OPEN ACCESS – CASE REPORT

Lipomatous Angiomyofibroblastoma in the Vulva: A Case Report with Review of Literature

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ABSTRACT

This is a case of a 55-year-old female, surgical menopause for nine years, not on hormone therapy with one year history of a progressively enlarging left vulvar mass, who sought consult and subsequently underwent excision of mass. Microscopic findings showed alternating hypo- and hypercellular areas composed of plump spindle to epithelioid cells distributed in an edematous stroma with varying amount of collagen. Tumor cells have bland chromatin, inconspicuous nucleoli, and absent mitoses. The cells surround small to medium-sized thin-walled, hyalinized vessels found in hypercellular areas. An adipocytic differentiation of more than fifty percent of tumor is observed further classifying this neoplasm as lipomatous angiomyofibroblastoma, making this an even rarer type of benign mesenchymal tumor of the vulva. Immunohistochemistry stains performed showed positive staining for vimentin, smooth muscle actin, BCL-2, ER, PR, and negative staining for desmin which supports the diagnosis. The clinical presentation, operative findings, histopathologic features and the various considerations are discussed. Literature review of vulvar angiomyofibroblastoma is also presented.

Key words: vulva, angiomyofibroblastoma, lipomatous angiomyofibroblastoma

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INTRODUCTION

Lipomatous angiomyofibroblastoma (LAMF) is a rare, benign soft tissue tumor with a prominent well-differentiated adipocytic component. Only 10% of angiomyofibroblastomas are classified as LAMF.[1] These tumors commonly present as painless vulvar masses. Histologically, they are composed of spindle cells and epithelioid cells surrounding vascular network in a background of loose and edematous stroma. Mature adipocytes comprise a significant portion of the tumor. LAMFs have a good prognosis, with conservative local excision as curative treatment, and uncommon local recurrence.[2]

To date, no known studies in the Philippines have reported cases of vulvar LAMF. This report aims to provide clinical and histopathologic data of this uncommon benign soft tissue tumor in the vulva in the local setting.

CASE

Patient is a 55-year-old female, G3P3 (3-0-0-3) previous normal spontaneous delivery, surgical menopause for nine years, status post total abdominal hysterectomy in 2019 for removal of uterine myomas in a private hospital, not on hormone therapy and consulted for left vulvar mass. One year prior, patient noted a 2.0 cm soft cystic mass on the left vulva, which was non-tender, and non-erythematous, with no other associated symptoms. She did not take any medications nor sought consult at this time. In the interim, patient noted the mass to be increasing in size but still with no medications nor consults done.

One month prior, patient observed that the mass was approximately 5.0 cm. Persistent progression in size of the mass prompted her to consult with an obstetrician-

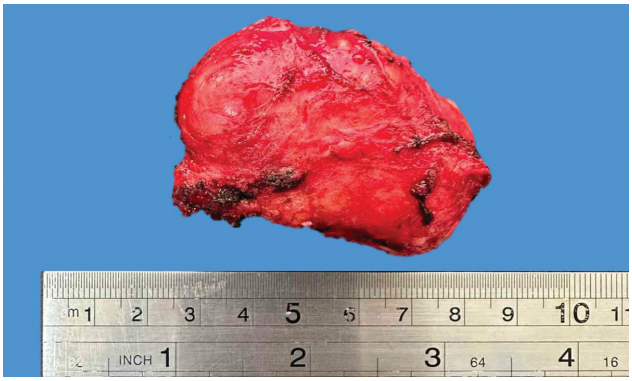


Figure 1. External surface of the mass, post-operatively.

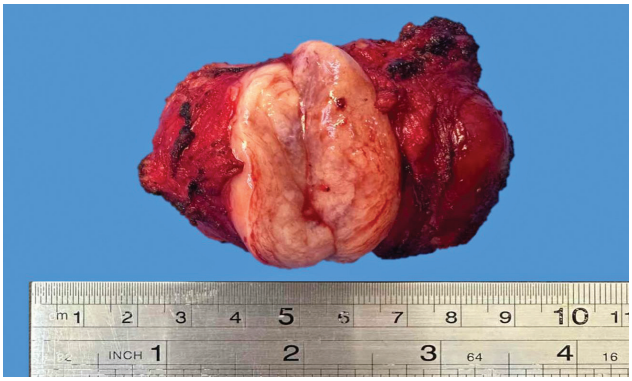


Figure 2. Cut surface of the mass, post-operatively.

gynecologist, with physical examination findings of a 5.0 cm cystic left vulvar mass. Transvaginal ultrasound showed surgically absent uterus, small ovaries, left adnexa, and a thin- walled cyst measuring 2.02 x 1.45 x 1.74 cm, volume of 2.68 ml. with no color on flow mapping suggestive of para-ovarian cyst.

