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Presenting patterns and management of aggressive angiomyxoma and cellular angiofibroma in the perineum and pelvic spaces

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ABSTRACT

Background: Angiomyxoma is a rare, slow-growing mesenchymal tumour occurring in the vaginal wall and perineum, but also in the deep spaces of the pelvis, commonly in premenopausal women. Cellular angiofibroma is also a slow-growing benign tumour, mostly superficial, and is commonly seen in men. Histopathology plays a crucial role in diagnosis between the two, and the mainstay treatment option is surgery in both. We aimed to understand the presentation features of this rare group of tumours occurring in the pelvic and perineal spaces.
Method: This is a retrospective analysis from a colorectal unit of a tertiary care teaching hospital. All consecutive patients diagnosed with angiomyxoma and angiofibroma of the pelvic spaces between April 2025 and March 2026 were included. Clinical presentation, diagnostic and treatment details were collected from electronic hospital records.
Results: Four patients were included. Three patients were symptomatic with a slow-growing swelling in the vagina or perineum, while one asymptomatic patient was diagnosed incidentally on a routine ultrasound scan. Magnetic Resonance Imaging of pelvis was the imaging modality of choice in all patients. Most patients underwent a preoperative biopsy to confirm the diagnosis. Two patients were diagnosed with deep infiltrating angiomyxoma on histopathology, one patient was diagnosed with cellular angiofibroma, and one patient had overlapping features of both angiomyxoma and angiofibroma on biopsy. Three patients underwent excision surgery, while one patient is planned for surgery.
Conclusion: Aggressive angiomyxoma and cellular angiofibroma are rare tumours with many overlapping clinical and histological features and should be considered in the differential diagnosis based on their presentation. Preoperative biopsy can be diagnostic and immunohistochemistry studies can help in reaching the diagnosis. Complete surgical excision is the definitive treatment.

Keywords: *Aggressive angiomyxoma, cellular angiofibroma, Immunohistochemistry, Perineum, Pelvic spaces, Mesenchymal tumour.*

INTRODUCTION

Aggressive Angiomyxoma is a rare, slow-growing mesenchymal tumour mostly found in the pelvic soft tissue spaces, vagina or perineum.It is shown to have a female preponderance with a male-to-female ratio of 1:6 and affects women of reproductive age(1). It presents as an insidious, painless growth with features of local infiltration, high recurrence, and rare distant metastasis(1,2). Cellular angiofibroma is another benign mesenchymal lesion affecting the genital regions, such as the vulval region in women and the inguinoscrotal region in men, but can also occur in other regions, such as the perineum(3). Although it shares some pathological features with angiomyxoma, the nature of this disease suggests a low risk of recurrence or metastasis(3). Histopathology plays a crucial role in the diagnosis of the two, and surgery is the mainstay treatment option for both. We aimed to understand the presentation features of this rare group of tumours occurring in the pelvic and perineal spaces and to compare their similarities and differences to better understand this uncommon disease.

METHODS AND MATERIALS

This was a retrospective analysis of data from the colorectal unit of a tertiary care teaching hospital. All consecutive patients diagnosed with angiomyxoma or angiofibroma of the pelvic spaces between April 2025 and March 2026 were included. Inclusion criteria were a diagnosis of either angiofibroma or angiomyxoma, based on biopsy report and pelvic or perineal location of the lesion. No cases were excluded. Clinical presentation features, diagnostic tests and treatment details were collected from electronic hospital records.

RESULTS

Four patients were included in the study. Two were male, and the other two were female patients. All patients presented in the fifth and sixth decades of life. Three patients were symptomatic with symptom duration ranging from 2 months to 4 years. One patient was asymptomatic and diagnosed on a routine gynaecology ultrasound scan. Magnetic Resonance Imaging (MRI) of pelvis was the imaging modality of choice in all patients. The largest dimension of the lesion ranged from 10 cm to 19 cm. Three patients underwent preoperative biopsy for diagnosis. One patient underwent fine needle aspiration cytology. The disease characteristics among study patients are tabulated in Table 1.

