

Case Report

Rare giant renal angiomyolipoma in a 2-year-old female: a case report

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ABSTRACT

Renal angiomyolipoma (AML) is a benign renal tumor that is uncommon in early childhood, particularly in giant form. We report a 2-year-old girl who presented with progressive abdominal distension, constipation, and vomiting for two months. Examination revealed pallor and a large left-sided abdominal mass, with hemoglobin of 8 g/dl (11-14g/dl). Contrast-enhanced CT showed a 15×12×11 cm [CC×AP×TR] mass arising from the left kidney with areas of fat attenuation and additional fat-density foci in the left renal sinus. Due to the patient's young age and the large size of the renal mass, a preoperative Tru-cut biopsy was performed to establish the tissue diagnosis and guide definitive management. Histopathology demonstrated thick-walled blood vessels, smooth muscle bundles, and nests of clear cells without mitotic figures or necrosis. Immunohistochemistry was positive for HMB-45 and Cathepsin K and negative for cytokeratin and Ki-67, supporting the diagnosis of AML. After correction of anemia and preoperative optimization, the patient underwent left nephrectomy with complete tumor excision and had an uncomplicated postoperative recovery. This case highlights the exceptional rarity of giant renal AML in a very young child and emphasizes considering AML in large pediatric renal masses, particularly when fat-containing components are identified on imaging.

Keywords: Angiomyolipoma, Biopsy, Kidney tumor, Smooth muscle

INTRODUCTION

Renal angiomyolipoma (AML) is an uncommon benign renal mesenchymal tumor characterized by a variable combination of mature adipose tissue, abnormal blood vessels, and smooth muscle components. It occurs predominantly in adults, whereas its occurrence in children and adolescents is relatively unusual.¹ Most renal AMLs are sporadic, although a recognized association exists with tuberous sclerosis complex. In the general population, renal AML has been reported with a relatively low incidence, while its prevalence among surgically resected renal tumors ranges from approximately 0.3% to 3.0%.^{2,3} These lesions are generally small, with most measuring between 2 and 8 cm; however, tumors exceeding 10 cm in diameter are uncommon and are classified as giant renal angiomyolipoma. Because of their considerable size, giant AMLs may pose substantial diagnostic and therapeutic

challenges and carry an increased risk of serious complications, particularly spontaneous hemorrhage and tumor rupture.⁴

CASE REPORT

A 2-year-old female child was brought to the hospital with progressive abdominal distension for approximately two months. The abdominal enlargement was associated with constipation and episodes of vomiting. There was no reported family history of malignancy. On general examination, the child appeared pale. Abdominal examination revealed marked distension with a large palpable mass occupying the left side of the abdomen. The mass extended from the left lumbar region inferiorly toward the left iliac fossa and medially approached the midline. Laboratory evaluation demonstrated anemia, with a hemoglobin level of 8 g/dl (11-14 g/dl).

Contrast-enhanced computed tomography (CT) of the abdomen demonstrated a large, approximately 15×12×11 cm [CC×AP×TR] enhancing mass arising from the left kidney. The lesion contained several small areas of fat attenuation. Additional tiny foci of fat density were identified within the left renal sinus (Figure 1). However, no calcification was observed. Multiple small fat-density lesions were also demonstrated within the hepatic

parenchyma, in association with hepatomegaly, as confirmed in the original radiological assessment. No pulmonary parenchymal nodules were identified. CT imaging of the chest and brain did not demonstrate any significant abnormality. Furthermore, the echocardiography was also normal. Based on the clinical and radiological findings, a left renal mass was considered as angiomyolipoma.

