

Cervical aggressive angiomyxoma – a rare case report

K Guruparan^a, A P Ginige^b, K Kowsikanth^a, K Muhunthan^a

Abstract

Aggressive angiomyxomas are rare tumours affecting the pelvic perineum of reproductive-age females with high recurrence rates. Cervical aggressive angiomas are always reported after histopathological confirmation of cervical polyps, which warrants long term follow up.

Introduction

Angiomyxoma is categorized as either superficial or aggressive. Superficial angiomyxomas usually present as nodular or polypoidal growths in the head and neck region among middle-aged adults. Aggressive angiomyxomas originate from the pelvic-perineal mesenchymal tissue of the reproductive age women. They are locally aggressive tumours of the perineum, and they occasionally metastasize to distant sites¹.

As they are aggressive locally and recur after excision, they were named aggressive angiomyxomas. Due to their local aggressive growth, they displace the adjacent tissues rather than invading¹. Steeper and Rosai firstly

described about aggressive angiomyxomas in a case series in early eighties². It has a female preponderance (Female: male – 6:1) and is often mistaken for benign growths such as skin tags, Bartholin cysts or other congenital cysts or cervical polyps³.

Aggressive angiomyxomas are oestrogen and progesterone responsive tumours and their growth is noted during pregnancy and females of reproductive age⁴.

Imaging techniques are helpful in cases of larger well defined pelvi-perineal aggressive angiomyxomas with their hypoechoic hypodense cystic and solid components⁵.

Sri Lanka Journal of Obstetrics and Gynaecology 2023; **45**: 138-140

DOI: <https://doi.org/10.4038/sljog.v45i3.8086>

^a Faculty of Medicine, University of Jaffna, Sri Lanka.

^b District General Hospital, Vavuniya, Sri Lanka.

Correspondence: KG, e-mail: kandiah.guruparan@yahoo.com

 <https://orcid.org/0000-0003-1146-0427>

Received 03rd September 2023

Accepted 26th October 2023



This is an open-access article distributed under the terms of the Creative Commons Attribution 4.0 International License, which permits unrestricted use, distribution and reproduction in any medium provided the original author and source are credited.

Macroscopically, they present as well-defined, bulky masses with a smooth external surface of glistening appearance and internally composed of homogenous tissue with focal areas of vascular congestion and bleeding.

Microscopically, there is a sparse population of small spindle cells with one or more thin cytoplasmic processes resulting in a stellate configuration seen on the background of loose myxoid stroma composed of wavy collagen fibrils and vascular tissue. The cells will have small, uniform nuclei with small, indistinct nucleolus. Cells show no cellular atypia or atypical mitosis or tumour cell necrosis. Blood vessels of varying sizes can also be observed within the myxoid stroma. Recurrence will also show similar histopathological characteristics⁶.

Uterine cervical aggressive angiomyxomas are rarely reported entities in the literature and are often diagnosed after histopathological confirmation.

We are reporting a case of cervical aggressive angiomyxoma following a regular cervical polypectomy for a benign lesion.

Case history

A 44-year-old multiparous woman was presented with increased vaginal discharge and heavy menstrual bleeding for three months duration. On speculum examination, she was found to have a small cervical polyp arising from the anterior lip of the cervix. Ultrasound imaging revealed a normal uterine outline with a thin endometrium measuring 4 mm.

A cervical polypectomy with endometrial sampling was performed under general anaesthesia, and the cervical polyp was reported as an aggressive angiomyxoma of the cervix. Given the higher probability of local recurrence and distant metastasis, the patient was under regular follow-up for one year at the reporting of this case, and no recurrence nor metastasis were noted.

