

AN UNUSUAL PRESENTATION OF AGGRESSIVE ANGIOMYXOMA AS CERVICAL POLYP – A CASE REPORT

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: The index patient presented with a growth protruding through her vagina. The preoperative diagnosis after the physical examination was mucinous cervical polyp. After the tissue was excised surgically, histology 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* in India in 2012, with diagnosis of Aggressive Angiomyxoma presenting as a cervical polyp.

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

This low-grade neoplasm most often presents in the pelvi-perineal region at a female to male ratio of 6:1 [1,6]. Its behaviour is typically locally aggressive but metastases are described [5,7]. The recommended treatment is radical surgery with wide margins [4] and long-term

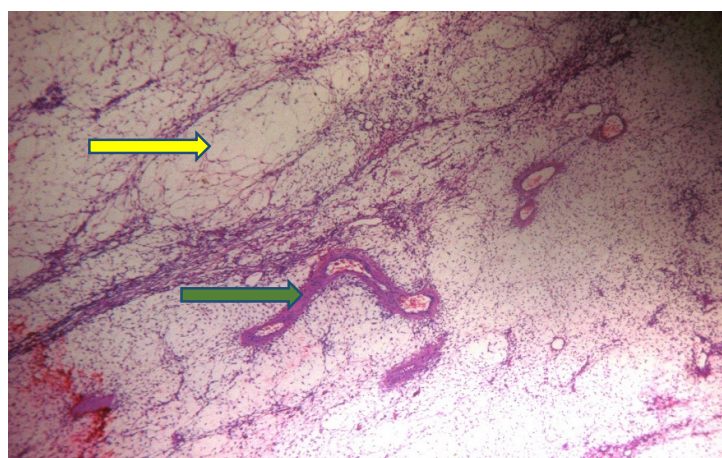
31 follow-up is advised. The recurrence rate is high, resulting in multiple operative procedures
32 with substantial morbidity [8].

33 CASE REPORT

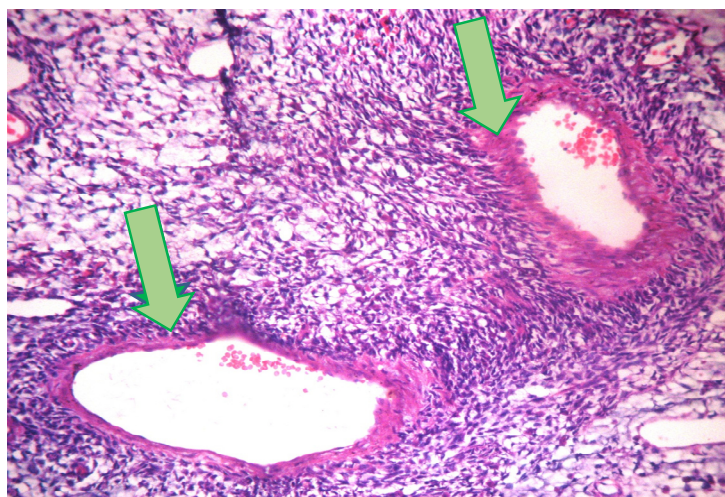
34 A 15-year –old girl presented to Gynaecologic clinic with complaint of a mass protruding per
35 vaginal since about 4 months. No history of changing in menstrual cycle, however, she was
36 anxious and there was associated dragging sensation and low back pain. No change in bowel
37 habit and no vagina discharge. The mass was removed by wide excision under general
38 anaesthesia and subjected to histological examination.

39 On gross examination, a gray-brown ulcerated lobular tissue aggregating 8x6x5cm was
40 received. It was bisected into five parts to reveal greyish brown solid cut surface and multiple
41 tan surfaces with soft myxoid appearances.

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



46
47 Figure 1 – Proliferating Vascular channels (green arrow) in a myxoid stroma (yellow arrow).
48 (H&E Mag. X100)



49

50 Figure 2 – Section showing dilated vascular channels with angiocentric arrangement of
51 tumour cells around vascular channels(green arrow)(H&E Mag. X400).

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DISCUSSION

53 Aggressive angiomyxomas are locally invasive connective tissue tumours presenting in about
54 90% of cases in women of reproductive age group with a peak incidence in the fourth decade
55 of life [6]. The initial presentation varies from an asymptomatic perineal or vulval
56 nodule/polyp or perineal hernia to a pelvic mass diagnosed on imaging studies. Occasionally,
57 the tumour can be cystic and mistaken for a Bartholin's, labial or Gartner's duct cyst. They
58 can exhibit a slow and insidious growth pattern and hence patients may be asymptomatic for
59 long time until they become conscious of the significant size of the tumour. Tumour
60 recurrences, like the primary ones, are often asymptomatic [9,10].

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

70 Macroscopically these tumours often have a smooth surface, partially or completely
71 encapsulated and cut surface gives a glistening, gelatinous appearance, bluish grey in colour,

72 with areas of haemorrhage and congestion. They are usually homogenous in consistency with
 73 no obvious nodularity [3]. Histologically they reveal a hypocellular mesenchymal lesion
 74 consisting of a sparse population of bland spindled and stellate cells scattered in a
 75 background of loose myxoid stroma composed of wavy collagen fibrils. The cells have
 76 abundant wispy pink cytoplasm with bland nuclei. There is no cytologic atypia, no atypical
 77 mitotic features or discernible mitotic activity and no evidence of coagulative tumour cell
 78 necrosis either. The lesion often also shows numerous blood vessels of varying calibre [3].

79 The stromal cells can show immunoreactivity to different combinations of vimentin, desmin,
 80 smooth muscle actin, muscle specific actin, CD 34, oestrogen and progesterone receptors
 81 [12]. Recurrent tumours usually show similar histological characteristics [9]. Oestrogen
 82 and progesterone receptors are commonly found in aggressive angiomyxoma, and so they
 83 may be hormone-dependent [10,13,14]. Cytogenetic studies reveal a HMGA2 rearrangement
 84 during embryogenesis [15] making it a sensitive but not specific biomarker for aggressive
 85 angiomyxoma [16].

86 Aggressive Angiomyxoma is a rare tumour [1-4] with extreme cases seen worldwide as
 87 evident in an 11-year old girl [17]; in young boys [18,19] and in pregnant women
 88 [10,20,21]. Adjuvant and complementary modalities for the treatment includes hormonal
 89 treatment, radiotherapy and angiography [22-27]. Its incidence masquerading as a cervical
 90 polyp is even rarer [28, 29].

91 CONCLUSION

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

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