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Cervical Aggressive Angiomyxoma.

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ABSTRACT

This is a rare case of cervical aggressive angiomyxoma who presented as a cervical polyp. This patient presented with a mass descending per vagina and was initially diagnosed to have infravaginal elongation of cervix with cervical polyp.Vaginal hysterectomy was done. Histopathology revealed cervical angiomyxoma. Out of 150 cases reported so far, only 2 patients presented as cervical polyp. Wide excision is the treatment with good prognosis. Long term follow up is necessary due to its high local recurrence rate.

Keywords: Aggressive angiomyxoma, cervical polyp, mesenchymal tumour

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INTRODUCTION

Aggressive angiomyxoma (AAM) is a rare mesenchymal neoplasm arising primarily in the soft tissue of the pelvis and perineum of adults. The tumour was first described by Steeper and Rosai in 1983 [1]. It occurs predominantly in women of reproductive age, with a peak incidence in the fourth decade of life and an age range of 11 to 77 years [2-4]. It is a slow-growing mass and has frequent local recurrence. In women, vulva is the most common site of involvement [5]. In men, AAM involves analogous sites, including the inguinoscrotal region and the perineum [6]. This patient was initially diagnosed to have infravaginal elongation of cervix with cervical polyp but postoperative histopathology revealed cervical angiomyxoma.

CASE REPORT

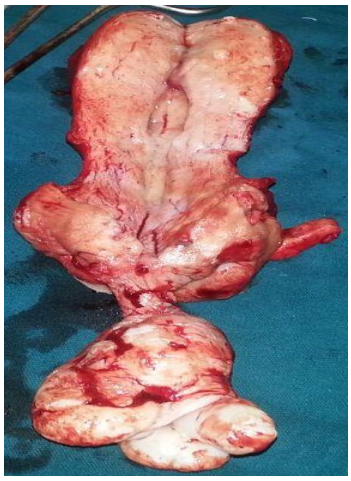
In June 2014, a fifty year old lady P2L2, both full term normal vaginal deliveries, sterilized, postmenopausal by 2 years attended our gynaecological clinic with complaints of mass descending per vagina, associated with backache since one year. She had difficulty in walking and sitting for the past twenty days.

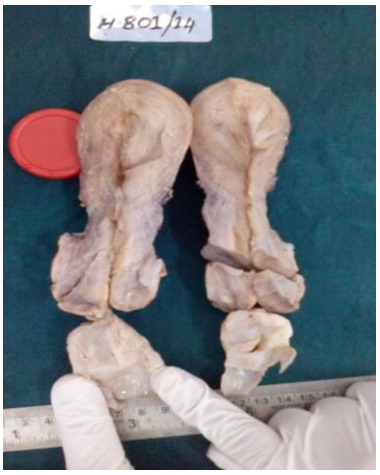
She attained menopause two years back. Regarding her obstetric history, both were home deliveries, with a pregnancy interval of three years.

She was moderately built and nourished. She was anaemic. On local examination, cervix was seen lying outside the introitus with a fleshy polypoidal growth of 5 x 3 cm arising from the anterior lip of cervix. There was no cystocoele or rectocoele on speculum examination. On per vaginal examination, a polypoidal mass was felt arising from the anterior lip of cervix. It did not bleed on touch. Infravaginal elongation of cervix was present with deep fornices and bulky uterus.

She was provisionally diagnosed to have infravaginal elongation of cervix with cervical polyp.As her haemoglobin was 8.1 g%, 1 unit of packed cells was transfused. Pap smear done showed low grade squamous intraepithelial lesion (LSIL). Colposcopy directed cervical biopsy showed chronic cervicitis with LSIL. Microscopy showed hyperplastic stratified epithelium with basal cells, hyperactive koilocytes and focal mild dysplastic changes. Sonography of abdomen: Bulky uterus 8.1 x 4.1 x 5 cm, endometrial thickness of 4mm, and normal ovaries.

Vaginal hysterectomy was done and the specimen was sent for histopathological examination. Gross examination showed the hysterectomy specimen measuring 12 x 5 x 3 cm with attached soft tissue mass attached to ectocervix measuring 5.5x 3.5 x 3 cm. Endometrial cavity measuring 6cm and cervix 2.5cm. Microscopic examination: Section from soft tissue mass attached to cervix showed stratified squamous epithelium enclosing dilated thick walled blood vessels in the background of myxoid stroma. Stromal cells are elongated and stellate with some showing dark stained nuclei. No mitosis or necrosis seen. Section from uterus showed endometrium in secretory phase with adenomyosis and showing chronic cervicitis with ulceration. Focal basal cell hyperactivity was seen.These findings were suggestive of Cervical Aggressive Angiomyxoma.





