Case Report

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Aggressive Angiomyxoma of the Vulva Mimicking a Benign Mass: A Case Report

Surgery Section

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ABSTRACT

Aggressive Angiomyxoma (AA) is a rare, slow-growing mesenchymal neoplasm predominantly affecting women of reproductive age, characterised by local infiltration and a high recurrence rate. The present case presents a 32-year-old woman with a vulvar mass persisting for three years, initially misdiagnosed as a Bartholin's cyst upon clinical examination. Magnetic Resonance Imaging (MRI) revealed a well-defined, hyperintense mass on T2-weighted imaging with a characteristic laminated internal architecture, displacing adjacent structures with limited invasion of the underlying musculature and without significant infiltration of the urogenital diaphragm or pelvic organs. Complete surgical excision was achieved with a margin of 1 cm. Histopathology demonstrated a sparsely cellular myxoid tumour composed of spindle and stellate cells in a loose matrix with prominent vasculature. Immunohistochemical analysis showed strong positivity for Oestrogen Receptors (ER) and Progesterone Receptors (PR), vimentin and focal desmin, while S-100 expression was negative. Given the hormone receptor profile, the patient was initiated on adjuvant Gonadotropin-Releasing Hormone (GnRH) agonist therapy to reduce recurrence risk, administered once a month for 12 months. She remained recurrence-free at six-month follow-up. The present case underscores the diagnostic challenge posed by AA, which can closely resemble benign lesions such as Bartholin's cysts and fibroepithelial polyps, as well as malignant neoplasms like myxoid liposarcoma, necessitating a high index of suspicion and thorough histopathological evaluation for accurate diagnosis. Accurate diagnosis requires integration of clinical, radiological and histopathological findings. The infiltrative nature of the tumour, despite its bland cytology, necessitates complete resection and prolonged surveillance. Hormonal therapy represents a valuable adjunct in receptor-positive cases, especially when surgical margins are uncertain or complete excision is infeasible.

Keywords: Immunohistochemistry, Mesenchymal tumour, Musculature, Vulvar neoplasms

CASE REPORT

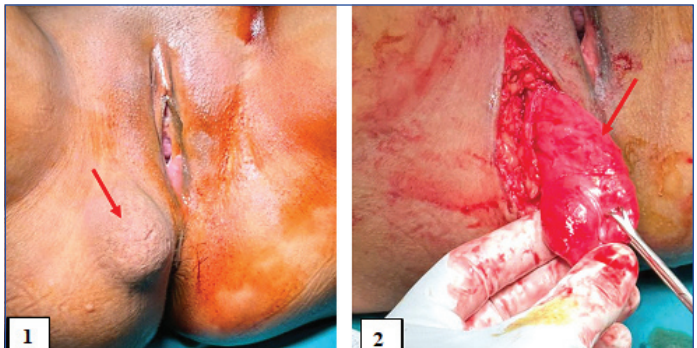
A 32-year-old unmarried woman from Southern India presented to the surgical outpatient department of a tertiary care hospital with a progressively enlarging swelling over the right labium majora that had been persisting for three years. The mass was initially painless; however, the patient began experiencing sharp, localised pain over the past two weeks, which prompted her to seek medical attention. The patient had no associated complaints and had ignored the swelling due to its indolent nature and financial constraints. She did not report any prior menstrual abnormalities or any similar swellings among her family members.

A well-defined, soft, non tender mass measuring approximately 5×6 cm was noted over the right labium majora at the 7 o'clock position, lateral to the vaginal introitus [Table/Fig-1]. The overlying skin was intact and showed no discolouration, ulceration, or signs of inflammation. There was no evidence of regional lymphadenopathy. The external genitalia were otherwise normal. Speculum and bimanual pelvic examinations revealed a normal cervix and a non tender, anteverted uterus of normal size and consistency, with no adnexal masses or tenderness. Clinical examination led to a differential diagnosis of Bartholin's cyst or epidermal inclusion cyst.

