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CASE REPORT

An Unusual Case of Vulvovaginal Mass: Aggressive Angiomyxoma

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ABSTRACT

Aim: To sensitize clinician about aggressive angiomyxoma as an unusual cause of vulvovaginal mass.

Background: Aggressive angiomyxoma (AA) is not a well known entity. Preoperative diagnosis is often incorrect, and most of the cases are detected on histopathological examination.

Case description: A 45-year-old female, presented with the complaint of slow-growing mass in perineal area for 2 years. A large irregular firm to cystic mass, arising from the posterior wall of the vagina and protruding out of introitus extending into right ischiorectal fossa. Contrast enhanced computed tomography (CECT) revealed a well-defined mass in right perineum arising from the right lateral vaginal wall with ischiorectal fossa extension. Wide local excision was done under general anesthesia. Histopathology showed AA.

Conclusion: Aggressive angiomyxoma (AA) is often misdiagnosed due to the rarity of this entity and absence of diagnostic features, but it should be considered in case of masses in the perianal region in reproductive age females. Complete surgical excision is the first line of management. Long-term follow-up is necessary.

Clinical significance: Correct diagnosis of aggressive angiomyxoma is important as these tumors are benign and does not need radical surgery but recurrence is common, so long-term follow up of the patient is required. So, we reported this case to sensitize clinician about this unusual entity.

Keywords: Aggressive angiomyxoma, Recurrence, Vulvovaginal mass.

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BACKGROUND

Aggressive angiomyxoma (AA) is a rare, slow-growing, mesenchymal tumor. It arises in the pelvic and perineal region of young females of reproductive age group, however, few cases outside pelvis has also been reported. Steeper and Rosai first described this tumor in 1983 and the term AA was coined by them only.[1] The word aggressive has been given to this tumor because of its locally infiltrative nature with the increased propensity of local recurrence, and does not reflect increased propensity of metastasis. The AA is not a well-known entity; available literature is in the form of case reports and small case series only. It is often misdiagnosed tumor, in most of the cases the preoperative diagnosis is often incorrect, and diagnosis is made on the histopathological report only. Correct diagnosis is important as these tumors recur and long-term follows up of the patient is needed. So we are presenting this case report to sensitize clinician about this unusual entity.

CASE DESCRIPTION

A 45 years old P3L3 perimenopausal female came with complaints of mass in the perineal region for 2 years along with difficulty in locomotion and discharge per vaginum since 1.5 years. It was not painful. The mass increased gradually in size. On examination, there was a 9 × 8 cm irregular firm to cystic mass, arising from the posterior wall of the vagina and protruding out of introitus with the bossellated surface. It was nontender and without any ulceration over the mass. The mass also extended into right ischiorectal fossa, 10 × 10 cm with cystic, smooth surface that was irreducible with no cough impulse (Fig. 1). The CECT abdomen and pelvis revealed a well-defined 12 × 10 × 8 cm mass in right perineum arising from right lower lateral vaginal wall with ischiorectal fossa extension. There was no extension into cervix, bladder or rectum. Biopsy taken from the mass was inconclusive. Wide local excision was done under general anesthesia wherein an ischiorectal and vaginal mass of size 30 × 10 cm was excised in toto (Fig. 2). The mass had a smooth surface. Cut section was homogenous in consistency, glistening, gelatinous, with areas of congestion. Her postoperative period was uneventful. Histopathology was suggestive of AA. The tumor showed positivity

