

INTRODUCTION

Aggressive angiomyxoma (AAM) is a rare, benign but locally infiltrative soft tissue tumour, typically arising in the female pelvis and perineum (1). With fewer than 450 reported cases, it is often misdiagnosed. This case report describes a woman with a vaginal bulge initially thought to be a rectocele, later identified as AAM after imaging and histological analysis. The report highlights diagnostic challenges and underlines the need for clinical awareness to ensure timely and appropriate management.

CASE PRESENTATION

A woman in her forties presented with vaginal bulge and associated discomfort. Key information is highlighted in Figure 1. Grade 2 rectocele was diagnosed but management with pessary failed. Upon review by physiotherapist, she was referred for surgical repair.

Further examination by a urogynaecologist revealed a 6-7 cm fluctuant mass arising from the posterior vaginal wall (Figure 2). Primary vaginal cyst was suspected and differential diagnoses are shown in Table 1.

MRI pelvis revealed a 9x6x3.5 cm unilocular lesion arising from the posterior vaginal wall/rectovaginal septum, with high T2 signal and low to isointense T1 and STIR signals, suggestive of developmental cyst (Figure 3). Colorectal multidisciplinary team (MDT) input was sought and their investigations showed no bowel involvement. Gynaecology MDT deemed the lesion benign.

History

42 years old
BMI 38
3 previous vaginal deliveries
Hypothyroidism on levothyroxine
Previous IIIA obstetric anal sphincter injury

Examination

6-7cm fluctuant mass arising from the posterior vaginal wall
No significant uterine descent
No rectovaginal septal defect on rectal examination

Investigations

MRI pelvis revealed a 9x6x3.5 cm unilocular lesion arising from the posterior vaginal wall/rectovaginal septum
Flexible sigmoidoscopy and defecating proctogram showed no bowel involvement

Figure 1 Key points summarised from patient's history, clinical examination and investigations performed



Figure 2 No communication with between lesion and rectum demonstrated on digital rectal examination

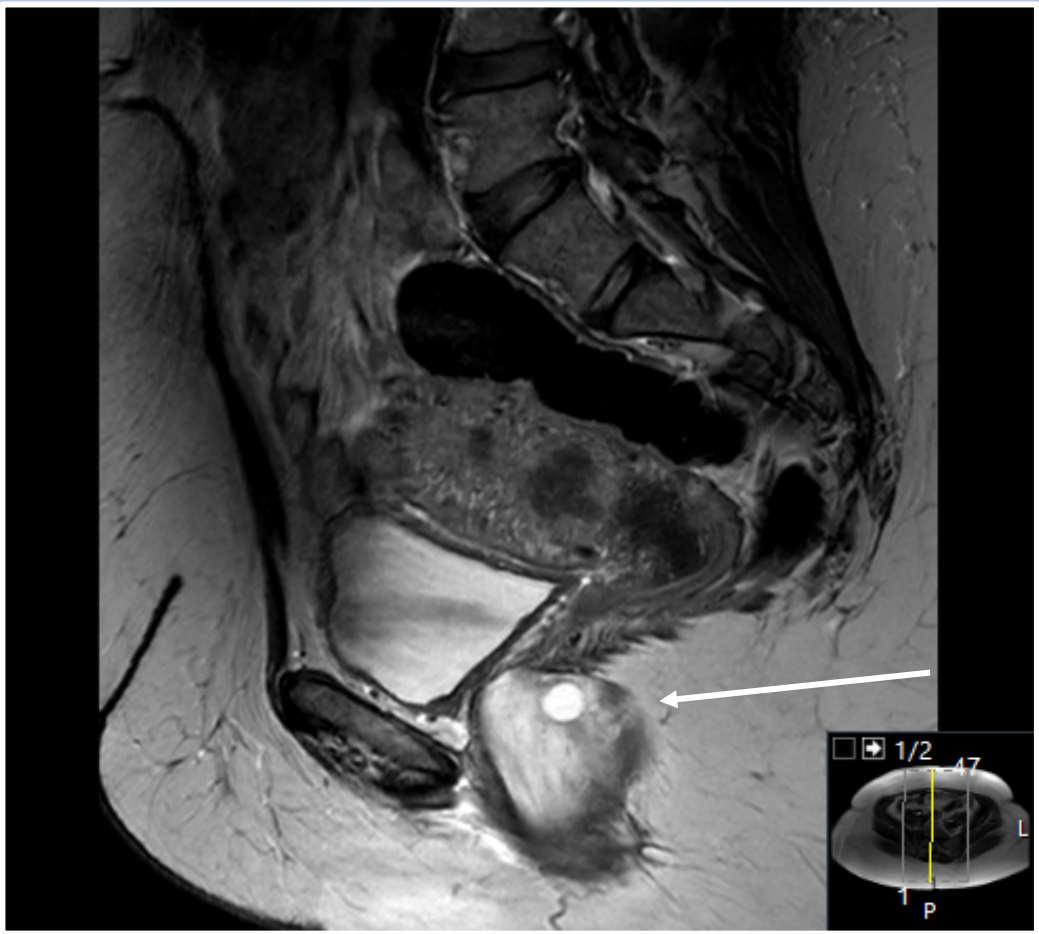


Figure 3 Coronal MRI image of pelvis with arrow identifying lesion

An Unusual Presentation Of Aggressive Angiomyxoma Arising From The Posterior Vaginal Wall