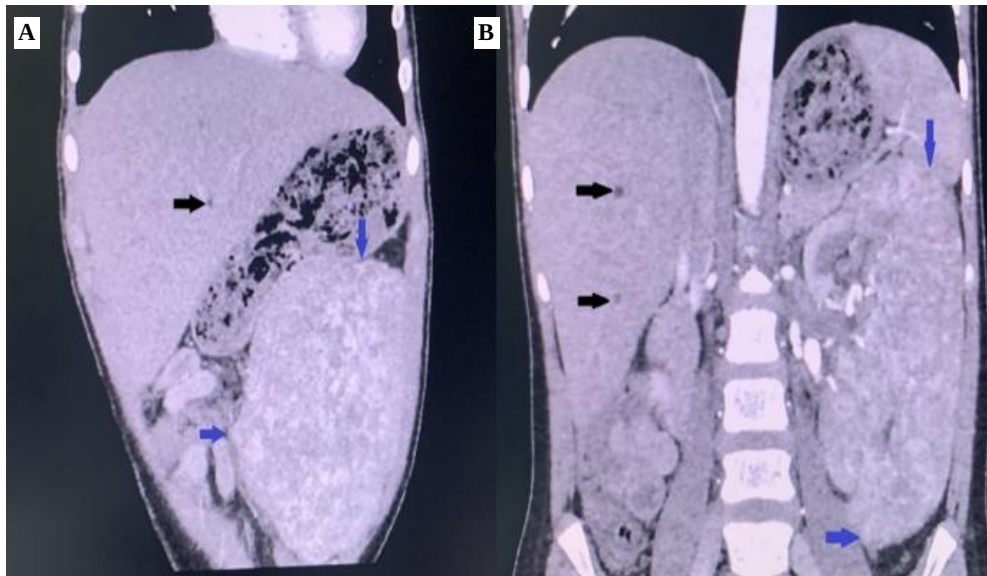


Figure 1: Contrast enhanced CT abdomen (coronal views). (A) This image demonstrates a giant angiomyolipoma arising from left kidney extending up to left iliac fossa crossing midline (blue arrows) with hepatomegaly and a tiny fat density lesion in hepatic parenchyma (black arrow). (B) This image demonstrates a giant angiomyolipoma arising from left kidney extending up to left iliac fossa crossing midline (blue arrows) with hepatomegaly and multiple tiny fat density lesions in hepatic parenchyma (black arrows).

To further characterize the renal lesion, a Tru-cut biopsy was performed. Histopathological examination revealed a lesion composed predominantly of numerous thick-walled blood vessels, with scattered nests of cells showing clear cytoplasm. No mitotic figures or areas of necrosis were identified. Bundles of smooth muscle fibers were also present. Immunohistochemical evaluation showed negativity for cytokeratin (CK) and Ki-67, while HMB-45 and Cathepsin K demonstrated positive staining. The morphological and immunohistochemical findings supported the diagnosis of renal angiomyolipoma.

The patient was optimized preoperatively, including correction of anemia, and subsequently underwent a left-sided nephrectomy with complete excision of the renal mass. The surgical procedure was completed without intraoperative complications, and the postoperative period was also uneventful. The child recovered well following surgery and was discharged with a plan for regular outpatient surveillance. She remains on routine follow-up, with continued clinical monitoring and assessment for any evidence of recurrence or other complications.

DISCUSSION

Renal AML is an uncommon benign mesenchymal renal tumor composed of variable proportions of adipose tissue, dysmorphic blood vessels, and smooth muscle cells. It is predominantly encountered in adults and is particularly uncommon in very young children.⁵

Renal AML may occur sporadically or in association with tuberous sclerosis complex (TSC). Sporadic lesions generally present as solitary renal masses, whereas TSC-associated AMLs are more often bilateral, multifocal, and may occur at a younger age.⁶ The occurrence of a giant renal AML in a 2-year-old child, as in the present case, is therefore an unusual presentation and represents an important diagnostic consideration when evaluating a large pediatric renal mass.

The clinical manifestations of renal AML are variable and depend largely on tumor size and associated complications. Small lesions are frequently discovered incidentally, whereas large tumors may produce abdominal distension, a palpable abdominal mass, pain,

vomiting, or gastrointestinal symptoms secondary to mass effect. Hematuria may occur, particularly in lesions containing abnormal vascular structures. In our patient, progressive abdominal distension was accompanied by constipation and vomiting, while examination demonstrated a large palpable mass occupying the left side of the abdomen. The patient's anemia, with a hemoglobin level of 8 g/dL (11-14 g/dl), was another clinically relevant finding, although there was no reported history of overt hematuria or acute hemorrhage.^{7,8}

Imaging plays a central role in the evaluation of renal AML. The identification of macroscopic fat within a renal lesion on CT is a particularly useful feature supporting the diagnosis of AML. In the present case, contrast-enhanced

CT demonstrated a large 15 × 12 × 11 cm enhancing mass arising from the left kidney with small areas of fat attenuation, together with additional fat-density foci in the left renal sinus.⁹ Multiple small fat-density lesions were also identified in the liver with associated hepatomegaly (Figure 2). The absence of pulmonary nodules and calcifications with normal CT examination of the brain along with normal echocardiography were additional findings in the radiological assessment favoring the diagnosis of angiomyolipoma. Furthermore, the presence of macroscopic fat strongly favors AML, but large or atypical fat-containing renal masses can create diagnostic difficulty, particularly in children, in whom other renal neoplasms must also be considered.¹⁰