Initial bloodwork was unremarkable except for an elevated total leukocyte count. Ultrasonography of the abdomen and pelvis showed no intra-abdominal or pelvic pathology. Ultrasound revealed a lobulated, hypoechoic soft tissue lesion in the right labium majora with heterogeneous echotexture and mild vascularity, suggestive of a benign soft tissue mass. MRI showed a well-defined, lobulated mass with high signal intensity on T2-weighted images and a characteristic "swirled" or layered internal architecture, consistent with AA. The mass exhibited intermediate signal intensity on T1-weighted images and heterogeneous but progressive enhancement following contrast administration. The lesion was noted to displace but not invade adjacent structures, including the urethra, vagina and rectum. The fat planes with the bladder and pelvic musculature were preserved and no signs of pelvic lymphadenopathy or distant extension were observed. There was no evidence of necrosis, haemorrhage, or calcification within the lesion.

Following a thorough preoperative evaluation, the patient was scheduled for surgical excision of the swelling under spinal anesthesia, with the patient in the lithotomy position. After aseptic preparation, a curvilinear incision was made over the lesion with a 1 cm margin of healthy tissue. The mass was irregular and moderately vascular, with limited invasion into adjacent musculature [Table/Fig-2]. No deeper extension was noted. Sharp and blunt dissection facilitated en-bloc removal. Haemostasis was secured and layered closure was achieved with absorbable sutures. The patient experienced an uneventful postoperative course and recovery.

Histology revealed a sparsely cellular lesion composed of spindle and stellate-shaped cells embedded in a loose myxoid matrix, interspersed with numerous thin-walled blood vessels, characteristic of AA based on the (2024) World Health Organisation

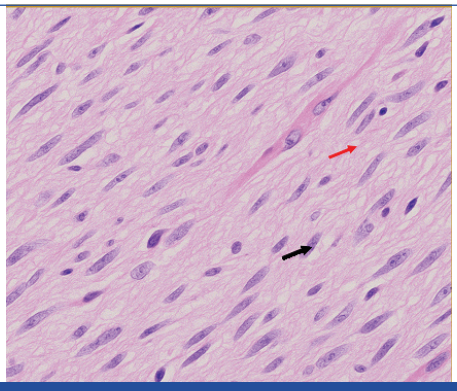


[Table/Fig-1]: Clinical photograph showing a 5*6 cm was noted on the right labium majora at the 7 o'clock position, lateral to the introitus (Red arrow).
[Table/Fig-2]: Intraoperative photograph showing a deep-seated lesion with significant vascularity arising from the labia majora (Red arrow).

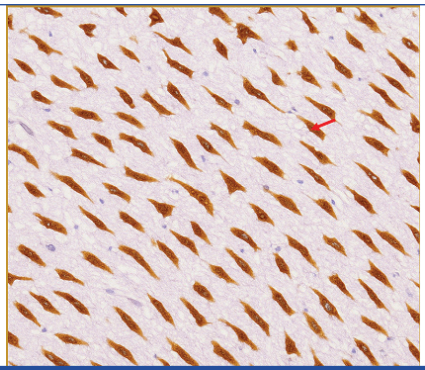
Diagnostic criterion	Present case findings
Deep-seated, slow-growing vulvoperineal mass in reproductive-age female	32-year-old woman with a 3-year history of a slowly enlarging vulvar mass located in the labium majora
Characteristic MRI features: swirled or laminated T2 hyperintense mass	MRI showed a well-defined, T2-bright mass with a swirled internal architecture and displacement of adjacent organs without invasion
Histopathology showing hypocellular myxoid stroma with spindle/stellate cells	Tumour composed of bland spindle and stellate-shaped cells in loose myxoid matrix with abundant vasculature
Immunohistochemistry (IHC) positive for ER, PR, vimentin, desmin; negative for S-100	IHC revealed strong ER and PR positivity; diffuse vimentin expression; focal desmin positivity; S-100 negative
Infiltrative margins and absence of atypia or mitoses despite large size	Tumour lacked mitotic activity or nuclear atypia but showed deep infiltration without encapsulation

[Table/Fig-3]: Correlation of present case findings with WHO Classification of Tumours: soft-tissue and bone tumours, 5th edition, 2024 update [1].

