Case Report

# The Incidental Aggressive Angiomyxoma of the Vulva: Looks can be Deceptive

**Akanksha Tripathi[1], Shikha Sharma[2]*, Sunil Kanvinde[3]**

[1]Department of Obstetrics and Gynaecology, GBH American Hospital, Udaipur, Rajasthan, [2]Department of Obstetrics and Gynaecology, Command Hospital, Chandi Mandir, Haryana, [3]Department of Pathology, Green Cross Pathology Laboratory, Ahmedabad, Gujarat, India

## Abstract

Aggressive angiomyxoma is a benign, slow-growing, locally aggressive tumor of mesenchymal origin primarily occurring in the pelvic–perineal regions of reproductive age group women and displays a high risk of local recurrence. Lack of specific symptomatology and overlap with other benign and malignant vulval masses makes it a diagnostic challenge. We describe the case of a 32-year-old nulliparous woman with a history of recurrent vulval abscess requiring multiple incision and drainage procedures before she presented to us with an actively draining abscess on the upper third of the left labia majora. She underwent excisional biopsy at our center, the histopathology of which revealed aggressive angiomyxoma with secondary pyogenic slough. The preoperative diagnosis of vulval aggressive angiomyxoma becomes challenging due to the absence of diagnostic features. It ought to be considered a differential in every perineal–pelvic region mass in adult women.

**Keywords:** Angiomyxoma, Bartholin's abscess, biopsy, mesenchymal, vulval

## Introduction

Aggressive angiomyxoma (AAM), first described by Steeper and Rosai in 1983, is a rare locally aggressive soft-tissue tumor of the mesenchymal origin with <250 cases reported in the literature till date.[1] The incidence among females is more than that of men in the ratio of 6:1 and is mostly seen in premenopausal women with a peak incidence in the third to fifth decades of life. The 2003 third edition and 2013 fourth edition of the World Health Organization Classification of Bone and Soft-Tissue Tumors classified it as a tumor of uncertain differentiation and named as deep (aggressive) angiomyxoma. It is a slow-growing, low-grade neoplasm involving the pelvis and perineum with a risk of local recurrences as high as 30%–72%, which may sometimes be seen even years later. Metastasis, however, is uncommonly seen.[2]

Since, AAM is rarely seen and has no specific clinical features, almost 80% of them are misdiagnosed as Bartholin's cysts, lipomas, or hernias preoperatively leading to delayed diagnosis in most cases. Diagnosis, mostly, is histopathological which reveals hypocellular mesenchymal neoplasm with minimal mitotic activity and scattered spindle cells with blood vessels of various caliber embedded in the myxoid matrix. Immunohistochemically, the neoplastic cells show strong positivity for vimentin and desmin, moderate positivity for CD34 as well as estrogen/progesterone hormone receptors, and usual negativity for S-100.[3]

Primary surgical resection is the mainstay of treatment, and the most effective too, although treatment with GnRH agonists is an emerging therapy.[4]

## Case Report

A 32-year-old, nulliparous woman, presented with a vulval swelling, purulent discharge, and low-grade fever for

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**Address for correspondence:** Dr. Shikha Sharma, Command Hospital, Chandi Mandir, Haryana, India.
E-mail: shikhalhmc@gmail.com



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1 week. There was a history of recurrent vulval abscesses for 8–9 months for which she had undergone incision and drainage up to 10 times at various centers, besides taking multiple courses of antibiotics. Examination revealed an irregular, tender, 4 cm × 2 cm abscess with a sinus draining purulent discharge over the upper 1/3 third of the left labia majora. There was no overlying skin redness, nodularity, puckering, or ulceration. No blood was expressed on additional pressure over the lesion. There were no associated menstrual irregularities, discharge per vaginum, any inguinal swelling, local itching or bladder/bowel complaints, or history of Koch's in self or exposure to the same. All routine investigations as well as ultrasonography (USG) abdomen and pelvis were normal. Although it had repeatedly been diagnosed with Bartholin's abscess previously, the anatomic location led us to keep differentials of labial abscess, Sexually transmitted diseases (STD), or even vulval carcinoma in mind. In view of the same, we proceeded with incision and drainage along with vulvar biopsy and postoperative antibiotics.