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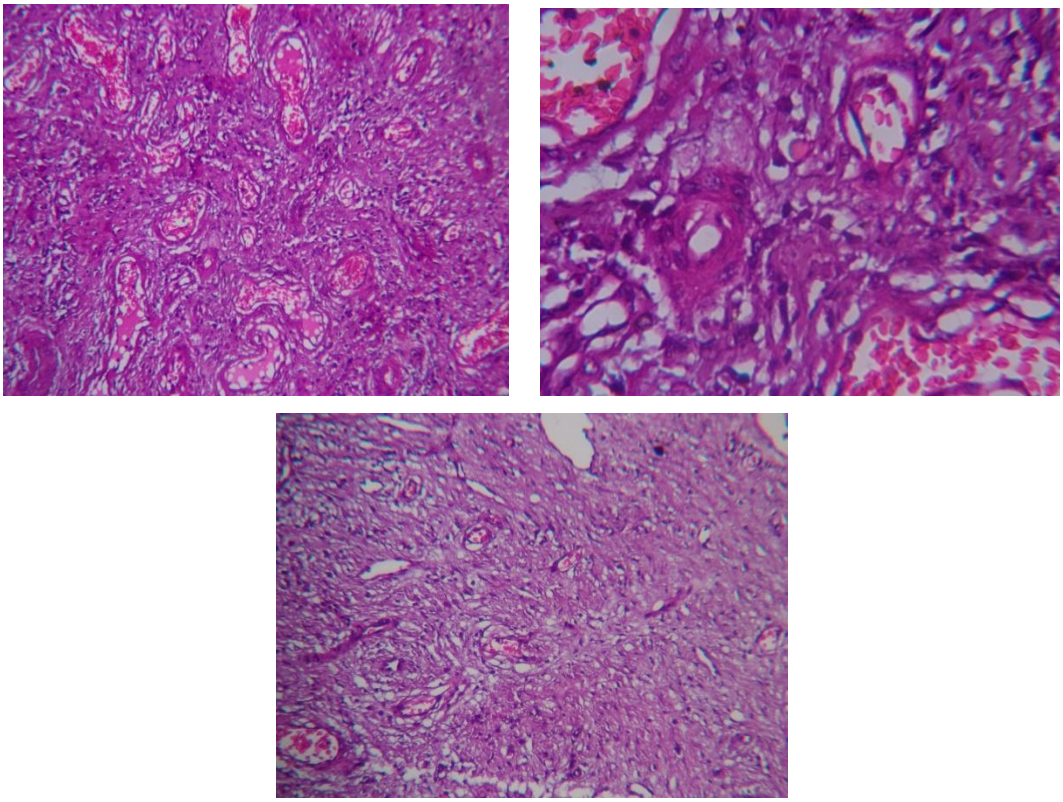

DISCUSSION

Aggressive angiomyxoma is a rare benign mesenchymal tumour occurring in the pelvis and perineum, predominantly in adult females, mostly in the third and fourth decades of life. In a review of more than 100 cases, the female to male ratio was 6.6:1 [7]. It may present as a palpable mass or heaviness and discomfort in lower abdomen or vulva. It is often clinically mistaken for more common entities, such as a Bartholin cyst, vaginal cyst, abscess, leiomyoma, lipoma, fibroepithelial polyp, and inguinal or perineal hernia [3, 8]. The term "aggressive" angiomyxoma was designated to the neoplastic nature of blood vessels, its locally aggressive nature and high risk of local recurrence after treatment[1].

On gross examination, aggressive angiomyxoma is unencapsulated, is poorly or vaguely circumscribed. Tumour size is highly variable and ranges from 1 cm to 60 cm [7]. The tumour is often tan-pink to tan-grey, bulky, and has a rubbery consistency with a glistening, gelatinous cut surface. Areas of congested blood vessels, hemorrhage, or fibrosis may be present [1, 11].

On microscopic examination a sparsely cellular tumour composed of pale to eosinophilic stroma studded with numerous haphazardly arranged blood vessels that stand out against the myxoid background and range in size from thin-walled capillaries and venules to larger muscular arteries [11, 12]. The stroma is distinctly myxoid with intermixed, collagen fibrils; scattered, smooth muscle bundles; and extravasated erythrocytes. The tumour cells are spindle, or stellate shaped. Nuclear chromatin is evenly dispersed with minimal to no cellular atypia. Necrosis or mitosis are usually not seen.

Immunohistochemistry shows focal positivity for vimentin, desmin, muscle specific actin, SMA, oestrogen receptor and progesterone receptor and stains negative for S100 and keratin [8]. There are no etiological factors. Metastasis is very uncommon [13]. Most cases are diagnosed only on histopathological examination after primary surgical excision.

Sonography shows hypoechoic or frankly cystic mass with no additional information. On angiography a hypervascular mass is seen. CT scan shows tumour with well-defined margins and iso- or hypo- attenuation compared to muscles with swirling appearance which is a diagnostic feature [9]. MRI has a swirled pattern in the angiomyxoma, and is more specific than CT. On T1 the tumour shows isosignal compared to the muscles,

similar to the presentation on CT, and moderate contrast enhancement on a gadolinium scan. On T2-weighed MRI, the tumour has high signal intensity. The attenuation on CT and high signal intensity on MRI is due to the loose myxoid matrix and high water content of angiomyxoma [10].

Complete surgical excision is the gold standard treatment. Other treatment options include use of hormonal manipulation such as tamoxifen, raloxifen or GnRH analogs, radiotherapy and arterial embolisation [14, 15]. External beam irradiation & intraoperative electronic beam radiotherapy decrease local recurrence. Prognosis is usually good. As recurrence rate is 30 to 72% [1], long-term follow up is recommended with careful clinical examination and MRI.

CONCLUSION

Aggressive angiomyxoma, though a rare condition, should be considered in the differential diagnosis of a uterine or cervical polyp. Histology is the gold standard for diagnosis. Wide excision is curative. Prognosis is good. Long term follow up is necessary and MRI is the preferred method for detecting recurrence.

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