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AGGRESSIVE ANGIOMYXOMA OF THE VULVA WITH TORSION: A RARE CASE REPORT

Obstetrics & Gynaecology

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ABSTRACT

Background: Aggressive angiomyxoma is a rare, benign but locally aggressive mesenchymal tumor commonly involving the pelvis and perineum in women of reproductive age. Vulval presentation as a pedunculated mass is uncommon and often misdiagnosed clinically. **Case Presentation:** A 40 year old parous woman presented with a rapidly enlarging vulval mass of four weeks duration. Examination revealed a 13 × 10 × 5 cm soft, non tender, pedunculated mass arising from the left labia majora with a broad pedicle. A clinical diagnosis of pedunculated lipoma was made and the mass was excised. Gross examination showed a soft mass with yellowish gelatinous cut surface. Histopathology revealed spindle to stellate shaped cells with mild nuclear atypia, congested blood vessels, hemorrhagic necrosis, dense mononuclear infiltrate and scattered collagen bundles, confirming aggressive angiomyxoma with torsion. **Conclusion:** Aggressive angiomyxoma should be considered in the differential diagnosis of rapidly growing vulval masses. Histopathological evaluation is essential for diagnosis, and surgical excision with long term follow up is recommended due to the risk of local recurrence

KEYWORDS

Aggressive Angiomyxoma, Vulva, Pedunculated Vulval Mass, Lipoma Mimic, Case Report

INTRODUCTION

Aggressive angiomyxoma is a rare, slow growing mesenchymal tumor that predominantly involves the vulvovaginal, pelvic and perineal regions.[1] It was first described by Steeper and Rosai in 1983.[2] The term aggressive reflects its tendency for infiltrative growth and frequent local recurrence rather than metastatic potential.[2] The tumor occurs most commonly in women of reproductive age and often expresses estrogen and progesterone receptors.[3] Consequently, it may increase in size during pregnancy and demonstrate responsiveness to hormonal therapy.[3] Given its locally aggressive behavior, appropriate surgical management and long term follow up are essential. Various treatment modalities have been employed for recurrent disease with variable outcomes, and no single approach has been established as clearly superior.[4]

Case Presentation

A 40 year old parous woman with a history of three full term home deliveries and tubectomy presented with a vulval growth of four weeks duration.

The mass was initially pea sized and showed rapid progression in size. There was no history of bleeding per vagina or menstrual irregularities. Local examination revealed a 13 × 10 × 5 cm soft, non tender, pedunculated mass arising from the left labia majora with a broad pedicle measuring 1.5 × 4 cm. The overlying skin was normal.

A provisional diagnosis of pedunculated lipoma was made, and the mass was excised at the base of the pedicle. Gross examination showed a soft mass with a yellowish gelatinous cut surface.

Histopathological examination revealed extensive areas of hemorrhagic necrosis, scattered spindle to stellate shaped cells with mild nuclear atypia arranged in fascicles, congested blood vessels, dense mononuclear inflammatory infiltrate and scattered collagen bundles, consistent with aggressive angiomyxoma with torsion.

DISCUSSION

Myxoid tumors constitute a vast array of lesions characterized by degrees of extracellular myxoid matrix.[5] In general, angiomyxomas are classified as either superficial (also referred to as cutaneous myxoma) or Aggressive angiomyxoma (Deep)[6]. Superficial angiomyxoma predominantly affects middle-aged adults and can manifest in superficial tissues throughout the body, but mostly it occurs in the trunk, lower extremities, and head and neck regions [7], The stroma is made up of mostly edema with little myxoid material[8]. On the other hand, Evidence suggests that AAM affects almost exclusively the genital, perineal, and pelvic regions in women of reproductive age, implicating especially the vulva[11] .

Aggressive angiomyxoma typically displaces adjacent organs rather than directly invading them however its locally infiltrative behavior accounts for the term aggressive reflecting its propensity for local invasion and frequent recurrence after excision. The tumor is generally regarded as non metastasizing although rare cases of multiple metastases have been reported in women initially treated with surgical excision who later succumbed to metastatic disease. Over time this infiltrative growth may result in a large mass occupying the abdominopelvic cavity and approximately one fourth of these tumors are pedunculated. [3,9,10]

The prevailing theory regarding AAM pathogenesis suggests that the lesion originates from a primitive multipotent mesenchymal cell found in the lower female genital tract, with the capacity for diverse differentiation pathways.[12] Molecular studies have identified a characteristic clonal aberration involving chromosome 12 at the 12q13–15 region with rearrangement of the HMGIC gene high mobility group protein isoform I C which has been associated with aggressive angiomyxoma.[29] These findings place aggressive angiomyxoma within the benign spectrum of mesenchymal tumors characterized by involvement of multiple chromosomal aberration regions[9,30]