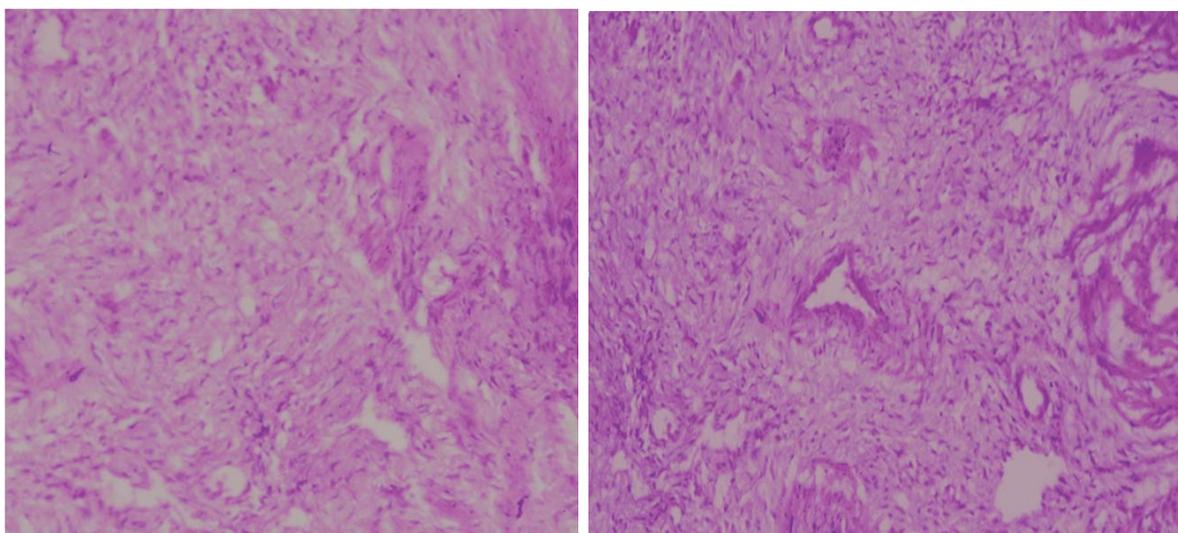


Figure 1. **Histopathological slide.**

(Description: small spindle cells with stellate configuration seen on the background of loose myxoid stroma composed of wavy collagen fibrils and vascular tissue).

Discussion

Aggressive angiomyxomas have a higher recurrence rate following surgical excision of local perineal tumours such as vulval or perineal lesions. This is due to incomplete excision of the aggressive angiomyxomas.

Though wide local excision with tumor-free margins was the treatment of choice, there are conflicting evidence in studies which suggest that even with clear margins, the risk of recurrence persists⁶.

Complete resection of the tumour is not possible due to the locally aggressive behaviour of the tumour.

As extensive surgeries would increase the morbidity, currently less radical surgery is recommended with a combination of raloxifene, tamoxifen, or GnRH agonists where the tumour is sensitive for estrogen and progesterone receptors^{7,8}.

There is minimal impact to radiotherapy and chemotherapy since the tumour has no significant mitotic activity.

In our case, the patient was evaluated for general gynaecological symptoms and underwent cervical polypectomy. As the cervical polyp was reported as an aggressive angiomyxoma, she is under regular follow-up.

Due to the risk of recurrence of aggressive angiomyxoma following surgery, our patient was offered the options of chemotherapy, and she preferred regular follow-up rather than adjuvant treatments. She is on a regular 6 monthly follow-up with no recurrence.

In the case of recurrence, a hysterectomy or wide local excision would be considered.

Conclusion

When managing female pelvic perineal masses, histopathological confirmation is always needed. Aggressive angiomyxoma must be considered as a differential diagnosis in cases of polypoidal growth.

References

1. Kura M, Jindal S, Khemani U. Aggressive angiomyxoma of the vulva: An uncommon entity. *Indian Dermatol Online*. 2012; 3(2): 128-30. <https://doi.org/10.4103/2229-5178.96712>
2. Steeper TA, Rosai J. Aggressive angiomyxoma of the female pelvis and perineum. *Am J Surg Pathol*. 1983; (7): 463-76. <https://doi.org/10.1097/0000478-198307000-00009>.
3. Fetsch JF, Lefkowitz M, Kindblom LG, Meis-Kindblom JM. Aggressive Angiomyxoma: A Clinicopathologic Study of 29 Female Patients. *Cancer*. 1996; 78(1): 79-90. [https://doi.org/10.1002/\(SICI\)1097-0142\(19960701\)78:1%3C79](https://doi.org/10.1002/(SICI)1097-0142(19960701)78:1%3C79)
4. Goyal LD, Garg P, Badyal R, Bhalla S. Aggressive (deep) angiomyxoma of the vulva: a case report. *BMC Journal of Medical Case Reports*. 2022; 16: 71-4. <https://doi.org/10.1186/s13256-022-03284-z>
5. Jeyadevan NN, Sohaib SAA, Thomas JM, Jeyarajah A, Shepherd JH, Fisher C. Imaging features of aggressive angiomyxoma. *Clin Radiol*. 2003; 58: 157-62. <https://doi.org/10.1053/crad.2002.1127>
6. Benson JC, Gilles S, Sanghvi T, Boyum J, Niendorf E. Aggressive angiomyxoma: case report and review of the literature. *Radiol Case Rep*. 2016; 11(4): 332-5. <http://dx.doi.org/10.1016/j.radcr.2016.10.006>
7. Bruce A, Fine, Munoz AK, Litz CE, Gershenson DM. Primary Medical Management of Recurrent Aggressive Angiomyxoma of the Vulva with a Gonadotropin-Releasing Hormone Agonist. *Gynecologic Oncology* 2001; 81: 120-2. <https://doi.org/10.1006/gyno.2000.6119>
8. Schwartz PE, Hui P, McCartney S. Hormonal therapy for aggressive angiomyxoma: a case report and proposed management algorithm. *J Low Genit Tract Dis*. 2014; 18(2): E55-61 <https://doi.org/10.1097/lgt.0b013e3182a22019>