

**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 16-year old girl.

CASE PRESENTATION: A 16-year old patient was referred on account of recurrent mass protruding per vagina 3 months after a repeat polypectomy. There were no other co-morbidities observed. She had polypectomy and subtotal hysterectomy. Histological examination showed a benign mesenchymal neoplasm that was poorly circumscribed which composed of spindle shaped cells widely spaced from each other in angimyxoid stroma with clustered small to medium sized vascular channels. These findings were consistent 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.* in Greece and by Bothale K. A. *et al* 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

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

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

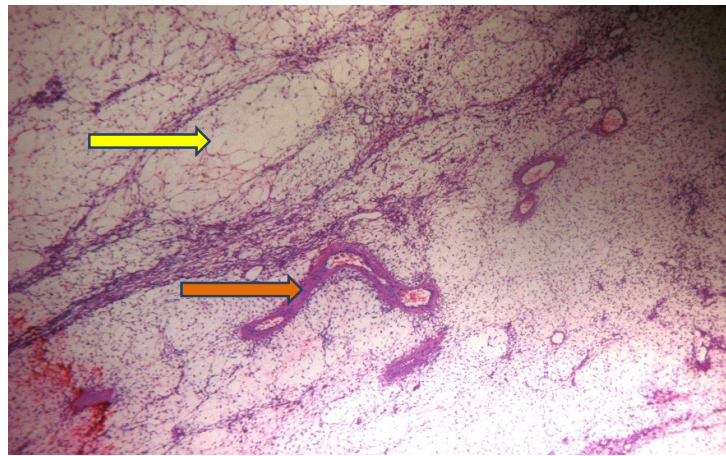
42 **2.0 CASE PRESENTATION**

43 A 16-year –old girl was referred to the Gynaecologic Clinic from another hospital on account
44 of a recurrent mass protruding per vagina for about 4 months. The mass appears to have
45 arisen from the posterior wall of the lower uterine segment and extending to the upper half
46 of the cervix, hence protruding through the cervical canal and inadvertently through the
47 vaginal introitus to the outside. She had a repeat polypectomy (first polypectomy done a year
48 ago and a repeat was done about 4 months prior to presentation). The mass measured about
49 16x17cm with a broad based sessile stalk. The origin and extent of the tumour was quite
50 difficult to ascertain due to risk of bleeding and distorted anatomy from previous surgery.
51 There was no history of changes in menstrual cycle or vaginal discharge. However, she had
52 associated dragging sensation and low back pain. She was anxious, with associated loss of
53 appetite and weight loss but no change in bowel habit. Abdominal Ultrasound revealed
54 features suspicious of a uterine soft tissue mass suspected to be Sarcoma botryoides.

55 The patient was prepared for surgery and the operative findings were that of a predominantly
56 uterine mass extending to the upper cervix that prolapsed below the vaginal introitus. It is
57 firm and measures about 16x16x12cm. It has a broad based sessile stalk and bleeds easily to
58 touch. The upper cervix and lower uterus was destroyed by the mass leaving a small fundus.
59 The round ligament, fallopian tubes and ovaries were grossly normal. She had polypectomy
60 and subtotal hysterectomy. The choice of subtotal hysterectomy as against total was because
61 the patient was young and the lower cervix was not affected.. Consequently because the

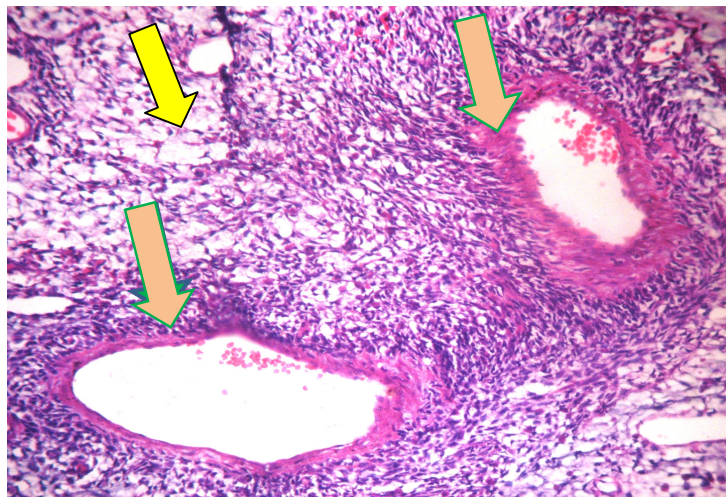
62 Cervix enhance sexual excitement during coitus a subtotal hysterectomy was a better option
63 for this young lady who eventually may consider having a partner to consummate marriage to
64 her satisfaction. Although spontaneous pregnancy cannot be achieved, her sexual function
65 have being preserved to some extent .Specimen was sent for histology.

66 On microscopic examination, sections show a mesenchymal neoplasm that is poorly
67 circumscribed and composed of spindle shaped cells that are widely spaced from each other
68 in a myxoidstroma. Within this stroma are seen numerous clustered small to medium sized
69 vascular channels. Features are those of Aggressive Angiomyxoma



70

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



73

74 Figure2– Section showing dilated vascular channels with angiocentric arrangement of tumour
75 cells (brown arrow) disposed in a myxoid stroma (yellow arrow) (H&E Mag. X400).

76 3.0 DISCUSSION

77 Aggressive angiomyxomas are locally invasive connective tissue tumours presenting in about
78 90% of cases in women of reproductive age group with a peak incidence in the fourth decade
79 of life [6]. The initial presentation varies from an asymptomatic perineal or vulval
80 nodule/polyp or perineal hernia to a pelvic mass diagnosed on imaging studies. Occasionally,
81 the tumour can be cystic and mistaken for a Bartholin's, labial or Gartner's duct cyst. They
82 can exhibit a slow and insidious growth pattern and hence patients may be asymptomatic for
83 long time until they become conscious of the significant size of the tumour. Tumour
84 recurrences, like the primary ones, are often asymptomatic [9,10].

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

94 Macroscopically these tumours often have a smooth surface, partially or completely
95 encapsulated and cut surface gives a glistening, gelatinous appearance, bluish grey in colour,
96 with areas of haemorrhage and congestion. They are usually homogenous in consistency with
97 no obvious nodularity [3]. Histologically they reveal a hypocellularmesenchymal lesion
98 consisting of a sparse population of bland spindled and stellate cells scattered in a
99 background of loose myxoidstroma composed of wavy collagen fibrils. The cells have
100 abundant wispy pink cytoplasm with bland nuclei. There is no cytologicatypia, no atypical
101 mitotic features or discernible mitotic activity and no evidence of coagulative tumour cell
102 necrosis either. The lesion often also shows numerous blood vessels of varying calibre
103 (fig.1&2)[3].

104 The stromal cells can show immunoreactivity to different combinations of vimentin, desmin,
105 smooth muscle actin, muscle specific actin, CD 34, oestrogen and progesterone receptors
106 [12]. Recurrent tumours usually show similar histological characteristics [9]. Oestrogen and
107 progesteronereceptors are commonly found in aggressive angiomyxoma, and so they may be

hormone-dependent [10,13,14]. Cytogenetic studies reveal a HMGA2 rearrangement during embryogenesis [15] making it a sensitive but not specific biomarker for aggressive angiomyxoma [16].

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

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 and ancillary diagnostic techniques such as Immunohistochemistry are needed to improve accurate 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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