Patient sought another consult which revealed a 6.0 x 5.0 cm cystic mass, non-tender and non-erythematous, located at the 5 o'clock position of the left vulvar area. Speculum exam and internal examination was deferred. She was advised excision of mass. Intra-operatively, a fluctuant vulvar mass admixed with blood, measuring 6.0 x 5.0 cm, was obtained (Figures 1 and 2). Patient tolerated the procedure without undue complications and was discharged several hours after recuperating from anesthesia. Follow-up consult two weeks after surgery was uneventful.

Submitted specimen at the histopathology laboratory showed a circumscribed, irregularly ovoid, lobulated, rubbery mass measuring 6.0 x 4.0 x 2.5 cm with a gray-pink external surface covered with focal fibrous tags. It has been previously opened along one side, and further sectioning revealed a pink gray, glistening soft cut surface.

Microscopic examination showed alternating hypo- and hypercellular areas composed of spindle to epithelioid myofibroblastic cells distributed in a loose and fibrous stroma (Figure 3). There was no involvement of the surgical margins. The cells surrounded small to medium-sized thin-walled, as well as hyalinized vessels, that were irregularly distributed (Figures 4 and 5). More than 50%

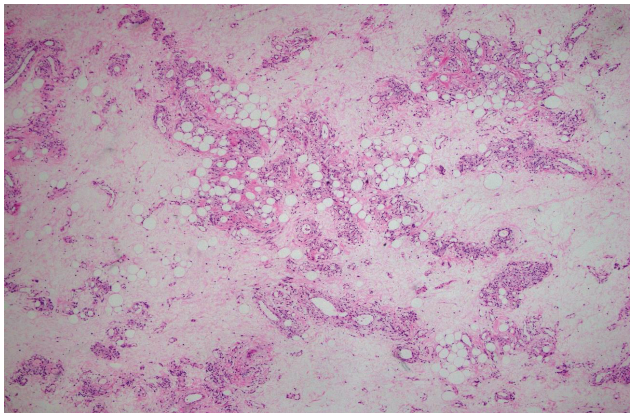
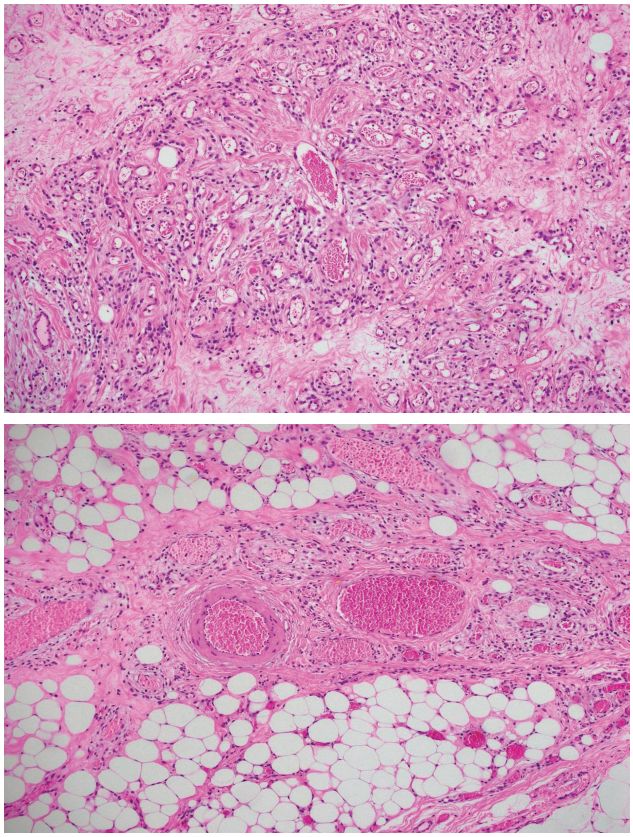


Figure 3. Alternating hypo- and hyper-cellular areas (H&E, 40x).



