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Case Report Article

AGGRESSIVE ANGIOMYXOMA OF VULVA IN YOUNG FEMALE

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ABSTRACT

Aggressive Angiomyxoma (AA) is an uncommon, locally invasive mesenchymal neoplasm predominantly affecting women of reproductive age. It is characterized by a notable potential for local recurrence, making it critical to distinguish AA from other mesenchymal tumors in the same anatomical regions. This tumor typically arises within the soft tissues of the pelvic region, perineum, and lower genital tract. While metastasis remains rare, the tumor's aggressive nature and propensity for local relapse highlight its clinical significance. The majority of cases occur in women during their reproductive years, underlining the age-specific risk. **Presentation of case** A 23-year-old married Libyan woman presented with a gradually enlarging mass on the right labia majora, which had been present for two years. The patient's primary concern was the increasing size of the mass, accompanied by ulceration and a clear discharge, which began in the final two months of her pregnancy and persisted for two months postpartum. There was no history of bleeding or sexual difficulties, and her menstrual cycles were regular with normal flow. Upon local examination, a well-circumscribed, pedunculated mass measuring 12 cm × 7 cm was observed (see Figures 1, 2, and 3). The mass was non-tender, with a fleshy, soft, and spongy consistency. The overlying ulcer measured 5 cm × 1 cm, with the floor showing unhealthy pale granulation tissue. Bilateral inguinal lymph nodes were not palpably enlarged. Following informed written consent, the patient underwent wide local excision of the mass along with the surrounding tissue under general anesthesia. Histopathological examination revealed the diagnosis of aggressive angiomyxoma. **Conclusion** Aggressive angiomyxoma constitutes a key differential diagnosis when evaluating vulvovaginal masses in female patients. Given the tumor's documented propensity for local recurrence, precise diagnosis and comprehensive management strategies utilizing a multidisciplinary approach are paramount in optimizing patient care.

Key words: Aggressive angiomyxoma, Mesenchymal tumor, Vulvar tumor.

INTRODUCTION:

The primary goal of this case report is to improve our ability to suspect aggressive angiomyxoma. This will, in turn, enhance our understanding of the tumor's characteristics and the appropriate follow-up after surgery.

The term "aggressive" refers to its ability to cause local invasion and infiltration of perivaginal and pararectal tissues. [1, 2, 3]It is an uncommon, slowly-growing benign vulvovaginal mesenchymal tumor. Studies have demonstrated a local recurrence rate ranging from 30% to 72% for this entity. [4, 5] The pathology of this tumor is undecided. [6]in the majority of cases, chromosomal abnormalities have often been discovered.

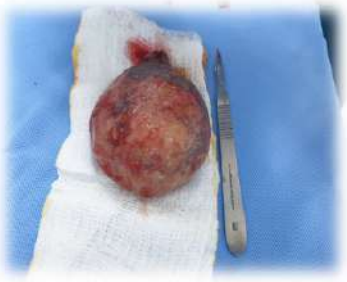
This neoplasm exhibits hormonal dependence, with anticipated growth during pregnancy and potential responsiveness to hormone modulation. Its occurrence is predominantly observed in women of reproductive age, demonstrating a peak prevalence within the third to fifth decades of life. [7] Patients with severe angiomyxoma have nonspecific symptoms that lead to misdiagnosis. [8]

Due to the infrequent occurrence of aggressive angiomyxoma (AA), approximately 80% of cases are initially misdiagnosed as alternative entities, including Bartholin's cysts, lipomas, hernias, fibroepithelial stromal polyps, or labial cysts. This diagnostic ambiguity often results in suboptimal and delayed therapeutic interventions, negatively impacting patient outcomes. [9] The magnetic resonance is an important tool for differential diagnosis. [10]

Histopathological findings in some cases indicate the presence of progesterone and estrogen receptors in these tumor cells, suggesting a hormone-dependent growth mechanism. Consequently, gonadotropin-releasing hormone (GnRH) agonists are being explored as adjuvant therapeutic options. Despite its infiltrative properties and potential for local recurrence, aggressive angiomyxoma (AA) generally has a favorable prognosis. [10, 11]Due to its locally aggressive behavior, aggressive angiomyxoma (AA) requires a cautious therapeutic strategy and long-term surveillance. Wide surgical excision with histologically clear margins is the most efficacious treatment option, but is often associated with substantial patient morbidity. Furthermore, adjuvant therapy employing gonadotropin-releasing hormone (GnRH) agonists has demonstrated efficacy in preventing recurrence. [8] We present a case of aggressive angiomyxoma (AA) involving the vulva, which was successfully managed via surgical excision.

Figures 1

Figures 2



Figures 3

Aggressive angiomyxoma of the right labia majora

DISCUSSION:

Although aggressive angiomyxoma (AA) is a benign neoplasm, it exhibits locally aggressive behavior similar to malignancies and is frequently misdiagnosed by clinicians prior to surgery. Thus, the presence of any spongy vulvar mass in women in their second to fourth decades of life requires a high index of clinical suspicion. The designation "aggressive" underscores its tendency for local recurrence and infiltrative nature, despite its benign pathology. [12]

Patients typically present with a progressively enlarging mass, often accompanied by symptomatic manifestations. These may include dull, aching pain, and urinary symptoms such as dysuria, urinary retention, or dyspareunia. The tumor's dimensions can range from 10 cm to 40 cm in its largest diameter. The pathogenesis of this neoplasm remains incompletely understood. Recent hypotheses suggest a correlation between translocation of chromosome 12 (12q13-15) and aberrant expression of the high-mobility group protein isoform 1-C (HMG1-C). [13]

Wide surgical excision with histologically clear margins is considered the most efficacious therapeutic intervention for aggressive angiomyxoma (AA). In addition, hormonal agents, including raloxifene, tamoxifen, and gonadotropin-releasing hormone (GnRH) agonists, have been suggested as adjuvant therapies for chronic and recurrent cases. The postoperative local recurrence rate for aggressive angiomyxoma (AA) is documented to range from 30% to 40%, with recurrences potentially occurring 10 to 15 years after the initial surgical procedure. [14] Long-term vigilant surveillance, utilizing computed tomography (CT) or magnetic resonance imaging (MRI), is recommended for high-risk cases.

CONCUSION:

This case illustrates that aggressive angiomyxoma (AA) can manifest across a broad spectrum of age groups, exhibiting variable tumor morphology and dimensions. The presentation of a painless vulvar mass should prompt consideration of aggressive angiomyxoma (AA) in the differential diagnosis. The primary therapeutic approach consists of wide surgical resection with histologically negative margins. Furthermore, adjuvant therapies may be indicated for residual or recurrent disease. Extended follow-up with magnetic resonance imaging (MRI) or computed tomography (CT) is advised in select cases. Enhanced understanding of this rare entity is essential for the development of more efficacious treatment strategies. Currently, a multidisciplinary approach is necessary for optimal patient management.

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