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RESEARCH ARTICLE

ANGIOMYXOMA OF VULVA: A SERIES OF 4 CASES

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ABSTRACT

Background: Angiomyxoma is a rare slow growing and soft and benign tumour that predominantly affects in the perineum of women in reproductive age group. It develops from myxoid cells of connective tissue. It is locally infiltrating and recurrent in nature. It is of It is of two types: superficial which grows near the surface and aggressive which involves the deeper structures.
Case report: We herein present a series of 4 cases of angiomyxoma. A 70yrs female presented with complaints of burning sensation and difficulty in passing urine and per vaginal pus discharge for last few days, A 45 yr female presented department with the complaints of a mass in introitus and something coming down per vagina for 5-6 months, A 40yr female came with mild pain in right iliac fossa. Vaginal examination showed a left labial polypoidal mass measuring 4 × 4 cm, and clinically thought to be a Bartholin gland cyst and 50 yr female showed up with a growth on vault.
Conclusion: It is important to accurately diagnose this rare tumor as it can be mistaken both clinically and on microscopy for several other conditions leading to loco-regional recurrence.

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INTRODUCTION

Angiomyxoma is a rare slow growing and soft and benign tumour that predominantly affects in the perineum of women in reproductive age. It is usually mistaken both clinically and microscopically for several other conditions. (Alam *et al*., 2014)

Case reports

Here we report 4 cases with different signs and symptoms;

Case 1

A 70yrs female presented with complaints of burning sensation and difficulty in passing urine and per vaginal pus discharge for last few days. Per speculum examination did not reveal any growth. Bilateral fornices were however tender, A vulval biopsy was taken and sent to our department. Histopathology revealed hypocellular tissue with spindle and stellate-shaped cells against myxoid background. There were plenty of capillary-sized blood vessels with perivascular infiltrate made up of lymphocytes, eosinophils, and a few neutrophils. Focally arborizing pattern of blood vessels were also seen. There was no mitotic activity. Features were suggestive of Cutaneous superficial angiomyxoma. On IHC, spindle cells were positive for Vimentin, SMA but negative for S-100, CK.

Case 2

A 45 yr female presented department with the complaints of a mass in introitus and something coming down per vagina for 5-6 months. There was no history of urinary or menstrual disorder. The mass was gradually increasing in size. She had one child, age about 17years. On examination there was a mass about 9x7x6 cm protruding through introitus, which was soft and irregular consistency but with a shiny smooth surface. It was not reducible, but it was pushed into introitus manually. Then the mass was removed per vaginal route by giving incision on anterior vaginal wall. It was en capsulated, solid nodular lobulated yellow-bluish lipoma like structure. Histopathological report revealed the mass as anxiomyxoma.

Case 3

A 40yr female came with mild pain in right iliac fossa. Vaginal examination showed a left labial polypoidal mass measuring 4 × 4 cm, and clinically thought to be a Bartholin gland cyst. The cervix and vagina were healthy. The uterus was normal in size, pushed to the left and posteriorly with restricted mobility. The microscopic examination of polyp proved to be a angiomyxoma.

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Case 4

A 50 yr female showed up with a growth on vault measuring approximately 3x2cm. Microscopy revealed it to be angiomyxoma.

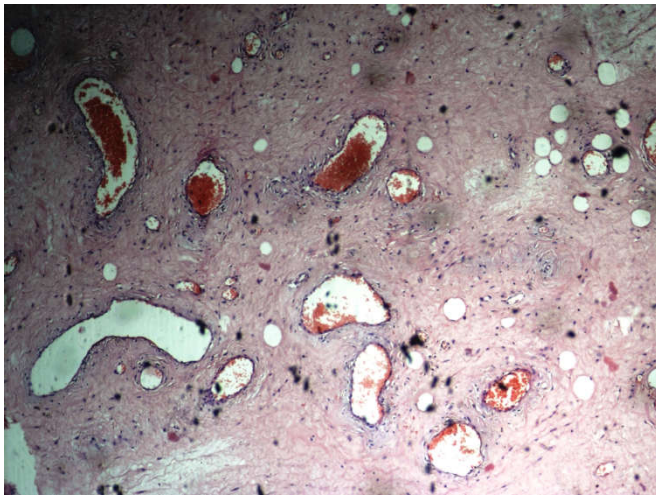

Fig. 1. Photomicrograph showing angiomyxoma vascular areas (H&E, 40x)

