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(CASE REPORT)

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Giant Retroperitoneal Angioleiomyoma, Case report and brief literature review

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Abstract

Angioleiomyoma (ALM), or vascular leiomyoma, is a rare, benign soft tissue tumor originating from the smooth muscle of vessel walls (tunica media). Retroperitoneal ALM is an exceptionally uncommon presentation, often creating substantial preoperative diagnostic uncertainty due to non-specific imaging findings that may overlap with malignant mesenchymal tumors. We report the case of a 45-year-old woman who presented with longstanding progressive abdominal distension, dull abdominal and lower back pain, early satiety, intermittent nausea, and exertional dyspnea. Cross-sectional imaging revealed a giant, well-defined, heterogeneous retroperitoneal mass measuring 17 × 12 cm, characterized by internal myxoid changes and prominent vascularity, which displaced the adjacent viscera without clear invasion. Because malignancy could not be excluded radiologically, a multidisciplinary tumor board reviewed the case, and a CT-guided core biopsy was performed to improve preoperative characterization and operative planning.

Exploratory laparotomy revealed a pedunculated retroperitoneal tumor attached by a fibrovascular pedicle, allowing complete en bloc excision with negative margins. Histopathology showed bland spindle smooth muscle cells surrounding thick-walled vascular channels, without atypia, necrosis, or mitotic activity. At the same time, immunohistochemistry (IHC) was positive for SMA, desmin, and h-caldesmon, confirming the diagnosis of retroperitoneal ALM. The postoperative course was uneventful, and no recurrence was detected at 12 months. This case highlights the diagnostic challenge, operative considerations, and favorable outcome associated with the complete resection of this rare benign retroperitoneal tumor.

Keywords: Angioleiomyoma; Smooth muscle; Benign soft-tissue tumor, Thick-walled vascular channels; Giant; Retroperitoneal

1. Introduction

ALM is a vascular type of benign soft-tissue tumor that arises from smooth-muscle proliferation, usually localized to the lower extremities. [1] In general, ALM is predominantly seen in females, with a mean age of 55.5 years. [2] For retroperitoneal ALM, the mean age is 46.3 years with a standard deviation of 13.2 years. [3] Clinical presentation is generally nonspecific until mass effect develops with signs of abdominal or flank pain, a palpable abdominal mass, early satiety, nausea, and/or constipation. [1] Complications include ureteral obstruction, spontaneous rupture or hemorrhage, and mismanagement due to diagnostic delay. [3]

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Diagnosis often begins with CT or MRI, followed by biopsy to rule out malignancy. [1] Biopsy classically features fascicles of spindle cells, blood vessels with smooth muscle, and well circumcised borders. [4] Treatment of ALM is with complete surgical excision. [3] ALM generally has an excellent prognosis, with a small potential for local recurrence after complete resection. [5] We report this rare case of ALM to raise clinicians' awareness of similar cases and to add to the limited data pool.

2. Case Presentation

2.1. Clinical Presentation

A 45-year-old female presented to the surgical outpatient clinic with a several-year history of progressive, dull, aching abdominal and lower back discomfort. She described a gradual increase in abdominal girth over the preceding 18 months, initially attributed to weight gain. She had previously been evaluated at a primary care level, where empirical management for irritable bowel syndrome and musculoskeletal back pain had been attempted without meaningful relief. More recently, she noticed early satiety, occasional nausea, and exertional dyspnea, prompting her to seek further evaluation. She denied any rectal bleeding, changes in bowel habits, fever, or unintentional weight loss. There was no prior history of trauma or abdominal surgery. The progressive nature of her symptoms and failure of conservative management ultimately drove her referral to a tertiary surgical center for comprehensive evaluation.

