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## Case Report

# Aggressive angiomyxoma of vagina: a rare entity

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### ABSTRACT

Aggressive angiomyxoma a soft tissue tumor arising in the pelvis and perineal regions of women in reproductive age group is a rare entity. It is slow growing locally aggressive myxoid mesenchymal tumor, with a marked tendency to local recurrence. Preoperative clinical diagnosis is usually difficult due to absence of diagnostic features as well as rarity of the disease. We describe a case of aggressive angiomyxoma of vagina in a 47-year-old para 4 woman with multiple fibromyoma (upto 22-week size of pregnant uterus) with 10\*10 cms posterior vaginal cyst. Total abdominal hysterectomy with surgical excision of vaginal wall cyst done. A retrospective diagnosis-Aggressive Angiomyxoma of the vagina was made after histological confirmation. Surgical excision with wide margins and long term follow up remains treatment of choice.

**Keywords:** Aggressive angiomyxoma, Retrospective diagnosis, Surgical excision

### INTRODUCTION

Aggressive angiomyxoma (AA) of female pelvis and perineum is a rare tumor, with less than 150 cases reported in the literature.<sup>1</sup> Steeper and Rosai first described this tumor of mesenchymal origin.<sup>2</sup> It usually occurs in reproductive age with peak incidence between the third and fourth decades of life, with higher (95%) preponderance in women.<sup>3-7</sup> AA is a rare, locally aggressive myxoid mesenchymal tumor, arising in the deep soft tissues of the vulva, vagina, perineum and pelvis of young adult women.

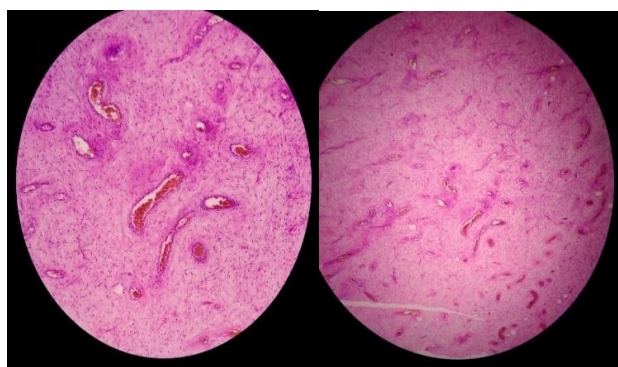
The term “aggressive” emphasises neoplastic nature of blood vessels, its locally infiltrative nature and high risk of local recurrences and does not indicate its malignant nature.<sup>2</sup> In men, the tumor is extremely rare appearing at an older age involving analogous sites including the scrotum and inguinal area.<sup>8-10</sup> A slow growing locally aggressive tumor with frequent local recurrences in 30-72%, sometimes occurring years later.<sup>1,9,11</sup> Diagnosis is usually made retrospective after histological examination

following surgical resection.<sup>8</sup> Complete surgical resection with wide margins, long term follow up, careful monitoring with imaging is essential. Recurrence rate in patients with narrow surgical excision margins is not higher than patients with wide surgical margins.<sup>12</sup> Metastases is very uncommon, and misdiagnosis is a very frequent problem.<sup>3,12</sup> We describe a rarest case of Aggressive Angiomyxoma of vagina, in fact first ever in 34 years of gynaecologic experience.

### CASE REPORT

A 47-year-old, Para 4 perimenopausal women presented with multiple fibromyoma uterus with large painless 10\*10 cms posterior vaginal wall cyst, in gynecology outpatient department, Government Medical College, Patiala, Punjab, India. On Per abdomen and bimanual pelvic examination there was a nontender mass, 22 week size of pregnant uterus, firm in consistency with irregular surface. A 10\*10 cms soft, cystic, nontender swelling with smooth surface, extending high upto posterior fornix was felt in posterior vaginal wall, incidental finding.

Ultrasound confirmed clinical diagnosis of multiple fibromyoma uterus with posterior vaginal wall cyst. Total abdominal hysterectomy, vaginal wall cyst excision with posterior colpoperineorrhaphy done under regional anaesthesia. Vaginal cyst was globular, soft to firm with smooth margins. Cut section presented homogenous mass with gelatinous consistency. On histopathological examination of vaginal cyst, monotonous, hypocellular stroma composed of small stellate fibroblast was seen. Stroma was myxoid with collagen fibres and prominent dilated thick walled blood vessels with some showing hyalinisation suggestive of aggressive angiomyxoma (Figure 1).