Histopathological examination revealed a tumor composed of myxoid stroma embedded in plenty of vascular spaces with heavy pyogenic inflammation suggestive of AAM with secondary pyogenic slough. She was counseled regarding the risk of early recurrence warranting a close-up with an onco surgeon, the need for more extensive surgery in case of recurrence and most importantly, to report as soon as any vulval mass appears and to not wait until it turns into an abscess. However, the couple opted to follow-up at our center for further management as our center was the most accessible and affordable to them. As expected, she reported after 2½ months with a 6 cm × 4 cm nontender, irregular swelling over the left labia majora with no purulent discharge [Figure 1]. All blood investigations as well as USG pelvis were normal.

After a written informed consent, the patient underwent wide local excision under spinal anesthesia wherein a 7 cm × 5 cm × 3 cm polypoidal mass was removed. The specimen on histopathology revealed a soft-tissue tumor having large myxoid areas interspersed with blood vessels showing arborization. There was no significant mitotic activity or nuclear atypia noted. The closest cutaneous margin was 0.5 cm and epithelial margin was 3 cm away from the lesion meaning that the tumor margins were free. Florid secondary pyogenic features were noted [Figure 2].

The patient was kept on 3 monthly follow-ups for 6 months followed by 6 monthly follow-ups for 1 year with clinical examination and USG. She conceived spontaneously within a year, had full-term normal delivery with no ante/intra/postpartum complications. She has had no relapse over a period of 5-year follow-up since then.

## Discussion

Angiomyxoma is a rare, benign neoplastic disease originating from mesenchymal tissues and characterized by extensive local invasiveness and a high recurrence rate of 30%–72%, sometimes even years later. It is mainly witnessed in the pelvic, perineal, and genital regions with a reported peak incidence among young women in the third and fourth decades of life.[2] The term "aggressive" was adopted considering its propensity for local aggression and recurrence after excision and not to indicate malignant potential. There are only two cases reported to have had metastasis in the literature. Owing to its anatomical location, there can be numerous differential diagnoses such as Bartholin's cyst, labial cyst or abscess, leiomyoma, lipoma, or hernia.[5,6] The preoperative rate of misdiagnosis can vary from 70% to 100%. Definitive diagnosis is possible only on histopathological examination of the excised specimen. Even in our case, the patient had, repeatedly, been managed as Bartholin's cyst at various centers before she presented to us. We took a vulvar biopsy, besides the incision and drainage, only

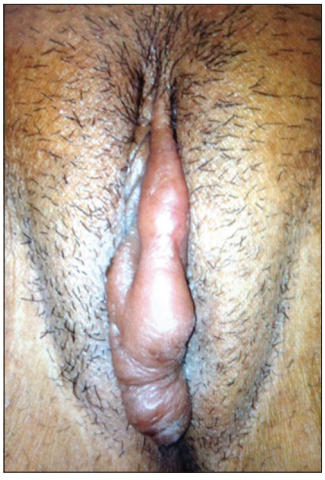

**Figure 1:** Image depicting the visible lesion on the left vulva

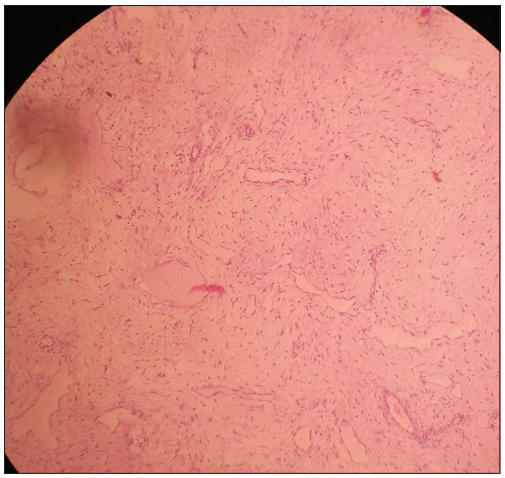

**Figure 2:** Histopathological view displaying large myxoid areas interspersed with blood vessels showing arborization

because it was not at the conventional location of a Bartholin's cyst leading us to rule out other possible causes.

Currently, there are records of 250 cases in the literature, with the largest case series from Fetsch *et al*. who reviewed 29 diagnosed cases of AAM in women from The Armed Forces Institute of Pathology database.[7] They found out that most tumors were larger than 10 cm in size with lobulated architecture, sharp margins and low-to-moderate cellularity, commonly positive for desmin, smooth muscle actin, vimentin, estrogen and progesterone, and negative for Ki-67 and S-100 n IHC staining. As per the available data, tumor grossly varies 1 cm to 60 cm in size, is often tan-pink to tan-gray, bulky, with a rubbery consistency, is glistening and gelatinous on cut surface, interspersed with congested blood vessels, and areas of hemorrhage or fibrosis.[8] Histologically, the tumor displays small, somewhat stellate, and spindle-shaped neoplastic cells with thin cytoplasmic processes that are scattered in a loose myxoid matrix composed of delicate wavy collagen fibrils and hyaluronic acid, rendering it a pale pink color on eosin staining. It is also accompanied by prominent vascular components, ranging from tiny capillary-like vessels to larger thick-walled vessels with a distinct smooth muscle layer but with no evidence of anastomosis or arborization. It is strongly positive for vimentin and desmin with moderate positivity for CD34 and estrogen and progesterone receptors on immunohistochemistry.[2]

