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SUPERFICIAL ANGIOMYXOMA OF THE LABIA MAJORA: A CASE REPORT AND REVIEW OF DIAGNOSTIC AND THERAPEUTIC APPROACHES

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Abstract

Superficial angiomyxoma (SAM) represents an uncommon benign mesenchymal tumor of cutaneous origin, with only sporadic cases reported in the genital region. This article presents a detailed case study of a 35-year-old female who developed a slow-growing, asymptomatic mass on the left labia majora over a one-year period. The clinical presentation, diagnostic workup, surgical management, and histopathological findings are comprehensively described. Microscopic examination revealed characteristic features of SAM, including a well-demarcated lesion composed of myxoid stroma with prominent vascular components and scattered adipocytes, without evidence of cellular atypia. The patient underwent complete surgical excision with no signs of recurrence at six-month follow-up. This case underscores the diagnostic challenges posed by vulvar SAM due to its clinical similarity to more common labial lesions and emphasizes the critical role of histopathological examination in establishing a definitive diagnosis. The discussion incorporates a thorough review of current literature regarding the epidemiology, pathological characteristics, immunohistochemical profile, and management strategies for SAM, with particular attention to its differentiation from morphologically similar entities such as aggressive angiomyxoma.

INTRODUCTION

Superficial angiomyxoma (SAM) is a rare, benign soft tissue tumor that was first characterized in the context of Carney complex, an autosomal dominant multiple neoplasia syndrome, but has since been recognized to occur more frequently as an isolated lesion [1]. The tumor typically presents as a slow-growing, painless cutaneous or subcutaneous nodule, with a predilection for the trunk, head and neck region, and extremities in male patients during their fourth to fifth decades of life [2]. Genital involvement, particularly in the vulvar region, remains exceptionally uncommon, with fewer than 50 documented cases in the medical literature [3].

The clinical significance of SAM lies in its potential to mimic both benign and malignant vulvar lesions, necessitating careful histopathological evaluation for accurate diagnosis. While SAM is histogenetically distinct from its more aggressive counterpart - aggressive angiomyxoma - the two entities share sufficient morphological overlap to pose diagnostic challenges [4]. The present case report contributes to the limited body of knowledge regarding vulvar SAM by providing a comprehensive clinical-pathological correlation and synthesizing current understanding of this rare entity through an extensive literature review.

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Case Report

Clinical Presentation
A 35-year-old, premenopausal female with no significant medical history presented to our outpatient department with a gradually enlarging mass on the left labia majora of one year's duration. The patient reported that the lesion had initially appeared as a small, pea-sized nodule that progressively increased in size without associated pain, pruritus, or discharge. There was no history of trauma, fever, or weight loss. The patient's menstrual history was unremarkable, and she had no symptoms suggestive of urinary or bowel dysfunction.

Physical Examination
Inspection revealed a solitary, well-circumscribed, oval-shaped mass measuring approximately 3 × 2 cm on the left labia majora. The overlying skin appeared slightly hyperpigmented but otherwise intact, without evidence of ulceration or telangiectasia. On palpation, the mass was soft to rubbery in consistency, mobile, and non-tender, with no local warmth or fluctuance. The contralateral labia and remaining external genitalia appeared normal.

Gynecological Examination

Speculum examination demonstrated normal vaginal mucosa with no evidence of discharge or bleeding. Bimanual palpation revealed free bilateral fornices, an anteverted uterus of normal size, and non-tender adnexa.

Diagnostic Workup
Routine haematological and biochemical investigations were within normal limits. Pelvic ultrasound showed no abnormalities of the internal genital organs or pelvic structures. A Papanicolaou smear revealed non-specific inflammatory changes with no evidence of intraepithelial malignancy or dysplasia.

Surgical Management
The patient underwent complete excisional biopsy under spinal anaesthesia. Intraoperatively, the mass was found to be well-encapsulated and easily separable from surrounding tissues, with no evidence of local infiltration. The surgical specimen measured 3.5 × 2.5 × 2 cm and was sent for histopathological examination.