**Figure 1: Aggressive angiomyxoma of vagina.**

## DISCUSSION

Aggressive angiomyxoma is a rare painless tumour with a peak incidence in the third and fourth decades of life in women majority of them being white.<sup>7</sup> The tumor usually involves pelvis, often located in the vagina, vulva, perineum or buttocks.<sup>3,4</sup> It is a slow growing tumour invading paravaginal and pararectal spaces occupying whole pelvis with displacement of adjacent structures. Women may take two months to seventeen years in reporting to hospital with a painless mass after initial discovery.<sup>5</sup>

Our patient 47 years old presented with large multiple fibromyoma uterus, a large aggressive angiomyxoma in posterior vaginal wall was incidental finding, patient being not aware of this painless tumour. Chen et al reported a large 57\*47\*23 cm aggressive angiomyxoma of pelvis, growing slowly in over eight years.<sup>13</sup> On gross examination, aggressive angiomyxoma is a soft, bulky with smooth surface and appears nonencapsulated. Cut section gives bluish grey hue, homogenous, gelatinous consistency with focal areas of congestion and hemorrhage. Tumour usually displaces rather than invading adjacent tissues and are rarely destructive. Similar finding was reported in our patient. Occasionally tumour can invade bladder, bowel and pelvic bone.<sup>3,8,10</sup> However there was no involvement of bowel, bladder or pelvic bone in our patient. Histological examination depicts, stellate and spindle-shaped neoplastic cells scattered in loose myxoid matrix with pale pink colour

after eosin stain. Prominent vascular component ranging from tiny capillaries to large thick walled vessels with no evidence of anastomosis or arborisation is also seen.<sup>3,5,8,10</sup>

Translocation at chromosome 12 with aberrant expression of high mobility group protein isoform I-C(HMGI-C) involved in DNA transcription has been demonstrated. Localisation of HMGI-C in neoplastic stromal cells might be applied as diagnostic tool in assaying margin status.<sup>3,5,8</sup> Aggressive angiomyxoma arises from specialised cells of lower genital tract. Tumor cells are positive for Vimentin, weakly positive for Desmin but not for myosin, suggesting a fibroblastic origin and differentiation of tumor.<sup>9,10</sup> Tumor usually expresses estrogen (ER) and progesterone receptors (PR). However dermal fibroblast and stromal cells in variety of vulvar lesions may also be positive. ER or PR immunoreactivity may not distinguish aggressive angiomyxoma from its histological mimics.<sup>10</sup> Distinctive vascular component of the tumor helps in differentiating it from other soft tissue tumor like Bartholin cyst, leiomyoma, vulvar lipoma, angiomyo-fibroblastoma, myxolipoma, myxoid neurofibroma, myxofibrosarcoma and botryoid rhabdomyosarcoma among others.<sup>3,10,14,15</sup> Rarely the tumour may show mild atypia. Though slow growing local recurrence are as high as 30-72%, but rarely undergo malignant change.<sup>2,10</sup> However literature does report two cases of pulmonary metastasis in 63 years old and in a young woman.<sup>11</sup>

Aggressive angiomyxoma is often misdiagnosed as it may have similar clinical presentation to common lesions such as Bartholin cyst or prolapse vaginal wall, gartner cyst or levator hernia. Misdiagnosis rate may be as high as 70 to 100%.<sup>5</sup> Diagnosis may be missed even after sonography, computerised tomography (CT) and MRI. Diagnosis is mostly histological after primary excision.<sup>8</sup> The swirling appearance remains diagnostic feature seen in 83% cases. MRI remains diagnostic modality of choice as swirled pattern is more specific than seen on CT. High signal intensity on MRI and attenuation on CT is due to loose myxoid matrix and high water content of angiomyxoma.<sup>3,4,10</sup> Preoperative imaging (CT or MRI) is of significant importance as it defines extent of tumour as well as involvement of surrounding structures thus helps to decide the operative route to be followed.<sup>4,7</sup> In our patient preoperative imaging with CT or MRI was not done as the clinical appearance was suggestive of vaginal cyst. Complete surgical resection with long term follow up and careful monitoring with imaging is essential in view of high risk of local recurrence.

Aggressive angiomyxoma has estrogen and progesterone receptors, thus tumour grows rapidly during pregnancy. Hence gonadotropin agonist (GnRh) remains a viable option in primary treatment of small tumours and in event of recurrence.<sup>3,6,8,10</sup> Oophorectomy may be a treatment modality in women past middle age who have completed their family as tumour is receptor positive.<sup>1,3</sup> Further research is needed. Radiotherapy or chemotherapy as an

adjunct to primary surgery does not offer any advantage as tumour has low mitotic activity.<sup>2,8,10</sup> Embolization or chemoembolization also does not offer any advantage as angiomyxoma has more than one feeding vessel.<sup>10</sup> Surgical excision with wide margin and long term follow up with imaging remains treatment of choice.

## CONCLUSION

Aggressive angiomyxoma is rarest of rare, locally aggressive mesenchymal tumor arising in deep soft tissue of vulva, vagina, perineum and pelvis of young women between third and fourth decade of life with a very high risk of recurrence. It should be kept in mind for every mass in genital or pelvic region in women of reproductive age for preoperative diagnosis. A complete preoperative imaging is mandatory to know extent of tumour and decide route of surgery. Surgical excision with wide margins remain treatment of choice. Long term follows up by clinical examination and MRI essential for early detection of local recurrence.

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