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Angioleiomyoma of the Kidney: Differentiating the Rare Benign Histology from the More Sinister Malignant Tumors of Kidney- A Case Report

Böbreğin Anjioleiomyomu: Nadir Görülen İyi Huylu Histolojinin Böbreğin Daha Tehlikeli Kötü Huylu Tümörlerinden Ayırt Edilmesi - Bir Olgu Sunumu

Parth Shah[1], Siddharth Yadav[1], Harshdeep Singh[1], Aravind T K[1], Sneha Sangwan[2]

[1] Department of Urology VMMC & Safdarjung Hospital, New Delhi, India
[2] ICMR-National Institute of Pathology, VMMC & Safdarjung Hospital, New Delhi, India

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Corresponding Author: Siddharth Yadav / VMMC & Safdarjung Hospital, Department of Urology, New Delhi, India / drsidyadav@gmail.com
ORCID ID: 0000-0002-5632-8967

Abstract

Renal angioleiomyoma is an exceptionally rare benign mesenchymal tumor, with fewer than five cases documented in the literature. Histologically, it is composed of proliferating smooth muscle cells intersected by branching, slit-like vascular channels and typically lacks significant epithelial components. Despite its benign nature, its morphological overlap with certain renal malignancies presents a diagnostic challenge. Notably, renal cell carcinoma with angioleiomyoma-like stroma, a recently recognized variant of renal cell carcinoma, demonstrates a histologically and immunohistochemically similar stromal component. Although it generally exhibits a more indolent clinical course, there have been reports of lymph node involvement, underscoring its malignant potential. In this report, we present a case of renal angioleiomyoma and discuss the importance of distinguishing it from its malignant counterpart. Histopathological evaluation, supplemented by immunohistochemistry, plays a vital role in achieving a definitive diagnosis. Accurate differentiation is crucial to avoid overtreatment and to ensure appropriate clinical management. The overall prognosis is excellent, reflecting the inherent benign nature of renal angioleiomyoma. No routine long-term surveillance is required once a diagnosis is accurately confirmed. However, pathological overlap with renal cell carcinoma with angioleiomyoma-like stroma may necessitate selective follow-up in cases of diagnostic ambiguity. Improved awareness of renal angioleiomyoma helps ensure correct diagnosis and prevents confusion with malignant or other benign mesenchymal tumors that involve the kidney.

Keywords: angioleiomyoma, renal cell carcinoma with angioleiomyoma like stroma, immunohistochemistry

Özet

Renal anjiyoleyomiyom, literatürde beşten az vakası belgelenmiş, son derece nadir görülen iyi huylu bir mezenkimal tümördür. Histolojik olarak, dallanan, yarık benzeri vasküler kanallarla kesişen prolifere düz kas hücrelerinden oluşur ve tipik olarak önemli epitelyal bileşenlerden yoksundur. İyi huylu yapısına rağmen, bazı renal malignitelerle morfolojik örtüşmesi tanısal zorluk yaratmaktadır. Özellikle, renal hücreli karsinomun yeni tanımlanmış bir varyantı olan anjiyoleyomiyom benzeri stromalı renal hücreli karsinom, histolojik ve immünohistokimyasal olarak benzer bir stromal bileşen göstermektedir. Genellikle daha yavaş bir klinik seyir göstermesine rağmen, malign potansiyelini vurgulayan lenf nodu tutulumu bildirilmiştir. Bu raporda, bir renal anjiyoleyomiyom olgusu sunulmakta ve onu malign muadilinden ayırmanın önemini tartışmaktayız. İmmünohistokimya ile desteklenen histopatolojik değerlendirme, kesin tanıya ulaşmada hayati bir rol oynamaktadır. Doğru ayrım, aşırı tedaviden kaçınmak ve uygun klinik yönetimi sağlamak için çok önemlidir. Genel prognoz, renal anjiyoleyomiyomun doğal iyi huylu yapısını yansıtacak şekilde mükemmeldir. Tanı doğru bir şekilde doğrulandıktan sonra rutin uzun süreli gözetim gerekmez. Ancak, anjiyoleyomiyom benzeri stromalı renal hücreli karsinom ile patolojik örtüşme, tanı belirsizliği olan vakalarda seçici takip gerektirebilir. Renal anjiyoleyomiyom konusunda farkındalığın artırılması, doğru tanıyı sağlamaya yardımcı olur ve böbreği tutan malign veya diğer iyi huylu mezenkimal tümörlerle karışıklığı önler.

