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Giant abdominopelvic angiolipoleiomyoma: Case report of a rare benign uterine tumor with an outstanding presentation

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Abstract

Angiolipoleiomyoma (ALLM) is a rare benign smooth muscle entity included in the group of Perivascular endothelial cell tumors (PEComas), not being currently classified within uterine mesenchymal tumors. It has low incidence reported (0.06%) and affects mostly women over 40 years.

We describe a case of a 34-year-old woman who presented with 5 months history of abdominal progressive distention. A computer tomography scan reported an abdominopelvic tumor of 30 cm diameter. Without previous biopsy and with suspected malignant soft tissue lesion, radical surgery was performed. Macroscopic examination revealed a well-delimited soft whitish tumor which arose from the uterus isthmus. On microscope slides, we observed a benign neoplasm consisting of 3 components: smooth muscle cells, mature adipose cells and thick and tortuous blood vessels. Immunohistochemistry was positive in muscle cells with Smooth Muscle Actin and Desmin and negative with HMB45 and MELAN-A, leading to the final diagnosis of ALLM.

The differential diagnoses that can be made against this clinical and histological features are diverse and may simulate other abdominopelvic pathologies, even malignant neoplastic lesions.

Keywords

Angiolipoleiomyoma; angiomyolipoma; leiomyoma.

Introduction

ALLM is a benign tumor that is included in the group of PEComas. These are mesenchymal neoplasms with distinctive morphological, immunophenotypic and molecular characteristics. They include angiomyolipoma (kidney as most frequent location), lung clear cell tumor and lymphangioleiomyomatosis. Most PEComas have mutations in the tuberous sclerosis complex gene (TSC1), which participates in the mTOR pathway, being sensitive to target therapies with immunomodulators specifically used in lesions with malignant behavior. The incidence of these lesions in the gynecological tract is low.

Unlike the one that originates at the kidney, uterine angiomyolipoma has differences in the immunophenotype for which the name of ALLM was designated.

This is a rare benign tumor with a smooth muscle phenotype, and it is not currently classified within uterine mesenchymal tumors. It has a low incidence reported (0.06%) and affects mostly women over 40 years of age. There is very little written in the literature and to date, less than 100 cases are published. Clinical presentation is asymptomatic or with unspecific and variable symptoms.

Because of the benign nature of the lesion, surgical treatment is usually performed. Conservative treatment is recommended in small and asymptomatic tumors in WHO Female Genital Tumors 5th Edition 2020. Neoplasms larger than 4 cm or in cases where a rupture have occurred, surgical resection is the best choice. On the other hand, it is reported that exceptionally atypical leiomyomatous proliferation with anaplastic, pleomorphic giant cells may arise from this tumor so complete resection of the uterus by hysterectomy it is much more appropriate.

Prognosis is excellent and to date no recurrences have been reportedso far with follow-up.

Case Report

A 34-year-old female with no relevant medical history presented with a 5 month history of generalized abdominal distention without pain or gastrointestinal symptoms. The physical examination revealed a great increase of the abdominal perimeter (Figure 1).

A computed tomography was performed in which a voluminous central abdominopelvic tumor with sharp borders and heterogeneous density of 30 cm maximum diameter was identified. It displaced the uterus ventrally and encompassed parametria and peri-uterine vascular structures. Without prior biopsy and with suspected diagnosis of malignant soft tissue lesion, radical surgical procedure was decided (anexohysterectomy).

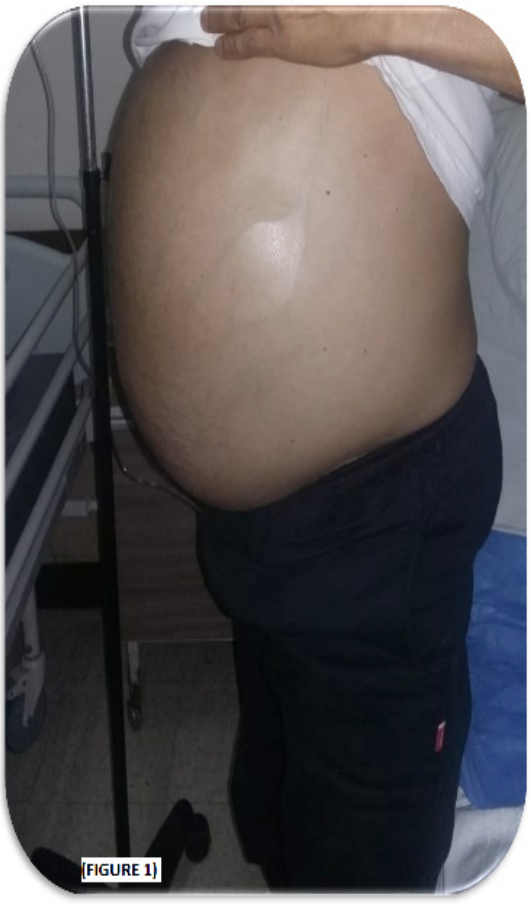
Macroscopically in the uterine isthmus a large well-defined nodular of 30 X 25 X 13 cm whitish tumor with congestive areas and a shiny pseudocapsule was noticed (Figure 2). Cuts into parallel slices showed solid and cystic parenchyma with swirling fibrous sectors, myxoid areas, and foci of hemorrhage (Figure 3).