2.2. Physical Examination, History, Laboratory & Imaging Findings

On physical examination, the abdomen was notably distended, with a palpable, firm, non-tender, smooth-surfaced mass occupying the left hemi-abdomen and extending toward the midline. The mass appeared relatively mobile on bimanual palpation. Peripheral lymphadenopathy and lower extremity edema were not observed. Personal and family history were unremarkable, with no history of neurofibromatosis, hereditary leiomyomatosis, or gastrointestinal stromal tumor syndromes. Laboratory investigations, including complete blood count, comprehensive metabolic panel, and tumor markers (CEA, CA-125, CA 19-9, AFP), were all within normal limits.

Contrast-enhanced CT of the abdomen and pelvis revealed a well-defined, heterogeneous, predominantly soft-tissue-density retroperitoneal mass measuring 17 × 12 cm, with internal areas of myxoid degeneration and prominent vascular channels. The mass appeared pedunculated, displacing adjacent bowel loops and the left kidney superiorly, without evidence of organ invasion. MRI further demonstrated a T2-hyperintense lesion with flow voids, consistent with intralesional vascularity. No regional lymphadenopathy or distant metastases were identified. Radiographic differential diagnosis included retroperitoneal leiomyosarcoma, solitary fibrous tumor, gastrointestinal stromal tumor with exophytic growth, schwannoma, and dedifferentiated liposarcoma.

2.3. Multidisciplinary Tumor Board Discussion

Given the radiographic inability to reliably distinguish a benign from a malignant mesenchymal process, the case was referred to the institutional multidisciplinary tumor board. Discussion centered on several critical points: the large size of the lesion, which statistically raised concern for malignancy; the apparent retroperitoneal origin and pedunculated morphology, which were atypical features complicating radiographic characterization; and the absence of definitive imaging hallmarks favoring any single diagnosis.

The board deliberated the role of preoperative tissue sampling. Given the lesion's size, vascularity, and retroperitoneal location, image-guided core needle biopsy under CT guidance was recommended as the next step to obtain histopathological material. This would allow for preliminary characterization, guide surgical planning, and inform intraoperative decision-making, particularly regarding the extent of resection and margin strategy, while minimizing the risk of unplanned violation of a potentially malignant capsule.

2.4. Management, Pathology, IHC, and Final Diagnosis

Following the tumor board recommendation, a CT-guided core needle biopsy was performed without complications. Preliminary histology revealed bland spindle cells arranged in fascicles with abundant thick-walled vascular channels, raising the possibility of a vascular smooth muscle neoplasm. Since these findings, surgical resection was planned.

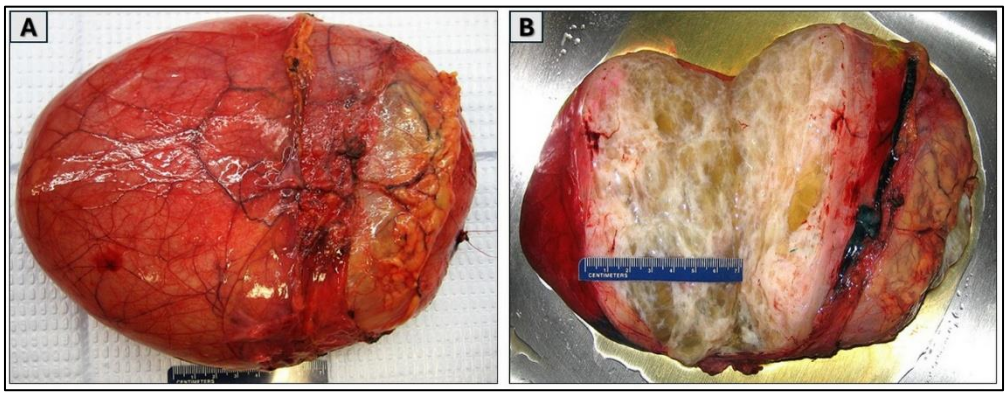
The patient underwent an elective exploratory laparotomy. Intraoperatively, the mass was found to arise from the retroperitoneum via a well-defined fibrovascular pedicle, without invasion of adjacent organs. Complete en bloc resection with ligation of the feeding pedicle was accomplished with negative margins.

