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Case Report

Fat-Poor Extra-Renal Retroperitoneal Angiomyolipoma: A Case Report and Review of Diagnostic Pitfalls

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Abstract: ***Background*:** Angiomyolipoma (AML) is a benign mesenchymal neoplasm belonging to the family of perivascular epithelioid cell tumors (PEComas). While most AMLs arise in the kidney, extra-renal occurrences are exceptionally rare. Fat-poor variants may pose significant diagnostic challenges because of their resemblance to malignant mesenchymal neoplasms. ***Case Presentation*:** We report the case of a 58-year-old woman presenting with progressive abdominal enlargement. Imaging revealed a giant retroperitoneal mass measuring approximately 40 cm, radiologically suggestive of sarcoma. Preoperative biopsies yielded discordant diagnoses, including dedifferentiated liposarcoma and spindle-cell neoplasm with smooth muscle differentiation. Complete surgical excision was performed. Histological examination demonstrated a predominantly smooth muscle proliferation lacking atypia, necrosis, or increased mitotic activity. Extensive sampling identified focal mature adipose tissue and dysmorphic blood vessels. Immunohistochemistry showed positivity for smooth muscle actin, desmin, HMB-45, and Melan-A, confirming the diagnosis of fat-poor extra-renal retroperitoneal angiomyolipoma. ***Conclusion*:** Extra-renal angiomyolipoma is an uncommon benign neoplasm that may mimic retroperitoneal sarcoma clinically, radiologically, and histologically. Adequate sampling and appropriate immunohistochemical investigations are essential for accurate diagnosis and prevention of overtreatment.

Keywords: Angiomyolipoma, PEComa, Retroperitoneum, Fat-Poor Angiomyolipoma, HMB-45, Melan-A.

INTRODUCTION

Angiomyolipoma (AML) is a mesenchymal tumor composed of variable proportions of mature adipose tissue, smooth muscle cells, and thick-walled dysmorphic blood vessels [1]. It belongs to the family of perivascular epithelioid cell tumors (PEComas), a group of neoplasms characterized by dual melanocytic and smooth muscle differentiation [2]. Although AML most commonly arises in the kidney, extra-renal angiomyolipomas represent an exceptionally uncommon subset of PEComas [3].

Retroperitoneal extra-renal AMLs are particularly rare and often constitute a diagnostic challenge. Their clinical presentation is usually nonspecific and largely related to mass effect [4]. Radiologically, lesions with minimal adipose tissue may mimic aggressive soft tissue sarcomas, especially dedifferentiated liposarcoma. Histologically, smooth muscle-predominant tumors may be confused with leiomyoma, leiomyosarcoma, or other spindle-cell neoplasms [5].

We report a giant retroperitoneal extra-renal angiomyolipoma with predominant smooth muscle differentiation in a 58-year-old woman, highlighting the importance of extensive tissue sampling and

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immunohistochemical analysis in establishing the diagnosis.

CASE PRESENTATION

A 58-year-old woman with no significant medical history was referred for evaluation of progressive abdominal enlargement evolving over several months. She denied urinary symptoms, digestive complaints, weight loss, fever, or constitutional manifestations.

Physical examination revealed a large palpable abdominal mass occupying the hypogastric region. The mass was firm, non-tender, and mobile relative to superficial planes. No signs of vascular compression or neurological involvement were observed.

Contrast-enhanced abdominal computed tomography demonstrated a huge retroperitoneal tissue mass measuring approximately 40 cm in maximum diameter. The lesion displayed heterogeneous enhancement and irregular contours while surrounding the right kidney (Figure 1). Radiological findings strongly suggested retroperitoneal sarcoma.

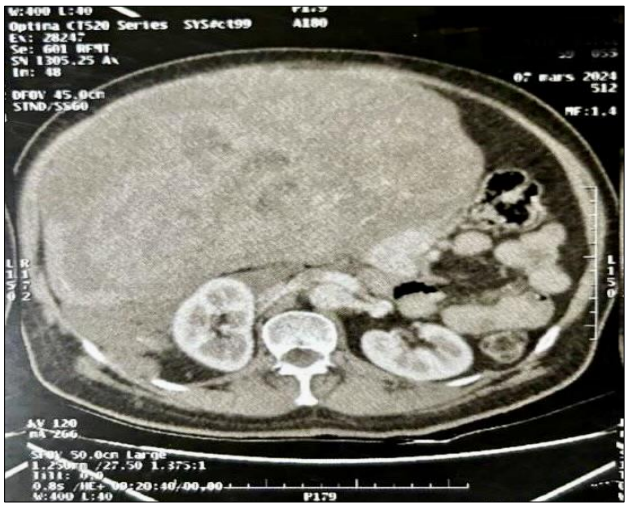

Figure 1: CT scan showing a right renal cortical mass with associated intraperitoneal tumor involvement.