(WHO) Classification of Soft Tissue and Bone Tumours [Table/Fig-3] [1]. There was no evidence of necrosis or mitotic figures. Immunohistochemistry (IHC) showed strong positivity for oestrogen and progesterone receptors, supporting the diagnosis. IHC also demonstrated diffuse cytoplasmic vimentin expression and focal desmin expression, while S-100 protein was negative [Table/Fig-4-6]. Due to the high risk of recurrence, the patient was started on monthly GnRH agonist therapy for three months post-surgery. She was prescribed Leuprolide, 3.75 mg, subcutaneously, once every 28 days for 12 months. At the six-month follow-up, the patient remained asymptomatic, with no clinical or radiological evidence of recurrence.



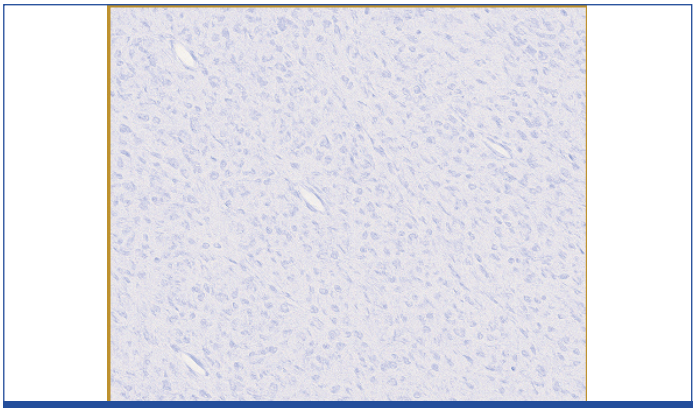
[Table/Fig-4]: Photomicrograph- revealed hypocellular, myxoid stroma (Red arrow) with loosely arranged spindle-shaped tumour cells (Black arrow) possessing elongated nuclei and bland chromatin. The cytoplasm appeared pale and eosinophilic and no significant atypia or mitotic activity was noted {Haematoxylin and Eosin (H&E) 400x}.



[Table/Fig-5]: Image demonstrating strong, diffuse cytoplasmic immunoreactivity for desmin in spindle-shaped stromal tumour cells (Red arrow) arranged in a myxoid background. The brown chromogen deposits indicate positive desmin expression, confirming myofibroblastic differentiation. Staining was performed with Monoclonal mouse anti-human Desmin antibody (Clone D33) and detection was performed using DAB (diaminobenzidine) chromogen and haematoxylin counterstain (IHC, 400x).

DISCUSSION

Vulvar masses are frequently encountered in clinical practice, with most presumed to be benign lesions such as Bartholin's cysts,



[Table/Fig-6]: Image showing negative immunoreactivity for S-100 protein in the tumour cells. The absence of brown chromogenic signal in the cytoplasm and nuclei of the spindle cells indicates lack of S-100 expression, excluding differential diagnoses such as neural tumours and myxoid liposarcoma, which typically exhibit S-100 positivity.Staining was performedusing Polyclonal rabbit anti-human S-100 antibody and detected based on DAB chromogen with haematoxylin counterstain (IHC, 400x).

lipomas, or epidermoid inclusions. However, on rare occasions, mesenchymal neoplasms with locally infiltrative potential may be mistaken for these common entities, leading to underdiagnosis and suboptimal treatment. AA is one such rare, benign mesenchymal tumour that predominantly affects women of reproductive age and is clinically significant for its deep local infiltration and high risk of recurrence despite its histologically bland appearance [2].

First described by Steeper TA and Rosai J in 1983, it was later reclassified by the World Health Organisation as "deep angiomyxoma" in 2003 [3]. With fewer than 350 cases documented in the literature, its incidence remains low and its slow-growing, indolent nature often leads to delayed or incorrect diagnoses [4,5]. An overview of reported cases of AA, highlighting variations in clinical presentation, diagnostic challenges, treatment approaches and follow-up outcomes is provided in [Table/Fig-7] [6-10].

