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Case Report

Challenges in Diagnosing Rare Retroperitoneal Tumors: A Case Report of Extrarenal Giant Angiomyolipoma

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Abstract
Introduction: Extrarenal angiomyolipomas are extremely rare tumors, with only isolated reports in the literature. Their diagnosis is challenging as these lesions are often misdiagnosed as more common retroperitoneal tumors. This report presents a case of a retroperitoneal extrarenal angiomyolipoma, emphasizing its clinical, imaging, and histopathological features to facilitate accurate diagnosis and avoid errors. ***Case Presentation:*** A 58-year-old male with a history of benign prostatic hyperplasia presented with progressive abdominal distension and weight gain. Imaging studies revealed a giant retroperitoneal mass displacing the left kidney and abdominal aorta. A core needle biopsy initially suggested a well-differentiated liposarcoma T4N0M0, leading to neoadjuvant chemotherapy with doxorubicin and ifosfamide. Surgical resection of a 20 × 30 × 25-cm mass was performed. Histopathological and immunohistochemical analysis of the specimen confirmed the diagnosis of retroperitoneal extrarenal angiomyolipoma. Chemotherapy was discontinued, and the patient remains stable under follow-up. ***Discussion and Conclusion:*** Retroperitoneal extrarenal angiomyolipomas are rare, presenting significant diagnostic challenges due to their resemblance to other retroperitoneal neoplasms such as liposarcomas. This case highlights the importance of comprehensive imaging,

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histopathological examination, and immunohistochemical studies for accurate diagnosis. Increased awareness of this entity can help clinicians avoid unnecessary treatments and ensure appropriate management of similar cases.

Introduction

Angiomyolipomas (AMLs) are one of the least common renal neoplasms, constituting less than 1% of kidney tumors. However, their extrarenal occurrence is extremely rare, and they are generally diagnosed as incidentalomas during imaging for other conditions. Historically, fewer than 60 cases of extrarenal AMLs have been reported since their first description in 1982 [1, 2]. Retroperitoneal occurrence varies between 6 and 30 cases according to various authors [1, 3, 4]. Anatomically and pathologically, this tumor behaves benignly and is composed of mesenchymal cells with different components: mature fat cells, smooth muscle cells, and thick-walled vascular vessels [5]. Due to this heterogeneous composition, clinical, imaging, and pathological diagnosis can be challenging [1].

This article presents a rare case of a giant retroperitoneal AML. The unusual size of the tumor and its ambiguous presentation show the diagnostic challenges faced in clinical practice. This case contributes to the scientific literature by highlighting the importance of considering extrarenal AML in differential diagnoses for abdominal masses, particularly when imaging suggests a different neoplastic process. Additionally, it emphasizes the need for comprehensive histopathological evaluation and the use of immunohistochemistry in achieving accurate diagnoses in similar cases.

Clinical Case

A 58-year-old mestizo male, a farmer from Samaniego, Nariño (southwest Colombia), with a history of benign prostatic hyperplasia under management, without other significant personal or family history, began with a 3-month history of weight gain and abdominal distension. A medical check-up revealed non-painful hepatomegaly without other associated symptoms; the rest of the physical exam was completely normal. Urinalysis was normal, with no evidence of hematuria. An abdominal ultrasound showed a giant retroperitoneal mass in the left hypochondrium, prompting an abdominal computed tomography (CT) scan that reported a giant left tumor displacing the kidney (shown in Fig. 1a, b). A tru-cut biopsy suggested a well-differentiated liposarcoma. Based on pathology reports, chemotherapy with doxorubicin 60 mg/m^2 and ifosfamide 2,000 mg/m^2 for 5 days was administered for the treatment of a T4N0M0 liposarcoma. The tumor was scheduled for surgical resection, removing a 20 cm × 30 cm × 25 cm mass displacing the left colon and lower pole of the left kidney. The lesion had irregular borders, a soft consistency with firm areas, mostly yellowish adipose appearance with purplish zones (shown in Fig. 2).