Image-guided biopsies performed at another institution resulted in discordant interpretations, including dedifferentiated liposarcoma and spindle-cell proliferation with smooth muscle differentiation. Following multidisciplinary discussion, complete surgical resection was recommended.

Pathological Findings

Gross examination revealed a resection specimen measuring 42 × 21 × 17 cm associated with a kidney measuring 10 × 6.5 × 4 cm. The tumor was well-circumscribed and finely encapsulated. On sectioning, it appeared homogeneous, firm, and whitish, with focal cystic degeneration. The lesion surrounded the renal parenchyma without evidence of direct invasion (Figure 2).

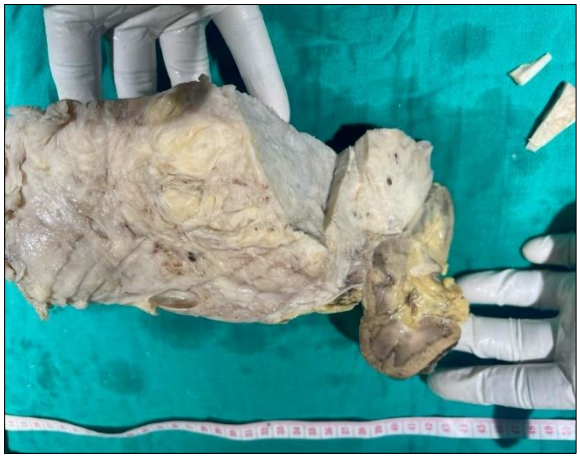

Figure 2: Gross appearance of the surgical specimen showing a firm, whitish lesion. The arrow indicates the residual kidney

Microscopically, extensive sampling showed that approximately 98% of the tumor consisted of intersecting fascicles of spindle-shaped smooth muscle cells. Tumor cells exhibited elongated nuclei with bland chromatin and abundant eosinophilic cytoplasm. No significant cytological atypia, coagulative necrosis, or increased mitotic activity was identified.

Careful examination of numerous additional sections revealed focal areas containing mature adipocytes admixed with thick-walled dysmorphic blood vessels. These findings suggested a smooth muscle-predominant angiomyolipoma (Figure 3).