The AA is believed to arise from pluripotent mesenchymal cells capable of myxoid matrix production and smooth muscle differentiation [11]. At the molecular level, a significant proportion of cases demonstrate chromosomal rearrangements involving 12q13-15, specifically involving the High Mobility Group AT-hook 2 (HMGA2) gene, which encodes a non histone architectural transcription factor implicated in mesenchymal tumourigenesis. Aberrant HMGA2 expression, typically due to gene rearrangement or amplification, promotes dysregulated growth through alteration of chromatin structure and downstream transcriptional control [12]. Immunohistochemically, AA frequently expresses oestrogen and progesterone receptors, supporting its hormone-dependent behavior and explaining its growth acceleration during pregnancy or hormonal shifts.

In the 2024 WHO Classification of Soft Tissue and Bone Tumours, AA remains categorised under the group of benign fibroblastic and myofibroblastic tumours, but with a more clearly defined clinical and molecular profile compared to the 2020 edition [13]. The updated classification highlights HMGA2 gene rearrangements, particularly those involving 12q14-15, as a consistent genetic hallmark of AA. This finding reinforces its mesenchymal origin and serves as a valuable diagnostic tool in morphologically ambiguous cases [14]. The 2020 edition described AA primarily by its histological characteristics, including a hypocellular myxoid stroma, bland spindle and stellate cells and prominent vasculature. In contrast, the 2024 revision broadens the diagnostic criteria by incorporating hormonal receptor positivity for oestrogen and progesterone, along with evidence of local invasiveness, to better distinguish it from superficial angiomyxoid tumours [15].

The AA typically presents as a slow-growing, painless soft tissue mass located in the vulva, perineum, or deep pelvic region. The clinical symptoms are often vague and non specific, including dull pelvic

Author and year	Age/ sex	Duration of swelling	Clinical features	Histopathological profile	Immunohistochemical (IHC) profile	Radiological profile	Earlier misdiagnosis and treatment	Recurrence/ follow-up/ outcome
Sethi P et al., 2023 [6]	32/F	18 months	Gradually enlarging painless vulvar mass; no discharge or systemic symptoms	Hypocellular myxoid stroma with abundant thin-walled blood vessels; spindle-shaped cells without atypia	Desmin+, CD34+, ER+, PR+; S-100–, SMA–.	MRI: Large, hyperintense mass on T2 with swirled pattern; no infiltration	Initially suspected as Bartholin's cyst; wide excision performed	No recurrence at 12-month follow-up
Kumar A et al., 2022 [7]	45/F	3 years	Soft, lobulated mass in the left labia majora; slowly progressive.	Myxoid matrix, numerous vessels with thick and thin walls, scattered stellate cells	ER+, PR+, CD34+; negative for S-100, cytokeratin.	MRI: Well-defined mass with high T2 signal intensity and internal septations	Treated initially as lipoma; incomplete excision led to recurrence	Local recurrence at 18 months; re-excised with clear margins
Liang Y et al., 2021 [8]	38/F	8 months	Mobile, painless mass in vulva; occasional discomfort on sitting	Loose myxoid stroma with spindle and stellate cells; no necrosis or mitosis	CD34+, desmin+, ER+, PR+; Ki-67 <1%.	Ultrasound: hypoechoic soft tissue mass; MRI: T2-bright well-circumscribed lesion	Provisional diagnosis of fibroepithelial polyp; local excision.	No recurrence at 10-month review
Patel Z et al., 2024 [9]	28/F	2 years	Firm vulvar mass; increasing size, pressure symptoms	Low-cellularity myxoid tumour with fibrovascular bundles and minimal atypia	ER+, PR+, CD34+; SMA focally+, S-100–	MRI: Lobulated T2-bright lesion with finger-like projections	Misdiagnosed as Aggressive Angiomyxoma (AA); managed with wide excision	Disease-free at 14 months; no evidence of metastasis
Yadav P et al., 2023 [10]	41/F	6 years	Recurrent swelling post local excision; mild pain	Hypocellular tumour with prominent vascularity, myxoid matrix, spindle cells	Desmin+, CD34+, ER+, PR+; Ki-67 <2%, S-100–	MRI: T2-hyperintense mass with infiltration of adjacent tissue planes	Initially excised as benign cyst; recurrence due to incomplete excision	Re-excised with clear margins; no recurrence at 2-year follow-up
Present case, 2025	32/F	3 years	Non tender soft swelling over right labium majora (5x6 cm), lateral to introitus; no regional lymphadenopathy; normal uterus and cervix	Sparsely cellular lesion with spindle and stellate-shaped cells in a myxoid matrix; numerous thin-walled blood vessels; no mitosis or necrosis	Desmin+, (Diffuse cytoplasmic) Vimentin+, ER+, PR+; S-100–, SMA–; Ki-67 <1%	MRI: Well-defined lobulated mass with high T2 signal; minimal infiltration	Initial clinical impression was benign vulvar soft tissue tumour; complete excision performed	No recurrence at 11-month follow-up; patient remains asymptomatic

