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Case report

DEEP AGGRESSIVE ANGIOMYXOMA IN THE PELVIC REGION: A CASE REPORT

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ABSTRACT

Introduction: Aggressive angiomyxoma is a rare soft tissue neoplasm that usually arises within perineum of woman of reproductive age. A mass in vulva, which clinically can be diagnosed as a Bartholin gland cyst, should have aggressive angiomyxoma in differential diagnosis. Rarely, cases reported in males and children. **Case report:** A case of 40yrs female, presented with complaints of pain in abdomen and distension of abdomen since 20 days, is being reported. On abdominal palpation: Tender, abdominal mass felt corresponding to 24-26 weeks. Per speculum examination revealed that cervix taken up. Ultrasound abdomen suggestive of 36x23x15cms neoplastic abdomino-pelvic mass, heterogenous, hypoechoic & solid. Uterus bulky with loss of endometrial-myometrial complex. Total abdominal hysterectomy with bilateral salpingoophorectomy done. Grossly, a huge 32x20x14cms tumor, weighing 2500gm, was received; whose histological diagnosis was deep aggressive angiomyxoma. **Conclusion:** Aggressive angiomyxoma is a rare, mesenchymal neoplasm, which infiltrates surrounding tissue. It is more common in women.

Keyword: Aggressive angiomyxoma, Soft tissue neoplasm.

INTRODUCTION

Aggressive angiomyxoma is a rare mesenchymal soft tissue neoplasm which has a high predilection to vulva, vagina and perineum of women. In 1983, Steeper and Rosai presented a case series of 9 female patients¹. At that time, tumor was first reported in literature. In that case series, they found the most common age group of presentation of the tumor was 21-38 years (young adult women) and size of the tumor was large (up to 60 X 20 cm), and seen infiltrating into the surrounding tissue and found to recur at the same site. For this reason, they labeled the tumor as aggressive¹. It occurs predominantly in woman of reproductive age in the second or third decade of life, but cases have been reported in children². Rarely this tumor has been described in

scrotal and inguinal region in males^{3,4}. The female to male ratio was found to be 6.6:1 in the literature⁵. When a patient presents to a clinician, a clinician may think of other superficial lesions of vulva, vagina and perinium such as vaginal or labial cysts and lipomas. On clinical examination, the size of tumor cannot be assessed properly, as the depth can only be seen by radiological examination. It is difficult to remove it completely by surgery because it infiltrate the surrounding tissue. Because of this reason, in ischiorectal and retroperitoneal spaces tumor recurrence occurs commonly². Distant metastasis is seen in 2 cases. Both were of lung metastasis^{6,7}.

CASE REPORT

We are presenting a case of 40 yrs female presented with complain of pain in abdomen, difficulty in passing stools and urine and feeling of abdominal mass since 2 months. There was no history of fever, nausea, vomiting, and bleeding per-vaginal, apparent weight loss.

Menstrual history was regular with cycle and flow and there were no clots and no dysmenorrhea. Last menstrual period was 10 days back. Obstetric history: Patient was Para 2 Live 2. Both were full term normal deliveries & history of tubal ligation done 15yrs back. On general examination, vitals were stable. No peripheral lymphadenopathy and other signs of malignancy found.

Per abdomen: Tender, abdominal mass felt corresponding to 24-26 weeks; oval, on Rt. side of lumbar spine, arising from pelvis, slightly mobile, firm in consistency, around 20x15cm. Rt. iliac fossa completely obliterated.

Per vaginal examination: Uterus normal size, firm, mobile, separately felt from the mass which was felt in Rt. Adnexa. Rt. Adnexa was fixed; boggy felt in Rt. fornix, Lt. Fornix was relatively free.

Per Rectal examination: large firm mass felt anteriorly free from rectal mucosa, POD was obliterated & mass compressing rectal lumen.

All hematological investigations were done and the investigation reports were within normal limits.

Ultrasound abdomen suggestive of 36x23x15cms neoplastic abdomino-pelvic mass, heterogenous, hypoechoic & solid. Bulky uterus with loss of endometrial-myometrial complex. Ovaries seen separately from tumor on Transvaginal sonography examination.

Exploratory laprotomy under General anaesthesia performed. Large whitish shiny tumor drawn out of peritoneal cavity which was arising from Rt. Adnexa of the uterus and its extension as peduncle in POD and also in the ischio-rectal fossa noted. Total abdominal hysterectomy with bilateral salpingo-oophorectomy done.

Gross examination (Figure1): Weight: 2500gm, 32x20x14 cms in dimension. External surface: Shiny, gray white in colour and rubbery in consistency. Cut surface: Smooth, gelatinous, and gray-white. Areas of haemorrhage seen.



Fig 1: Gross examination and cut surface.

Microscopic examination (Figure2&3): A hypocellular myxoid stroma with numerous small to large blood vessels. At places, perivascular rings of condensed collagen are noted. The tumor cells are oval, spindle shaped, and stellate in appearance. The cells are bland and benign in appearance with ill-defined cytoplasmic borders.