Table 1: Disease presenting patterns among study patients

Patients	Patient 1	Patient 2	Patient 3	Patient 4
Age at presentation	41	58	53	45
Sex	Female	Female	Male	Male
Presenting symptom	Swelling in vagina	Asymptomatic	Perineal swelling and pus discharge	Perineal swelling
Duration of symptoms	10 months	Zero	2 months	4 years
Imaging done	Magnetic Resonance Imaging	Magnetic Resonance Imaging	Magnetic Resonance Imaging	Magnetic Resonance Imaging
Location of lesion	Vulva	Left paravesical space	Left ischiorectal fossa	Left ischiorectal fossa and perineal fat
Largest dimension of lesion	10 cm	19 cm	11 cm	15 cm
Preoperative Biopsy or fine needle aspiration report	Fine needle aspiration - Inadequate smears	Hypocellular spindle cell neoplasm with myxohyaline matrix and increased vascularity suggestive of aggressive angiomyxoma	Spindle cell neoplasm suggestive of cellular angiofibroma	Benign spindle cell neoplasm with histologic and immunoprofile more in favour of cellular angiofibroma over deep aggressive angiomyxoma
Surgery done	Excision of labial lesion	Transabdominal excision	Excision in prone position	Planned for excision
Final histopathology	Deep aggressive angiomyxoma	Deep aggressive angiomyxoma	Cellular angiofibroma	Not appliable

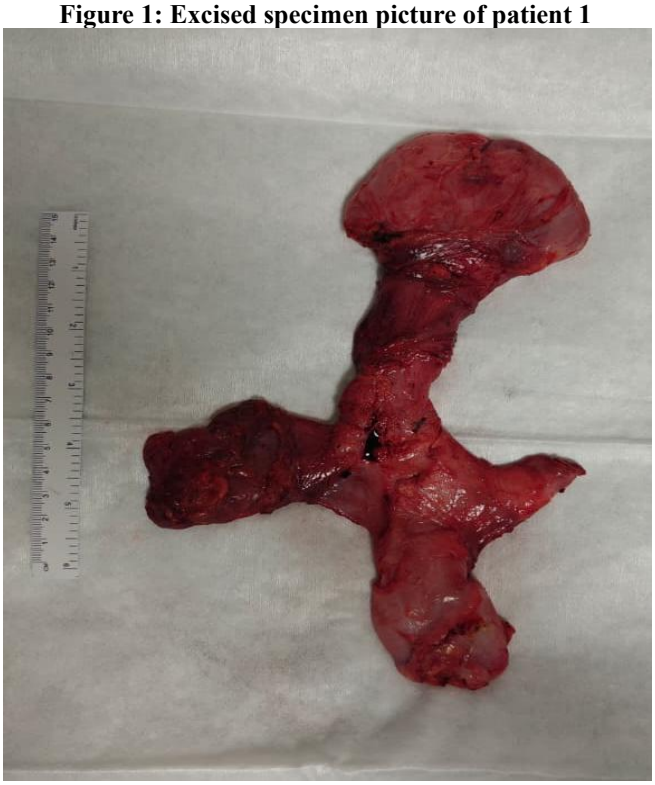
Three patients underwent excision surgery and one patient was advised surgery and is planned for the same. Two patients had final histopathology reported as deep aggressive angiomyxoma, and one patient was diagnosed with cellular angiofibroma. The last patient had a preoperative biopsy with features more in favour of cellular angiofibroma over deep aggressive angiomyxoma. He was advised excision surgery and planned for the same. A concise review of each study patient is given below.

Patient 1

A 41-year-old woman presented with swelling in the right labia for 10 months. It was an insidious onset, painless, gradually progressive swelling with no prior history of trauma and no associated symptoms. She had no diagnosed comorbidities. On clinical examination, she had two swellings in the right labia pushing against the vaginal introitus. The superior swelling was 10 cm by 8 cm in size, soft in consistency, and the inferior swelling was 6 cm by 4 cm and also soft in consistency. Vaginal exam and rectal exam were normal.

She was evaluated with an MRI Pelvis, which showed a dumbbell-shaped lesion in the right labia with extension into the right gluteal region. The largest dimension of the lesion was 10 cm. Fine needle aspiration cytology was not contributory to the diagnosis. She underwent an excision biopsy in lithotomy position under general anaesthesia. Intraoperatively, both the swellings seen clinically were part of a single dumbbell-shaped lesion with lateral extension, which was excised as a single specimen. The excised specimen is shown in Figure 1, and the final histopathology was reported as a deep aggressive angiomyxoma. She was doing well at the 6-month follow-up.