Histopathological Findings
Gross examination revealed a nodular, gelatinous mass with a smooth external surface. Sectioning demonstrated a glistening, myxoid cut surface with scattered small haemorrhagic foci. Microscopic examination showed:

1. An intact epidermis with unremarkable squamous epithelium
2. A well-circumscribed dermal lesion with partial encapsulation
3. Moderately cellular myxoid stroma with abundant mucin deposition
4. Numerous small to medium-sized, thin-walled blood vessels exhibiting variable dilatation and congestion
5. Scattered mature adipocytes within the stroma
6. No evidence of nuclear atypia, mitotic activity, or necrosis

These features were diagnostic of superficial angiomyxoma

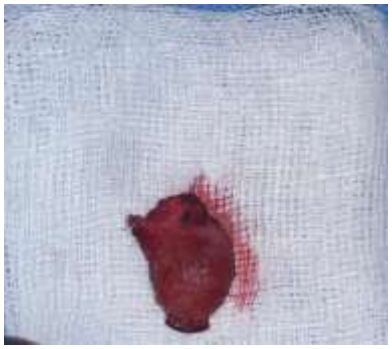

Figure 1

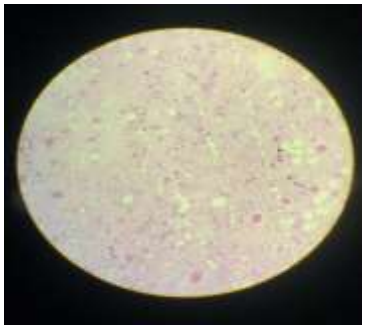

Figure 2

Postoperative Course
The patient recovered uneventfully and was discharged on postoperative day 2. At six-month follow-up, there was no evidence of local recurrence, and the surgical site had healed well.

DISCUSSION

Epidemiology and Pathogenesis
SAM represents a rare mesenchymal neoplasm of uncertain histogenesis, with an estimated incidence of 0.1 cases per million population [5]. While originally described as a cutaneous manifestation of Carney complex, the majority of SAM cases occur sporadically without association with this syndrome [6]. The tumor shows a male predominance (M:F ratio 2:1) and typically affects adults in their 30s to 50s, making our case of a 35-year-old female somewhat unusual [7].

The pathogenesis of SAM remains incompletely understood. Some authors propose that it arises from primitive mesenchymal cells capable of multidirectional differentiation, while others suggest a vascular or fibroblastic origin [8]. The consistent presence of stromal myxoid change likely reflects altered hyaluronic acid metabolism within the tumor microenvironment.

Clinical Features
Vulvar SAM typically presents as a slow-growing, painless mass ranging from 1 to 5 cm in diameter [9]. The clinical differential diagnosis includes:

1. Bartholin gland cyst or abscess
2. Epidermal inclusion cyst
3. Lipoma
4. Angiomyofibroblastoma
5. Aggressive angiomyxoma
6. Superficial myxofibrosarcoma

The indolent growth pattern and lack of systemic symptoms often lead to delayed presentation, as seen in our patient who waited one year before seeking medical attention.

Histopathological Characteristics
SAM demonstrates several distinctive microscopic features:

1. Well-circumscribed but non-encapsulated architecture
2. Hypocellular to moderately cellular myxoid stroma
3. Prominent vascular component with thin-walled vessels
4. Variable inflammatory infiltrate (predominantly neutrophils)
5. Frequent presence of entrapped adipocytes

These characteristics help distinguish SAM from its principal histological mimics:

1. **Aggressive angiomyxoma:** Infiltrative margins, larger vessel size, and deeper location
2. **Angiomyofibroblastoma:** More cellular stroma with plump stromal cells
3. **Myxoid neurofibroma:** S100 positivity and characteristic wavy nuclei

Immunohistochemical Profile
SAM typically shows:

- Strong diffuse positivity for vimentin
- Variable CD34 expression (30-40% of cases)
- Negative for:

- Smooth muscle actin
- Desmin
- S100 protein
- Cytokeratins

This immunoprofile helps exclude other myxoid neoplasms with overlapping morphology [10].

Management and Prognosis
Complete surgical excision with clear margins represents the treatment of choice. Local recurrence rates range from 30-40%, primarily due to incomplete initial excision [11]. Long-term follow-up is recommended to monitor for recurrence, particularly in cases with positive margins. There are no reported cases of malignant transformation or metastasis.

CONCLUSION

This case highlights the diagnostic challenges posed by superficial angiomyxoma of the vulva, emphasizing the importance of histopathological examination in distinguishing this rare benign entity from more common vulvar masses. While SAM carries an excellent prognosis following complete excision, clinicians should be aware of its significant recurrence potential. The present report contributes to the limited literature on genital SAM and underscores the need for increased awareness of this diagnostic possibility among both surgeons and pathologists.

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Figures:

- **Figure 1:** Gross appearance of excised specimen
- **Figure 2:** High-power view demonstrating myxoid stroma with vascular proliferation (H&E, ×200)