Case Report

Fat Deficient Renal Angiomyolipoma Mimicking Renal Cell Carcinoma — A Diagnostic Challenge

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Abstract

Background : Renal Angiomyolipomas (AMLs) are benign tumors usually diagnosed on imaging by their typical appearance due to their high fat content. Atypical or fat poor renal AMLs are difficult to diagnose on imaging and mimics Renal Cell Carcinoma (RCC). We report a case of 50-year-female who underwent laparoscopic right partial lower pole nephrectomy for right RCC and which proved to be an atypical AML on histopathology and immunohistochemistry.

Key words : Angiomyolipoma, Renal Cell Carcinoma, HMB45, Renal Tumor, Adipose Tissue.

A benign kidney tumour called a renal Angiomyolipoma (AML) has varied quantities of smooth muscles, mature adipose tissue and dysmorphic blood vessels[2]. The majority of renal AMLs are classified as "Classical AML" because they are typically fat-rich and have a distinctive look on CT or MRI scans. Some renal AMLs can be mistaken for renal cell carcinoma because they have very few adipocytes, are difficult to identify on imaging and are rare Renal Cell Carcinoma (RCC). We present a case of a 50-year-old female who underwent laparoscopic partial right lower pole nephrectomy due to right RCC and was later diagnosed with a fat deficient AML based on histology and immuno-histochemistry.

CASE REPORT

50-year-female presented to us with on and off dull aching abdominal pain for 1 year. There was no history of lower urinary tract symptoms, loss of weight or appetite. On abdominal examination there was no significant finding. Complete Blood Count, renal and liver function were also within normal limits. An ultrasound of whole abdomen was ordered which showed a 6x5 cm right perirenal soft tissue mass reaching up to the Morrison pouch with internal vascularity. Contrast enhanced MRI revealed a 5.2x2.8x3.8 cm heterogenous signal intensity lesion seen in right perirenal space appearing hyperintense in T1 and

> **Editor's Comment :**
> ■ Fat deficient AMLs appear similar to RCC on imaging and can be misdiagnosed as RCC. Whenever in doubt, one may order a core tissue biopsy supported by immuno-histochemistry to clinch the diagnosis that may affect the subsequent management.

hypointense in T2/DW1 with enhancement on post contrast study. Ultrasound guided Fine Needle Aspiration Cytology (FNAC) from the perirenal mass was done and came out to be positive for malignant cells (Fig 1). Based on imaging and FNAC findings a diagnosis of RCC was made and laparoscopic partial lower pole nephrectomy was performed on the right side.

On histopathology the tumor was seen focally invading the perinephric fat at lower pole of right kidney with resection margins being free of the tumor. Microscopically the tumor display spindle shaped cells with many intervening thin and thick-walled blood vessels. These cells focally exhibit epithelioid morphology and only few areas showed scanty adipocytes. On immuno-histochemistry the tumor cells show immuno-reactivity for HMB-45, SMA, AMACR andfocal positivity for EMA and negative for CK-7 (Fig 2). Based on immunohistochemistry and histopathology finding angiomyolipoma was the final diagnosis.