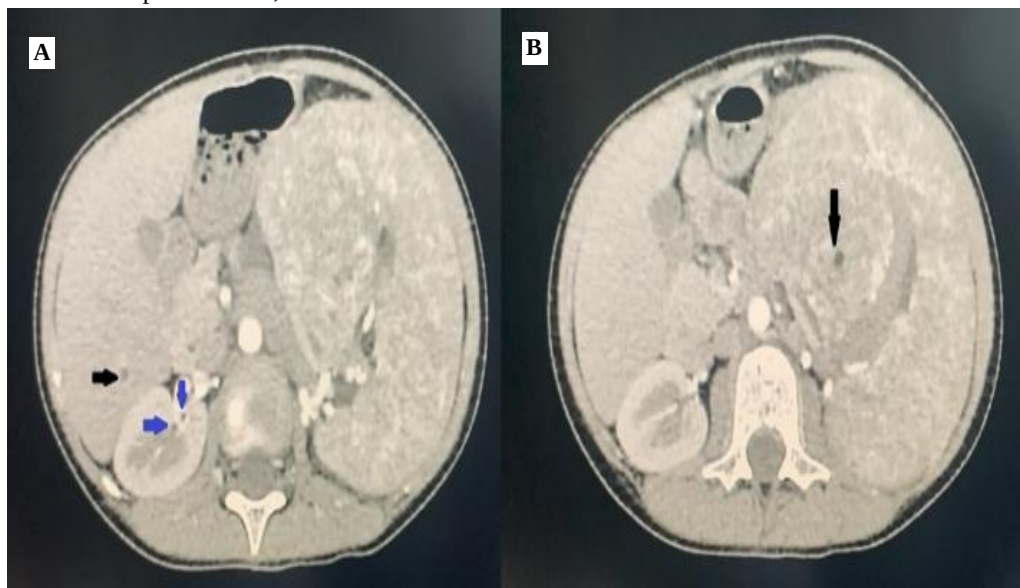


Figure 2: Contrast enhanced CT abdomen (axial views) (A) This image demonstrates few tiny fat density foci in right renal sinus (blue arrows) with tiny fat density lesion in hepatic parenchyma (black arrow). (B) This image demonstrates giant angiomyolipoma arising from left kidney with a tiny focus of fat density (black arrow).

Histopathological examination remains important when the radiological appearance is atypical or when differentiation from other pediatric renal tumors is required. Classical AML demonstrates a characteristic triphasic composition consisting of mature adipose tissue, smooth muscle elements, and thick-walled blood vessels. In our patient, Tru-cut biopsy demonstrated numerous thick-walled blood vessels, nests of cells with clear cytoplasm, and bundles of muscle fibers, without mitotic activity or necrosis.¹¹ The immunohistochemical profile further supported the diagnosis. The tumor cells were positive for HMB-45 and Cathepsin K and negative for cytokeratin and Ki-67. HMB-45 positivity is particularly useful in supporting a perivascular epithelioid cell tumor such as AML, whereas cytokeratin negativity helps distinguish it from epithelial renal neoplasms. Cathepsin K expression can provide additional support for the diagnosis in diagnostically challenging cases.¹²

An important consideration in a pediatric patient with renal AML is the possibility of an underlying syndromic

association, particularly TSC. TSC-associated AMLs commonly demonstrate multifocal or bilateral renal involvement and may occur at a younger age than sporadic tumors. In the present case, the renal lesion was unilateral, and there was no reported family history of malignancy. The available clinical information did not describe other characteristic manifestations of TSC. Nevertheless, given the exceptionally young age at presentation and the presence of additional fat-density hepatic lesions on CT, appropriate clinical assessment for an underlying syndromic association is important. The hepatic findings should be interpreted in conjunction with the final radiological and pathological assessment rather than being assumed to represent malignant metastatic disease.¹³

Management of renal AML depends on tumor size, symptoms, growth pattern, vascular characteristics, risk of hemorrhage, renal function, and the presence of syndromic disease. Active surveillance is generally appropriate for small, asymptomatic lesions, whereas intervention is considered for symptomatic or substantially enlarged

tumors and those at increased risk of hemorrhage.¹⁴ Selective arterial embolization may be useful for controlling or preventing hemorrhage and can preserve functioning renal tissue. Nephron-sparing surgery may also be considered when technically feasible, particularly when preservation of renal function is important. However, very large tumors may require complete renal excision when conservative approaches are unsuitable or when the extent of the lesion makes renal preservation impractical.¹⁵

In this child, the unusually large tumor size, significant abdominal distension, gastrointestinal symptoms, and uncertain nature of a large pediatric renal mass supported definitive surgical management. The patient's anemia was corrected before surgery, following which a left nephrectomy with complete tumor excision was performed. No intraoperative or postoperative complications were encountered, and the child subsequently entered routine follow-up. This favorable immediate outcome emphasizes that, despite its rarity in this age group, giant renal AML can be successfully managed when appropriately evaluated and treated.¹⁴