Anahtar kelimeler: anjiyoleyomiyom, anjiyoleyomiyom benzeri stromalı renal hücreli karsinom, immünohistokimya

ORCID ID:
P. Shah 0009-0008-2013-2744
H. Singh 0009-0009-8227-856X
A. TK 0000-0002-6501-5014
S. Sangwan 0009-0000-4510-5082

Introduction

Approximately 20% of renal masses clinically suspected to be malignant are ultimately identified as benign on final histopathological examination following surgical resection [1]. Angioleiomyomas are benign smooth muscle tumors that most commonly arise in the skin and subcutaneous tissue, while their occurrence in visceral organs, including the kidney, is exceedingly rare [2]. Despite their rarity, angioleiomyomas represent the most common benign mesenchymal tumors of the kidney. To date, fewer than five cases of renal angioleiomyoma have been reported in the literature. This highlights the rarity and diagnostic challenge posed by this entity. Herein, we report an unusual case of renal angioleiomyoma in a young female. We emphasize the importance of distinguishing it from its malignant mimics, particularly renal cell carcinoma with angioleiomyoma-like stroma (RCC-AMLSt), as well as other morphologically similar renal tumors. Accurate diagnosis is crucial to prevent unnecessary aggressive treatment and ensure effective patient management.

Case

A 34-year-old female presented with a two-year history of dull, aching pain localized to the right flank. A contrast-enhanced computed tomography (CECT) scan of the whole abdomen revealed a 5.2x4.6x3.8 cm heterogeneously enhancing, predominantly exophytic mass arising from the upper pole of the right kidney, with angiography suggestive of the presence of two renal arteries and two renal veins on the right side **(Figure 1a, 1b)**. Given the radiologically confirmed solid enhancing renal mass, a preliminary diagnosis of renal cell carcinoma (RCC) was made, and the patient underwent robot-assisted laparoscopic right partial nephrectomy using the Da Vinci Xi robotic system. Intraoperatively, a 6x5 cm well-defined, irregularly lobulated, exophytic tumor measuring approximately was identified on the posterior aspect of the upper pole of the right kidney **(Figure 1c, 1d)**. The lesion demonstrated preserved fat planes with adjacent structures and had extensive peritumoral neovascularization. The tumor was excised with an adequate surgical margin, followed by renorrhaphy. The warm ischemia time was 25 minutes, and the total operative duration was approximately 3 hours. Estimated intraoperative blood loss was minimal. The postoperative course was uneventful, and the patient was discharged on postoperative day 4. On the cut section, the tumor appeared tan to light brown, homogenous, and lacked areas of necrosis or cystic degeneration. Histopathological evaluation of the tumor revealed oval to spindle-shaped cells arranged in interlacing fascicles and bundles, interspersed with branching, slit-like blood vessels **(Figure 2a, 2b)**. Tumor cells exhibited mild to moderate nuclear atypia, with no evidence of mitotic activity or necrosis **(Figure 2c)**. The capsular margin was involved; however, the surgical resection margin was free of tumor infiltration. Immunohistochemically, the tumor cells were positive for Smooth Muscle Actin (SMA) and negative for HMB-45, STAT6, and CD34. These findings confirmed the diagnosis of renal angioleiomyoma **(Figure 2d)**. Postoperative follow-up consisted of clinical evaluation at 1 month, 6 months, and at 1 year, as a part of individualized institutional protocol. No new symptoms or complications were noted during follow-up. Ultrasound performed at 1 year showed no evidence of recurrence.

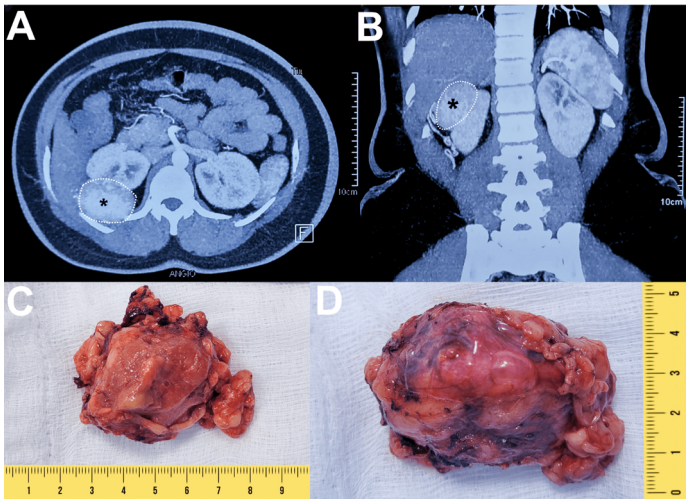

Figure 1. A,B,C,D: Cross sectional imaging & specimen
1A & 1B - Axial and Coronal sections of CECT abdomen depicting an exophytic right renal mass arising from the upper pole extending to the Perinephric fat (tumor demarcation engraved by dotted lines and epicenter marked by * mark)
1C & 1D - Resected partial nephrectomy renal mass specimen with 2-dimensional scaling measuring approximately 5x4 cm