Gross pathology demonstrated a well-encapsulated, rubbery tan-white mass with focal areas of hemorrhage and myxoid change. (Figure 1 A, B) Microscopy showed many well-differentiated smooth muscle cells with thick-walled, dilated blood vessels, no atypia, necrosis, or mitoses, indicating ALM. (Figure 2 A, B, C) Immunohistochemistry (IHC) showed strong, diffuse positivity for smooth muscle actin (SMA), desmin, and h-caldesmon, with negativity for S-100, CD117 (c-KIT), DOG1, and CD34. Ki-67 proliferative index was low (< 2%). No molecular studies were required. Pathological differential diagnosis included leiomyosarcoma, vascular leiomyoma variants, and angiomyolipoma. The final diagnosis was retroperitoneal angioleiomyoma (ALM), a benign vascular smooth muscle tumor.

2.5. Outcome, Follow-Up, and Surveillance

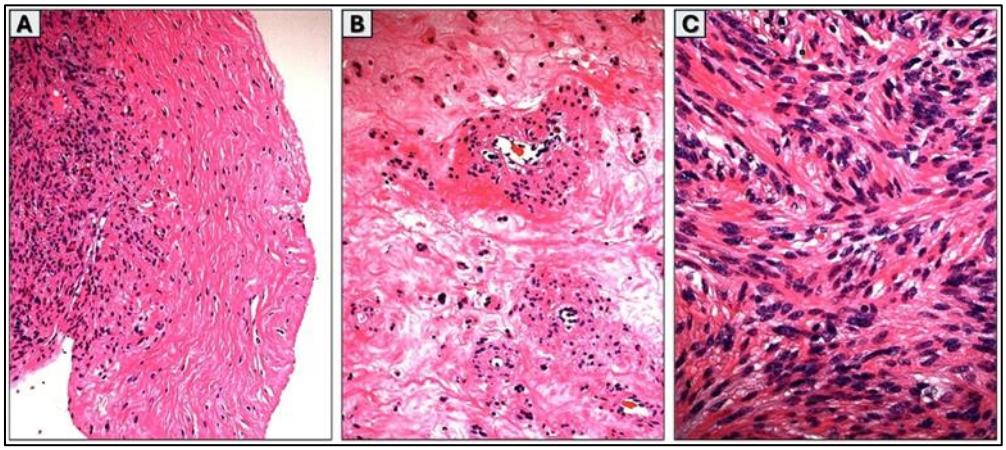
The postoperative course was uneventful. The patient was discharged on postoperative day four without complications. At her six-week checkup, she shared that all her abdominal discomfort, back pain, and symptoms caused by compression had fully disappeared. Follow-up cross-sectional imaging at three and twelve months demonstrated no residual or recurrent disease.

Given the benign nature of the final diagnosis and the achievement of complete resection with clear margins, no adjuvant therapy was indicated. A surveillance protocol consisting of annual clinical examination and imaging for 3 years was nevertheless established, given the lesion's unusual size and retroperitoneal location, which fall outside the classical presentation of ALM. At 1 year following surgery, the patient remained in excellent health, with no evidence of recurrence, and reported full return to her baseline functional status.



1A: Intact 17 cm giant angioleiomyoma, a well-circumscribed, encapsulated giant mass; 1B: Bisected 17 cm giant angioleiomyoma with a whorled appearance and prominent myxoid content

Figure 1 Gross features of the giant angioleiomyoma (ALM)



2A**:** Low power view showing prominent, densely packed fascicles and bundles of spindle-shaped smooth muscle cells weaving between the vascular channels. (H&E X20).; 2B**:** Intermediate power view showing a highly vascular mass, displaying a "bizarre" mix of thick-walled vessels, and venous channels, in the background of hyalinized edematous stroma (H&E X40); 2C**:** High power view showing distinct whorled arrangement of muscle bundles (H&E X60).