[Table/Fig-7]: Summary of reported cases of Aggressive Angiomyxoma (AA) based on clinical characteristics, management and outcomes [6-10].

discomfort, dyspareunia, or pressure-related urinary complaints, which may be attributed to mass effect [16]. Unlike superficial lesions such as Bartholin's cysts, which are usually fluctuant, tender and localised near the posterior vaginal introitus, AA tends to be firm, non tender and deeply seated with poorly defined borders. Similarly, while lipomas and epidermoid cysts are well-circumscribed and mobile, AA is often ill-defined and infiltrative, lacking a discrete capsule [17]. In the present patient, the unusually prolonged duration, coupled with progressive enlargement, prompted surgical referral after several years of symptomatic neglect.

Histologically, AA presents as a paucicellular neoplasm composed of bland spindle-shaped and stellate cells embedded within a loose, gelatinous myxoid background. The stroma is rich in delicate collagen fibres and typically traversed by numerous thin-walled, branching blood vessels. Mitotic figures are rare and cellular atypia is generally absent, contributing to its benign histologic appearance despite its locally aggressive behavior. Occasionally, areas of edema and entrapped adipose tissue may also be observed [18]. Superficial angiomyxoma is a histological variant that closely resembles AA but remains clinically distinct, as it demonstrates benign behavior, well-defined margins and lacks significant infiltrative capacity [Table/Fig-8] [19].

The AA consistently demonstrates strong positivity for vimentin, desmin, and estrogen and progesterone receptors, supporting its myofibroblastic origin and hormonal responsiveness. It is typically negative for S-100, cytokeratins, and CD31, helping to exclude neural, epithelial, and vascular tumours [20]. Immunohistochemical staining plays a critical role in differentiating AA from histologically similar mesenchymal tumours of the vulvoperineal region [Table/Fig-9] [21-23].

Imaging is essential in the preoperative evaluation of AA, particularly for delineating tumour extent and guiding surgical planning. Ultrasonography may reveal a well-defined, hypoechoic soft tissue

Parameters	Superficial angiomyxoma	Aggressive Angiomyxoma (AA)
Typical location	Usually involves the dermis or subcutis of trunk, head, neck, or genital region; often superficial.	Arises in deep soft-tissues of the pelvis, perineum and vulva in reproductive-age women.
Clinical behaviour	Benign, non infiltrative, with low recurrence risk after complete excision.	Locally aggressive, infiltrative growth with high recurrence rate even after wide excision.
Histopathology	Myxoid stroma with spindle to stellate cells, neutrophilic infiltrates and thin-walled vessels.	Hypocellular myxoid matrix with spindle and stellate cells, prominent thick and thin-walled blood vessels, lacking inflammatory infiltrates.
Immunohistochemistry (IHC)	Often positive for CD34 and vimentin; variable S-100; hormone receptor negative.	Positive for desmin, CD34, vimentin, ER and PR; negative for S-100.
Recurrence	Low; typically curative with local excision.	High (30–72%) due to infiltrative margins; long-term surveillance required.
Radiological features	Well-circumscribed superficial lesion; low T1 and high T2 signal on MRI.	Deep-seated mass with a swirled or laminated pattern on MRI; T2 hyperintense with ill-defined borders.

