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# Aggressive Angiomyxoma of the Cervix: A Case Report and Literature Review

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**Case Report**

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## Abstract

Aggressive angiomyxoma (AA) is a rare mesenchymal, locally aggressive tumour that mainly involves the vulval region of young females. It is characteristically a soft unencapsulated and poorly circumscribed mass, with a smooth external surface. The microscopic appearance is characterized by a myxoid, glistening cut surface and indistinct outlines. Although it is a slow-growing tumour, AA has a marked tendency to local recurrence. Surgical resection is the main treatment modality with a good prognosis, however long-term follow-up is necessary due to its high local recurrence rate. Alternative treatment options include hormonal treatment, due to the presence of estrogen and progesterone receptors in tumour cells. Hormonal therapies with tamoxifen, raloxifene or GnRH analogues have been shown to reduce the tumour size and help to achieve near-complete resection in large tumours. There are no guidelines in the postoperative management of AA. However, due to the high recurrence rate and potential morbidity associated with undiagnosed recurrences, several authors have recommended periodic evaluations up to 15 years after initial treatment.

**Keywords:** Tumour cells; Aggressive angiomyxoma; Anaesthesia; Mesenchymal; Vulvo-perineal

**Abbreviations:** AA: Aggressive Angiomyxoma; CGIN: Cervical Glandular Neoplastic Changes; MDT: Multidisciplinary Meeting; MRI: Magnetic Resonance Imaging.

## Introduction

Aggressive angiomyxoma (AA) is a rare mesenchymal, locally aggressive tumour that mainly involves the vulvo-perineal region of young female patients and was first described by Steeper and Rosai in 1983 [1,2]. Although it is a slow-growing tumour, AA has a marked tendency to local recurrence with a low metastasis capacity [3]. Pre-operative diagnosis is difficult due to the absence of diagnostic features [4]. Surgical resection is the main treatment modality with a good prognosis. Long-term follow-up is necessary due to its high local recurrence rate [5]. Various adjuvant treatment modalities have also been tried to reduce tumour recurrence

but have not yet been fully established [6].

## Case report

AB, a 46-year-old lady presented to the gynaecological outpatient department with a history of abnormal menstrual bleeding for 2 years and postcoital bleeding for the last 6 months. Abdominal examination was unremarkable. Pelvic examination revealed a vaginal mass which was 5cm, broad-based, arising from the cervix. The rest of the examination was normal. She was listed for urgent examination under anaesthesia, hysteroscopy and endometrial biopsy. Examination under anaesthesia revealed a 5cm cervical growth protruding into the vagina, which was excised; vulva, vagina and endometrial cavity were normal. An endometrial biopsy was obtained. Histology of the cervical mass confirmed an aggressive angiomyxoma of the cervix; there were no cervical glandular neoplastic changes (CGIN) or invasive malignancy. The endometrial biopsy revealed early secretory phase changes. The histology was referred for an expert opinion in a tertiary centre due to the rarity of the lesion, which confirmed the same.

The case was discussed at the gynaecological oncology multidisciplinary meeting (MDT) following a review of her magnetic resonance imaging (MRI) scans of her abdomen and pelvis; as well as a chest X-ray, which were confirmed to be normal. She was offered a simple hysterectomy to ensure complete resection of the tumour as she had completed her family. She had a total laparoscopic hysterectomy, bilateral salpingo-oophorectomy and omental biopsy. Findings at surgery include a 12-week size uterus; normal fallopian tubes, ovaries, omentum and bowels. She was discharged home within 24hrs of her operation. The postoperative histology reports confirmed aggressive angiomyxoma of the cervix with no definite morphological evidence of residual angiomyxoma.

## Discussion

Aggressive angiomyxoma is a rare, benign, locally invasive soft tissue tumour occurring mainly in the pelvis. About 90% of these cases present in women of reproductive age with a peak incidence in the fourth decade of life and a female-to-male ratio of 6.6:1 [7]. It is described as aggressive angiomyxoma due to the neoplastic nature of the blood vessels, its locally infiltrative nature and the high recurrence rate [8]. Patients usually complain of a vaginal mass, typically about 5-10cm, although its size is highly variable (1-60cm) [7]. It is rarely painful [9]. AA characteristically grows slowly and may occupy the whole pelvis invading paravaginal and para-rectal spaces, and displacing pelvic structures before the patient seeks any medical advice [10]. It is a polypoid or cystic lesion with infiltrating margins. It is misdiagnosed in more than 80% of cases because of its rarity, non-specific radiological findings, and vast differential diagnosis which include Bartholin's abscess, Gartner's duct cyst, hernia and vaginal prolapse [6,11]. Some women may present with abnormal vaginal or post-coital bleeding as described in this case report.

Histologically, AA should be differentiated from angiomyofibroblastoma, fibroma, myxofibrosarcoma, leiomyoma, lymphangioma, neurofibroma, malignant mesenchymoma, malignant fibrous histiocytoma, myxolipoma, sclerosing hemangioma, botryoid pseudosarcoma, myxoid smooth muscle and nerve sheath tumours, mixed mesodermal tumour, leiomyosarcoma, and embryonal rhabdomyosarcomas [12-15]. On gross examination, it is characteristically a soft unencapsulated and poorly circumscribed mass, with a smooth external surface. The microscopic appearance of these neoplasms is characterized by a myxoid, glistening cut surface and indistinct outlines. It consists of a mixture of spindle cells in a loosely myxoid stroma. The stroma contains collagen fibers and a prominent vascular component containing large, thick-walled vessels with exceeding low mitotic activity [12]. It is possible that it is a hormonally responsive neoplasm, as it tends to occur in women of reproductive age [16].

The surgical treatment is wide local excision with tumour-free margins [6], which are difficult to obtain because the tumour is locally invasive and has the same consistency as the surrounding connective tissue. Furthermore, in a review of over 100 cases, there was no statistically significant difference in the chance of remaining disease-free between cases of complete and incomplete tumour resection (50% vs. 40% in 10 years). Alternative treatment options include hormonal treatment, as tumour cells are usually estrogen and progesterone positive [15]. Hormonal therapies with tamoxifen, raloxifene or GnRH analogues have been shown to reduce the tumour size and help achieve near-complete resection in large tumours. It can also be used to treat recurrence. [17] Radiotherapy and chemotherapy are considered less suitable due to the low mitotic activity of this tumour [6,13]. Han-Guerts, et al. [18] propose the following guideline in treating AA: (a) complete excision of the lesion when possible and avoiding mutilating surgery, (b) adjunct therapy when partial resection is performed (arterial embolization, hormonal treatment), and (c) radiotherapy is reserved to cases that are resistant to adjunct therapy and where patient is still symptomatic.

There are no guidelines in the postoperative management of AA. Due to the high recurrence rate (recurrence rates range from 25% to 47%, 85% of these occurring within 5 years of initial surgery [7]) and potential morbidity associated with undiagnosed recurrences, several authors

have recommended periodic evaluations with physical examination and MRI up to 15 years after treatment [19,20]. Aggressive angiomyxoma can present with a broad range of symptoms, including those that are the most common presenting complaint in a gynaecological outpatient and this is something that the clinician should bear in mind.

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