Microscopic examination revealed a benign neoplasm consisting of 3 components: smooth muscle cell proliferation, without cytological atypia (Figure 6), interspersed with clusters of mature adipose cells (Figure 4) and thick blood vessels (Figure 5). Smooth Muscle Actin and Desmin were positive in the muscle cells (Figures 7 and 8) and HMB45 and MELAN-A were negative. The proliferation index evaluated with ki67 was 3%. These histological and immunohistochemical findings were compatible with Angiolipoleiomyoma. Surgical resection was sufficient and the patient remains currently without recurrences.

Discussion

Large abdominopelvic masses often represent a challenge to clinicians, imaging specialists, surgeons and pathologists on finding the correct way to approach the patient and to reach the correct diagnosis. Most common differential diagnoses are liposarcoma-like mesenchymal tumors.

Documented clinical and imaging presentation are nonspecific and diverse, therefore they can be easily confused preoperative and misdiagnosed. Biopsy of this lesions are usually not profitable due to its histological heterogeneity and because of the need to obtain representative material since immunohistochemistry in mandatory for diagnosis. It is of utmost importance an interdisciplinary evaluation to make the right decision on how to approach the patient, achieve the correct diagnosis and avoid unnecessary additional treatments that can generate unwanted adverse effects. An exhaustive morphological and immunohistochemical study will provide us the key to understand the nature and behavior of the tumor, to assume the correct medical practice and appropriate follow-up for prompt recovery of the patient.



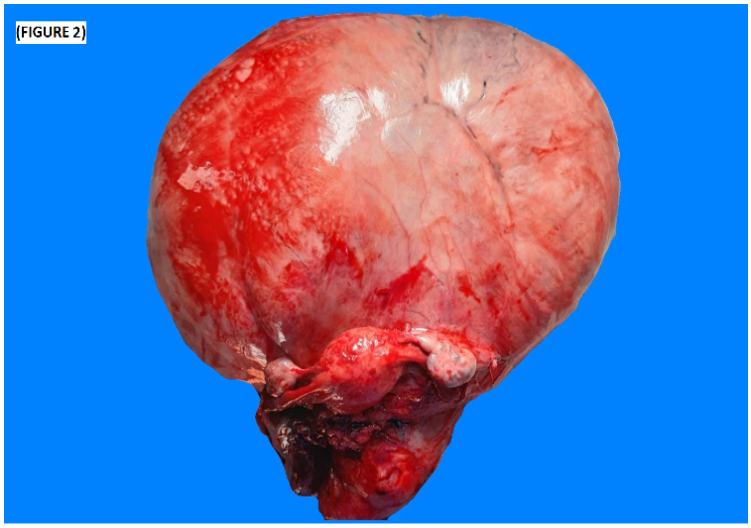


Figure 2: Large well-defined nodular whitish tumor with congestive areas and a shiny pseudocapsule protruding from the postero-inferior sector of the uterus (isthmus).

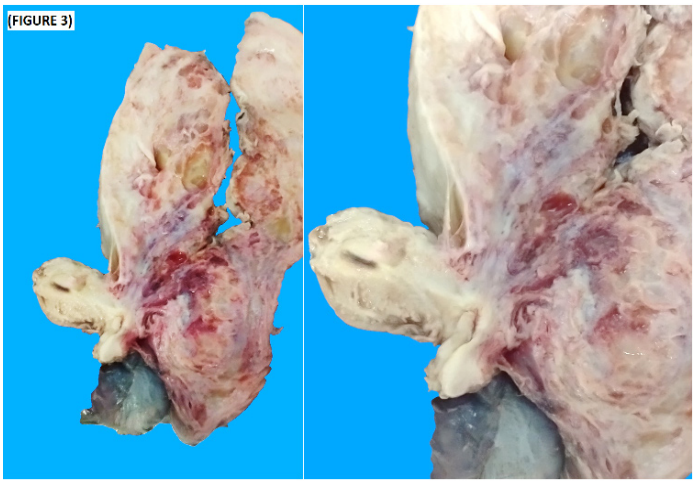


Figure 3: Cuts into parallel slices showed olid and cystic parenchyma with swirling fibrous sectors, myxoid areas, and foci of hemorrhage.

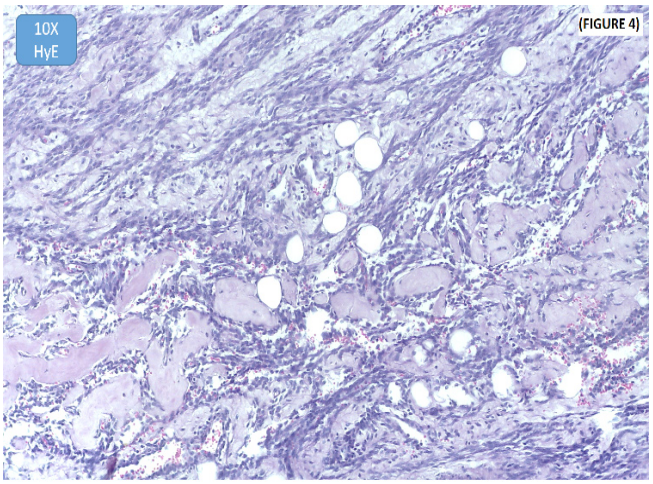


Figure 4: Smooth muscle cell profileration interspersed with clusters of mature adipose cells.