[Table/Fig-8]: Clinico-pathological profile of superficial angiomyxoma and Aggressive Angiomyxoma (AA) vulva [19].

mass, but its sensitivity in assessing infiltration is limited [24]. Magnetic Resonance Imaging (MRI) is considered the imaging modality of choice, as it offers superior soft tissue resolution and characteristically demonstrates a laminated or "swirled" pattern caused by intersecting fibrous stroma within a myxoid matrix. This appearance, typically seen on T2-weighted sequences, can aid in differentiating AA from more superficial or encapsulated vulvar lesions [25]. Moreover, MRI allows precise mapping of deep pelvic extension and involvement of adjacent structures, which is critical for surgical margin assessment

Feature	Aggressive Angiomyxoma (AA)	Bartholin's cyst	Fibroepithelial polyp	Angiomyofibroblastoma (AMFB)	Myxoid liposarcoma
Nature	Benign but locally aggressive mesenchymal tumour	Benign cystic lesion	Benign reactive lesion	Benign mesenchymal tumour	Malignant soft-tissue sarcoma
Age group (in years)	Reproductive-age females (20-40 years)	Any age, common in reproductive age	Reproductive age	30-50 years	40-60 years
Location	Vulva, perineum, pelvis	Posterolateral vulva (Bartholin's gland area)	Vulva, cervix, vagina	Superficial vulvovaginal region	Deep soft tissues of extremities, rare in vulva
Growth pattern	Slow-growing, large, infiltrative, locally recurrent	Rapid, fluctuant, small to moderate	Small, pedunculated	Well-circumscribed, small	Infiltrative, rapidly growing
Clinical presentation	Painless, soft, ill-defined vulvar mass	Painful or painless swelling; may become tender if infected	Soft, polypoid mass	Well-circumscribed painless mass	Deep, painless enlarging mass
Consistency	Soft to gelatinous	Cystic	Soft, mobile	Soft, well-circumscribed	Soft to firm
Imaging (MRI/CT)	Hypo- to isointense on T1, hyperintense on T2; swirled/streaky pattern; infiltrative margins	Well-defined cystic lesion	Not routinely imaged	Well-defined, homogenous	Myxoid matrix with septa, 'pseudocapsule'
Histology	Hypocellular, myxoid stroma, spindle to stellate cells, thick-walled vessels, infiltrative	Cyst lined by squamous or transitional epithelium	Loose fibrovascular stroma, surface squamous epithelium	Alternating hyper/hypocellular areas, thin-walled capillaries, plump stromal cells	Myxoid background, lipoblasts, delicate vasculature ('chicken wire')
Immunohistochemistry (IHC)	+Vimentin, +Desmin, +ER/PR, +CD34 (variable), SMA+, S-100–	Negative or non specific	Vimentin+, Desmin–, ER/PR–	+Desmin, +Vimentin, +ER/PR, +SMA, CD34 variable	+Vimentin, ±S-100, MDM2+, CDK4+
Margins	Poorly defined, infiltrative	Well-circumscribed	Well-defined	Well-circumscribed	Infiltrative
Recurrence	High local recurrence (30-70%) even after excision	Rare if drained or excised completely	Rare	Rare	High recurrence if not excised widely
Metastatic potential	Rare to none	None	None	None	High (haematogenous spread)
Management	Wide local excision; long-term follow-up due to recurrence; hormonal therapy (GnRH agonists) may help	Drainage or marsupialisation; antibiotics if infected	Simple excision	Local excision is curative	Wide excision ± chemotherapy/ radiotherapy
Prognosis	Good with long-term follow-up; recurrence common	Excellent	Excellent	Excellent	Variable; depends on grade and treatment

[Table/Fig-9]: Comparative features of Aggressive Angiomyxoma (AA) and its common benign and malignant differentials [21-23].

and long-term follow-up. Recent advances in imaging, including diffusion-weighted MRI and dynamic contrast-enhanced MRI, have improved the characterisation of AA by highlighting its low cellularity and gradual vascular enhancement, which help distinguish it from malignant myxoid tumours. Emerging tools such as radiomics and texture-based MRI analysis are also being explored for non-invasive prediction of recurrence risk and treatment response, although their clinical use remains investigational [26].