Figures 4 and 5. Spindle to epithelioid cells surrounding small to medium-sized, thin-walled, as well as hyalinized vessels (H&E, 100x).

of the tumor consisted of mature fat cells (Figure 6). The tumor cells were monomorphic with bland chromatin and inconspicuous nucleoli, and absent mitoses and necrosis (Figures 7 and 8). Immunohistochemistry stains performed showed positive staining for vimentin, smooth muscle actin, BCL-2, ER, PR, and negative staining for desmin (Figure 9 A-F).

DISCUSSION

An angiomyofibroblastoma is a benign, well-circumscribed myofibroblastic neoplasm, usually arising in the pelviperineal region, especially the vulva, and composed

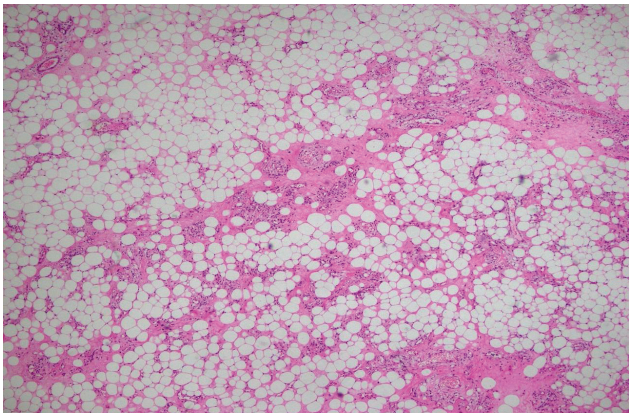
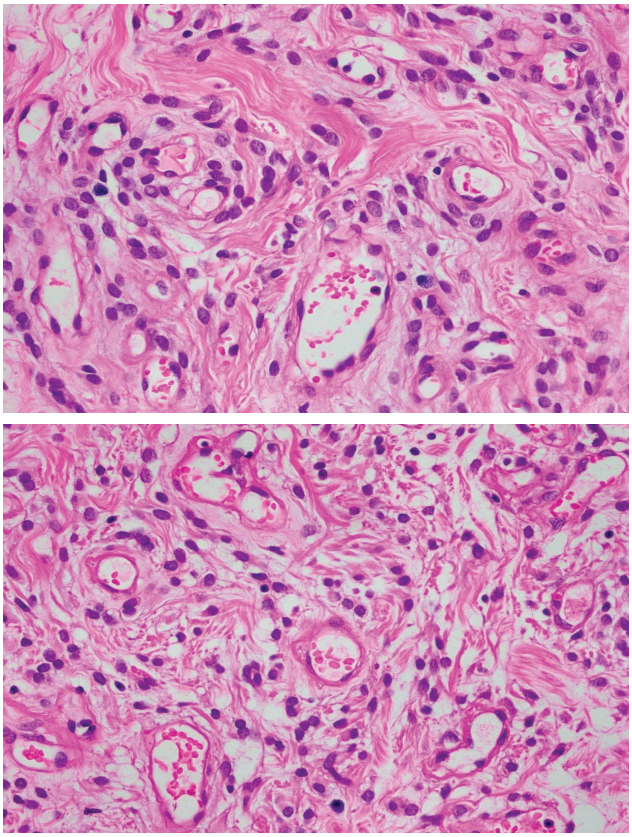


Figure 6. Numerous fat cells within the tumor (H&E, 40x).



Figures 7 and 8. Monomorphic spindle to epithelioid cells with bland chromatin, inconspicuous nucleoli, and absent mitoses and necrosis surrounding small to medium-sized dilated vessels (H&E, 400x).

of stromal cells distinctive of this anatomical region.[1] Angiomyofibroblastoma is uncommon, having an incidence comparable to that of deep (aggressive) angiomyxoma, which is unknown due to its rarity. These tumours arise predominantly in adult females, between menarche and menopause. About 10% of patients are postmenopausal. Convincing examples have not been described before puberty, and rare cases occur in males.

Pathophysiology is uncertain but may arise from subepithelial stroma or a perivascular stem cell.[1] In tumors with a significant adipocytic component, said components can possibly be caused by adipocytic metaplasia or fatty differentiation of tumor cells.[3] It has also been postulated that genital angiomyofibroblastomas may arise from precursor cells of hormonally responsive stroma, capable of multidirectional mesenchymal differentiation, which could be either be fibroblastic, myofibroblastic, or lipomatous.[2]

Common clinical features include a slowly enlarging, painless, circumscribed mass. It may be present for weeks to years before diagnosis, and multifocal cases are rare. The most frequent preoperative diagnosis is Bartholin gland cyst. Grossly, lesions are unencapsulated but well circumscribed, with a tan/pink cut surface and a soft consistency. Necrosis is not seen. Most cases measure <5 cm in maximum diameter, although rare examples as large as 10 cm have been recognized. The clinical presentation of this case is a one-year history of a slowly enlarging, non-tender mass which was initially observed to be 2.0 cm, with no other associated symptoms.