The histopathological examination of the resected specimen showed a triphasic mesenchymal neoplasm with mature adipose tissue, prominent vessels, and myoid spindle cells. The blood vessels exhibited dismorphic architecture with notable wall thickening. No presence of lipoblasts or chicken-wire vascular pattern was shown in Figure 3a–c.

Considering the hematoxylin-eosin histopathological findings, immunohistochemistry was performed to confirm the diagnosis (Fig. 4). Tumor myoid cells located in perivascular

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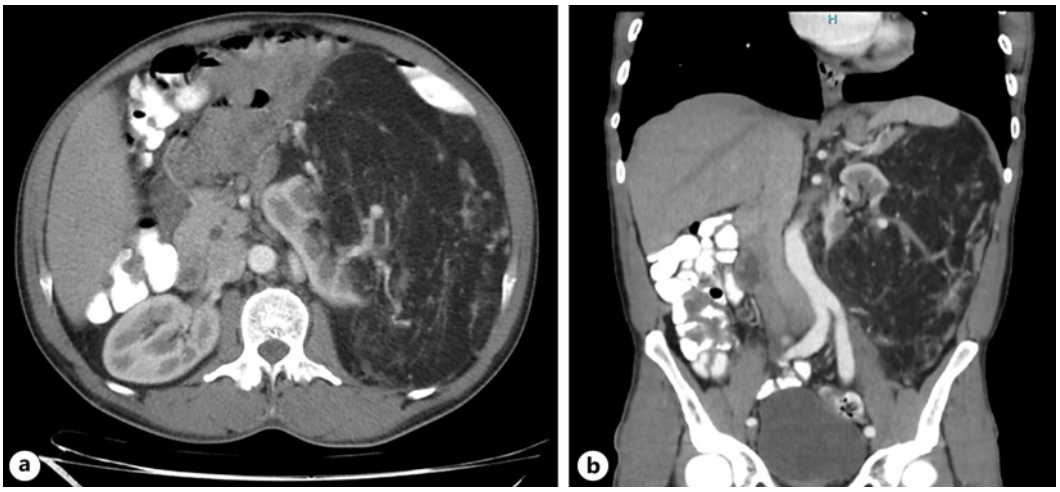

Fig. 1. a, **b** Abdominal computed tomography (CT) scan showing a retroperitoneal tumor lesion with adipose density and vascular tracts, approximately 24 × 21 × 20 cm, causing displacement of adjacent organs.

and interstitial regions showed positive immunoreactivity for HMB-45, a marker consistent with perivascular epithelioid cell tumors. With this immunophenotype, the diagnosis of retroperitoneal AML was made, leading to the discontinuation of chemotherapy, continued surveillance, and genetic testing to identify any association with other conditions such as tuberous sclerosis. The genetic test requested during follow-ups was negative, and the patient is currently stable, no post-operative complications and is now under medical follow-up.

Discussion

AML is a benign tumor composed of blood vessels, smooth muscle, and adipose tissue, belonging to the family of perivascular epithelioid cell tumors [6, 7]. These have shown association with other diseases such as tuberous sclerosis, although genetic study in this patient was negative for it [8, 9]. However, this association is usually described in renal tumors but not in extrarenal lesions, possibly due to their rarity, consistent with the findings of this case report [4]. Retroperitoneal AMLs reported in the literature have various locations, including the liver, spleen, fallopian tubes, or uterus, with the liver being the most frequent extrarenal location [10]. Epidemiologically, little information exists about retroperitoneal AML. Of the 30 cases in Venyo et al. [3] review, only 16% were in men, as in this case. Most AMLs are incidentalomas found during radiological examination; other manifestations can be more severe, such as compressive symptoms, intra-abdominal hemorrhage, and acute abdomen, yet there are no specific symptoms for these tumors [11–15]. Given the association with tuberous sclerosis in renal AMLs, a search for a history of seizures, neurological alterations, or skin tumors was also conducted [8–14].