When the tumour is well-circumscribed and confined to superficial soft tissues with minimal invasion, wide local excision with clear margins is feasible and often curative [27]. However, deeply infiltrative lesions involving the pelvic floor, perineal musculature, or adjacent viscera may preclude complete excision without significant functional compromise. In such cases, a more conservative debulking approach may be adopted, particularly when the tumour is hormone receptor-positive. Laparoscopic or combined abdominoperineal approaches are occasionally employed for high pelvic or retroperitoneal extension. There are currently no universally accepted consensus on a specific safe margin for excising AA, due to its ill-defined, infiltrative nature and the rarity of the tumour [28]. The surgical strategy is thus individualised based on tumour location, depth of invasion and hormonal responsiveness, balancing oncological control with preservation of continence, sexual function and quality of life.

Hormonal therapy has emerged as an important adjunct in the management of AA, particularly in tumours that express oestrogen and progesterone receptors. Gonadotropin-Releasing Hormone (GnRH) agonists, such as leuprolide and goserelin, have been used effectively in both the neoadjuvant and adjuvant settings [29]. In the neoadjuvant context, these agents can induce tumour regression by downregulating gonadotropin secretion and reducing circulating oestrogen levels, which in turn decreases the hormonal stimulation of the tumour. This reduction in size may facilitate less extensive surgical resection and improve preservation of surrounding structures. In the adjuvant setting, GnRH agonists are considered in cases with positive or close surgical margins, residual disease, or high risk of recurrence. They may also serve as an alternative in select patients who are poor surgical candidates or in whom complete excision is not feasible [30].

Current guidelines, including the National Comprehensive Cancer Network (NCCN) and the International Federation of Gynaecology and Obstetrics (FIGO), do not recommend routine use of systemic chemotherapy or radiotherapy for AA, as these modalities have demonstrated minimal efficacy in reducing tumour burden or preventing recurrence. Radiotherapy is considered only in exceptional cases where hormonal therapy fails and surgery is contraindicated [31]. AA is considered histologically benign but clinically unpredictable due to its high propensity for local recurrence. Reported recurrence rates range from 30% to 72%, often occurring within the first five

years following surgery, particularly in cases with positive or close surgical margins or incomplete resection [32]. The tumour's deeply infiltrative nature and lack of encapsulation contribute to this recurrence risk, even in patients with apparently complete excision. Although metastatic transformation is exceedingly rare, long-term surveillance is essential. Current follow-up protocols recommend pelvic examinations every 6 to 12 months, supplemented by annual or biennial pelvic MRI, especially in hormone receptor-positive tumours or those managed conservatively [32]. Overall, the prognosis is favourable in terms of survival, with no mortality reported from the tumour itself, but lifelong follow-up is advised due to its chronic, relapsing potential.

CONCLUSION(S)

The AA, though infrequent, must be included in the differential diagnosis of vulvovaginal masses, especially in women of reproductive age. Its slow, infiltrative growth and frequent clinical resemblance to benign entities often lead to diagnostic delays. The definitive diagnosis hinges on histopathological evaluation supported by immunohistochemical profiling, while cross-sectional imaging is essential for assessing tumour extent and surgical planning. A multidisciplinary treatment strategy combining conservative surgical excision with hormone-modulating therapy in receptor-positive cases can yield favourable outcomes. However, given the tumour's notorious propensity for delayed local recurrence, long-term clinical and radiological surveillance is imperative.

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