Histologically, tumor cells are round to spindle-shaped with eosinophilic cytoplasm, concentrated around vessels. Binucleated and multinucleated tumor cells are common. Mitoses are rarely seen. In postmenopausal women, stroma is more fibrous rather than edematous, with hyalinization of vessel walls. Essential criteria include the following: well-circumscribed, prominent stromal vessels, and round to spindle-shaped cells (often multinucleated) in a perivascular distribution.[1] Ten percent of cases have a prominent well-differentiated adipocytic component,[1] which are classified as lipomatous variants of angiomyofibroblastomas (LAMF) in review of literature.[3,4] While angiomyofibroblastomas are rare, LAMFs are even rarer, with no reported cases in the local setting.

In a 20-year literature review and case report done in 2015, there were a total of ten reported cases of angiomyofibroblastoma, four of which were the lipomatous variant.[4] Age at time of diagnosis ranged from 23 to 50 years old, and reported symptom was a painless mass or lesion. All LAMFs occurred in the vulva, and size varied from 1.1 to 11 cm with an average of 5.5 cm. In six of the cases, mature adipocytes comprised 30% to 80% of the mass. In all ten cases, LAMFs were histologically similar, being described as having hypercellular and hypocellular areas composed of spindle and epithelioid cells, in a background of loose and edematous material. Condensed spindle cells surrounding vascular networks were dispersed throughout the tumor, with mature adipocytes comprising a significant portion of the tumor volume. Features of malignancy like high mitotic rate, cytologic atypia, and necrosis were absent.

Another literature review and case report were done in 2016.[2] They reported 16 cases of LAMF, wherein the fatty component comprised 30% to >90% of the tumor. In this study, they state that LAMF may be used when fat cells constitute more than or equal to 30% of the tumor. Tumor size ranged from 1.8 to 11.0 cm, while patient age ranged from 23 to 69 years of age. Histologically, the case report revealed an abundance of fat cells involving around 85% of the tumor, with numerous medium and small-sized vessels, and multifocal fibrous areas and pseudoangiomatous spaces. Similar to the study done in 2015, there was a

