not been included in this MIFAML series, since they have no fat and they have malignant potential.

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**Fig. 11:** In (a) and out (b) of phase GRE MRI images. Tiny amounts of fat are sometimes (as in this case) but not always found at MIFAML MRI studies. MRI was performed in rare occasions in our study, so it has not been possible to clarify its diagnostic profitability.

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Fig. 12: Percutaneous CT-guided MIFAML biopsy. Giving the overlapping CT features of MIFAML and kidney malignant lesions, our data lead us to think that percutaneous biopsy should be done more often, trying to avoid the unnecessary resection of these benign lesions.

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Conclusion

This is the largest reported MIFAML series, composed by 86 solid kidney lesions incidentally found at imaging studies, resected because a renal cell carcinoma suspicion. Being a retrospective study, imaging of these incidental lesions was performed with different protocols, even at the same institution, making difficult to evaluate the imaging findings.

Many of the MIFAML studied could have avoid surgery if a complete solid renal lesion protocol would have been performed and a thorough analysis of the existing fatty components would have been carried out. It must be encouraged to always include a baseline study in the CT protocol of a solid kidney lesion, to particularly look for the low attenuation pixels that indicate the presence of fat, and to demonstrate the possible basal high attenuation of a MIFAML. Corticomedullary and nephrographic CT phases and a MRI study can add further information.

There are many image data orienting in favour of a diagnosis of MIFAML, but they are not specific nor frequent. Given the MIFAML imaging findings overlapping with those of renal cell carcinoma we can not label a lesion as a MIFAML without doing a biopsy (Fig. 12), which if possible should be performed percutaneously, reducing the morbidity of an open or laparoscopic surgery.

In the near future artificial intelligence techniques and machine learning with CT texture analysis could achieve a further insight on these lesions, with the ultimate aim of avoiding the need to biopsy a lesion characterized as MIFAML by imaging findings.
Personal information

María Jesús Díaz Candamio
Complejo Hospitalario Universitario de Ferrol (CHUF).
A Coruña, Spain.
Maria.Jesus.Diaz.Candamio@sergas.es

José Martel Villagrán
Hospital Universitario Fundación Alcorcón (HUFA).
Madrid, Spain.
jmartel@fhalcorcon.es
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