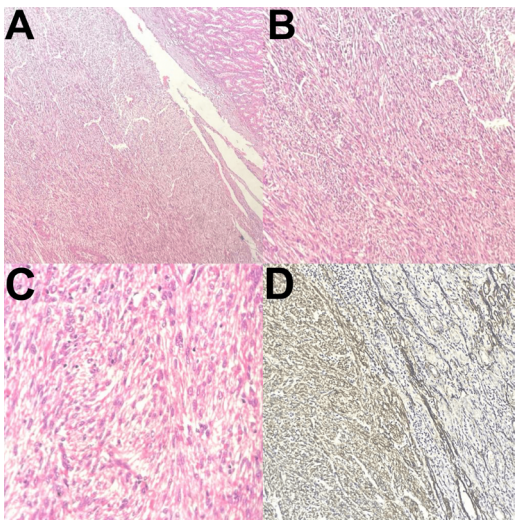

Figure 2. A,B,C,D: Tumor histology
2A - The tumor histology depicting clear demarcation from the adjacent unremarkable normal renal parenchyma, without any infiltrative edges [10x Magnification, Hematoxylin and Eosin (H&E)]
2B - The tumor is composed of interlacing fascicles of spindle cells arranged in a whorled pattern, with numerous thin, branching, slit-like blood vessels dispersed throughout the stroma (10x Magnification, H&E)
2C - Higher magnification demonstrates bland spindle cell morphology with elongated nuclei and eosinophilic cytoplasm. No atypical mitotic figures, nuclear pleomorphism, or necrosis are identified in 40X magnification
2D - Immunohistochemistry for Smooth Muscle Actin demonstrating diffuse, strong cytoplasmic positivity in tumor cells, supporting smooth muscle differentiation, with adjacent renal tubules serving as internal negative controls (10x magnification)

Discussion

Cross-sectional imaging is an indispensable part of the evaluation of renal masses. However, it often lacks sufficient specificity to reliably distinguish benign from malignant lesions, except in cases of simple renal cysts and lipid-rich angiomyolipoma (AML) [3]. Consequently, the management of renal masses is frequently guided by the presumed risk of RCC. The histopathological confirmation, therefore, remains essential for establishing an accurate diagnosis and guiding appropriate treatment planning.

Angioleiomyoma is a purely mesenchymal tumor composed of well-differentiated smooth muscle cells and thick-walled blood vessels of varying calibers. It most commonly occurs in the skin as subcutaneous nodules, whereas its development within the renal parenchyma is exceedingly rare [2]. Renal angioleiomyoma must be carefully distinguished from its more aggressive and relatively more common counterpart, RCC-AMLSt. Other important histological differential diagnoses include conventional RCC, AML, renal leiomyoma, mixed epithelial and stromal tumor, solitary fibrous tumor, and smooth muscle–predominant or adenoma-like renal tumors. RCC-AMLSt is a recently recognized entity within the spectrum of renal neoplasms. Histologically, it is characterized by a biphasic pattern, comprising areas with mixed epithelial proliferation intermingled with a prominent smooth muscle-rich stromal component. The epithelial component consists of elongated neoplastic cells arranged in sweeping fascicles within a richly vascular stroma, which often exhibits a concentric perivascular arrangement of tumor cells. Immunohistochemically, it is typically positive for carbonic anhydrase IX (CAIX), high molecular weight cytokeratin, cytokeratin 7 (CK7), and CD10, while negative for alpha-methylacyl-CoA racemase (AMACR). Notably, these tumors lack 3p25 deletions and von Hippel–Lindau (VHL) gene alterations, which are commonly seen in clear cell RCC [4-7].

While both entities share overlapping stromal features and immunohistochemical profiles, the presence of epithelial proliferation is the key distinguishing feature favoring a diagnosis of RCC-AMLSt over a benign angioleiomyoma. Given the potential for diagnostic confusion, careful correlation of histological patterns with immunohistochemical and, where available, molecular findings is crucial to avoid misdiagnosis and overtreatment. Although RCC-AMLSt is considered less aggressive than conventional RCC, it still carries malignant potential, making the distinction from angioleiomyoma clinically essential.

