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VULVAL ANGIOMYXOMA : A CASE REPORT

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KEYWORDS :

INTRODUCTION :

Vulvar diseases make up only a small portion of cases seen in gynecological practice, with benign tumors comprising 22% of all vulvar conditions[1]. Angiomyxomas are rare benign, slow-growing tumors that arise from mesenchymal cells and are distinguished by a prominent myxoid matrix of connective tissue and abundant blood vessels[2]. There are two phenotypes of angiomyxomas: superficial (also known as cutaneous myxoma) and aggressive angiomyxoma[3].

Superficial angiomyxoma (SAM) is an uncommon benign soft tissue tumor infrequently observed in the genital region, with only a handful of cases involving the vulva reported to date[4]. Due to its rarity, the prevalence in the population is unknown, complicating management and counselling efforts. SAM are more frequently found in middle-aged patients, typically presenting as a single nodule or a polypoidal mass in the head and neck region, trunk, and lower extremities. They are often mistaken for skin tags, cysts, or neurofibromas and are commonly associated with Carney's complex (a triad of cardiac myxomas, spotty pigmentation, and endocrine overactivity)[5].

Aggressive angiomyxoma (AAM) is known for its localized aggressiveness, primarily affecting the connective tissue of the perineum or lower pelvis. It predominantly occurs in women of reproductive age[6]. The term "aggressive" emphasizes its potential for infiltrative behavior. However, AAM is not inherently aggressive, as it rarely metastasizes and its histologic features are less aggressive compared to the more common vulvar carcinoma, resulting in a considerably better prognosis[7]. The most frequently involved anatomical sites include the pelvis, perineum, vulva, vagina, and bladder[8].

2. Presentation of case :

A 30-year-old, Female, married since 8 years, Parity 1,living issue 1 with Previous 1 LSCS and last child birth 5 years back came to Gynecology Out Patient Department (OPD) with a swelling on the left labium majora for last 4 years, slowly growing in size not associated with any ulceration or discharge. Past medical and family history was unremarkable. On general examination, the patient was moderately built and afebrile. There was no evidence of jaundice, anemia, cyanosis, lymphadenopathy, clubbing, weight loss, or any bowel and bladder function alterations. Her menstrual cycles were regular with normal flow. Her abdomen was soft and nontender.

On local examination, a well-circumscribed, 6.8□x 5.9 x 4.8 cm pedunculated mass seen arising from left labium majora at 2 o'clock position with a stalk of 6 x 2 cm. On palpation, the mass was nontender, nonreducible, and soft in consistency(Fig. 1). No inguinal lymphadenopathy was seen. On gynecological examination, the uterus was normal in size with a healthy cervix and vagina. Her baseline investigations were normal.

In Ultrasonography (USG) abdomen and pelvis, no significant abnormality detected. USG Local done suggestive of "a polypoidal outgrowth arising from vulva measuring 4.9 x 5.6 x 5.8 cm, seen attached to vulva by a stalk. The stalk shows presence of internal vascularity within. There are few anechoic cystic spaces within the polypoidal outgrowth. There is a presence of internal vascularity within the lesion. Features are suggestive of neoplastic etiology, HPR correlation for further evaluation". A CECT scan or Magnetic resonance Imaging (MRI) could not be performed due to the patient's financial constraints.

3. Surgical Procedure:

After taking written valid informed consent, local excision of the mass and surrounding margins was done under spinal anesthesia. Hemostasis checked and achieved. The specimen was sent for histopathological examination.

On gross examination; the mass was greyish brown, soft and on cut section, it was homogenous, whitish with myxoid areas. Histopathology revealed an unencapsulated hypocellular myxoid tumor. The tumor cells were spindle to stellate with delicate eosinophilic bipolar cell processes and bland ovoid to rounded nuclei. The tumor stroma was myxoid with scattered delicate collagenous fiber with scattered medium to large sized blood vessels with focal hyalinization of the vessel wall. No evidence of atypia/mitosis/ necrosis ,(Fig. 2). Her postoperative recovery was uneventful. On the third day post-surgery, the patient was sent home in a vitally stable condition.