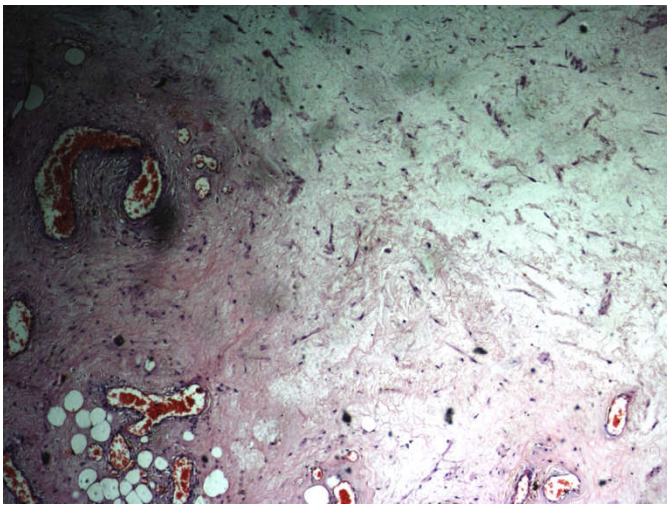

Fig. 2. Photomicrograph showing angiomyxoma; myxoid areas (H&E, 40x)

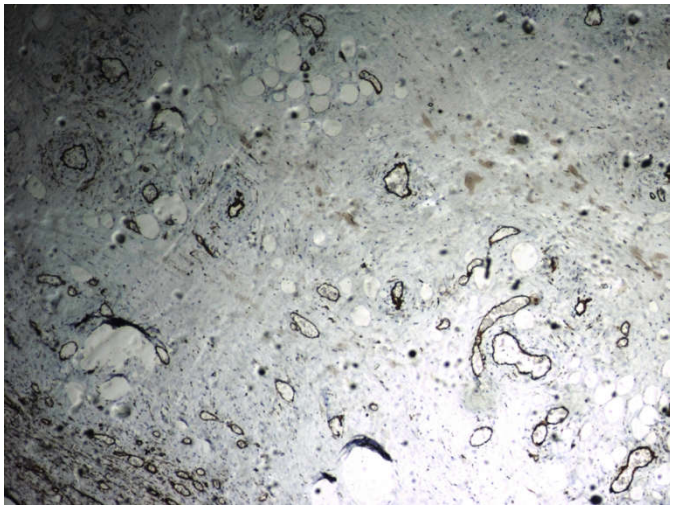

Fig. 3. IHC showing CD34 positivity (IHC, 40x)

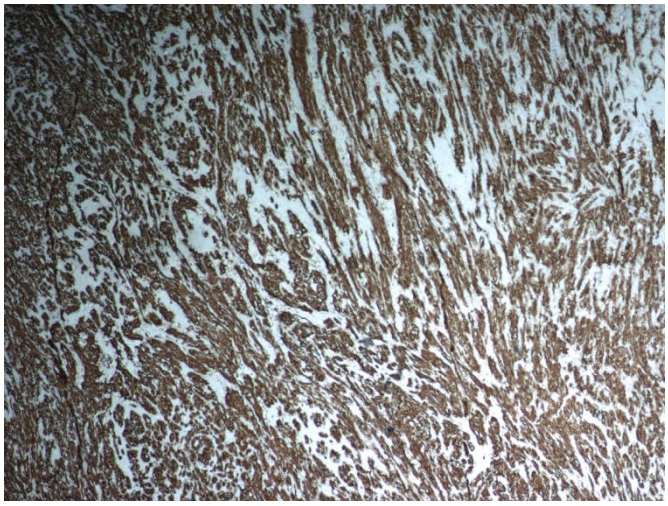

Fig. 4. IHC showing SMA positivity (IHC, 40x)