In a study by Williamson et al. [8], 11 patients diagnosed with RCC-AMLSt were followed for a period ranging from 26 to 58 months. All patients remained alive without evidence of residual, recurrent, or metastatic disease. Notably, one patient presented with a synchronous tumor in the contralateral kidney, which was successfully resected four months later. Conversely, Verkarre et al. reported a clinicopathological analysis of 17 patients, confirming the neoplasm's malignant potential through cases with lymph node involvement [9]. The contrasting findings underscore the diagnostic and prognostic importance of differentiating benign angioleiomyoma from malignant RCC-AMLSt. These divergent outcomes highlight the biological variability of RCC-AMLSt and the necessity for long-term follow-up in affected patients.

Among other renal tumors with overlapping histologic features, AML is an important differential diagnosis. This tumor is readily distinguishable on imaging due to the presence of macroscopic fat, which appears as areas of low attenuation ranging from -15 to -20 Hounsfield units (HU) on unenhanced CT scans. Histologically, they contain a variable mixture of smooth muscle, fat, and abnormal blood vessels. Immunohistochemical positivity for melanocytic markers such as HMB-45 and Melan-A further supports the diagnosis of AML [10]. This distinction is diagnostically significant because, unlike angioleiomyoma, they may occasionally be associated with tuberous sclerosis complex and can demonstrate aggressive growth or spontaneous hemorrhage.

Other rare mesenchymal tumors of the kidney, such as leiomyoma and smooth muscle–predominant or adenoma-like renal tumors, can also resemble angioleiomyoma. However, these entities generally lack the characteristic thick-walled vessels seen in angioleiomyoma, aiding in their distinction on histological grounds [11,12].

From a clinical standpoint, the differentiation between RCC, AML, and angioleiomyoma profoundly impacts management strategies and patient outcomes. RCC, being malignant, requires surgical resection, either partial or radical nephrectomy, followed by close surveillance for recurrence or metastasis. In contrast, AMLs, depending on their size and symptomatology, can often be managed conservatively. Small, asymptomatic AMLs are typically monitored with serial imaging, while larger or symptomatic lesions may be treated with selective arterial embolization or nephron-sparing surgery to prevent hemorrhage [13]. Renal angioleiomyoma, however, being entirely benign, does not necessitate aggressive intervention if correctly diagnosed preoperatively. Complete local excision is usually curative, and recurrence is exceptionally rare. Hence, an accurate preoperative and histopathological diagnosis is paramount to avoid unnecessary radical nephrectomy and to preserve renal function. This distinction underscores the importance of a multidisciplinary approach involving radiologists, urologists, and pathologists for accurate diagnosis and individualized patient management.

No standardized follow-up protocol currently exists for renal angioleiomyoma. Owing to its benign biological behavior, routine long-term surveillance is generally considered unnecessary once the diagnosis is confirmed. However, in cases where any diagnostic uncertainty persists, even after final histopathological evaluation, periodic follow-up imaging may be prudent to ensure timely detection of a potentially misclassified or coexisting malignant component.

Despite its indolent nature, renal angioleiomyoma represents an important histopathological diagnosis that poses a significant diagnostic challenge due to its extreme rarity and morphological similarity to RCC-AMLSt. To date, fewer than five cases have been documented in the literature. [14-15]. While numerous reports of RCC-AMLSt exist [4-9], meaningful outcome comparisons for benign angioleiomyoma remain limited because of the scarcity of reported cases and the lack of long-term follow-up data in published reports. The present case aims to contribute to the existing literature by providing insights into postoperative outcomes, follow-up considerations, and the clinical importance of distinguishing renal angioleiomyoma from RCC-AMLSt. The tumor's overlapping similarity to RCC-AMLSt, coupled with limited familiarity among clinicians and pathologists, underscores the need for careful histopathological and immunohistochemical evaluation. Accurate distinction between these entities is critical to guide appropriate clinical management and to avoid both misclassification and unnecessary aggressive treatment.

Renal angioleiomyomas are extremely rare benign tumors that closely mimic RCC-AMLSt, a neoplasm with malignant potential and comparatively poorer prognosis. Increased awareness of renal angioleiomyoma is essential for ensuring accurate diagnosis and for distinguishing it from both malignant and other benign mesenchymal renal tumors. Timely recognition can prevent overtreatment and support more tailored, conservative management when appropriate.

