

**AGGRESSIVE ANGIOMYXOMA PRESENTING AS A RECURRENT CERVICAL
POLYP – A CASE REPORT**

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ABSTRACT

BACKGROUND: In 1983, Steeper and Rosai first described Aggressive Angiomyxoma. It is a mesenchymal neoplasm of the soft tissue that grows slowly in the pelvis and perineum and infiltrates locally. We present a case of an unusual presentation of Aggressive Angiomyxoma of the cervix uteri in a 15-year old girl.

CASE PRESENTATION: A 16-year old patient was referred on account of recurrent mass protruding per vagina 3 months after undergoing 2 polypectomies. No other co-morbidities were observed. The patient underwent polypectomy plus subtotal hysterectomy. Histological examination showed a mesenchymal neoplasm that was poorly circumscribed which composed of spindle shaped cells widely spaced from each other in a myxoid stroma with clustered of small to medium sized vascular channels. These findings were in consistence with Aggressive Angiomyxoma.

CONCLUSION: After thorough review of the literature, this index case is the second case reported in Africa and third in the world; other two were reported by Paplomata *et al.* (2010) in Greece and by Bothale K. A. *et al* (2012) in India, with diagnosis of Aggressive Angiomyxoma presenting as a cervical polyp. Aggressive angiomyxoma is rare, so case reports are needed to improve diagnosis and treatment.

Keywords – aggressive angiomyxoma, cervical polyp, rare

1.0 INTRODUCTION

Aggressive angiomyxoma (AA) is a rare, locally aggressive myxoid mesenchymal neoplasm preferentially arising in the pelvic and perineal regions of young adult females, most

31 commonly in the reproductive age group [1]. In men, the tumour involves analogous sites
32 including the scrotum and inguinal area and usually appears at an older age [2]. AA was first
33 described in 1983 [3] and since then a few hundred cases have been reported in literature,
34 mostly in the form of small case series or isolated case reports. Though most of these tumours
35 are locally aggressive occasional distant metastasis has also been reported [4,5].

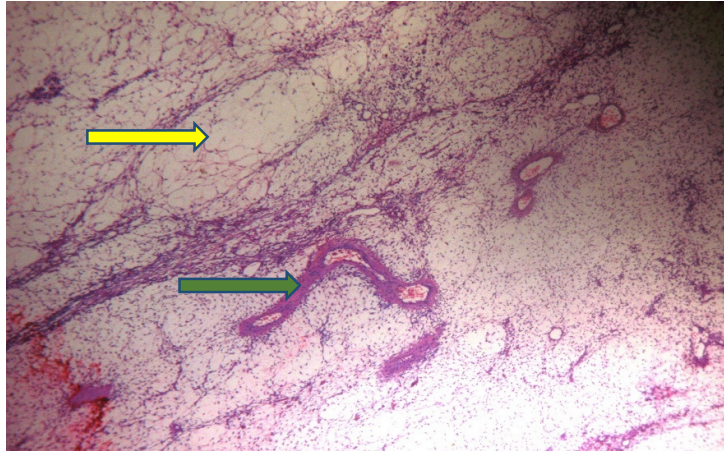
36 This low-grade neoplasm most often presents in the pelvi-perineal region at a female to male
37 ratio of 6:1 [1,6]. Its behaviour is typically locally aggressive but metastases are described
38 [5,7]. The recommended treatment is radical surgery with wide margins [4] and long-term
39 follow-up is advised. The recurrence rate is high, resulting in multiple operative procedures
40 with substantial morbidity [8].

41 **2.0 CASE PRESENTATION**

42 A 15-year –old girl was referred to the Gynaecologic from another hospital on account of a
43 recurrent mass protruding per vaginal since about 4 months. She underwent polypectomy
44 twice (a year ago and a recurring mass 3 months prior to presentation). The mass measured
45 about 16x17cm with a stalk of about 2.5cm thick. The extent of the tumour was not
46 ascertained due to risk of bleeding. No history of changes in menstrual cycle, however, she
47 was anxious and there was associated dragging sensation and low back pain. No change in
48 bowel habit and no vagina discharge.

49 The patient was prepared for surgery and the operative findings were those of a uterine mass
50 that prolapsed below the introitus. It is firm and measures about 16x16x12cm. it has no stalk
51 and bleeds to touch. The upper cervix and lower uterus was destroyed by the mass leaving a
52 small fundus. The round ligament, fallopian tubes and ovaries were normal. She had
53 polypectomy plus subtotal hysterectomy. Specimen was sent for histology.

