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Case Report

Aggressive angiomyxoma of the vulva presenting as a massive perineal mass

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ABSTRACT

Aggressive angiomyxoma is a rare benign but locally infiltrative mesenchymal tumour that predominantly affects women of reproductive age. It commonly arises in the pelvis and perineal regions and is characterized by slow growth and a high recurrence rate. We report a case of a 39-year-old multiparous woman presenting with a progressively enlarging vulval swelling over 12 years. Imaging revealed a well-defined soft tissue lesion without infiltration of adjacent structures. Complete surgical excision was performed, and histopathological examination confirmed aggressive angiomyxoma. This case highlights the importance of considering aggressive angiomyxoma in the differential diagnosis of large vulval masses and emphasizes the role of complete surgical excision and long-term follow-up.

Keywords: Aggressive angiomyxoma, Vulval mass, Perineal tumour, Rare vulval tumour, MRI pelvis, Surgical excision, Myxoid tumour, Case report

INTRODUCTION

Aggressive angiomyxoma is a rare soft tissue tumour first described by Steeper and Rosai in 1983.[1] It typically occurs in women of reproductive age and arises in the pelvis, perineum, and vulvovaginal region.[2] Despite being histologically benign, it is locally aggressive and has a high recurrence rate.[3] Because of its slow-growing nature and nonspecific presentation, it is often misdiagnosed as Bartholin cyst, lipoma, or vulval abscess.[4,5] We present a case of a massive vulval aggressive angiomyxoma in a multiparous woman with a long history of progressive swelling.

CASE REPORT

A 39-year-old female, para 4, live issue 4, presented with complaints of progressively enlarging swelling in the vulval region for the past 12 years. The swelling was slow-growing, painless, and gradually increased in size over time. The patient reported discomfort and heaviness due to the mass but denied urinary or bowel complaints.

There was no significant menstrual, medical, surgical, or family history. Her uterus and ovaries were normal on evaluation.

Clinical examination

On examination, a large cystic mass measuring approximately 18×20 cm was noted arising from the right vulva. The swelling had both cystic and solid components, with areas of necrosis and ulceration. The mass was non-tender and slow growing (Figures 1a and b).

Imaging findings

Magnetic resonance imaging (MRI) of the pelvis revealed a large, well-defined soft tissue lesion arising from the right vulval and perineal region. The mass appeared predominantly hyperintense on T2-weighted images with internal heterogeneous areas suggestive of myxoid content and focal solid components. The lesion measured approximately 18×20 cm and extended inferiorly into the perineal region, producing a large exophytic mass (Figure 2-5).

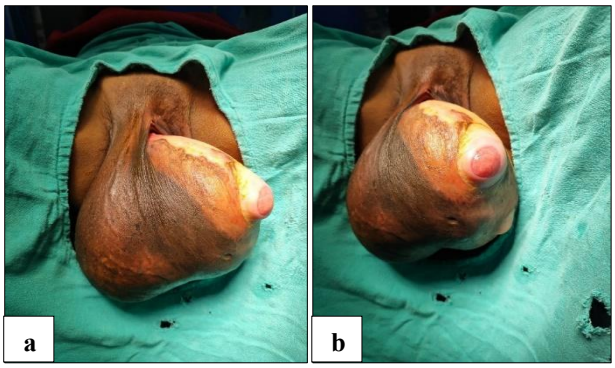

Figure 1 (a and b): Pre-operative picture during clinical examination.

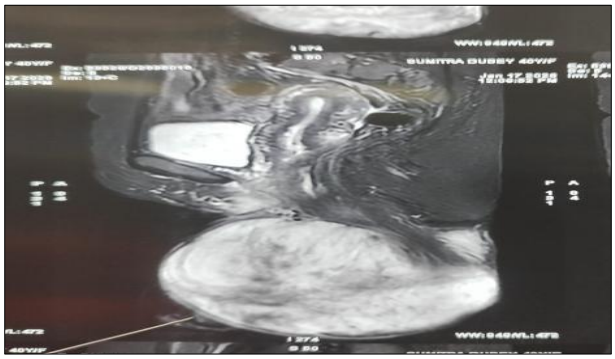

Figure 2: MRI image with pointer pointing towards mass with no evidence of infiltration into surrounding structures.