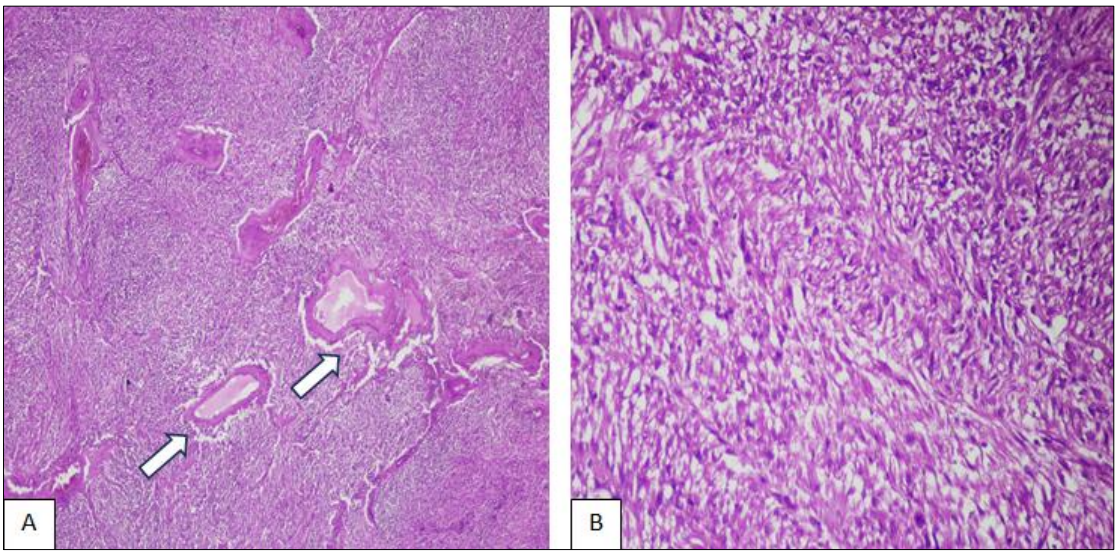

Figure 3: Histopathological findings (H&E staining). (A) Low-power view showing a spindle cell tumor composed predominantly of intersecting fascicles of smooth muscle cells admixed with thick-walled dysmorphic blood vessels (Arrow). (B) High-power view demonstrating bland spindle cells with elongated nuclei and abundant eosinophilic cytoplasm, with focal mature adipocytes

Immunohistochemical studies demonstrated diffuse expression of smooth muscle actin (SMA) and desmin. Tumor cells also exhibited positivity for HMB-45 and Melan-A, confirming melanocytic differentiation.

Based on the morphological and immunophenotypic findings, a diagnosis of extra-renal retroperitoneal angiomyolipoma with predominant smooth muscle differentiation was established (Figure 4).

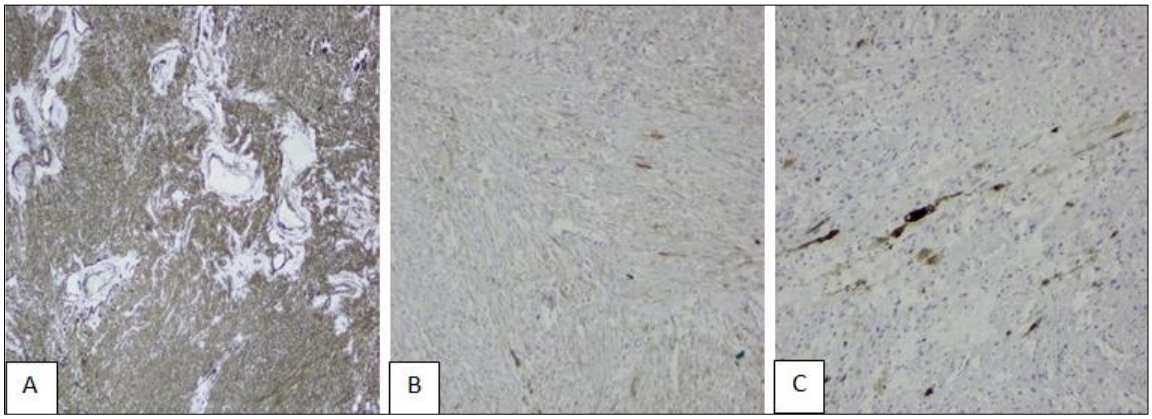

Figure 4: Immunohistochemical findings. (A) Diffuse SMA positivity in the spindle cell component, (B) Melan-A positivity, (C) HMB-45 positivity

DISCUSSION

Angiomyolipoma (AML) is the most common mesenchymal neoplasm of the kidney and belongs to the family of perivascular epithelioid cell tumors (PEComas) as defined in the WHO 2022 classification of urinary and male genital tract tumors [1, 2]. Histologically, conventional AML is characterized by a triphasic admixture of mature adipose tissue, smooth muscle cells, and dysmorphic thick-walled blood vessels. Although renal AMLs are well recognized, extra-renal angiomyolipomas (ERAMLs) remain exceptionally rare, particularly in the retroperitoneum [6–8].

The retroperitoneum represents one of the rarest locations for ERAML. Literature data suggest that fewer

than 40 well-documented primary retroperitoneal cases have been reported [7, 8]. ERAMLs predominantly affect middle-aged women and may reach very large dimensions before diagnosis due to the expansile capacity of the retroperitoneal space. Giant lesions exceeding 20 cm have been reported, although tumors of extreme size remain exceptional [6, 7].

Clinically, ERAMLs usually present with nonspecific symptoms such as abdominal pain, distension, palpable mass, or compression-related symptoms. In our patient, progressive abdominal enlargement was the only clinical manifestation despite the massive size of the lesion (42 cm), which is among the largest reported in recent literature [9, 10]. Similar giant retroperitoneal AMLs have been described, often diagnosed incidentally or after prolonged evolution due to their indolent growth pattern [6-9].