Figure 2 Microscopic features of the giant angioleiomyoma (ALM)

3. Discussion

3.1. Background (History, epidemiology, risk factors, and WHO classification)

The term ALM, or "vascular leiomyoma," was first used by Stout in 1937 to describe a benign tumor of vascular origin, to distinguish it from other cutaneous leiomyomas. [6,7] In 1973, Morimoto categorized tumors based on their location and histological features into three types: solid, due to their closely compacted smooth muscle appearance; venous, due to their distinguishing muscular walls; and cavernous, due to dilated vascular channels with less smooth muscle. Additionally, he grouped them into a larger group of solid types, which are often painful, and a smaller group of venous types, which are usually painless or less painful. [8,9,10] Our case represents the solid type, which is often found in the extremities; however, in our case, it was found in the retroperitoneal area.

The WHO classifies soft tissue tumors based on their distinct line of differentiation. ALM is classified as “Tumors of uncertain differentiation.". ALM is characterized by a "well-circumscribed tumor composed of cytologically bland smooth muscle cells showing concentric growth around vascular channels of varying caliber." [11] ALM is often described as a firm, well-circumscribed, slow-growing, benign tumor with an unknown etiology. In terms of genetic dispositions, there are no consistent driver mutations, though a few karyotypic abnormalities have been observed, including 6p, 13q, and 21q rearrangements, and t(X; 10) (q22; q23.2) translocations have been described. However, it is not consistent in all cases. [1,8] Minor injury and trauma, as well as venous stasis and hormonal changes, have been proposed as a potential risk factor. [8]

In terms of occurrence, it most commonly presents in the subcutis or dermis of the extremities but can also occur in the head and neck and the trunk, albeit much rarer, as in this case. It is painful in approximately 60% of cases and is more prevalent in females, who usually have the solid type, than in males, who usually have the venous and cavernous types. ALM can occur at any age, with a peak in the 4th-6th decades of life, and accounts for approximately 5% of all benign neoplasms; giant retroperitoneal angioleiomyoma is much rarer. [8,11]

3.2. Pathogenesis and Pathophysiology

Retroperitoneal ALMs are uncommon, benign, well-demarcated, often solitary, slowly growing neoplasms. ALMs originate from the smooth muscle of the tunica media of vascular walls, with some literature suggesting a potential origin from perivascular cells (pericytes) or from cells that differentiate into vascular structures. [1] The pathogenesis of ALMs is unknown because the disease is relatively rare. There is no known genetic or chromosomal abnormality associated with the neoplasm; however, a study found that mutations in the MED12 gene are present in some smooth muscle tumors arising outside the uterus, including retroperitoneal ALMs. [12] These neoplasms strongly stain with smooth muscle antigen (SMA) and desmin. [4] Because of the tumor's highly vascular nature, smooth muscle cells proliferate around the dense vascular spaces without local invasion or metastasis. The retroperitoneum offers a large space within which a tumor can grow to a considerable size before diagnosis.

The clinical features of this tumor relate to its size and the typical properties of smooth muscle. The majority of symptoms are caused by the mass's space-occupying effect and by secondary involvement of adjacent structures, such as the kidneys, gastrointestinal tract, and ureter. [13] The ureter can be obstructed, leading to failure of the affected kidney and a rise in creatinine levels. The intestines can also be involved, leading to gastrointestinal symptoms. [14] The smooth muscle cells of ALM have abundant, mature-appearing eosinophilic cytoplasm and a rich blood supply, consistent with a vascular lesion. The lack of cellular atypia and low mitotic activity suggests a benign nature. [15]

3.3. Comparative Analysis of Our Case with Existing Literature. (Clinical, radiology, pathology, Lab, Diagnosis, management, and outcome)