Figure 1: Excised specimen picture of patient 1



Patient 2

A 58-year-old female was diagnosed with a pelvic mass on a routine ultrasound exam during a gynaecology evaluation. She was further evaluated with an MRI Pelvis, which revealed a 19 cm solid lesion in the left paravesical space, with no areas of restricted diffusion within. Posteriorly, the lesion was piercing the left mesorectal fascia and traversing the left mesorectal space. Ultrasound-guided biopsy was done, which was reported as a hypocellular spindle cell neoplasm with myxo-hyaline matrix and increased vascularity. These features favoured aggressive angiomyxoma. She underwent transabdominal excision, and final histopathology was confirmed to be deep aggressive angiomyxoma. She is on surveillance follow-up.

Patient 3

A 53-year-old male presented to the outpatient department with swelling in the perineal region associated with pus discharge for 2 months. It was associated with dull aching pain in the region. He had no prior surgeries. Clinical exam revealed an 8 cm by 5 cm soft swelling in the left ischiorectal fossa with an extra mucosal bulge palpable on rectal exam. MRI Pelvis revealed a solid mass in the left ischiorectal fossa, likely originating from the left urogenital diaphragm, with loss of plane with the left side of the external anal sphincter and left puborectalis muscle. He underwent excision of the lesion in prone position under general anaesthesia. Surgical histopathology was reported as cellular angiofibroma, and the patient was advised to be on follow-up.

Patient 4

A 45-year-old male presented with an insidious onset, gradually progressive perineal swelling for 4 years. He had no associated symptoms or discharge from the swelling. On clinical examination, he had a 20 cm by 10 cm swelling in the left ischiorectal fossa extending into the perineal region. Digital rectal examination was normal. MRI pelvis revealed a dumbbell-shaped mass with the largest dimension of 15 cm, with epicentre in the left puborectalis. The swelling extended superiorly to the left ischiorectal fossa and inferiorly into the perineal fat. Core biopsy revealed benign spindle cell neoplasm. The spindle cells were focally positive for desmin, diffuse positive for Cluster of Differentiation 34 (CD 34) and estrogen receptor (ER) and negative for Signal Transducer and activator of Transcription 6 (STAT 6) and Smooth Muscle Actin(SMA). The histologic features and immunoprofile were more in favour of cellular angiofibroma over deep aggressive angiomyxoma. He was advised excision surgery and is planned for the same.

DISCUSSION

The term "aggressive angiomyxoma" was first used by Steeper and Rosai (4) to describe this rare, slow-growing mesenchymal tumour with a predilection for the female pelvis. They commonly present in vulva, perineum, gluteal region, retroperitoneum and deep pelvic spaces such as paravaginal, perirectal and ischiorectal spaces. This tumour has been recorded in patients with ages ranging from 16 to 70 years, with a peak incidence in females in the fourth decade(5). The cells exhibit fibroblastic and myofibroblastic features and are hormonally responsive(5).

Clinical presentation symptoms may be non-specific, and a high index of suspicion is required while treating such lesions. It presents as a slow-growing swelling, sometimes asymptomatic, while otherwise accompanied by tenderness, dyspareunia, or urinary complaints(6). Commonly mistaken differentials include lipoma, myxoma, Bartholin cyst, and vulval abscess (1). MRI of the pelvis gives information on the origin, extent of the lesion and local invasion.

A biopsy is performed to confirm the diagnosis. In superficial swelling, a true cut biopsy is feasible, and in deep-seated lesions, image-guided biopsy (ultrasound or computed tomography) can be performed. The cut surface can be translucent or congested, myxoid to fibrous or gelatinous, with cystic degeneration, focal yellow areas resembling adipose tissue, and haemorrhagic areas (6). Microscopic examination shows spindled stromal cells and a prominent vascular component set in a myxoid matrix. The lesional spindle cells may show eosinophilic cytoplasm and round to ovoid nuclei. Medium to large-sized vessels may be dispersed throughout the tumour, and perivascular hyalinization may be present. Immunohistochemistry(IHC) markers such as desmin, vimentin, SMA, CD 34, Cyclin-dependent kinase 4(CDK 4), Progesterone receptor (PR), ER can be positive but mostly negative for IHC marker Soluble in 100% ammonium sulfate(S-100), and Cluster of Differentiation 68(CD 68), which was similar to the histopathology reports seen in our study patients(7).

