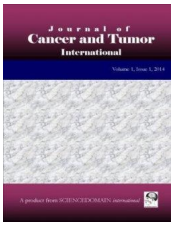

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Aggressive Angiomyxoma Presenting as a Recurrent Cervical Polyp – A Case Report

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Authors' contributions

This work was carried out in collaboration between both authors. Author SMS designed the study, wrote the protocol and wrote the first draft of the manuscript. Author AUA managed the analysis of the study and literature search. Both authors read and approved the final draft of the manuscript.

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Case Study

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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 composed of spindle shaped cells widely spaced from each other in a myxoid 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

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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. INTRODUCTION

Aggressive angiomyxoma (AA) is a rare, locally aggressive myxoidmesenchymal 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 ata 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 follow-up is advised. The recurrence rate is high, resulting in multiple operative procedures with substantial morbidity [8].

2. CASE PRESENTATION

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

The patient was prepared for surgery and the operative findings were that of a predominantly uterine mass extending to the upper cervix that prolapsed below the vaginal introitus. It is firm and measures about 16x16x12cm. It has a broad based sessile stalk and bleeds easily to touch. The upper cervix and lower uterus was destroyed by the mass leaving a small fundus. The round ligament, fallopian tubes and ovaries were grossly normal. She had polypectomy and subtotal hysterectomy. The choice of subtotal hysterectomy as against total was because the patient was young and the lower cervix was not affected. Consequently, because the Cervix enhance sexual excitement during coitus a subtotal hysterectomy was a better option for this young lady who eventually may consider having a partner to consummate marriage to her satisfaction. Although spontaneous pregnancy cannot be achieved, her sexual function have been preserved to some extent .Specimen was sent for histology.

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

3. DISCUSSION

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 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].

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

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

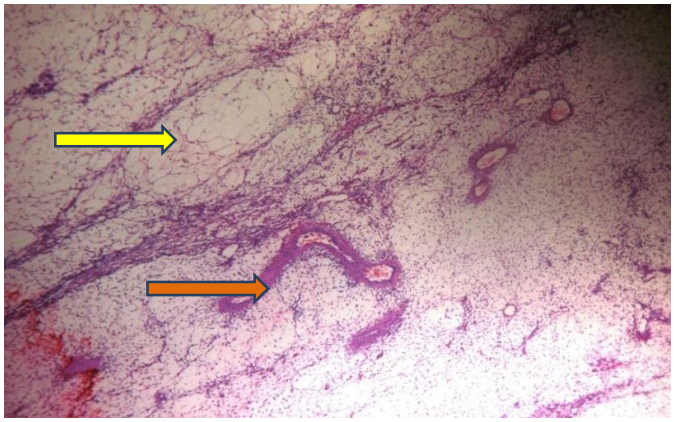

Fig. 1. Proliferating Vascular channels (red arrow) in a myxoid stroma (yellow arrow). (H&E Mag. X100)

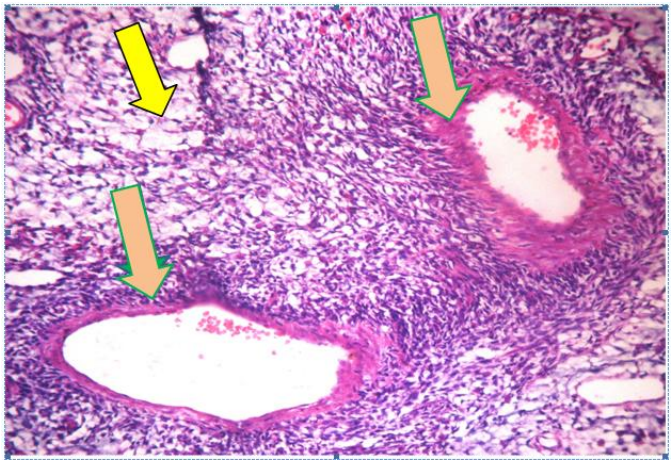

Fig. 2. Section showing dilated vascular channels (brown arrow) disposed in a myxoid stroma (yellow arrow) (H&E Mag. X200)

Table 1. Comparison of index case with other reported cases

	Index case	Case 1 [28]	Case 2 [29]
Age (years)	16	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

The stromal cells can show immunoreactivity to different combinations of vimentin, desmin, smooth muscle actin, muscle specific actin, CD 34, oestrogen and progesterone receptors [11,12]. Recurrent tumours usually show similar histological characteristics [9]. Oestrogen and 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].

4. 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.

The limitation of this study is that Immunohistochemistry was not done to support the histological diagnosis and was because the markers for this technique are currently out of stoke.

CONSENT

All authors declare that written informed consent was obtained from the patient (or other approved parties) for publication of this paper and accompanying images.

ETHICAL APPROVAL

All Authors hereby declare that all experiments have been examined and approved by the appropriate ethics committee and have therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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