DISCUSSION

Angiomyxoma is a rare tumor developing from myxoid cells found in body's connective tissue. It has predilection for the pelvis and perineal regions in females mainly and occur rarely in men.1 It is locally infiltrative and has recurrent nature perhaps due to incomplete excision because of misdiagnosis and rarity of tumor. The age distribution is wide, with the peak incidence at 31 to 35yrs. It is of two types:

(1) Superficial which grows near the surface and
(2) Aggressive which involves the deeper structures.

There is paucity in its complete description in available literature since the time it was first reported in 1983. (Chandra Gupta *et al*., 2007) Superficial angiomyxoma presents as subcutaneous nodule of 3-4 cm and mostly involves the genitalia, trunk and head and neck. It presents as polypoid mass and is mostly associated with Carney's complex which is a triad of spotty pigmentation, cardiac myxomas, and endocrinal over activity. As seen in light microscope, the lesion has lobular or multinodular appearance at low magnification. It is poorly circumscribed with extension into subcutaneous tissue. A sparse proliferation of spindle and stellate-shaped cells is seen in extensive myxoid stroma. There is often a prominent vasculature that is focally arborizing. A mixed cellular infiltrate particularly of neutrophils is a feature. (Chandra Gupta *et al*., 2007) Clinically it should be differentiated from cyst, skin tag, and neurofibromatosis. Complete surgical removal is the treatment of choice, but it has a recurrence rate of 30-40%. (Chandra Gupta *et al*., 2007) The other variety named aggressive angiomyxoma is distinctive slowly growing locally infiltrative tumor. It occurs chiefly in genital, perineal, and pelvic regions in women of childbearing age group. Size ranges from a few centimeters to more than 20 cm. It is not associated with Carney's complex. Grossly they are soft and partly circumscribed. On cross section they have gelatinous appearance. Microscopically the tumor is composed of widely scattered stellate and spindle-shaped cells with variable-sized thick and thin-walled vascular channels in myxoid stroma. Clinically it needs to be differentiated from bartholin cyst, periurethral cyst, and hernia. It needs complete surgical removal but has a local recurrence rate of 30-40%. The differential diagnoses ranges from benign tumours such as myxolipoma, myxoid neurofibroma, and myxoid leiomyoma to myxofibrosarcoma, myxoid liposarcoma, and botryoid

rhabdomyosarcoma, vaginal polyps, myxoma, spindle cell lipoma. The distinctively striking vascular component in aggressive angiomyxoma helps ruling out most of the above mentioned neoplasms as differentials. Aggressive angiomyxoma in the perineum may also be misdiagnosed as angiomyofibroblastoma. However, the latter has circumscribed margins, higher cellularity, plumper stromal cells, and many more vascular channels. Another important entity, especially the case of multifocal tumours, is disseminated peritoneal leiomyomatosis, which can be associated with multiple leiomyomas in the uterus and pelvis. Our present cases had no uterine leiomyomas on the pelvic scan. (Kaur *et al*., 2000) Cytogenetic and molecular analysis revealed clonal karyotypic abnormalities including chromosomal translocation involving chromosome 12 associated with rearrangement of the HMGIC gene and HMGIC expression in aggressive angiomyxoma is of value in the distinction of difficult cases from its histological mimics. (Akbulut *et al*., 2006)

Conclusion

Aggressive angiomyxoma is a rare, benign neoplasm that can be mistaken both clinically and on microscopy for several other conditions. It is important to diagnose this condition because the tumour is locally infiltrative and requires wide excision and follow up.

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