Optimal management involves surgical excision with clear margins. The surgical approaches can be transvaginal or transperineal. Large tumours, such as those seen in patient 4 in our study, may require abdominoperineal resection (8). Adjacent pelvic organ resection, such as rectal resection, pelvic side wall or bladder dissection, may be required in large infiltrating lesions. The risk of local recurrence is 30%, with positive surgical margins being an important risk factor to predict recurrence (9). Late recurrence up to 15 years after initial presentation has been reported, and though rare, distant metastasis has also been reported (2,4).

Cellular angiofibroma is another benign mesenchymal tumour occurring in the genital and perineal region, described in 1997 under the term "cellular angiofibroma," emphasising the cellular spindle cell component and prominent blood vessels (10). It is reported to have equal incidence in both men and women, with a peak in the seventh decade in men and earlier in the fifth decade in women (3).

It usually presents as a painless, slow-growing swelling in the vulva, inguinoscrotal or perineal region, but has also been reported in extra-genital sites such as the retroperitoneum, anus, prostate, and even the hypopharynx (11,12). MRI Pelvis was the diagnostic imaging performed in our study patients. Preoperative biopsy confirmation is useful in planning treatment and was performed in our study patients.

The tumour is well circumscribed on cut section, with gelatinous and myxoid appearance. Microscopic examination shows spindle cells arranged in sheets or loose fascicles, as seen in our patient 3. Stroma can be oedematous with conspicuous vascularity and can also have hyalinized vessels, and adipose tissue seen in parts of the well-circumscribed tumour. Mitotic activity can be high. The stromal cells were reported in the literature to be positive for vimentin and negative for S-100 protein, actin, epithelial membrane antigen and desmin, suggesting fibroblastic differentiation (10). In our patient 3, CD34 showed diffuse positivity, with patchydesmin positivity.

In contrast to aggressive angiomyxoma, this tumour has a low recurrence rate and little to nil metastatic risk (3). The presence of atypical or sarcomatous features has been reported in the literature, but does not correlate with recurrence risk or aggressive biological behaviour (13). Complete surgical excision is the recommended treatment. Due to the well-circumscribed nature, the need for extensive adjacent organ resection or damage to adjacent structures is less.

This study highlights the clinical presentation and diagnostic challenges in evaluating these two rare mesenchymal tumours. Both aggressive angiomyxoma and cellular angiofibroma have overlapping clinical and histological features, with both lesions occurring in the pelvic space, the genital and perineal regions. There are differences in sex predilection, aggressiveness of the lesion, recurrence risk, and radicality of surgery required for treatment. Although both entities share similar morphological features, IHC can help differentiate between them. Both show positivity for vimentin, but differ in the expression of hormone receptors, muscle markers, and CD34. The role of preoperative biopsy in prognostication and surgical planning is also highlighted in this study. This also helps counsel patients on the extent of surgical resection required, acceptable morbidity, and recurrence risk. This, in turn, will help improve patient care, manage expectations, and enhance patient satisfaction.

LIMITATIONS

This study included a small number of patients because of the rarity of the disease. At the time of publication, the study patients had completed a six-month follow-up, and longer-term follow-up may be useful in understanding the nature of the disease and surgical outcomes.

CONCLUSION

Aggressive angiomyxoma and cellular angiofibroma are two distinct clinical entities with overlapping clinical and histological features but different biological behaviour, which, in turn, has implications for surgical planning. Both lesions present as indolent, slow-growing swellings in the genital, perineal or pelvic spaces, which can even have an asymptomatic course. MRI Pelvis is the preferred imaging modality for assessing the extent of the lesion. Preoperative biopsy, either as a true cut biopsy or an image-guided biopsy, in deeper pelvic lesions is recommended to obtain a diagnosis and plan treatment. IHC studies help differentiate the entities pathologically. Complete surgical excision is the definitive treatment for both lesions.

Author Contributions: RPS, RR and MRJ were involved in the conception and design of the study. RPS contributed to data acquisition and the drafting of the article. RPS and RR contributed to the analysis of acquired data. RR and MRJ contributed to the revision of the manuscript for critically important intellectual content. RPS, RR and MRJ were the consultants involved in the care of study patients. MRJ was the head of the treating surgical unit at the time of the study and the overall facilitator of the study.

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