Clinically, aggressive angiomyxoma may be mistaken for a Bartholin cyst, lipoma, labial cyst, Gartner duct cyst, levator hernia, or sarcoma. In addition, fibro epithelial stromal polyp, superficial angiomyxoma, angiomyofibroblastoma, cellular angiofibroma, and smooth muscle tumors should be considered in the differential diagnosis of a polypoidal mass in the perineum. Aggressive angiomyxoma is characteristically infiltrative, whereas angiomyofibroblastoma is well circumscribed, a distinction that can also be appreciated on magnetic resonance imaging. Furthermore, aggressive angiomyxoma typically shows fewer but thick walled vessels, compared with the numerous thin walled vessels seen in angiomyofibroblastoma.[20]

Microscopically the tumor is composed of spindle and stellate shaped cells embedded in a myxoid stroma containing delicate wavy collagen fibrils. Numerous vascular channels of varying calibers are present. The tumor cells show abundant wispy eosinophilic cytoplasm with bland nuclei and there is no cytological atypia atypical mitotic activity or evidence of coagulative tumor cell necrosis[14].

Recurrent tumors usually show similar histological characteristics.

Diagnostic problems may arise when the pathologist is dealing with uncommon morphological features, deposition with or without hyalinized blood vessels, or neurofibroma-like appearance [15]

The vast majority of these neoplasms exhibit positivity for estrogen and progesterone receptors, indicating that AA is likely a hormone-dependent tumor. This is supported by observations of rapid growth and recurrence during pregnancy[15] .Hormonal correlation suggests that antihormonal therapy (e.g., Tamoxifen), gonadotrophin-releasing hormone (GnRH) agonist, or aromatase inhibitors could be considered feasible emerging options in AA treatment[17]. Moreover, a neo-adjuvant treatment to reduce tumor size before surgery, facilitating complete excision, or an adjuvant approach for incompletely resectable/residual mass could be investigated[9]. However, the adverse effects of long-term use of the GnRH agonist (e.g., menopausal symptoms and bone loss) and tumor regrowth after drug interruption do not allow us to approve it as a best choice of treatment[17]

Wide surgical excision with tumor free margins remains the cornerstone of management however long term follow up is essential because of the high recurrence[16].

Imaging plays a crucial role in defining the true extent of the tumor guiding surgical planning and detecting recurrence. Contrast enhanced MRI particularly with gadolinium is the preferred imaging modality for diagnosis especially on T2 weighted sequences where the lesion typically appears hyperintense relative to muscle,ultrasound can be useful in follow-up monitoring as well [18]

Preoperative angiographic embolization may also aid subsequent surgical resection by reducing tumor size and facilitating clearer delineation of the lesion from surrounding normal tissues[19]

In summary aggressive angiomyxoma should be considered in the differential diagnosis of soft tissue tumors arising in the vulvovaginal region perineum or pelvis as early recognition and appropriate surgical management (wide excision possibly with free margins) key determinants of prognosis.

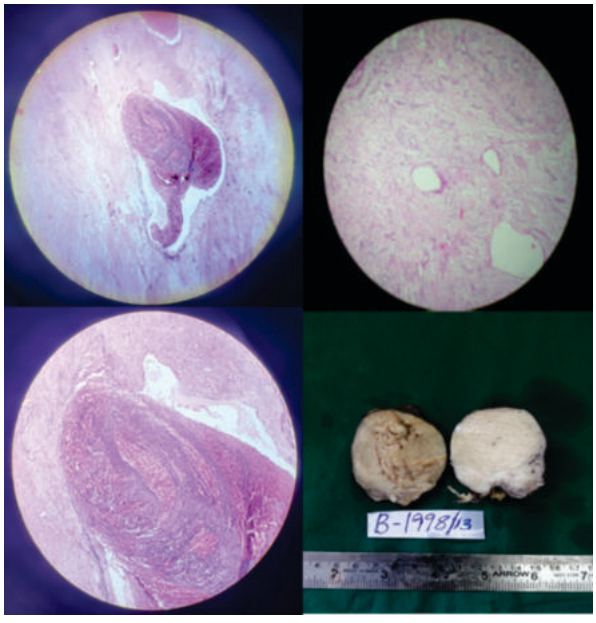

Figure - 1

- Microscopy (multiple sections) showed:
- Extensive areas of hemorrhagic necrosis
- Scattered spindle to stellate-shaped cells with mild nuclear atypia arranged in fascicles
- Congested blood vessels, dense mononuclear infiltration, and scattered collagen bundles

CONCLUSION

Aggressive angiomyxoma should be considered in the differential diagnosis of rapidly enlarging vulval masses. Definitive diagnosis relies on histopathological examination, as clinical features may mimic benign lesions. Early surgical excision and long term follow up are essential due to the locally aggressive nature and risk of recurrence.

Informed Consent

Written informed consent was obtained from the patient for publication of this report and associated images.

Conflict of Interest

The authors declare no conflicts of interest.

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