International Journal of Reproduction, Contraception, Obstetrics and Gynecology
*Jayaram S et al. Int J Reprod Contracept Obstet Gynecol. 2020 Dec;9(12):5131-5133*
www.ijrcog.org
pISSN 2320-1770 | eISSN 2320-1789

DOI: https://dx.doi.org/10.18203/2320-1770.ijrcog20205261

**Case Report**

# Aggressive angiomyxoma vulva

**Surya Jayaram*, Sarojini Mayadevi**

Women's Health, Aster Medcity, Ernakulam, Kerala, India

**Received:** 07 September 2020
**Accepted:** 17 October 2020

***Correspondence:**
Dr. Surya Jayaram,
E-mail: suryaj1@yahoo.co.in



## ABSTRACT

Aggressive angiomyxoma is a mesenchymal tumour which is locally invasive and seen in women of reproductive age group. We present a case of 46-year-old post hysterectomy lady with swelling in left side of vulva.

**Keywords:** Aggressive angiomyxoma, Mesenchymal tumour, Vulval tumour

## INTRODUCTION

Angiomyxomas are a rare type of soft tissue tumours which arise from myxoid cells of body's connective tissue. They can be superficial or aggressive. Aggressive angiomyxoma, first described by Steeper and Rosai in is a rare, locally, invasive mesenchymal tumour occurring usually in female pelvic and perineal regions in women of reproductive age group.[1-3] It has a marked tendency for local recurrence, but low tendency to metastasize. Treatment is complete resection if possible, with adjuvant chemotherapy and radiotherapy when necessary. Long-term follow up is necessary because of its locally aggressive nature.

## CASE REPORT

46-year-old para 2, live 2, post hysterectomy patient was referred to us with complaints of swelling in left side of vulva noticed 2 months back and gradually increasing in size. The swelling was otherwise asymptomatic. It was diagnosed as a case of lipoma from local hospital and she was referred to us for excision of the lesion. She had undergone total abdominal hysterectomy for fibroid uterus 4 years back. She was diabetic and hypertensive.

On examination there was a large cystic, non-tender lesion of size 7×6 cm in size in the left labia majora in subcutaneous plane.

Ultrasound examination showed a well-defined hypoechoic lesion 6.29×3.40 cms in left vulva with no internal cystic degeneration or calcification. Impression: Lipoma. Patient underwent excision of vulvar lesion. Lesion of 6×4.5×3 cms was excised by enucleating the same. There was difficulty in getting a plane of excision. The defect was closed by approximating the skin edges after excising the excess tissue.

Histopathology reports showed aggressive angiomyxoma with CD 34, Desmin, smooth muscle actin (SMA) and estrogen (ER) Positive in tumour cells. There were no conspicuous mitosis or necrosis.

Since margins could not be assessed in the specimen sent, a wide local excision was done after 2 months and margins were found to be free of the disease.

She was followed up at 4 weeks, 2 months, 6 months and 1 year and found to be disease free.

## DISCUSSION

Angiomyxomas are classified either as superficial (also called as cutaneous myxoma) or aggresive angiomyxomas.

Superficial angiomyxomas may occur in the setting of carney complex.[4] This lesion is observed predominantly in male middle-aged adults and can arise anywhere in the superficial tissues, but mostly it involves the trunk, lower extremities, and head and neck. Clinically, most lesions appear as slowly growing polypoid cutaneous lesions and are easily confused with a cyst, skin tag, or neurofibroma. The stroma is mostly edematous with little myxoid material.

The aggressive angiomyxoma tumour commonly presents as an asymptomatic mass in the genital area of women in their reproductive life, but is occasionally reported in men (male-to-female ratio being 1:6). Estrogen and progesterone receptors are commonly found in aggressive angiomyxoma . It is thus likely to grow during pregnancy and respond to hormonal manipulation. It involves mainly the pelvis, vulva, perineum, vagina and urinary bladder in adult women in the reproductive age.[5] The term “aggressive” refers to the locally infiltrative nature of this lesion and its tendency to recur.[6]

Clinically, aggressive angiomyxoma may be misdiagnosed as bartholin cyst, lipoma, labial cyst, gartner duct cyst, levator hernia or sarcoma. Fibro-epithelial stromal polyp, superficial angiomyxoma, angiomyofibroblastoma, cellular angiofibroma and smooth muscle tumors also need to be considered in the differential diagnoses of a polypoidal mass in the perineum. Aggressive angiomyxoma is an infiltrative tumor, whereas angiomyofibroblastoma is well circumscribed (this characteristic can also be identified on magnetic resonance imaging [MRI]). Also, aggressive angiomyxoma has thick-walled vessels, which are less numerous than the thin-walled vessels in angiomyofibroblastoma. On computed tomography (CT) scan, these tumours have a well-defined margin with attenuation less than that of the muscle. On MRI, these tumours show high signal intensity on T2-weighted images. The attenuation on CT and high signal intensity on MRI are likely to be related to the loose myxoid matrix and high-water content of angiomyxoma.[7]

On gross examination aggressive angiomyxoma, is unencapsulated, is poorly or vaguely circumscribed, and may blend imperceptibly with surrounding soft tissue. Tumor size is highly variable and ranges from 1 to 60 cms.[8] The tumor is often tan-pink to tan-gray, bulky, and has a rubbery consistency with a glistening, gelatinous cut surface. Areas of congested blood vessels, hemorrhage, or fibrosis may be present.[1,9]

