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CASE REPORT

A RARE CASE OF ANGIOMYXOMA

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ABSTRACT: Angiomyxoma is a rare mesenchymal neoplasm. It mainly presents in female. It is generally a benign tumor and does not invade the neighboring tissues. It has a tendency to recur after surgical excision so it is termed as aggressive. The commonest site is labia majora.
KEYWORDS: Angiomyxoma, puberty, female, polypoidal growth.

INTRODUCTION: First described by Steeper and Rosai in 1983, angiomyxoma is a mesenchymal tumor found mainly in the pelvis and perineum. There is a strong female predominance, with a female-to-male ratio of approximately 6: 1. It is a slow-growing tumor with a high rate of local recurrence. Because of its rarity, it is often initially misdiagnosed, frequently as a gynecological malignancy.

CASE REPORT: A 24 years old married primigravida female with 4 months amenorrhea presented to our tertiary care hospital with a large growth on the left labia majora since the age of 12 years. Initially the lesion was as small as a pea size and then gradually increased to a large cauliflower size. Her personal history included menarche at 15 yrs. of age with regular cycles. There was no h/o pain, swelling, bleeding or difficulty in carrying out daily routine activities. Patient denied any h/o similar lesions in the family.

Dermatological examination showed a large pedunculated polypoidal skin coloured growth approximately 12cms x 15cms arising from left labia majora (photo: 1). The growth was soft in consistency.

The differential diagnosis considered was Giant achrochorodon and Plexiform neurofibroma.

The basic blood investigations were within normal limits. The patient was referred to gynaec deptt. and complete excision of the tumor was done.

Histopathology revealed many thick walled vessels of varying sizes in a loose myxoid and collagenous stroma with spindle and stellate shaped neoplastic cells (photo: 2).

Based on the clinical and histopathological examination a final diagnosis of angiomyxoma was made.

DISCUSSION: Angiomyxoma was 1st described by Steeper and Rosai in 1983.[1]

It mostly occurs in women of child bearing age and progresses rapidly during pregnancy suggesting estrogen may stimulate its growth. It has a peak incidence in the reproductive age group, although cases have been reported from 16 to 70 years.[2]

It generally involves the genital, perineal, and pelvic region, with vulva being the most common site. Patients present with a painless, slowly growing soft tissue mass in the pelvi-perineal region, which may reach a large size by the time the patient seeks medical advice.

Our case also had a similar course. A minority of cases have been reported in males, occurring in the scrotal, perineal, and inguinal regions.[3] It may simulate inguinal hernia, testicular neoplasm, spermatic cord neoplasm, hydrocele, or spermatocele.[4, 5]

This tumor arises from fibroblasts or myoblasts. Microscopically, the tumor is composed of spindle and stellate-shaped cells in a myxoid matrix. The tumor cells express vimentin, desmin, and smooth muscle antigen (SMA) and are negative for S-100.[3] Angiomyxoma is termed aggressive because of its propensity for local recurrences.[4]

Cytogenetic and molecular genetic analyses have revealed rearrangements of chromosomal region 12q13-15, resulting in altered expression of the HMGIC gene, a transcription factor belonging to the high mobility group of proteins.[3]

The differential diagnosis of aggressive angiomyxoma includes fibroepithelial stromal polyp, angiomyofibroblastoma, myxomas, superficial angiomyxoma, myxoid neurofibroma, myxoid liposarcoma, and myxofibrosarcoma.[5] Superficial angiomyxoma differs clinically as they are superficially located and often polypoid. Histologically, they commonly involve the dermis and subcutaneous tissue, unlike AA, which is more deeply located.

Wide local excision is the therapy of choice. The excision of these tumors is difficult as they have the same consistency as that of normal connective tissue and therefore have a propensity for local recurrence.

Systemic metastases have been reported to occur.[6] Long-term follow-up is necessary for early diagnosis of local recurrence and metastases. Imaging studies such as CT scan and MRI help in preoperative evaluation and postoperative follow-up as this tumor is ill defined clinically.[7]

A retrospective review has shown that patients having positive margins were as likely to have recurrence as those with negative margins.[8]

The presence of estrogen receptors in the tumor and its enlargement in pregnancy suggest the possibility of hormone dependence of this neoplasm. Gonadotropin-releasing hormone agonists have been suggested for those cases that are not amenable to surgical excision.[8]

A regular follow-up is important to evaluate local recurrence, if any.[9, 10]

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Photo 1: Pedunculated mass arising from labia majora.

Photo 1

Photo 2: Histopathology: Thick walled vessels of varying sizes in a loose myxoid and Collagenous stroma with spindle and stellate shaped neoplastic cells.

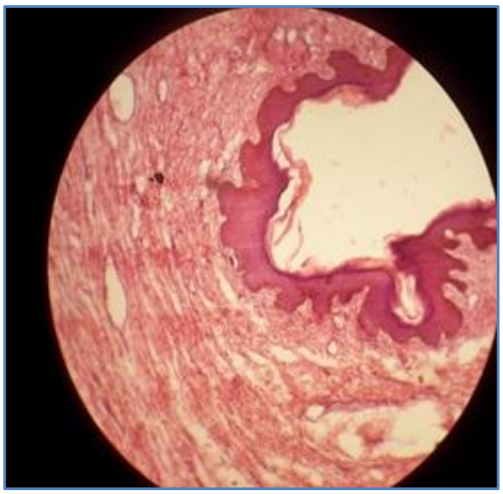

Photo 2

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