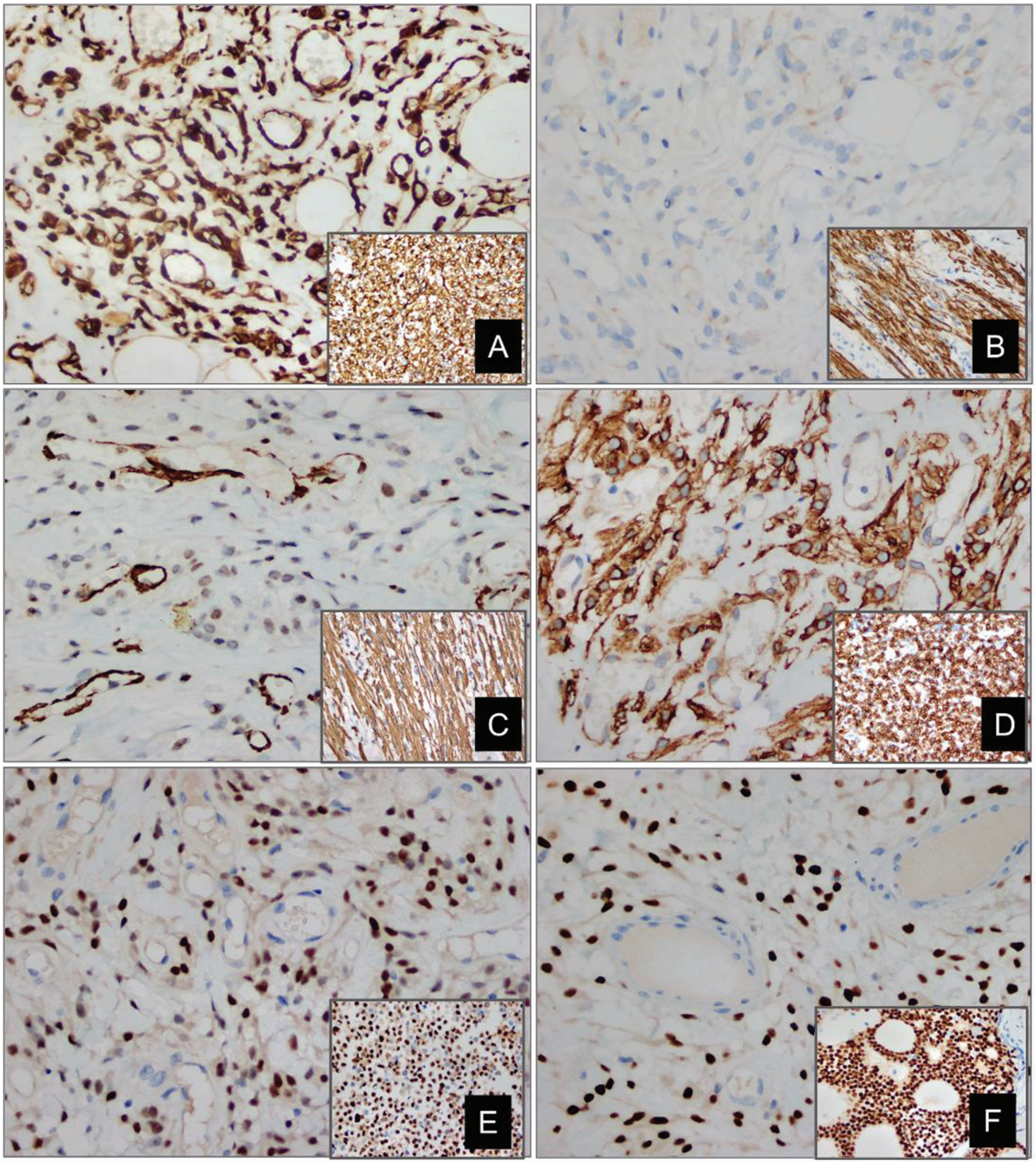


Figure 9. Immunohistochemistry stains (Horseradish peroxidase method, 400x) with respective positive controls *(inset)*: **(A)** Vimentin, **(B)** Desmin, **(C)** SMA, **(D)** BCL-2, **(E)** ER and **(F)** PR.

proliferation of spindle-shaped, rounded or epithelioid tumor cells in small nests and cords within perivascular fibrous tissue located in both fatty and fibrous areas. No mitotic figures were seen.[4]

Immunohistochemical studies of the tumor cells based on the aforementioned studies would show positive staining for vimentin, BCL-2, estrogen and progesterone receptors. Occasionally, tumor cells would be positive in CD34, desmin, CD10, CD99, muscle-specific actin, smooth muscle actin, and S100. Negative staining is seen for cytokeratin, EMA, c-kit, HMB45, GFAP, CD68, and factor VIII.[2,4] This is in comparison to the WHO Classification of Female Genital Tumors, which states that majority of cases would show strong and diffuse positive staining for desmin, and focal positivity for SMA or pan-muscle actin. However, it was also acknowledged that desmin staining may be reduced or absent in postmenopausal cases.[1]

Differential diagnoses for LAMF include benign and malignant mesenchymal neoplasms of the female genital tract. Benign mesenchymal neoplasms include superficial and aggressive angiomyxoma, solitary fibrous tumor, cellular angiofibroma, and spindle cell lipoma. Liposarcomas fall under the category of malignant mesenchymal neoplasms.

Superficial angiomyxomas present as multinodular dermal or subcutaneous myxoid lesions that consist of bland spindle cells. They are seen on the vulva, but they are more commonly seen on the lower limbs, head, and neck.[5] They occur more frequently in males, but in females, they usually present at reproductive age. Compared to LAMF, they have no perivascular congregation of cells, with prominent stromal mucin and conspicuous stromal neutrophils, with the last feature not seen in LAMF. Immunohistochemical studies are not useful in distinguishing between the two.[4] Superficial angiomyxoma was ruled out due to patient's age and the histomorphologic features of the tumor, which had the presence of perivascular congregation of said cells and lack of stromal mucin and conspicuous stromal neutrophils.