The present case is noteworthy because of the combination of extremely young age and a giant renal AML presenting as a large abdominal mass. Most renal AMLs are encountered in adults, making a lesion measuring 15 cm in a 2-year-old particularly unusual. The case highlights the importance of considering AML among the differential diagnoses of a large pediatric renal mass, especially when CT demonstrates even small areas of macroscopic fat. Careful correlation of imaging, histopathology, and immunohistochemistry is essential for establishing the diagnosis and distinguishing AML from other renal neoplasms. Early recognition and individualized management can facilitate complete tumor control while minimizing the risk of hemorrhagic and other mass-related complications.⁹⁻¹¹

CONCLUSION

Giant renal angiomyolipoma is an exceptionally uncommon renal tumor in early childhood. This case of a 15 cm renal AML in a 2-year-old girl demonstrates that AML should be considered in the differential diagnosis of a large pediatric renal mass, particularly when fat-containing components are identified on CT. Accurate diagnosis through radiological assessment, histopathology, and immunohistochemistry is essential for appropriate management. In this patient, correction of anemia followed by complete surgical excision resulted in an uncomplicated postoperative course. Individualized treatment and continued follow-up are important in children with giant AML to monitor for recurrence and other potential complications.

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REFERENCES

- Faraji H, Nguyen BN, Mai KT. Renal epithelioid angiomyolipoma: a study of six cases and a meta-analytic study. Development of criteria for screening the entity with prognostic significance. *Histopathology*. 2009;55(5):525-34.
- Çalışkan S, Gümrükçü G, Özsoy E, Topaktaş R, Öztürk MI. Renal angiomyolipoma. *Rev Assoc Med Bras* (1992). 2019;65(7):977-81.
- Schieven LW, Smedts F, Hopman AH, van der Wijk J, Nijman RJ, de Jong IJ. Fine needle aspiration using improved agar microbiopsy is highly concordant with renal mass final diagnosis and subclassification. *J Urol*. 2009;182(6):2590-3.
- Nelson CP, Sanda MG. Contemporary diagnosis and management of renal angiomyolipoma. *J Urol*. 2002;168(4 Pt 1):1315-25.
- Zhang JQ, Fielding JR, Zou KH. Etiology of spontaneous perirenal hemorrhage: a meta-analysis. *J Urol*. 2002;167(4):1593-6.
- Earthman WJ, Mazer MJ, Winfield AC. Angiomyolipomas in tuberous sclerosis: subselective embolotherapy with alcohol, with long-term follow-up study. *Radiology*. 1986;160(2):437-41.
- Svec A, Velenská Z. Renal epithelioid angiomyolipoma—a close mimic of renal cell carcinoma. Report of a case and review of the literature. *Pathol Res Pract*. 2005;200(11-12):851-6.
- Garg PK, Jain BK, Kumar A, Bhatt S, Vibhav V. Fat poor angiomyolipoma with lymphadenopathy: diagnostic dilemma. *Urol Ann*. 2012;4(2):126-9.
- Minja EJ, Pellerin M, Saviano N, Chamberlain RS. Retroperitoneal extrarenal angiomyolipomas: an evidence-based approach to a rare clinical entity. *Case Rep Nephrol*. 2012;2012:374107.
- Fejes Z, Santa F, Jenei A, Király IE, Varga L, Kuthi L. Angiomyolipoma of the kidney—clinicopathological analysis of 52 cases. *Pathol Oncol Res*. 2023;28:1610831.
- Song S, Park BK, Park JJ. New radiologic classification of renal angiomyolipomas. *Eur J Radiol*. 2016;85(10):1835-42.
- Aydin H, Magi-Galluzzi C, Lane BR, Sercia L, Lopez JJ, Rini BI, et al. Renal angiomyolipoma: clinicopathologic study of 194 cases with emphasis on

- the epithelioid histology and tuberous sclerosis association. *Am J Surg Pathol.* 2009;33(2):289-97.
13. Gaikwad AB, Madathil MB, Kothari AS. Giant renal angiomyolipoma with fatal hemorrhage due to a large pseudoaneurysm. *J Clin Ultrasound.* 2008;36(3):174-6.
14. Singla A, Arudra SKC, Bharti N. Giant sporadic renal angiomyolipoma treated with nephron-sparing surgery. *Urology.* 2009;74(2):294-5.
15. Bishay VL, Crino PB, Wein AJ, Malkowicz SB, Trerotola SO, Soulen MC, et al. Embolization of giant renal angiomyolipomas: technique and results. *J Vasc Interv Radiol.* 2010;21(1):67-72.

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