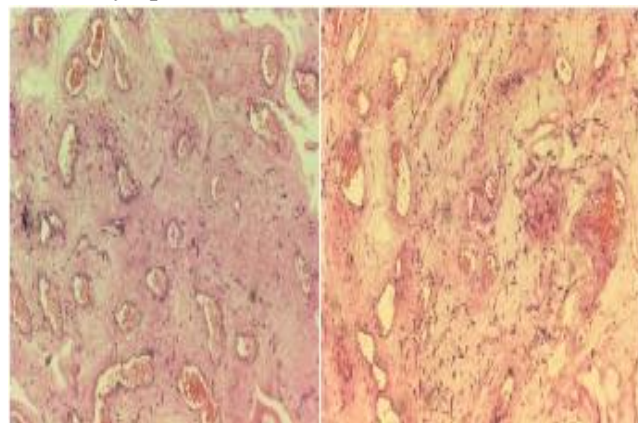


Fig 2: Microscopic examination showed hypocellular myxoid stroma with numerous small to large blood vessels; x200 (H& E Stain).

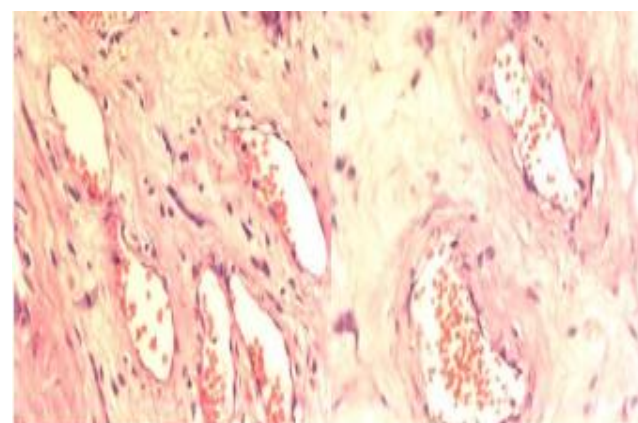


Fig 3: Microscopic examination show cytologically bland, spindle, ovoid and stellate shaped cells. Perivascular rings of condensed collagen are noted; x400 (H& E Stain).

DISCUSSION

Aggressive angiomyxomas are locally invasive connective tissue tumors presenting in about 90% of cases in women of reproductive age group with a peak incidence in the fourth decade of life⁸. Locally infiltrative, may present as a vulval mass, vaginal polyp, Bartholin or a vaginal cyst, ovarian cyst, etc. Cases presented as retroperitoneal mass⁹ and gluteal mass¹⁰ have been reported. These lesions are characterized as soft, non-encapsulated tumors with finger-like projections infiltrating the surrounding soft tissues. These tumors are mostly benign, as we have seen histologically, composed of benign and bland cells. Their growth is slow, and generally do not metastasize. However, it usually tends to recur locally. The rarity of this condition makes the preoperative diagnosis fairly difficult. It has also been also related to hormonal activity which explains female dominance.

Immunohistochemistry: The stromal cells can show immunoreactivity to different combinations of vimentin, desmin, smooth muscle actin, muscle specific actin, CD 34, estrogen and progesterone receptors¹¹.

Angiomyxoma genetics: Chromosomal abnormality involving chromosome 12, associated with rearrangement of HMGIC gene, has been reported in cases of aggressive angiomyxoma¹². HMGIC expression seen in the spindled stromal cells in high proportion but in blood vessels not as high and consistent. This fact indicates that these stromal cells are neoplastic in nature rather than vascular component. Probably, after stromal cell proliferation, blood vessels genesis occurs.

By immunohistochemistry, neoplastic stromal cells show HMGIC expression. As, this is specific to neoplastic cells, the margins can be assessed, which is difficult on clinical ground and also histologically.

Differential diagnosis: Myxolipoma, myxoid variant of liposarcoma, myxoid neurofibroma, myxoid leiomyoma, leiomyosarcoma, and myxofibrosarcoma, fibroepithelial stromal polyp, angiomyofibroblastoma, cellular angiofibroma, massive vulvar edema, fibrous histiocytoma and botryoid rhabdomyosarcoma.

The blood vessels (small and large) and perivascular rings of condensed collagen are helpful in

distinguishing aggressive angiomyxoma from other neoplastic lesions.

Treatment: As, we have discussed that this tumor has high tendency for recurrence at the site of origin, wide local excision is considered the treatment of choice. The margins should be tumor free, otherwise, reexcisions will be the next step during follow up of patient. Sometimes, marked operative morbidity has been reported because of involvement of surrounding organs, as bladder, large intestine, uterus, cervix and bone. So tumor free margins are necessary, and this can be confirmed by immunohistochemical examination.

As, stromal cells show immunoreactivity to estrogen and progesterone receptors, so hormonal therapy can influence the proliferation of tumor cells. GnRH analogues have been used in a few instances in premenopausal women with aggressive angiomyxoma^{13,14}.

Radiotherapy or chemotherapy are not as good treatment options, because tumor proliferation rate is slow and mitotic activity is low.

CONCLUSION

Aggressive angiomyxomas are rare, locally aggressive tumors, arising mainly in the female pelvis with a high likelihood of recurrence even after complete surgical excision. Preoperative diagnosis is difficult due to its rarity and lack of characteristic features. Few management options are available and multimodal therapy may be a good option.

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Conflict of interest: Authors declare that they have no conflict of interests.

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