Radiological diagnosis remains particularly challenging in fat-poor AMLs. While macroscopic fat on CT or MRI is highly suggestive of AML, fat-poor or smooth muscle–predominant variants lack this hallmark feature and frequently mimic retroperitoneal sarcomas, particularly dedifferentiated liposarcoma [5-12]. In the present case, imaging revealed a large heterogeneous retroperitoneal mass surrounding the kidney, strongly suggestive of sarcoma, illustrating a well-known diagnostic pitfall in this entity [5-12].

Preoperative core needle biopsies may also be misleading. In smooth muscle–predominant AMLs, the classical triphasic architecture is often absent in limited samples, leading to misinterpretation as spindle-cell neoplasms or sarcomas [2-5]. In our case, biopsies yielded discordant diagnoses, including dedifferentiated liposarcoma and spindle-cell proliferation. This highlights the limitation of sampling in large heterogeneous retroperitoneal tumors and the need for caution in preoperative interpretation [5-11].

Histologically, the main differential diagnoses include leiomyoma, leiomyosarcoma, dedifferentiated liposarcoma, solitary fibrous tumor, inflammatory myofibroblastic tumor, and other spindle-cell mesenchymal neoplasms [3-5]. The absence of significant cytologic atypia, atypical mitoses, coagulative necrosis, and infiltrative growth supported a benign lesion in the present case; however, morphology alone was insufficient due to the overwhelming smooth muscle predominance [3-11].

Extensive tumor sampling is therefore crucial to identify focal diagnostic components such as mature adipose tissue and dysmorphic blood vessels. In smooth muscle–predominant AML, these elements may be extremely focal and easily missed if sampling is limited [4-12].

Immunohistochemistry remains the cornerstone of diagnosis. The co-expression of melanocytic markers (HMB45, Melan-A) and smooth muscle markers (SMA, desmin) is highly characteristic of PEComas and allows definitive diagnosis even in fat-poor lesions [1-3]. In our patient, diffuse positivity for HMB45, Melan-A, SMA, and desmin confirmed the diagnosis despite the misleading morphological appearance [3-11].

An additional important point in this case is the extra-renal origin of the tumor. Although the lesion surrounded the kidney, careful macroscopic examination showed no renal parenchymal invasion, supporting a primary retroperitoneal origin rather than an exophytic renal AML [6-9]. Distinguishing these entities is essential because large exophytic renal AMLs may also mimic retroperitoneal sarcomas both radiologically and pathologically [6-12].

Complete surgical excision remains the treatment of choice for symptomatic or diagnostically uncertain ERAMLs. Prognosis is generally excellent after complete resection, with very low recurrence rates [3-7]. However, rare cases of recurrence or aggressive behavior have been reported, justifying long-term follow-up in selected cases, particularly large or histologically atypical tumors [2-7].

A review of recently reported cases is summarized in Table 1, confirming the rarity of this entity and the predominance of middle-aged women presenting with large retroperitoneal masses often initially misdiagnosed as sarcomas [9–11].

Our case illustrates several important diagnostic lessons. First, retroperitoneal AML should be included in the differential diagnosis of giant retroperitoneal masses, even in the apparent absence of fat on imaging. Second, extensive pathological sampling is mandatory in smooth muscle-predominant tumors. Finally, immunohistochemistry remains indispensable for distinguishing fat-poor AML from malignant spindle-cell neoplasms and preventing overtreatment [1-11].

CONCLUSION

Extra-renal retroperitoneal angiomyolipoma is a rare benign PEComa that may clinically and radiologically mimic retroperitoneal sarcoma, particularly in fat-poor variants. Histological examination with extensive tissue sampling and immunohistochemical characterization remains essential for diagnosis. Awareness of this uncommon entity may prevent misdiagnosis and unnecessary aggressive therapeutic strategies.

Declarations

Ethics Approval and Consent to Participate: Not applicable.

Consent for Publication: Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Competing Interests: The authors declare no competing interests.

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Author Contributions: All authors contributed to the conception, pathological evaluation, manuscript preparation, and final approval of the submitted version.

Data Availability: All data generated or analyzed during this study are included in this published article.

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