Histologic examination shows a sparsely cellular tumor composed of pale to eosinophilic stroma studded with numerous haphazardly arranged blood vessels that stand out against the myxoid background and range in size from thin-walled capillaries and venules to larger muscular arteries. The stroma is distinctly myxoid with intermixed, wispy collagen fibrils; scattered, smooth muscle bundles; and extravasated erythrocytes.[7,10] Immunohistochemically, most aggressive angiomyxomas express different combinations of estrogen and progesterone receptors, vimentin, desmin, smooth-muscle actin, SMA, CD34, and CD44, but all are invariably negative for S-100, CEA, and keratin.[11,12] Cytogenetic and molecular analysis revealed clonal karyotypic abnormalities including chromosomal translocation involving chromosome 12 associated with rearrangement of the HMGIC gene and it was proposed that aggressive angiomyxoma molecularly belonged to the benign group of mesenchymal tumors showing multiple aberration region involvement. HMGIC expression in aggressive angiomyxoma is of value in the distinction of difficult cases from its histological mimics.[13]

The first line of therapy for aggressive angiomyxoma is surgery, although achieving negative resection margins is difficult because of the infiltrative nature of the tumour and the absence of a defined capsule. Smaller, more-superficial tumours of the vulva or vagina may be removed with wide, local excision, but larger, deep-seated tumours of the pelvis may require more extensive surgery with partial or complete resection of some pelvic organs, conferring a higher risk of morbidity.[10] A review of 111 cases of aggressive angiomyxoma called into question the necessity of such radical resections by comparing patient’s risk of recurrence based on margin status. The data showed that there was no statistical difference in remaining disease-free between groups of patients with positive and negative resection margin results (40% and 50% in 10 years, respectively).[14] Even though complete surgical resection is the desired goal, incomplete removal is acceptable when significant operative morbidity is anticipated or when preservation of fertility is a concern.[14]

## CONCLUSION

Vulval masses are usually detected on routine gynaecological examination. Angiomyxoma should be kept in mind as differential diagnosis when evaluating a painless vulval mass. Treatment options are surgical resection followed by chemotherapy or local radiation if necessary. Hormonal therapy with GnRH agonists should be considered to prevent recurrence.

*Funding: No funding sources*
*Conflict of interest: None declared*
*Ethical approval: Not required*

## REFERENCES

1. Steeper TA, Rosai J. Aggressive angiomyxoma of female pelvis and perineum.Report of nine cases a

distinctive type of gynecologic soft tissue neoplasm. Am J Surg Pathol. 1983;7:463-75.
2. Fetsch JF, Laskin WB, Lefkowitz M, Kindblom LG, Meis-Kindblom JM. Aggressive angiomyxoma: A clinicopathologic study of 29 female patients. Canc 1996;78:79-90.
3. Granter SR, Nucci MR, Fletcher CD. Aggressive angiomyxoma: Reappraisal of its relationship to angiomyofibroblastoma in a series of 16 cases. Histopathol. 1997;30:3-10.
4. Carney JA, Headington JT, Su WD. Cutaneous myxomas: a major component of the complex of myxomas, spotty pigmentation, and endocrine overactivity. Arch Dermatol. 1986;122(7):790-8.
5. Han-Geurts IJ, van Geel AN, van Doorn L, M den Bakker, Eggermont AM, Verhoef C. Aggressive angiomyxoma: Multimodality treatments can avoid mutilating surgery. Eur J Surg Oncol. 2006;32:1217-21.
6. Kiran G, Yancar S, Sayar H, Kiran H, Coskun A, and Arikan DC. Late recurrence of aggressive angiomyxoma of the vulva. J Lower Genital Tract Dise. 2013;17(1):85-7.
7. Outwater EK, Marchetto BE, Wagner BJ, Siegelman ES. Aggressive angiomyxoma: Findings on CT and MR imaging. AJR Am J Roentgenol. 1999;172:435-8.
8. Siassi RM, Papadopoulos T, Matzel KE. Metastasizing aggressive angiomyxoma. N Engl J Med. 1999;341(23):1772.
9. McCluggage WG. A review and update of morphologically bland vulvovaginal mesenchymal lesions. Int J Gynecol Pathol. 2005;24(1):26-38.
10. Haldar K, Martinek IE, Kehoe S. Aggressive angiomyxoma: a case series and literature review. Eur J Surg Oncol. 2010;36(4):335-9.
11. Dahiya K, Jain S, Duhan N, Nanda S, and Kundu P. "Aggressive angiomyxoma of vulva and vagina: a series of three cases and review of literature," Archiv Gynecol Obstetr. 2011;283(5)1145-8.
12. Dierickx I, Deraedt K, Poppe W, and Verguts J. Aggressive angiomyxoma of the vulva: a case report and review of literature. Archiv Gynecol Obstet. 2008;277(6):483-7.
13. Nucci MR, Fletchr CD. Vulvovaginal soft tissue tumors: Update and review. Histopathol. 2000;36:97-108.
14. Chan YM, Hon E, Ngai SW, Ng TY, Wong LC. Aggressive angiomyxoma in females: is radical resection the only option? Acta Obstet Gynecol Scand. 2000;79(3):216-20.

**Cite this article as:** Jayaram S, Mayadevi S. Aggressive angiomyxoma vulva. Int J Reprod Contracept Obstet Gynecol 2020;9:5131-3.