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TREATMENT

The patient consented to surgical excision of the lesion whilst prioritising preserving rectal integrity despite the potential for incomplete excision. Intraoperatively, a transverse incision was made on the vaginal wall and the lesion was dissected whilst taking care to avoiding bowel injury (Figure 4). The lesion's stalk was difficult to delineate but it was excised as much as possible (Figure 5). The patient recovered well post-operatively and was discharged with analgesia and antibiotics. The lesion was sent for histological analysis (Figure 6).

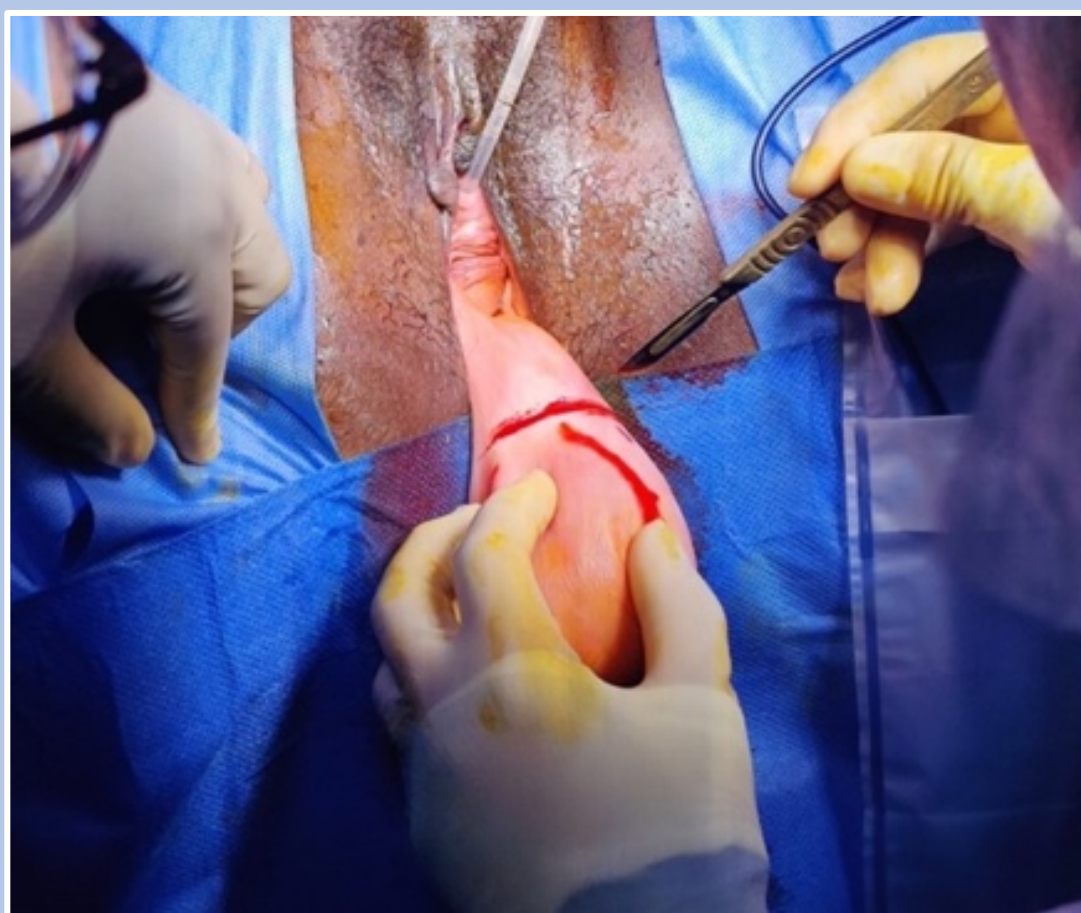


Figure 4 Transverse elliptical incision made on posterior vaginal wall overlying the lesion