Aggressive angiomyxomas (AAM) occur in deep soft tissues of the perineum, pelvis, and genitalia of women of reproductive age. They are typically large, with most tumors measuring more than 10 cm, and have a more uniformly hypocellular myxoid stroma. Tumor cells are more uniformly spindled, with infiltrative borders and no perivascular congregation of spindle cells. Despite the word "aggressive" in its name, the term mainly refers to the tumor's predilection for local recurrence and is considered a benign tumor. They have a high local recurrence rate. AAM was ruled out as well, due to the patient's age, the histomorphologic features of the patient's tumor, which presented with perivascular congregation of tumor cells and no presence of infiltrative borders. Immunohistochemical studies overlap among LAMF and AAM,[3] but it has been noted that tumor cells of AAMs have a positive staining for HMGA2.[6] Despite this, HMGA2 is non-specific for AAM as positive staining can also be seen in a subset of leiomyomas. Correlation between immunohistochemical and histomorphologic features are therefore essential.

Solitary fibrous tumors are grossly well-circumscribed masses, usually unencapsulated, that consist of ovoid to fusiform spindle cells with indistinct cell borders arranged haphazardly or in short, ill-defined fascicles. Compared to LAMF, they are more commonly seen in extragenital sites and have prominent staghorn vessels with thick keloid-like stromal collagen bands. CD34 and STAT6 are positive on immunohistochemical stain, but like superficial angiomyxomas, these studies are not useful in distinguishing between the two. Solitary fibrous tumors were ruled out due to the site of predilection of these tumors being extra-genital, as well as the patient's tumor not exhibiting prominent staghorn vessels with thick stromal collagen bands.

Cellular angiofibromas are also superficial soft tissue tumors, but compared to LAMF, the tumor is more highly cellular, with an abundance of vessels and the cells that proliferate are primarily spindled. Stroma is also more collagenous and/or hyalinized. In terms of immunohistochemistry stains, cellular angiofibromas are more frequently positive for Desmin, CD34, ER, and PR.[3,4] Cellular angiofibromas were ruled out as the patient's tumor also had presence of hypocellular areas, and the cells were not exclusively spindle-shaped, as it also included epithelioid cells.

Spindle cell lipomas are similar to LAMFs in that there are proliferating spindle cells, however these are the only cells that the tumor consists of, and they do not form nests. They are also usually seen as a component of leiomyomas and tend to be located within the myometrium instead of the vulva.[4] ER positivity has also not been reported in spindle cell lipoma.[3] Spindle cell lipomas were ruled out due to the site of predilection, as well as the presence of epithelioid cells and formation of nests of cells in the patient's tumor.

Malignant mesenchymal neoplasms include liposarcoma. Liposarcomas can also arise in the soft tissue of the vulva in middle-aged women, with tumor cells exhibiting significant nuclear atypia.[7] In cases where there is minimal nuclear atypia, as seen in well-differentiated liposarcoma, the presence of lipoblasts would favor liposarcoma over LAMF. Local recurrence in liposarcomas of the vulva have been reported. In the case of the patient's tumor, no nuclear atypia, no mitoses and no lipoblasts were seen.

The clinical presentation of a slowly growing vulvar mass over a course of one year not associated with any other symptoms, the gross appearance of a circumscribed, lobulated, soft to rubbery mass with defined borders, the microscopic features composed of edematous stroma having hypo- and hypercellular areas containing spindle to epithelioid cells with scattered thin-walled small to medium sized vessels amongst a background of abundant mature adipocytes, absence of nuclear atypia, necrosis, and mitoses are features compatible with lipomatous angiomyofibroblastoma. The positive stains for vimentin, smooth muscle actin, BCL-2, ER, and PR although non-specific also support our histologic diagnosis.

While there are multiple differential considerations for LAMF, immunohistochemical studies are not useful in distinguishing between the multiple soft tissue tumors as most have similar staining profiles.[8] Ultimately, differentiation among the tumors is based on clinical history and histomorphologic characteristics. In terms of treatment, conservative local excision is considered curative, and local recurrence is rare.[2]

CONCLUSION

Lipomatous angiomyofibroblastoma (LAMF) is a rare phenomenon, with few reported cases on literature review and none in the local setting. While immunohistochemistry studies are similar among the soft tissue tumors located in the vulva, diagnosis of LAMF is mainly based on clinical history, morphologic and histopathologic characteristics. This matters since differential diagnoses include a whole gamut of benign mesenchymal as well as aggressive angiomyxomas and liposarcomas of the lower genital tract.

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ETHICAL CONSIDERATION

Patient consent was obtained before submission of the manuscript.

STATEMENT OF AUTHORSHIP

All authors certified fulfillment of ICMJE authorship criteria.

DATA AVAILABILITY STATEMENT

No datasets were generated or analyzed for this study.

AUTHOR DISCLOSURE

The authors declared no conflict of interest.

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