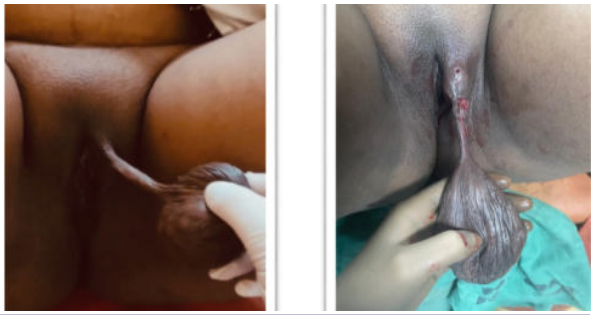

Fig. 1 Local examination of pedunculated soft mass from left labia majora (a) Pre operative (b) Intra operative

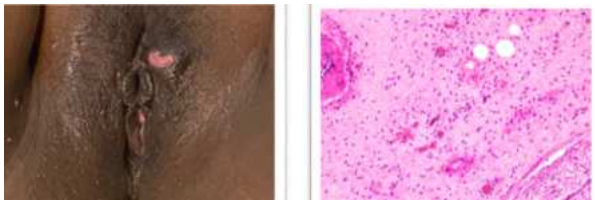

Fig. 2 (a) Post operative follow up after local excision of mass (b) HPR findings seen in angiomyxoma[8]

DISCUSSION :

Vulvar angiomyxomas are uncommon and often misdiagnosed as their clinical, radiographic, and histologic features are nonspecific.

The prevailing theory regarding the pathogenesis of angiomyxoma is that it originates from a primitive, multipotent perivascular mesenchymal cell in the lower female genital tract, which has the potential to differentiate in various ways and give rise to the tumor. Molecular studies have identified a recurrent clonal abnormality on chromosome 12, specifically in the 12q13-15 region, linked to rearrangements involving the high mobility group protein isoform I-C.[9]

There are two phenotypes of angiomyxoma: superficial and aggressive. Due to the rarity of angiomyxomas and the lack of distinct signs and symptoms, both Superficial and Aggressive phenotypes are often diagnosed only on histological examination following surgical resection. A different vascular pattern and involvement of the deeper tissues differentiate Aggressive angiomyxoma from the Superficial phenotype.[10]

Macroscopically, aggressive angiomyxoma appears as a soft, well-defined, occasionally polypoid mass, ranging from a few centimeters to over 20 cm in size and is characterized by a shiny, homogeneous, gelatinous appearance on gross examination. Microscopically, the tumor consists of widely scattered spindle to stellate-shaped cells with poorly defined cytoplasm and small, round to oval, hyperchromatic nuclei with centrally located nucleoli, all set within a myxoid stroma. This stroma is rich in collagen and often contains hemorrhagic areas. A distinctive feature of this tumor is the presence of variably sized blood vessels, ranging from small, thin-walled capillaries to large vessels with secondary changes, such as perivascular hyalinization and medial hypertrophy.[11]

Aggressive angiomyxoma predominantly affects the pelvic-perineal region with the vulva being the most frequently involved site. These often express estrogen and progesterone receptors, making them particularly prevalent in women of reproductive age, and they can grow rapidly during pregnancy.[12] Aggressive angiomyxoma can affect deeper tissues of the pelvis and retroperitoneum, so the full extent of the tumor may be underestimated during an initial physical examination. While CT imaging is commonly used due to its availability, MRI offers superior spatial resolution for better delineation of soft tissue and anatomical details. MRI is more effective than CT for evaluating perineal and vulvar angiomyxomas, particularly in assessing the tumor's depth and its relationship to the pelvic floor, which can aid in distinguishing between superficial and aggressive angiomyxomas.[13]

Treatment of aggressive angiomyxoma typically involves wide local excision with clear margins due to its potential for local infiltration. In cases of deep invasion or positive margins following initial excision, radical surgery may be necessary, though it is associated with significant operative morbidity, including persistent discomfort, groin pain, urinary complications, and sexual dysfunction[12]. Adjuvant therapy with agents such as raloxifene, tamoxifen, or GnRH agonists like leuprolide acetate and goserelin has shown benefits in cases where the tumor is sensitive to estrogen and progesterone receptors.[14] Recurrences of aggressive angiomyxoma cannot be predicted based on tumor size or cellular activity. They are usually rare and have been reported to occur from several months to several years after initial excision.

CONCLUSION :

Genital angiomyxomas are rare and aggressive angiomyxomas which is a locally aggressive neoplasm should be considered in the differential diagnosis when a patient presents with a slow growing mass in the vulvovaginal region, perineum, or pelvis. Due to its tendency for local recurrence, early diagnosis and management including surgical excision and adjuvant therapy are crucial for effective treatment.

Conflicts Of Interest : There are no conflicts of interest

Abbreviations:

SAM : Superficial Angiomyxoma
AAM : Aggressive Angiomyxoma
LSCS : Lower Segment Caesarean Section
CECT : Contrast Enhanced Computed Tomography
MRI : Magnetic Resonance Imaging
USG : Ultrasonography

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