ALM in the retroperitoneum is very rare, with most cases in the extremities. [14] The patient in this study was within the typical age range, but the tumor was larger than commonly reported, especially in extremity cases. [1,14] Similar symptomology and size were seen with other retroperitoneal ALMs, with patients experiencing abdominal pain and distension. [16,17] The MRI findings of T2-hyperintense lesions with adjacent vasculature are consistent throughout the literature, as well as in this case. [1,14]

ALMs can only be diagnosed by histology, with previous reports demonstrating diffuse positivity for SMA, muscle-specific actin, and calponin, and variable positivity for h-caldesmon & desmin. [**1,18**] Our case followed a similar IHC profile with SMA, desmin, and h-caldesmon being positive. This case deviated from the standard diagnosis process in the tumor board’s decision to proceed with CT-guided biopsy before surgery. This decision was focused on the abnormal

location, the size of the tumor, increasing the risk for malignancy, and inconclusive imaging findings. Surgical intervention was the preferred approach in other reported instances. [17,19]

The duration of follow-up assessments varied, ranging from 1 year to more frequent evaluations at 1, 3, and 6 months; nevertheless, all cases exhibited complete symptom resolution and no recurrence. [17,19] Although one instance of recurrence was documented nineteen years post-surgical excision, the case was identified as a leiomyoma; the absence of a pathology report precluded definitive identification, and the recurrence was subsequently diagnosed as an ALM. [14]

4. What Have We Learned from This Case?

This case reinforces several practical clinical lessons. First, a slowly enlarging retroperitoneal mass with compressive symptoms, preserved laboratory studies, and absence of overt invasion should not be presumed benign or malignant on imaging grounds alone, because angioleiomyoma in this location may closely mimic leiomyosarcoma, solitary fibrous tumor, gastrointestinal stromal tumor, schwannoma, or dedifferentiated liposarcoma. [4,16] Second, progressive abdominal girth, early satiety, back pain, and exertional dyspnea represented important red flags for mass effect rather than functional gastrointestinal disease, emphasizing the need to revisit an initial benign explanation when symptoms persist or evolve.

This case also illustrates the value of structured multidisciplinary decision-making. In a large and vascular retroperitoneal lesion, preoperative core biopsy may be useful when the result is likely to refine surgical planning and reduce the risk of an unplanned or inadequate operation. Definitive diagnosis remains histopathological, supported by a characteristic IHC and a very low proliferative index. [4] From a management perspective, complete surgical excision remained both diagnostic and therapeutic, with generally a favorable prognosis after clear-margin resection. [4,16]

For future practice, this case suggests that even histologically benign retroperitoneal ALMs may warrant interval surveillance when presenting as unusually large, anatomically deep, or outside the classical clinicopathologic profile.

Abbreviations:

- Angioleiomyoma (ALM);
- Immunohistochemistry (IHC)

5. Conclusion

We present this case of a giant retroperitoneal angioleiomyoma, which remains an exceptionally rare entity whose clinical and radiologic features can closely simulate those of a malignant retroperitoneal neoplasm. The case adds value by demonstrating persistent compressive symptoms, careful cross-sectional imaging, multidisciplinary review, selective image-guided biopsy, and definitive surgical excision, which together allowed accurate diagnosis and curative treatment. For the medical community, this report can broaden awareness of an important benign differential diagnosis for large retroperitoneal masses and underscores that histopathology with IHC remains essential when imaging is indeterminate. In practical terms, the case challenges clinicians to balance oncologic caution with recognition that complete resection of an unusual but benign vascular smooth muscle tumor may lead to excellent functional recovery and durable disease control.

Compliance with ethical standards

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All authors make the following declarations:

- Payment/services information: All authors have declared that they received no financial support from any organization for the submitted work.
- Financial relationships: All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might be interested in the submitted work.

Statement of informed consent

This study was conducted as a retrospective review of archival pathology material collected during routine clinical care. All data were fully de-identified prior to analysis. No patient contact or intervention was involved, and informed consent was not required in accordance with institutional policies.

Data access statement

All relevant data are included in the paper.

Author contributions

All authors contributed equally to producing this manuscript.

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