For AML, CT scanning and CT angiography are the most used imaging methods. Wang et al. [16] showed the most common imaging characteristics of these tumors: aneurysmal dilation of intratumoral vessels, intratumoral linear vascularization, bridging veins, hematomas, and discrete intra/extrarenal fatty tumors, but none of these findings are pathognomonic [2, 16]. Some studies indicate that when this pathology is found, radiological

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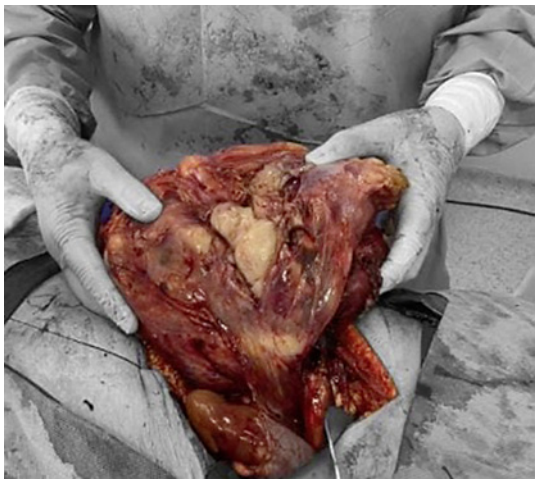

Fig. 2. Surgical resection of the retroperitoneal tumor.

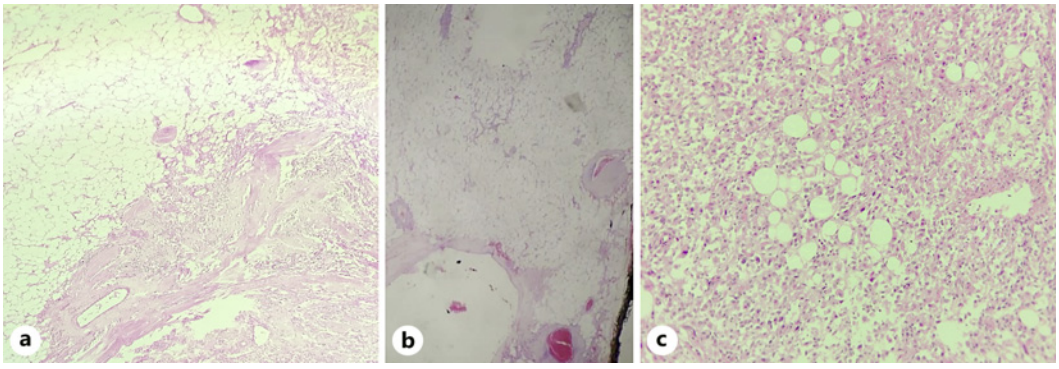

Fig. 3. a The image shows a mesenchymal neoplasm with a triphasic pattern, characterized by mature adipocytes, blood vessels with mural thickening, and myoid cells with eosinophilic cytoplasm. Hematoxylin-eosin (H&E) stain, ×4 magnification. **b** H&E, ×4 magnification. Mature adipose tissue with prominent blood vessels exhibiting wall thickening. **c** The image shows myoid cells with eosinophilic cytoplasm and fusocellular and epithelioid morphology, distributed among mature adipocytes. H&E stain, ×40 magnification.

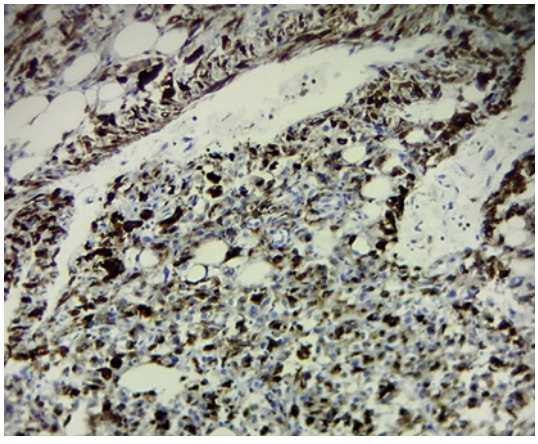

Fig. 4. Immunohistochemistry for HMB-45 shows positive staining in myoid tumor cells located in perivascular and interstitial regions, confirming the diagnosis of retroperitoneal AML. Magnification: ×40.

studies should be expanded, with a cranial CT scan being necessary due to the association of tuberous sclerosis with AML [2]. However, this recommendation was not found in the case of retroperitoneal AML.