54 On microscopic examination, sections show a mesenchymal neoplasm that is poorly
55 circumscribed which composed of spindle shaped cells widely spaced from each other in a
56 myxoid stroma. Within this stroma are seen numerous clustered of small to medium sized
57 vascular channels. Features are those of Aggressive Angiomyxoma

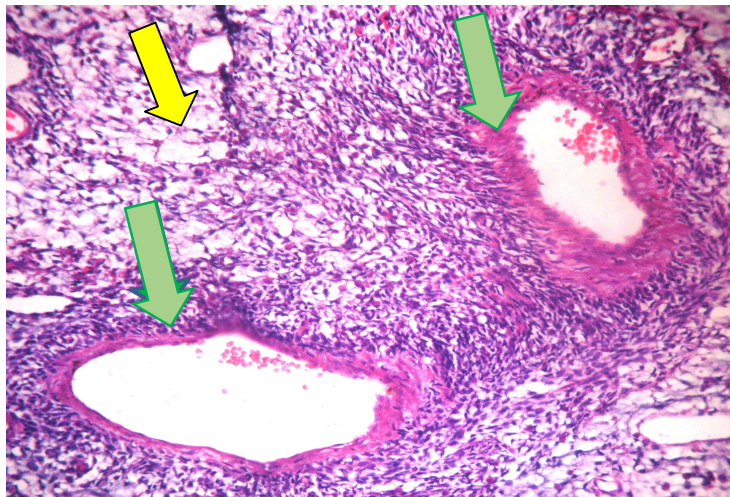


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Sections showing proliferating vascular channels disposed in a hypocellular myxoid stroma with angiocentric arrangement of tumor cell around vascular channels



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Figure 3 – Section showing dilated vascular channels with angiocentric arrangement of tumour cells (green arrow) disposed in a myxoid stroma (yellow arrow) (H&E Mag. X400).

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3.0 DISCUSSION

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Aggressive angiomyxomas are locally invasive connective tissue tumours presenting in about 90% of cases in women of reproductive age group with a peak incidence in the fourth decade of life [6]. The initial presentation varies from an asymptomatic perineal or vulval nodule/polyp or perineal hernia to a pelvic mass diagnosed on imaging studies. Occasionally, the tumour can be cystic and mistaken for a Bartholin's, labial or Gartner's duct cyst. They can exhibit a slow and insidious growth pattern and hence patients may be asymptomatic for long time until they become conscious of the significant size of the tumour. Tumour recurrences, like the primary ones, are often asymptomatic [9,10].

73 Aggressive angiomyxoma (synonymously referred to as deep angiomyxoma by the World
74 Health Organization (9) is distinct among other benign vulvar neoplasms (including
75 fibroepithelial polyp, superficial angiomyxoma, angiomyofibroblastoma, cellular
76 angiofibroma, myxoid leiomyoma, myxoid neurofibroma, and nodular fasciitis) as it has
77 poorly defined margins, deeply infiltrates the vulvar/perineal soft tissues, and may recur in up
78 to 30% of patients. Pre-operative diagnosis is often a problem because of the rarity of these
79 neoplasms and lack of typical features and most cases are diagnosed on histology after
80 primary surgical excision. Accurate diagnosis before surgical excision is important in
81 planning extent of excision and patient counselling [3].

82 Macroscopically these tumours often have a smooth surface, partially or completely
83 encapsulated and cut surface gives a glistening, gelatinous appearance, bluish grey in colour,
84 with areas of haemorrhage and congestion. They are usually homogenous in consistency with
85 no obvious nodularity [3]. Histologically they reveal a hypocellular mesenchymal lesion
86 consisting of a sparse population of bland spindled and stellate cells scattered in a
87 background of loose myxoid stroma composed of wavy collagen fibrils. The cells have
88 abundant wispy pink cytoplasm with bland nuclei. There is no cytologic atypia, no atypical
89 mitotic features or discernible mitotic activity and no evidence of coagulative tumour cell
90 necrosis either. The lesion often also shows numerous blood vessels of varying calibre [3].