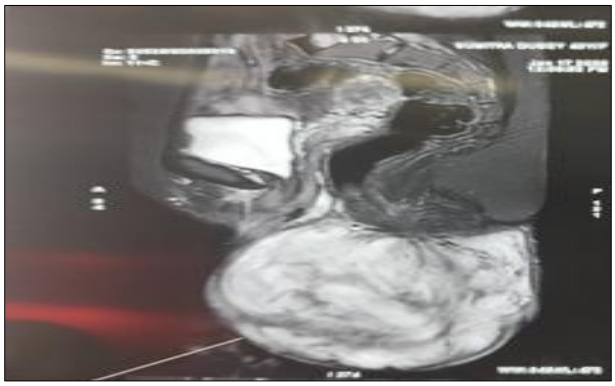

Figure 3: MRI findings.

The mass caused displacement of adjacent pelvic structures including the vagina, urinary bladder, and rectum without evidence of invasion or infiltration. The lesion demonstrated well-defined margins with preserved fat planes surrounding the mass.

No involvement of pelvic musculature, pelvic organs, or bony structures was noted. There was no evidence of lymphadenopathy. These imaging features were suggestive of a benign but locally expansive lesion, with aggressive angiomyxoma considered as a likely differential diagnosis.

Figure 4: MRI findings.

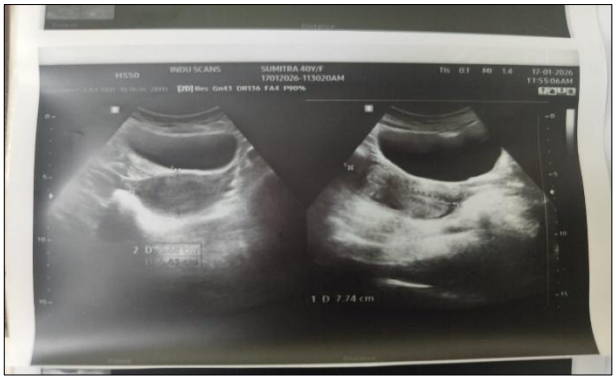

Figure 5: USG showing mass.

Management

The patient underwent complete surgical excision of the vulval mass under appropriate anesthesia. Intraoperatively, a large, well-defined mass arising from the right vulval region was identified, measuring approximately 18×20 cm, with both cystic and solid components and areas of necrosis and ulceration. Careful dissection was performed to separate the tumour from surrounding tissues, preserving adjacent structures.

The mass was excised completely, and haemostasis was achieved. The excised specimen weighed approximately 1.8 kg and was sent for histopathological examination. The postoperative period was uneventful, and the patient recovered well with symptomatic relief from discomfort and heaviness (Figures 6 and 7).

Histopathological findings

Gross examination revealed an unoriented, single, white, globular, partially skin-covered firm piece of tissue measuring 20.0×17.0×8.0 cm, along with a skin flap measuring 17.5×14.0 cm. All resected skin margins appeared grossly uninvolved; however, the base was seen to be formed by the tumor. A white area measuring 3.5×2.0 cm and an ulcerated area measuring 2.0×2.0 cm was identified. On serial sectioning, the cut surface of the mass appeared homogenous white with focal myxoid areas (Figure 8).

Figure 5: Post operative picture after reconstruction.

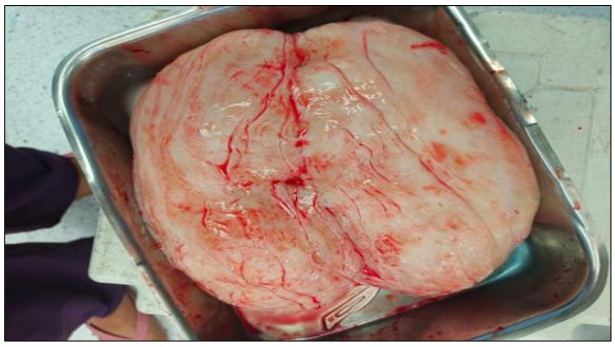

Figure 6: Gross image after cutting the perineal mass.