From a histopathological standpoint, the first diagnostic possibility for an adipose-appearing retroperitoneal tumor is liposarcoma. Nevertheless, it is noteworthy that tru-cut biopsies can be only minimally representative of the lesion since sometimes they do not show the typical triphasic component of AMLs, only the adipose component, leading to diagnostic errors, as occurred in this case, where initially a liposarcoma diagnosis was made. To limit the possibility of error, broad sampling of the lesion, strict correlation with imaging findings, and consideration of an immunohistochemical panel for myoid cells, including melanocytic and smooth muscle markers like S-100, AML, and HMB-45, are suggested. While these markers are typically expressed in myoid cells, they are also expressed to a lesser extent in adipocytes, which can aid in directing the diagnosis. Moreover, well-differentiated liposarcomas exhibit lipoblasts and blood vessels in a chicken-wire pattern, distinct from the mature adipocytes and dismorphic, thick blood vessels typical of AML [17–19].

Treatment options include minimally invasive techniques such as radiofrequency ablation, cryoablation, microwave ablation, selective angioembolization, and surgery [3, 11, 20]. Reviewed literature shows that angioembolization and surgery are the most used methods, with the former being preferred in patients with active bleeding, large retroperitoneal hemorrhage, staged surgical resection plan, or large tumors with the advantage of short recovery time and preserved renal function [11]. Favorable results have been shown in some studies using hormonal or targeted therapies, mainly as neoadjuvant therapy, reducing tumor size before surgical treatment, with mTOR inhibitors and sirolimus being highlighted for this indication [9, 11].

This case presents several strengths, including the unique presentation of a giant retroperitoneal AML and a comprehensive diagnostic approach involving imaging studies and histopathological analysis, which highlights the importance of thorough evaluation in complex cases. Additionally, the management insights gained may guide clinicians in similar situations and improve understanding of treatment outcomes for extrarenal AMLs.

Conclusions

AMLs are rare tumors, with retroperitoneal occurrences being particularly uncommon. This case highlights the diagnostic challenges posed by these tumors and underscores the importance of considering extrarenal AML, especially in patients presenting with ambiguous abdominal masses that may initially suggest more common tumors or incidental findings. A thorough histopathological evaluation, supported by immunohistochemistry, is essential to avoid misdiagnosis and prevent unnecessary treatments. The case also demonstrates the need for multidisciplinary collaboration to ensure effective management and optimal patient outcomes.

Statement of Ethics

This study was approved by the Ethics Committee of Fundación Hospital San Pedro de Pasto on August 11 of 2021 (Approval Reference No. R003764-2021) for the publication of the case and complied with current regulations on human research, outlined in the Declaration of Helsinki of the World Medical Association and Resolution 8430 of 1993 of the Ministry of Health of Colombia. Written informed consent was obtained from the patient for

publication of this case report and any accompanying images. The CARE Checklist has been completed by the authors for this case report, attached as online supplementary material (for all online suppl. material, see https://doi.org/10.1159/000543128).

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

Darío Sebastián López Delgado is the main author and is responsible for initial case identification, data collection, primary manuscript drafting, and overall coordination of the study. Andres Ricaurte-Fajardo assisted in writing the manuscript, literature review, and translation to English. Carlos Stiven Mueses-Tulcán assisted in data collection, contributed to the literature review, and helped draft sections of the manuscript. Omar Eduardo Benavides Ayala provided oncological expertise, participated in patient management and treatment planning, and contributed to manuscript review and corrections. Yeison Harvey Carlosama-Rosero is the senior author, provided critical revisions to the manuscript, contributed to the study design, and ensured the accuracy and integrity of the work. Each author has read and approved the final manuscript.

Data Availability Statement

All data generated or analyzed during this study are included in this article and its online supplementary material files. Further inquiries can be directed to the corresponding author.

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