91 The stromal cells can show immunoreactivity to different combinations of vimentin, desmin,
92 smooth muscle actin, muscle specific actin, CD 34, oestrogen and progesterone receptors
93 [12]. Recurrent tumours usually show similar histological characteristics [9]. Oestrogen and
94 progesterone receptors are commonly found in aggressive angiomyxoma, and so they may
95 be hormone-dependent [10,13,14]. Cytogenetic studies reveal a HMGA2 rearrangement
96 during embryogenesis [15] making it a sensitive but not specific biomarker for aggressive
97 angiomyxoma [16].

98 Aggressive Angiomyxoma is a rare tumour [1-4] with extreme cases seen worldwide as
99 evident in an 11-year old girl [17]; in young boys [18,19] and in pregnant women [10,20,21].
100 Adjuvant and complementary modalities for the treatment includes hormonal treatment,
101 radiotherapy and angiography [22-27]. Its incidence masquerading as a cervical polyp is even
102 rarer [28, 29].

103 Table 1 – Comparison of Index case with other reported cases.

	INDEX CASE	CASE 1 [28]	CASE 2 [29]
AGE (years)	15	29	45
GENDER	Female	Female	Female
ONSET – DIAGNOSIS INTERVAL	4 months	Routine health care visit	2 months
OTHER SYMPTOMS	Low back pain, recurrent mass	Mild suprapubic pain, dysuria	Yellowish discharge
TUMOUR SIZE	16x16x12cm	5x4x2cm	6.5x5.5x4cm
TREATMENT	Polypectomy and subtotal hysterectomy	Surgical Excision	Surgical Excision
PATIENT STATUS	Doing well 6 months post-operatively	Doing well 2 years post-operatively	Doing well 6 months post-operatively

4.0 CONCLUSION

Aggressive angiomyxoma is a rare vulvo-vaginal tumour affecting females of reproductive age. More extreme cases are even rarer such as in pregnant women. Treatment of choice is surgery complemented with radiotherapy, hormonal therapy and angiography. Since late recurrences are known, patients should be adequately counselled on the need for periodic follow up so that recurrences can be identified early and treatments offered. The patient presented is still on regular follow up. Aggressive angiomyxoma is rare, so case reports are needed to improve diagnosis and treatment.

CONSENT DISCLAIMER

As per international standard or university standard written patient consent has been collected and preserved by the author(s).

REFERENCES

1. Fetsch JF, Laskin WB, Lefkowitz M, Kindblom LG. Aggressive angiomyxoma: a clinicopathologic study of 29 female patients. Cancer 1996;78:79–90.
2. Idrees MT, Hoch BL. Aggressive angiomyxoma of male genital region. Report of 4 cases with immunohistochemical evaluation including hormone receptor status. Ann Diagn Pathol 2006 Aug;10(4): 197–204.

3. Steeper TA, Rosai J. Aggressive angiomyxoma of the female pelvis and perineum. Report of 9 cases of a distinctive type of gynaecologic soft tissue neoplasm. *Am J Surg Pathol* 1983;7:463–75.
4. Siassi RM, Papadopoulos T. Metastasizing aggressive angiomyxoma. *The NEJM* 1992 Dec;341:1772.
5. Blandamura S, Cruz J. Aggressive angiomyxoma: a second case of metastasis with patient's death. *Human Pathol* 2003 Oct;34: 1072.
6. Chan IM, Hon E, Ngai SW, Ng TY, Wong LC. Aggressive angiomyxoma in Females: is radical resection the only option? *Acta Obstet Gynaecol Scand* 2000;79:216–20.
7. Siassi RM, Papadopoulos T, Matzel KE. Metastasizing aggressive angiomyxoma. *N Engl J Med* 1999;2:1772.
8. Be'gin LR, Clement PB, Kirk ME. Aggressive angiomyxoma of pelvic soft parts: a clinicopathologic study of nine cases. *Hum Pathol* 1985;16:621–8.
9. Halder K, Martinek IE, Kehoe S. Aggressive angiomyxoma: a case series and literature review. *European Journal of Surgical Oncology (EJSO)*. 2010 Apr 30;36(4):335-9.
10. Han-Geurts IJ, Van Geel AN, van Doorn L, den Bakker MD, Eggermont AM, Verhoef C. Aggressive angiomyxoma: multimodality treatments can avoid mutilating surgery. *European Journal of Surgical Oncology (EJSO)*. 2006 Dec 31;32(10):1217-21.
11. Kempson RL, Teixeira MR, Hendrickson MR. Tumors of the vulva. World Health Organization classification of tumours. In: Tavassoli FA, Devilee P, eds. *Pathology and Genetics of Tumours of the Breast and Female Genital Organs*. Lyon: IARC Press, 2003:329.
12. Smith HO, Worrell RV, Smith AY, Dorin MH, Rosenberg RD. Aggressive angiomyxoma of the female pelvis and perineum: review of the literature. *Gynecol Oncol* 1991;42:79–85.
13. McCluggage WG, Patterson A, Maxwell P. Aggressive angiomyxoma of pelvic parts exhibits oestrogen and progesterone receptor positivity. *Journal of clinical pathology*. 2000 Aug 1;53(8):603-5.
14. McCluggage WG, Jamieson T, Dobbs SP, Grey A. Aggressive angiomyxoma of the vulva: dramatic response to gonadotropin-releasing hormone agonist therapy. *Gynecologic oncology*. 2006 Mar 31;100(3):623-5.