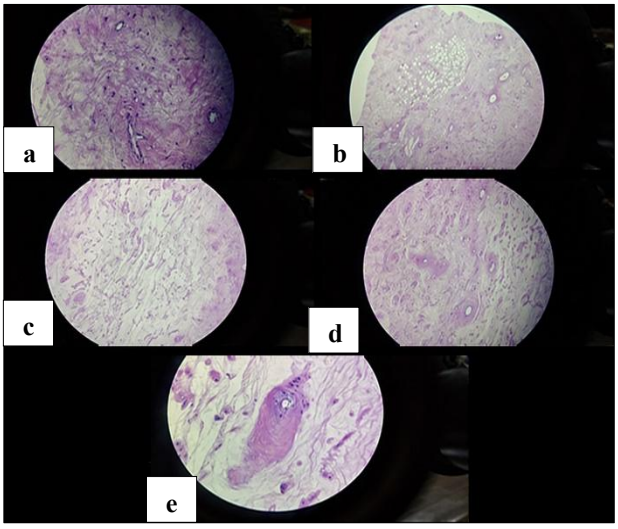

Figure 8: (a) Low-power view showing ill-defined subepithelial myxoid lesion with hypocellular stroma (H and E), (b) low-power view (40×) demonstrating infiltrative margins and focal fat entrapment (H and E), (c) tumour cells dispersed within loose myxoid background with delicate collagen fibers (H and E), (d) intermediate magnification (100×) showing numerous thin-walled blood vessels of varying caliber within myxoid stroma (H and E), (e) high-power view (200×) demonstrating spindle-to-stellate tumour cells with bland nuclear features embedded in abundant myxoid stroma (H and E).

Microscopic examination showed tissue lined by stratified squamous epithelium with an ill-defined subepithelial lesion composed of scattered spindle to stellate-shaped tumor cells embedded in abundant myxoid stroma. The tumor cells exhibited oval to spindle-shaped nuclei with bland chromatin, inconspicuous nucleoli, and moderate amounts of delicate cytoplasm. The stroma demonstrated delicate collagen fibers with numerous interspersed thin-walled blood vessels of variable size. Deeper sections showed focal fat entrapment. No areas of hemorrhage, necrosis, lymphovascular invasion, or perineural invasion were identified. Sections from the white and ulcerated areas showed similar tumor morphology. Tumor cells were present at the base, while sections from skin margins were free of tumor cells.

These histomorphological features were suggestive of aggressive angiomyxoma of the right labia. Immunohistochemistry with Desmin, ER, PR, HMGA2, and CD34 was advised for confirmation.

DISCUSSION

Aggressive angiomyxoma is a rare mesenchymal tumor that predominantly affects women of reproductive age and typically arises in the pelvis, perineum, and vulvovaginal region.[6] Although histologically benign, it is characterized by locally infiltrative growth and a high risk of recurrence, making early diagnosis and appropriate management essential.[7,8] Due to its slow-growing nature and nonspecific clinical presentation, aggressive angiomyxoma is frequently misdiagnosed as more common benign lesions such as Bartholin cyst, lipoma, vulval abscess, or hernia.[9,10]

The typical presentation includes vulval swelling, perineal mass, slow progressive growth and minimal pain.

In the present case, the patient presented with a progressively enlarging vulval mass over 12 years, consistent with the indolent nature of this tumor. The absence of pain and systemic symptoms often contributes to delayed presentation, allowing the lesion to attain a considerable size before diagnosis.[11] Our patient developed a massive perineal mass measuring approximately 18×20 cm, which caused discomfort and heaviness but no urinary or bowel symptoms. Such presentations have been reported in the literature and highlight the importance of maintaining a high index of suspicion in cases of long-standing vulval swelling.

Imaging plays a crucial role in the evaluation of aggressive angiomyxoma. MRI is considered the imaging modality of choice, typically demonstrating a well-defined mass with high signal intensity on T2-weighted images due to the myxoid matrix.[12,13] In this case, MRI revealed a large, well-defined lesion displacing adjacent pelvic structures without infiltration, supporting a benign but locally expansive lesion. These findings were helpful in surgical planning and assessment of tumor extent.

Histopathological examination remains the gold standard for diagnosis. The characteristic features include spindle to stellate cells embedded in a myxoid stroma with numerous thin-walled blood vessels and minimal cytological atypia.[14] These findings were consistent with aggressive angiomyxoma in the present case. Immunohistochemistry is often useful for confirmation, with many tumors showing positivity for estrogen and progesterone receptors, supporting hormonal responsiveness.

Complete surgical excision remains the treatment of choice. However, recurrence rates ranging from 30% to 70% have been reported due to the infiltrative nature of the tumor.[15] Hormonal therapy using GnRH analogues has also been used in selected cases, particularly in recurrent or unresectable tumors. Therefore, long-term follow-up is essential to detect recurrence early.

CONCLUSION

Aggressive angiomyxoma is a rare tumour of the vulva that may grow to a large size before diagnosis. Early recognition and complete surgical excision are essential. Due to the high risk of recurrence, long-term follow-up is necessary.

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