

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

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ABSTRACT

BACKGROUD: 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].

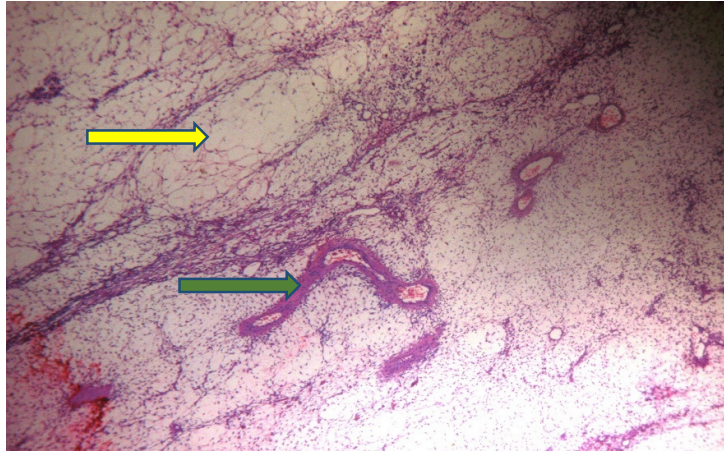
36 This low-grade neoplasm most often presents in the pelvi-perineal region at a female to male
37 ratio of 6:1 [1,6]. Its behaviour is typically locally aggressive but metastases are described
38 [5,7]. The recommended treatment is radical surgery with wide margins [4] and long-term
39 follow-up is 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 vaginal discharge. Abdominal Ultrasound revealed features suspicious of
49 left sided soft tissue mass ? due to Sarcoma botryoides.

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

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

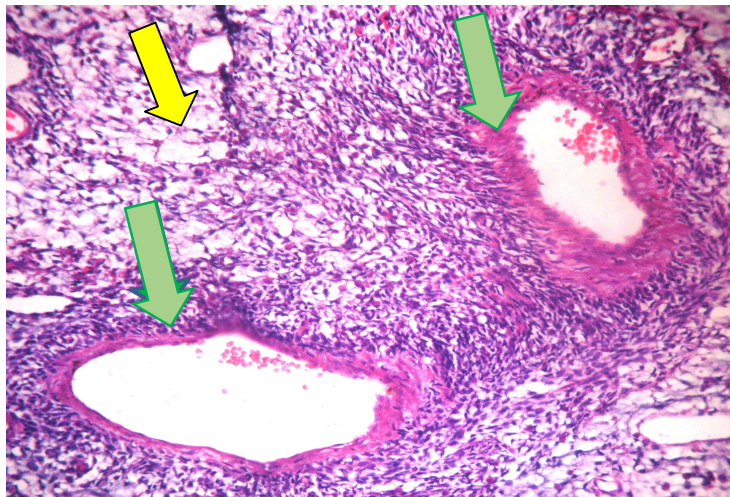


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Sections showing proliferating vascular channels disposed in a hypocellular myxoid stroma with angiocentric arrangement of tumor cell around vascular channels



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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].

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

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

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

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

104 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 11 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).

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