15. Rabban JT, Dal Cin P, Oliva E. HMGA2 rearrangement in a case of vulvar aggressive angiomyxoma. *International journal of gynecological pathology*. 2006 Oct 1;25(4):403-7.
16. McCluggage WG, Connolly L, McBride HA. HMGA2 is a sensitive but not specific immunohistochemical marker of vulvovaginal aggressive angiomyxoma. *The American journal of surgical pathology*. 2010 Jul 1;34(7):1037-42.
17. White J, Chan YF. Aggressive angiomyxoma of the vulva in an 11-year-old girl. *Pediatric Pathology*. 1994 Jan 1;14(1):27-37.
18. Kim HS, Park SH, Chi JG. Aggressive angiomyxoma of childhood: two unusual cases developed in the scrotum. *Pediatr Dev Pathol* 2003;6(2):187–91.
19. Carlinfante G, De Marco L, Mori M. Aggressive angiomyxoma of the spermatic cord. Two unusual cases occurring in childhood. *Pathol Res Pract* 2001;197:139–44.
20. Bagga R, Keepanasseril A, Suri V, Nijhawan R. Aggressive angiomyxoma of the vulva in pregnancy: a case report and review of management options. *Medscape General Medicine*. 2007;9(1):16.
21. Panti AA, Tunau KA, Sahabi SM, Umar M. Aggressive Angiomyxoma Of The Vulva In Pregnancy: A Case Report And Literature Review. *BOMJ* ,Vol. 10, No. 2, July-December 2013
22. Brooks SE, Balidimos I, Reuter KL, Khan A. Virtual consult: aggressive Angiomyxoma of the vulva: impact of GnRH agonists. *Medscape Women's Health* 1998;3:1–5.
23. Fine BA, Munoz AK, Litz CE, Gershenson DM. Primary medical management of Recurrent aggressive angiomyxoma of the vulva with a gonadotropin-releasing hormone agonist. *Gynecol Oncol* 2001;81:120–2.
24. Shinohara N, Nonomura K, Ishikawa S, Seki H, Koyanagi T. Medical management of recurrent aggressive angiomyxoma with gonadotropin- releasing hormone agonist. *Int J Urol* 2004;11(6):432–5.
25. Poirier M, Fraser R, Meterissian S. Case 1: aggressive angiomyxomaof the pelvis: response to luteinizing hormone-releasing hormone agonist.*J Clin Oncol* 2003;21(18):3535–6.
26. Rhomberg W, Jasarevic Z, Alton R. Aggressive angiomyxoma: irradiation for recurrent disease. *Strahlenther Onkol* 2000;176(7):324–6.
27. Nyam DC, Pemberton JH. Large aggressive angiomyxoma of the perineum and pelvis: an alternative approach. *Dis Colon Rectum* 1998; 41(4):514–6.

- 189 28. Paplomata E, Fotas A, Balaxis D, Filindris TH, Charalambous ST, Rombis VA.
190 Aggressive angiomyxoma mimicking cervical polyp. Pelviperineology. 2010
191 Mar;29:30-1.
- 192 29. Bothale KA, Mahore SD, Joshi AM. A rare presentation of aggressive Angiomyxoma
193 as a cervical polyp. Journal of Basic and Clinical Reproductive Sciences. 2012;1(1-
194 2):65-7.