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Aggressive Angiomyxoma of the Vulva in a Postmenopausal Woman Presenting as a Necrotic Pedunculated Mass: A Rare Case Report

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Abstract: ***Aggressive angiomyxoma is a rare, slow-growing mesenchymal tumor predominantly affecting the pelvis and perineum of women in the reproductive age group. It is characterized by a high propensity for local recurrence despite its benign histology. We report a case of a 49-year-old postmenopausal woman who presented with a painless mass over the right labia majora for two months, associated with foul-smelling discharge. She had attained menopause two years prior and had a history of type 2 diabetes mellitus without regular treatment. Clinical examination revealed a large pedunculated mass measuring 10 × 5 cm with areas of necrosis and discharge. Right inguinal lymph nodes were palpable. Per speculum examination showed a hypertrophied cervix and vagina. Surgical excision of the mass was performed, and histopathological examination confirmed the diagnosis of aggressive angiomyxoma. This case is notable due to its occurrence in a postmenopausal woman and its atypical presentation with necrosis and foul-smelling discharge, mimicking malignancy. Early diagnosis and complete surgical excision are crucial to reduce recurrence and ensure favorable outcomes.***

Keywords: Aggressive angiomyxoma, vulvar mass, postmenopausal, mesenchymal tumor, vulval tumor, recurrence

1. Introduction

Aggressive angiomyxoma is a rare benign mesenchymal tumor first described by Steeper and Rosai in 1983. It predominantly affects women in the reproductive age group and commonly arises in the pelvis, perineum, and vulvovaginal region. Despite being histologically benign, it is termed "aggressive" due to its infiltrative nature and high risk of local recurrence.

Clinically, aggressive angiomyxoma presents as a slow-growing, painless mass, often mistaken for more common conditions such as Bartholin cyst, lipoma, vulvar abscess, or malignancy. The tumor is hormonally responsive, often expressing estrogen and progesterone receptors, which explains its prevalence in reproductive-age women.

Diagnosis is frequently delayed due to its indolent growth and nonspecific presentation. Imaging modalities such as MRI can aid in preoperative diagnosis, showing a characteristic "swirled" appearance. However, definitive diagnosis is based on histopathological examination.

This case report highlights an unusual presentation of aggressive angiomyxoma in a postmenopausal woman with necrosis and discharge, features that clinically mimicked malignancy. The report underscores the importance of considering this rare entity in the differential diagnosis of vulvar masses.

2. Methodology

A 49-year-old postmenopausal woman presented with a painless swelling over the right labia majora for the past two months. The swelling was gradually increasing in size and was associated with foul-smelling discharge.

Menstrual History- Attained menopause 2 years ago
Obstetric History- Para 4, Live 4/ All normal vaginal deliveries

Clinical Examination

Local Examination:
A large pedunculated mass measuring approximately 10 × 5 cm was present over the right labia majora. The surface showed areas of necrosis with foul-smelling discharge.
Lymph Nodes: Right inguinal lymph nodes were palpable.
Per Speculum Examination (P/S):
Hypertrophied cervix and vagina were noted.

Based on clinical findings, provisional diagnoses included vulvar malignancy, infected fibroepithelial polyp, and soft tissue tumor.

The patient was planned for surgical excision of the mass. The excised specimen was sent for histopathological examination.

3. Results and Discussion

Aggressive angiomyxoma is a rare tumor with a predilection for the pelvic and perineal regions. It is characterized by locally infiltrative growth and a high recurrence rate, ranging from 30–70%.

In the present case, the patient was postmenopausal, which is an uncommon age group for this tumor. Additionally, the presence of necrosis and foul-smelling discharge raised suspicion of malignancy, making preoperative diagnosis challenging.

Gross Findings
Pedunculated mass measuring 10 × 5 cm

Soft, gelatinous consistency
Areas of necrosis

Histopathological Findings
Microscopic examination revealed:
Hypocellular lesion with spindle-shaped and stellate cells
Abundant myxoid stroma
Numerous thin- and thick-walled blood vessels
Minimal cellular atypia and low mitotic activity
These findings were consistent with aggressive angiomyxoma.