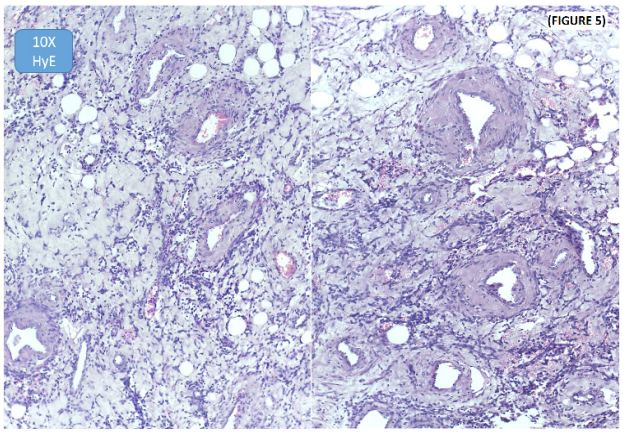


Figure 5: Thick and tortuous blood vessels.

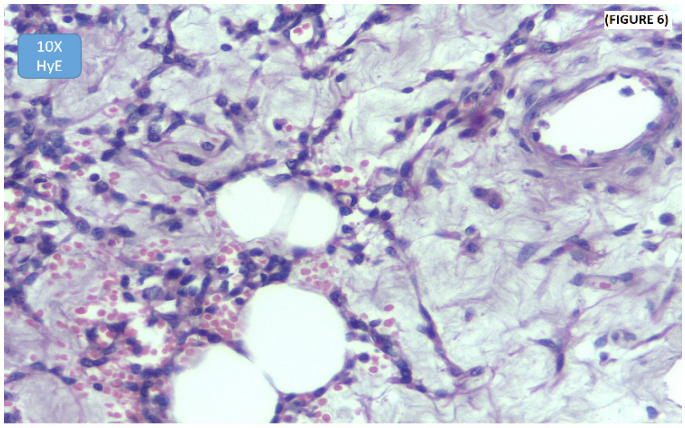


Figure 6: Bland smooth muscle cells with no atypia immersed in myxoid stroma.

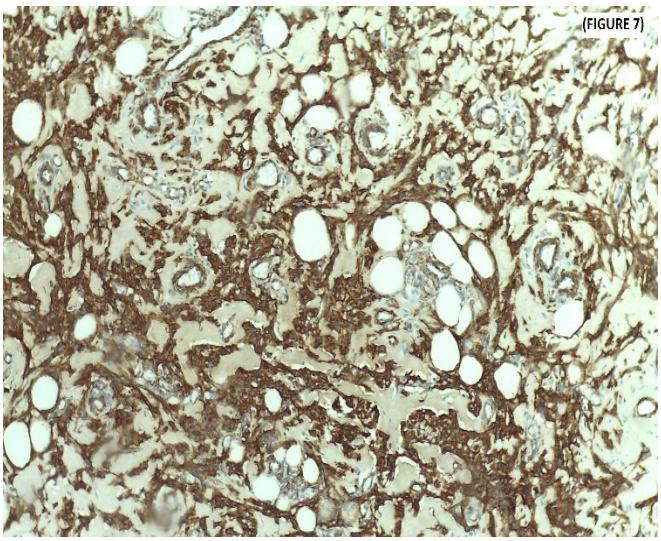


Figure 7: Smooth muscle actin.

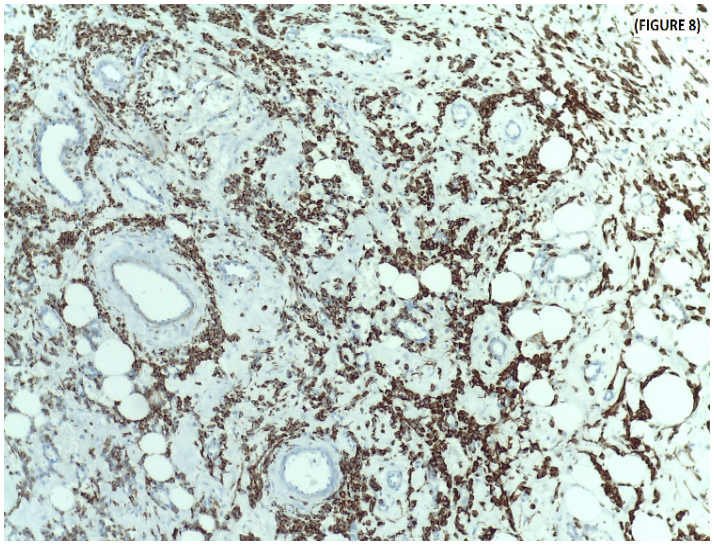


Figure 8: Desmin.

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