DISCUSSION

AML is a benign tumor composed of varying amount of fat, smooth muscles and blood vessels. It can occur at various other sites like the skin, appendix, colon, liver, lung, kidney and the smooth muscle fibers. It occurs sporadically or as a part of syndrome of Tuberous Sclerosis. Most AMLs are easily diagnosed because the high fat content gives them a characteristic appearance on imaging. On immuno-histochemistry AML are positive for HMB-45, Melan-A, CD68, CD117 and Ki-67[3].
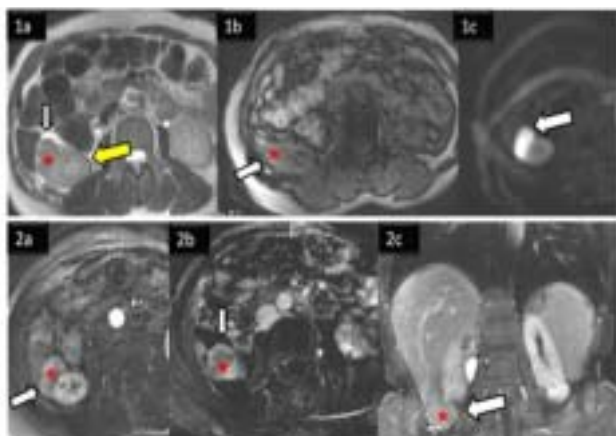
Fig 1 — 1(a) T2 weighted image of the upper abdomen shows a rounded solid signal intensity mass lesion (*) projecting exophytically from the lower pole of the right kidney (bold yellow arrow) showing intermediate-low signal intensity. 1(b) The exophytic mass (*) does not show any signal drop on (out of phase) chemical shift imaging (white bold arrow) sequence depicting absence of macroscopic fat within it. 1(c) Lesion (white arrow) shows restriction on Diffusion weighted sequence suggestive of tumor component. 2(a) the exophytic renal mass (*) shows mild enhancement on the arterial phase of the dynamic contrast study of the abdomen (white arrow). 2(b) the renal mass (*) shows peripheral enhancement on subsequent phase without washout. 2(c) Coronal sequence shows the renal mass (*) with enhancement on subsequent phase without washout suggestive of benign origin.

Recently a variant of renal AML which are fat deficient have been recognized and present as a diagnostic challenge. These lesions due to their low fat content have appearance similar to RCC on imaging and are misdiagnosed as RCC.
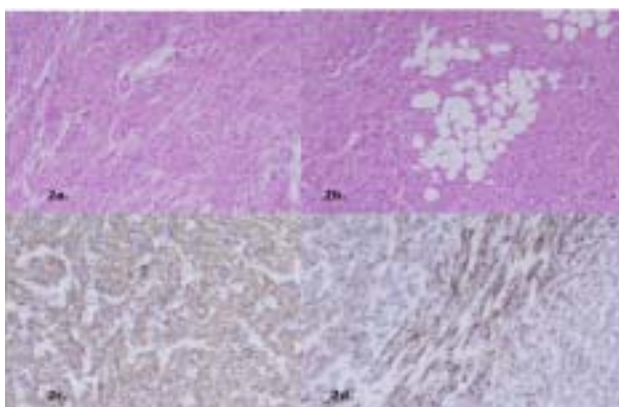

Fig 2 — Renal angiomyolipoma (2a) Smooth muscle component composed of interdigitating fascicles of spindle cells with intervening thick-walled blood vessels. (H&E stain, 10X). (2b) Fat component in the form of mature adipose tissue (H&E stain, 10X). (2c)Tumor cells showing diffuse strong positivity for SMA. (2d) Tumor cells displaying HMB45 positivity.

In our case as the patient had right renal lower pole mass which appeared to be RCC on imaging and was positive for malignant cells on ultrasound guided FNAC, he underwent laparoscopic right partial lower pole nephrectomy for the diagnosis of right RCC. Histopathology and immuno-histochemistry proved it to be a renal AML. The contradictory ultrasound guided FNAC report may be attributed to the spread out of atypical spindle cells and limited material in the smears.

The phrase "lipid-poor AML" or "minimum fat AML" has been used to describe AMLs with very little fatand these lesions account for approximately 5% of all AMLs[4].

Because of the pathophysiology of these lesions, Jinzaki, *et al* classified AMLs as fat-poor AMLs and clarified the existence of many subtypes of fat-poor AMLs to clear up any confusion among readers[1]. These lesions present a diagnostic challenge and there are reports supporting the role of FNAC and tissue biopsy if such fat deficient lesions are encountered on imaging studies and diagnosis is in doubt[5].

CONCLUSION

Atypical or fat poor renal AMLs are difficult to diagnose on imaging and mimics Renal Cell Carcinoma (RCC). The fat deficient lesions mimicking RCC are encountered on imaging and we should not hesitate to order a tissue biopsy immunohistochemistry to reach a final diagnosis and proper treatment and management of the patient.

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Conflict of Interest : None.

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