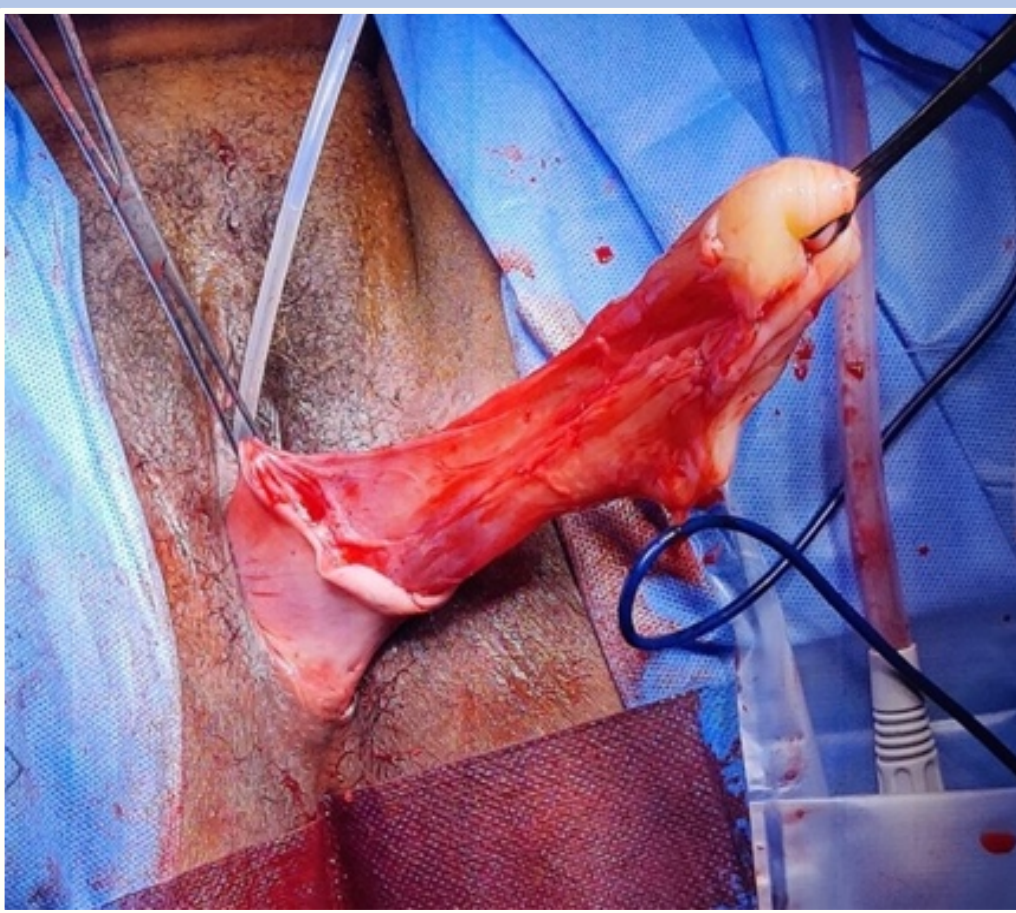


Figure 5 Lesion not clearly demarcated at its stalk



Figure 6 Excised lesion

OUTCOME AND FOLLOW-UP

The patient recovered well at follow-up at 1 week and 4 weeks. Histological examination confirmed the lesion as an un-encapsulated, locally infiltrative mesenchymal benign tumour, consistent with AAM. The diagnosis was shared with the patient and a 4-month follow-up with a repeat MRI is planned to check for recurrence or residual tumour.

LEARNING POINTS

1. Differentiating between a cystic vaginal wall structure and uterovaginal prolapse can be challenging.
2. Rectal examination and MRI help clarify the origin of the bulge.
3. Multidisciplinary discussions with colorectal and oncology teams aid in planning investigations and surgery.
4. Post-operative management of AAM is as important as the surgical procedure.

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DISCUSSION

AAM primarily affects premenopausal women, with a female-to-male ratio of 6.6:1 and peak incidence between ages 30-40 (2). These tumours are most commonly found in the vulva but can also occur in the vagina, pelvis, and groin (1). Although AAM is generally benign, it often shows local infiltration (1), and while rare, distant metastasis can occur (3-4) .

Imaging plays a critical role in diagnosis. Ultrasonography is often the first-line tool but may underestimate tumour extent (5). CT and MRI are more accurate for assessing tumour depth and its relation to nearby structures. On histopathological analysis, AAM is un-encapsulated, lobulated, and locally infiltrative, with a myxoid stroma and prominent vasculature (1). Tumour cells are positive for desmin, vimentin, smooth muscle actin, CD34, and oestrogen and progesterone receptors (1). The HMGA2 gene translocation is often present, though its diagnostic use has limitations (6).

There is no established consensus for AAM management due to its rarity. Surgical excision with clear margins is preferred, but achieving tumour-free margins can be challenging (7). Incomplete resections may be required to avoid complications or preserve fertility (8, 9), although recurrence remains common. Hormonal therapies, particularly GnRH agonists, have growing support, used both pre- and post-operatively to reduce tumour burden and recurrence (7). However, their duration and side effects, like bone demineralisation, need careful management. In some cases, a watchful waiting approach is adopted for slow-growing, asymptomatic tumours (8). Local recurrence rates are between 30-70%, especially in the first five years (2). Treatment at tertiary centres may improve outcomes and facilitate standardized care, research, and guideline development.

Benign		Malignant	
• Vaginal cyst	• Lipoma	• Squamous cell carcinoma	• Extra-gastrointestinal stromal tumour
• Mesenchymal lesion	• Abscess	• Malignant melanoma	• Sarcoma
• Pelvic organ prolapse		• Metastasis	

Table 1 Differential diagnoses for vaginal lesion