Ethics Committee Approval: Not Applicable.
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References

[1] Vijay V, Vokshi FH, Smigelski M, Nagpal S, Huang WC. Incidence of benign renal masses in a contemporary cohort of patients receiving partial nephrectomy for presumed renal cell carcinoma. Clin Genitourin Cancer 2023;21(3):e114-8. https://doi.org/10.1016/j.clgc.2022.11.006

[2] Bernard M, Le Nail LR, de Pinieux G, Samargandi R. Angioleiomyoma: An Update with a 142-Case Series. Life (Basel) 2024;14(3):338. https://doi.org/10.3390/life14030338

[3] Canvasser NE, Kay FU, Xi Y, Pinho DF, Costa D, de Leon AD, et al. Diagnostic accuracy of multiparametric magnetic resonance imaging to identify clear cell renal cell carcinoma in cT1a renal masses. J Urol 2017;198(4):780-6. https://doi.org/10.1016/j.juro.2017.04.089

[4] Kuhn E, De Anda J, Manoni S, Netto G, Rosai J. Renal cell carcinoma associated with prominent angioleiomyoma-like proliferation: report of 5 cases and review of the literature. Am J Surg Pathol 2006;30:1372-81. https://doi.org/10.1097/01.pas.0000213277.45715.82

[5] Yeh YA, Constantinescu M, Chaudoir C, Tanner A, Serkin F, Yu X et al. Renal cell carcinoma with leiomyomatous stroma: a review of an emerging entity distinct from clear cell conventional renal cell carcinoma. Am J Clin Exp Urol 2019;7(5):321-6. https://pubmed.ncbi.nlm.nih.gov/31763363/

[6] Rajaian S, Krishnamurthy K, Murugasen L, Narasimhachar SC. Clear renal cell carcinoma with angioleiomyomatous stroma in a noninherited renal cancer syndrome: Is it a different entity? Urol Sci 2020;31(4):194-6. https://doi.org/10.4103/UROS.UROS_27_20

[7] Shah RB, Stohr BA, Tu ZJ, Gao Y, Przybycin CG, Nguyen J et al. Renal cell carcinoma with leiomyomatous stroma harbor somatic mutations of TSC1, TSC2, MTOR, and/or ELOC (TCEB1): Clinicopathologic and molecular characterization of 18 sporadic tumors supports a distinct entity. Am J Surg Pathol 2021;45(1):45-58. https://doi.org/10.1097/PAS.0000000000001422

[8] Williamson SR, Cheng L, Eble JN, True LD, Gupta NS, Wang M, et al. Renal cell carcinoma with angioleiomyoma-like stroma: clinicopathological, immunohistochemical, and molecular features supporting classification as a distinct entity. Mod Pathol 2015;28:279-94. https://doi.org/10.1038/modpathol.2014.105

[9] Verkarre V, Mensah A, Leroy X et al. A clinico-pathologic study of 17 patients with renal cell carcinoma associated with leiomyomatous stroma identifies a strong association with tuberous sclerosis. Lab Invest 2015;95:266A.

[10] Saleem A, Narala S, Raghavan SS. Immunohistochemistry in melanocytic lesions: Updates with a practical review for pathologists. Semin Diagn Pathol 2022;39(4):239-47. https://doi.org/10.1053/j.semdp.2021.12.003

[11] Papke DJ. Mesenchymal neoplasms of the kidney and perinephric soft tissue. Surg Pathol Clin 2025;18(1):209-27. https://doi.org/10.1016/j.path.2024.08.006

[12] Panda M, Vangapandu S, Mandal S, Ayyanar P. Smooth muscle and adenoma-like renal tumor (SMART)- A Rare biphasic renal tumor with a review. Int J Surg Pathol 2025;33(5):1240-5. https://doi.org/10.1177/10668969241300962

[13] Fernández-Pello S, Hora M, Kuusk T, Tahbaz R, Dabestani S, Abu-Ghanem Y, et al. Management of sporadic renal angiomyolipomas: a systematic review of available evidence to guide recommendations from the european association of urology renal cell carcinoma guidelines panel. Eur Urol Oncol 2020;3(1):57-72. https://doi.org/10.1016/j.euo.2019.04.005

[14] Huang Y, Xiong Z. Angioleiomyoma of the kidney parenchyma. Indian J Pathol Microbiol. 2013; 56(3):318-9. https://doi.org/ 10.4103/0377-4929.120412

[15] Iakirevich AL. Angioleiomyoma of the kidney. Urol Nefrol (Mosk). 1967;32(3):47-8. https://pubmed.ncbi.nlm.nih.gov/5612369