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A Rare Case of Recurrent Aggressive Vulvar Angiomyxoma

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Abstract: ***Aim and Background: Aggressive angiomyxoma is a rare tumor which is locally aggressive and is typically seen in pelvic and perineal regions of women. Case Description: A 45-year-old woman presented with a 6-year history of a gradually increasing vaginal mass, with previous history of Aggressive Angiomyxoma and local excision done in 2006 and 2013, is currently diagnosed as Recurrent Aggressive Vulvar Angiomyxoma. Histopathological Examination confirmed the diagnosis of Recurrent Aggressive Vulvar Angiomyxoma. Conclusion: This case emphasizes the necessity of including Aggressive Angiomyxoma in the differential diagnosis for vaginal masses and necessitates an assertive treatment approach to minimize the risk of recurrence. Clinical Significance: Aggressive Angiomyxoma is an uncommon tumor that necessitates swift identification and intervention to avert recurrence.***

Keywords: Recurrent aggressive vulvar Angiomyxoma, Aggressive Angiomyxoma, Recurrent vulvar tumor, vaginal mass, histopathology, Clinical management

1. Introduction

Aggressive Angiomyxoma is an uncommon tumor known for its local aggressiveness, primarily found in the pelvic and perineal areas of females [1]. It is known for its elevated recurrence rate and the possibility of causing local damage.

2. Case Description

A 45 year old primiparous women came with complains of mass per vagina since 6 years which was gradual in onset and progressive in nature. Initially the size was of a pea and over the years has grown to the current size. Size of the mass increases during defecation and strenuous exercises. No h/o pain abdomen/ menstrual irregularities/ inter menstrual spotting. Menstrual history- Nothing significant. Obstetric history- p1l1 1 previous vaginal delivery. History of similar complaints and was diagnosed as Aggressive angiomyxoma in 2006 and 2013 and local excision was done for the same. On examining the patient, vitals were stable, Per abdomen was soft and non tender. Local examination- 8*7*5 cm globular mass arising from the lateral vaginal wall and vulva, soft in consistency, Not reducible / compressible swelling, 4*4 cm keratinisation seen over inferior and right lateral surface of the mass (Figure-1). Per vaginal examination- uterus normal size, b/l fornices free and non tender. Blood investigations done and were normal. Usg abdomen and pelvis showed uterus of normal size, Endometrial Thickness- 10mm. Patient underwent Wide Local excision under spinal anaesthesia and sample was sent for histopathological examination (Figure-2).