Differential Diagnosis
Vulvar carcinoma (due to necrosis and lymphadenopathy)
Bartholin gland cyst/abscess
Lipoma
Fibroepithelial polyp
Angiomyofibroblastoma

Discussion
The tumor's infiltrative nature explains its high recurrence rate. Although benign, it requires wide local excision with clear margins. Hormonal therapy (e.g., GnRH analogs) may be useful in recurrent or unresectable cases due to hormone receptor positivity.

The presence of inguinal lymphadenopathy in this case was likely reactive rather than metastatic, as aggressive angiomyxoma rarely metastasizes.

Long-term follow-up is essential due to the risk of recurrence, which may occur even years after initial treatment.

Operative Photographs

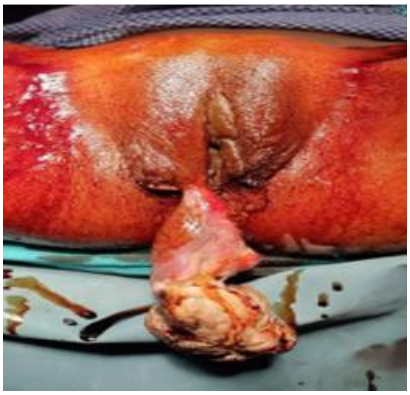


Figure 1: Clinical Image

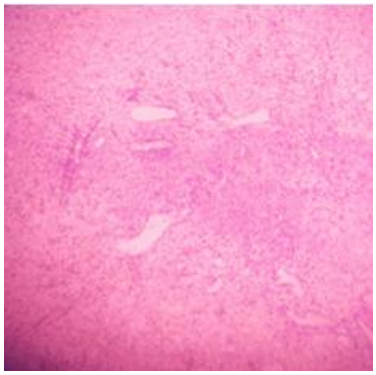


Figure 2: Histopathological image

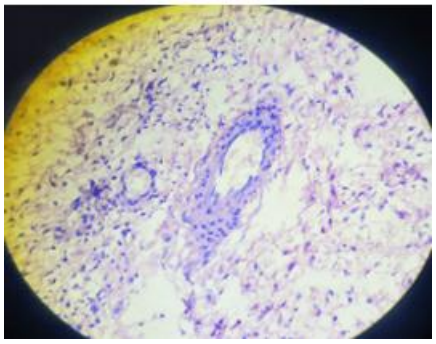


Figure 3: Histopathology magnified image

4. Conclusion

Aggressive angiomyxoma is a rare soft tissue tumor that should be considered in the differential diagnosis of vulvar masses, even in postmenopausal women. Its atypical presentation with necrosis and discharge can mimic malignancy, leading to diagnostic dilemmas.

Histopathological examination remains the cornerstone of diagnosis. Complete surgical excision is the mainstay of treatment, and long-term follow-up is essential due to the high risk of recurrence.

Early recognition and appropriate management can significantly improve patient outcomes and reduce morbidity.

References

[1] Kura MM, Jindal SR, Khemani UN. Aggressive angiomyxoma of the vulva: an uncommon entity. Indian dermatology online journal. 2012 May 1;3(2):128-30.
[2] Steeper, Theresa A. M.D.; Rosai, Juan M.D.. Aggressive angiomyxoma of the female pelvis and perineum: Report of nine cases of a distinctive type of gynecologic soft-tissue neoplasm*. The American Journal of Surgical Pathology 7(5):p 463-476, July 1983.
[3] Schwartz, Peter Edward MD1; Hui, Pei MD, PhD2; McCarthy, Shirley MD, PhD3. Hormonal Therapy for Aggressive Angiomyxoma: A Case Report and Proposed Management Algorithm. Journal of Lower Genital Tract Disease 18(2):p E55-E61, April 2014.
[4] Haldar K, et al. Aggressive angiomyxoma: a case report and literature review. Eur J Surg Oncol.
[5] Sutton BJ, et al. Aggressive angiomyxoma: a multidisciplinary approach. Obstet Gynecol.
[6] Chan YM, et al. Aggressive angiomyxoma in females: MRI features. AJR Am J Roentgenol.
[7] Behranwala KA, et al. Aggressive angiomyxoma: a distinct clinical entity. Int J Gynecol Cancer.