Although the pathogenesis of AA is poorly understood, few cytogenetic has shown that the architectural transcription factor HMGA2, located on chromosome 12q13-15, is sometimes rearranged in this tumor which may result in aberrant HMGA2 protein expression. HMGA2 is a member of high mobility group proteins, which are architectural transcription factors expressed primarily during embryogenesis. However, HMGA2 expression is also found in other vulvovaginal mesenchymal lesions (for example, leiomyomatous neoplasm) and hence, is not a specific marker of AAM. However, it could have a possible role in assessing tumor margins and detection of residual or recurrent tumor foci in excised specimens.[9] A novel translocation HMGA2-YAP fusion has also been described in a woman with AAM, who responded to estrogen antagonists which could lead us to newer therapies.[10]

In suspected cases, preoperative imaging whether computed tomography or magnetic resonance imaging (MRI) is crucial to determine the size of the lesion, involvement with surrounding structures, and subsequently, plan the surgical procedure. Currently, radical resection with negative margins appears to be the best modality of treatment for AA. However, a review of over 100 cases observed that those with positive margins are as likely to have recurrences as those with negative margins.[2,11] Radiotherapy and chemotherapy are not useful due to the low mitotic activity of this tumor. Hormone therapies with GnRH agonists have shown to reduce tumor burden where surgical resection was not feasible. However, long-term studies on its use and side effects are lacking.

In view of known late recurrences, all patients need to be in long-term surveillance, usually with clinical examinations, with MRI being the preferred modality for the detection of early recurrences. Options for the treatment of recurrence include repeat surgery, radiotherapy, and hormonal therapy, but no single modality is clearly beneficial over the others.

It is a good clinical practice to keep differential of angiomyxoma for nonpeculiar vulval masses. A better understanding of this pathology is the key to better treatment strategies with multidisciplinary approach being integral to optimal management.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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## Conflicts of interest

There are no conflicts of interest.

## References

1. Abu Saadeh F, Galvin D, Alsharbaty MJ, Gleeson N. Paravaginal aggressive angiomyxoma. BMJ Case Rep 2015;2015:bcr2014207287.
2. Dahiya K, Jain S, Duhan N, Nanda S, Kundu P. Aggressive angiomyxoma of vulva and vagina: A series of three cases and review of literature. Arch Gynecol Obstet 2011;283:1145-8.
3. Fatušić J, Hudić I, Fatušić Z, Čićkušić E, Šarkanović G. Giant aggressive angiomyxoma of the vulva: Case report. Acta Clin Croat 2015;54:220-2.
4. Deshmukh BD, Kulkarni MP, Momin YA, Sulhyan KR. Aggressive angiomyxoma of vulva: A case report and review of literature. Arch Med Health Sci 2015;3:88-90.
5. Sukgen G. A case report: The third-largest case in the literature of a vulvar lipoma. Gynecol Minim Invasive Ther 2020;9:234-6.
6. Handler SJ, Truong C, Yazdany T. Vulvar angioleiomyoma. Gynecol Minim Invasive Ther 2022;11:119-20.
7. Fetsch JF, Laskin WB, Lefkowitz M, Kindblom LG, Meis-Kindblom JM. Aggressive angiomyxoma: a clinicopathologic study of 29 female patients. Cancer. 1996;78:79-90.
8. 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;79:216-20.
9. McCluggage WG, Connolly L, McBride HA. HMGA2 is a sensitive but not specific immunohistochemical marker of vulvovaginal aggressive angiomyxoma. Am J Surg Pathol 2010;34:1037-42.
10. Lee MY, da Silva B, Ramirez DC, Maki RG. Novel HMGA2-YAP1 fusion gene in aggressive angiomyxoma. BMJ Case Rep 2019;12:e227475.
11. Foust-Wright C, Allen A, Shobeiri SA. Periurethral aggressive angiomyxoma: A case report. Int Urogynecol J 2013;24:877-80.