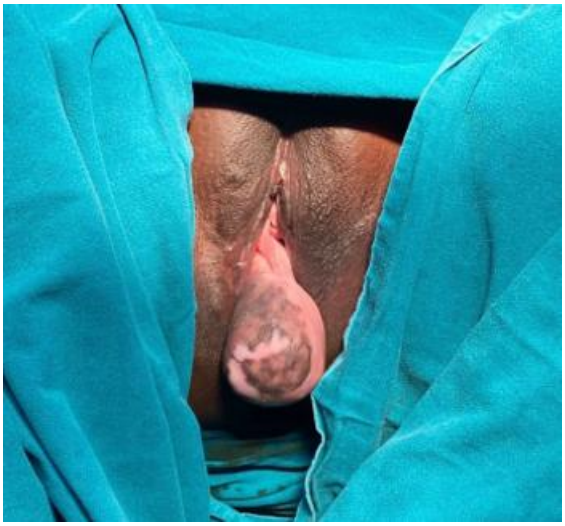

Figure 1

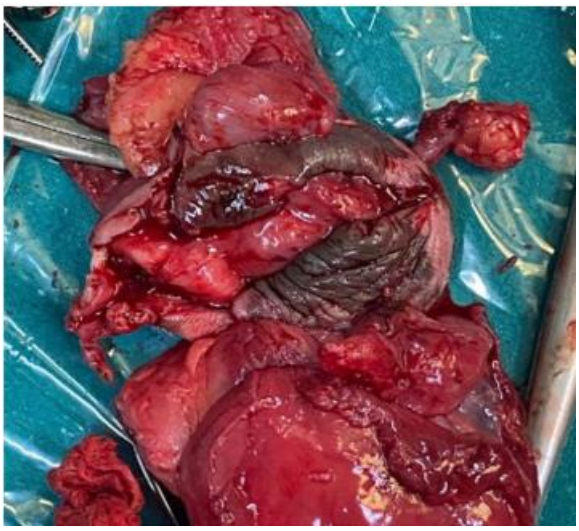

Figure 2

HistoPathological Examination reports showed- lesion which is not encapsulated and composed of predominantly spindle to stellar cells in a myxoid matrix with multi nuclear giant cells interspersed with many dilated vascular channels of venous caliber suggestive of Aggressive Vulvar

Angiomyxoma. Margins: free from the tumor (Figure- 3, 4, 5).

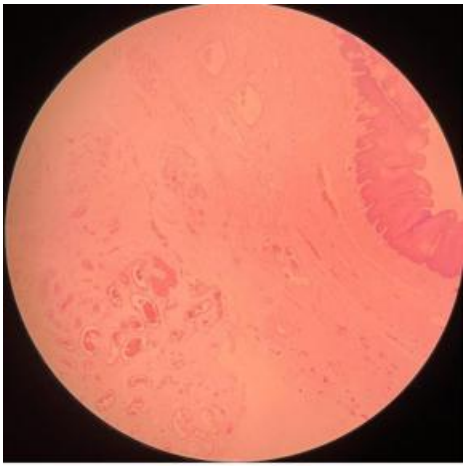

Figure 3 (4x)

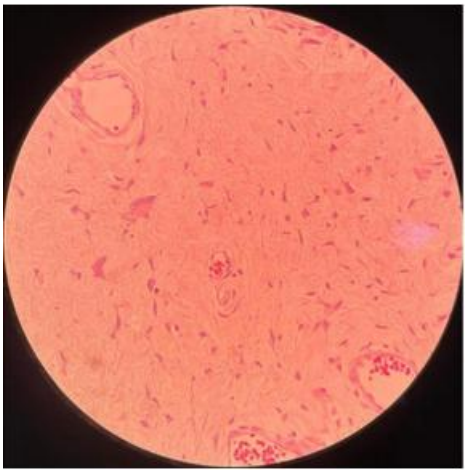

Figure 4 (10x)

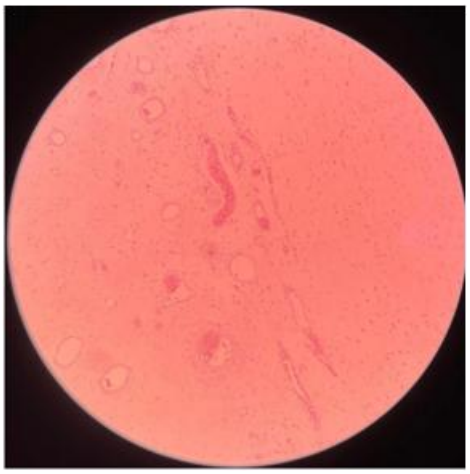

Figure 5 (40x)

Patient withstood the procedure and was discharged on 5th post operative day. This lady is under follow up and there is no recurrence till date.

3. Discussion

Aggressive angiomyxoma is an uncommon, slowly progressing yet locally invasive, non-encapsulated, estrogen-sensitive myxoid tumor [1, 2]. Initially described by Steeper and Rosai in 1983, it appeared as a vulval polyp in clinical evaluation and was confirmed through histological diagnosis. This condition arises in the genital, perineal, and pelvic areas of adult females, predominantly in their 30s, with the highest occurrence noted between the ages of 31 and 35 [1-2]. The tumor may recur as in our case.

