A Case Report of Aggressive Angiomyxoma of Vulva: An Often Misdiagnosed Neoplasm

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ABSTRACT
Aggressive angiomyxoma of vulva (AAM) is a very rare, slow growing, locally aggressive, and rarely metastasizing soft tissue neoplasm. Its preoperative diagnosis is often difficult. It has a high recurrence rate and requires prolonged follow-up. We report a case of a 36 years old female, who presented with a swelling on her left labium majus. Imaging studies could not confirm the diagnosis. She underwent wide local excision of the lesion and her final histopathology report was aggressive angiomyxoma of vulva. Patient has been in follow-up for the past four years with serial MRI without any recurrence.

CASE REPORT
A 36-year-old female presented with a slow growing painless swelling on her left labium majus of two years duration. Other tumour related history and obstetric, menstrual, social and family histories were unremarkable. On examination, a 6 cm x 5 cm soft compressible globular swelling was observed. Regional lymph nodes were clinically negative [Table/Fig-1]. Ultrasound of the genital region detected a hypoechoic lesion with ill-defined margins and without intraperitoneal communication. Doppler imaging showed randomly dispersed blood vessels. MRI detected a hyperintense lesion on T2 weighted MRI extending into the ischiorectal fossa. FNAC was deferred as haemangioma was an imminent possibility. The differential diagnoses at this juncture were Bartholin’s cyst, lipoma of the labia, and haemangioma. The tumour was excised with adequate margins. A solid, fleshy mass 8 cm x 4 cm x 10cm in dimensions extending into the ischiorectal fossa was removed [Table/Fig-2,3]. Histopathological examination was suggestive of aggressive angiomyxoma, showing hypocellular population of spindle shaped cells in a loose myxoid matrix with no cellular atypia. Thick walled vessels of varying calibre were noted. The cells stained positive for Oestrogen Receptor (ER) and vimentin. Since all margins were negative, no further treatment was given. She is under active surveillance with serial MRI for the past four years with no recurrence.

DISCUSSION
AAM is a very rare benign myxoid tumour. The challenge of diagnosing AAM, even histopathologically, cannot be emphasised enough. A wide range of other mesenchymal...
lesions occur commonly in this region and mimic AAM to a variable extent. The term “Aggressive Angiomyxoma” is of recent coinage, first reported in 1983 by Steeper and Rosai [1]. “Aggressive” denotes the local aggressiveness of the tumour. It has been classified by WHO as deep angiomyxoma and is considered a tumour of uncertain origin. Around 250 cases have been reported to date [2].

They have an indolent course occurring commonly in women of reproductive age during the 3rd decade of life, although rare cases have been reported in perimenopausal women, children and men [3,4]. A few cases have been reported during pregnancy with the tumour growing rapidly thus indicating their hormone dependency [5]. They commonly occur in the perineum and pelvis; in perineum they present as a soft painless mass. The other symptoms are dull aching pain, paraesthesia of the overlying skin, urinary retention and dyspareunia.

Preoperatively, they are misdiagnosed in > 80% of patients. Extensive imaging is necessary prior to surgery not only to aid in diagnosis, but also to find the extent of disease. On ultrasound, these lesions appear as hypoechoic cystic or solid masses. Plain CT shows a low density mass with attenuation less than surrounding muscle. Contrast enhanced CT shows a mildly enhancing mass with an internal swirling pattern [6]. MRI can delineate the perineal and pelvic extent of the tumour. Diffusion weighted MRI is both diagnostic and prognostic. In T2 weighted imaging, the tumour shows a swirled architecture and high signal intensity; the loose myxoid stroma with high water content is responsible for this appearance [7]. MRI is even more useful during follow-up for detecting early recurrence.

On gross examination, they appear as soft bulky masses with smooth external surface. Histological examination shows monotonous, sparsely arranged spindle or stellate shaped fibroblasts or myofibroblasts in a myxoid stroma. Mitotic figures are rare, indicating low proliferation activity. The margins can be infiltrative extending into adjacent muscles, adipose tissue, and rarely vessels. Vessel infiltration explains the isolated cases of metastasis.

**Table/Fig-4:** Differential diagnoses of aggressive angiomyxoma of vulva

<table>
<thead>
<tr>
<th>Clinical features</th>
<th>Macroscopic appearance</th>
<th>Microscopy</th>
<th>Other</th>
<th>Malignant potential</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aggressive angiomyxoma</td>
<td>Most common in 3rd decade of life;</td>
<td>Soft, bulky masses with smooth surface</td>
<td>Spindle or stellate shaped fibroblasts or myofibroblasts in a myxoid stroma</td>
<td>High recurrence rate; CD34, ER, PR, vimentin positive; S100 negative</td>
</tr>
<tr>
<td>Bartholin's cyst</td>
<td>All age groups; painful if large; Swelling adjacent to opening of Bartholin’s duct</td>
<td>Retained cyst material</td>
<td>Cysts lined by squamous or urothelium with inflammatory infiltrates; Residual mucinous glands seen</td>
<td>Treated by marsupialisation</td>
</tr>
<tr>
<td>Lipoma</td>
<td>More common in adults; rare in children</td>
<td>Bright yellow homogenous appearance; greasy cut surface</td>
<td>Well encapsulated adipose tissue without atypia</td>
<td>1-4% recur</td>
</tr>
<tr>
<td>Superficial angiomyxoma</td>
<td>More common in males than females; 4th decade of life</td>
<td>Greyish white, sometimes haemorrhagic appearance</td>
<td>Thin walled vessels; non-infiltrative margins; Otherwise similar to AAM</td>
<td>Associated with Carney’s syndrome; rarely recurrent</td>
</tr>
<tr>
<td>Angiomyofibroblastoma</td>
<td>Females, 4th &amp; 5th decade</td>
<td>Pedunculated mass with pink grey cut surface; no necrosis</td>
<td>Well encapsulated, numerous hylanized vessels with plump stromal cells around vessels</td>
<td>Vimentin, desmin, ER &amp; PR positive; Very weak CD34 positive</td>
</tr>
<tr>
<td>Myxoid liposarcoma</td>
<td>3rd or 4th decade</td>
<td>Fleshy opaque white nodules</td>
<td>Stellate, fusiform, round or spindle shaped cells depending on differentiation</td>
<td>S100 positive; Either well or poorly differentiated; High Ki-67 indicates poor prognosis</td>
</tr>
<tr>
<td>Myxofibrosarcoma</td>
<td>Elderly females</td>
<td>Multiple nodules sometimes with necrosis</td>
<td>Multinodular, with pleomorphic spindle cells</td>
<td>Grade doesn’t predict behaviour</td>
</tr>
</tbody>
</table>

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CONCLUSION

Just a few hundred cases of AAM of vulva have been reported worldwide. This, coupled with the vast differentials, makes preoperative diagnosis very difficult with a misdiagnosis in > 80%. Excision with adequate margins is the treatment of choice. Hormonal manipulation and arterial embolization have been tried with varying results. The prognosis is good, but the major issue to be considered is recurrence; they require long term follow-up with serial MRI. Rearrangements in HMGA2 gene is a sensitive marker which can be used in the post-op period for detecting residual and early recurrent lesions.

Compliance with Ethical Standards

Informed consent: Informed consent was obtained from the patient included in the report.

Ethical Approval: For this type of study formal ethical clearance is not required.

REFERENCES

Barath Raj Kumar et al., A Case Report of Aggressive Angiomyxoma of Vulva: An Often Misdiagnosed Neoplasm

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