The underlying cause of the condition remains unclear; however, a translocation has been identified in the 12q15 area of chromosome 12, resulting in abnormal expression of the high-mobility group protein isoform I-C (HMGI-C), which plays a role in DNA transcription [3]. Overall, these tumors present as soft, polyp-like or partially defined lesions. When cut, they exhibit a gelatinous look. At a microscopic level, they consist of numerous thick-walled blood vessels of different sizes arranged in a loose collagenous and myxoid stroma, interspersed with neoplastic cells that are spindle and stellate in shape. It demonstrates the presence of ER (oestrogen receptor) and PR (progesterone receptor), along with vimentin, desmin, SMA, and, to a lesser extent, CD34 [4]. Tumors that develop during pregnancy exhibit a quicker growth rate, which reinforces the link between estrogen and progesterone and this phenomenon [4-5]. The increase in tumor size during pregnancy and the positive nuclear staining for progesterone receptors indicate a potential hormonal (progesterone) dependency in certain instances of aggressive angiomyxoma [6].

The primary method of diagnosis is histopathological, although a CT scan or MRI might provide some indications. In the CT scan, angiomyxoma exhibits a clearly defined boundary and has lower attenuation than that of muscle. High signal images are seen in T2 weighted images of MRI. This phenomenon can be explained by the presence of a loosely organized myxoid matrix and the high-water content present in angiomyxoma [7, 8].

The recommended approach for treatment is extensive surgical removal. [8]. However, issues arise since there are recorded instances of recurrence, particularly after inadequate resections, as demonstrated in our case. In practice, fully removing the tumor is challenging since the texture of angiomyxoma closely resembles that of normal connective tissue, and they are also not encapsulated [9].

The reappearance of this tumor presents a complex issue. The rate of local recurrence ranges from 36% to 72% [1, 9.10]. Therapeutic approaches such as angiographic embolization of the tumor, hormonal treatments with Tamoxifen, Raloxifen, and Gonadotropin-releasing hormone agonists (GnRH-A) may be necessary in these situations, although they may have limited effectiveness [9, 10]. In certain documented instances, pre-operative GnRH treatment has been utilized to facilitate surgery and reduce the chance of recurrence. However, there is no agreement on their effectiveness in preventing recurrence. Therefore, an extensive local excision of the tumor should be performed during the initial procedure, disregarding cosmetic considerations, as we executed in our case to minimize the risk of further recurrence. Since recurrence can happen anywhere from 2 months to 15 years after the first resection, it is recommended to have long-term follow-up, with MRI being the preferred method of investigation [10, 11]. The patients should receive proper

counseling and encouragement for ongoing follow-up care. We have been monitoring our patient for the past year, and fortunately, she has not experienced a recurrence. Typically, the tumor does not spread to other parts of the body; however, there have been two documented instances where it resulted in metastasis to the lungs, peritoneum, and lymph nodes (including those in the aortic and iliac regions), ultimately causing the woman's death [12].

4. Conclusion

Aggressive angiomyxoma requires assertive treatment to avert recurrence. Wide surgical resection is the treatment of choice till date. GnRH analogues could be an option if there is another recurrence. The patient should be encouraged to commit to long-term follow-up, as recurrences can happen even after a decade to 15 years.

Clinical significance:
Aggressive Angiomyxoma is an uncommon tumor that necessitates swift identification and prompt intervention to avert recurrence.

List of abbreviations:
RAA- Recurrent Aggressive Angiomyxoma HPE- Histopathological Examination HMGIC- High Mobility Group protein gene Isoform C
SMA- Smooth Muscle Actin
CD 34- Cluster of Disfferentiation 34
GNRH- Gonadotropin-releasing hormone
GNRH A- Gonadotropin-releasing hormone agonists

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Figures and figure legends:

Figure 1: Local Examniation of Recurrent Aggressive Vulvar Angiomyxoma

Figure 2: Post operative image of Recurrent Aggressive Vulvar Angiomyxoma, Post wide local excision

Figure 3: Microscopic images of Recurrent Aggressive Vulvar Angiomyxoma in 4x magnifcation

Figure 4: Microscopic images of Recurrent Aggressive Vulvar Angiomyxoma in 10x magnifcation

Figure 5: Microscopic images of Recurrent Aggressive Vulvar Angiomyxoma in 40x magnifcation

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