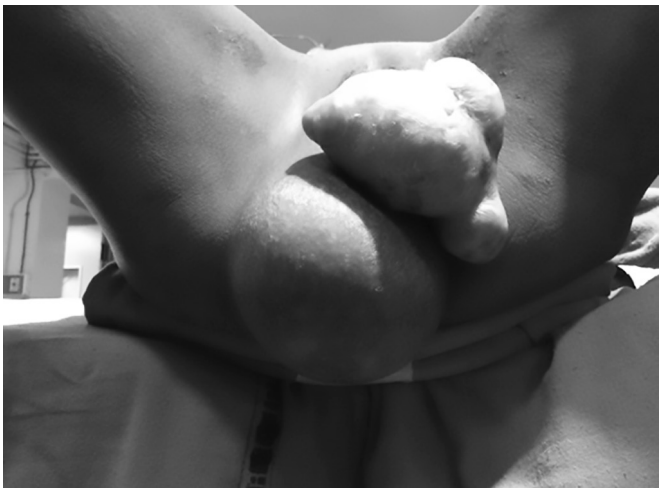

Fig. 1: Cystic vaginal and perineal mass with smooth surface

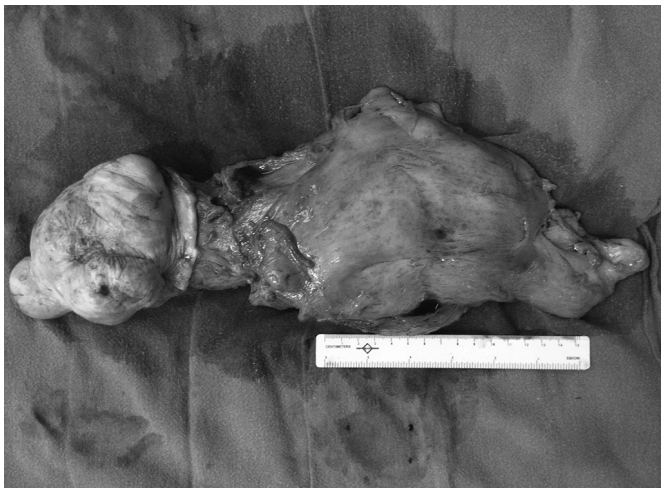

Fig. 2: Specimen after wide local excision, mass of 30 × 10 cm

to desmin and CD34. The patient is currently under follow up, and at 1 year follow-up there is no recurrence at present.

DISCUSSION

An AA is a benign soft tissue tumor. In recent WHO classification of tumors, this has been classified in the tumor of uncertain differentiation and the name given is deep AA.[2] It occurs more commonly in the female with a female to male ratio 6.6:1. It is seen in women in their 3rd to 6th decades, most commonly in the 4th decade.[3] It is rare in males and postmenopausal females. The patients present with the asymptomatic mass in the perineal region. These entities are not seen commonly by the clinician and often misdiagnosed as Bartholin gland cyst, vaginal cyst, hernia or lipoma. For imaging ultrasonography, computed tomography (CT) scan and magnetic resonance imaging (MRI) all are useful. But MRI is best to access tumor relation to pelvic structures and plan surgery accordingly. It is also best in cases of recurrence. Pathogenesis of this tumor is not known. Histopathology shows scant cellularity of spindled and stellate cells in the background of loose collagenous, myxoedematous matrix, and mitotic figures are absent. Immunohistochemistry shows diffuse positivity for vimentin, estrogen, and progesterone. Most of them show positivity for desmin. It shows the variable level of immunoreactivity to actin and CD34.[2] Studies have shown cytogenetic abnormality involving chromosome 12, which were monosomy, structural rearrangements of 12q13–15. On chromosome 12 rearrangement of the gene, called high mobility group protein isoform IC (HMGIC) might have a role in the pathogenesis of this tumor.[4] This gene encodes a protein which is involved in transcription regulation. These tumors are estrogen (ER) and progesterone receptor (PR) positive.[5] The ER and PR positivity can be the reason for its frequent recurrence in pregnancy. Treatment modality of choice is surgical excision with wide tumor-free margins. Various other treatment modalities have been tried. Based on ER and PR positivity hormonal treatment like raloxifene, and gonadotropin-releasing hormone agonist have been tried. Haldar et al.[6] tried GnRH agonist in two patients, one patient responded well but a tumor recurred after discontinuation of the drug and second patient did not respond satisfactorily. But it can be tried in large tumors for preoperative shrinkage so that less radical surgery can be done.[7] Radiotherapy and chemotherapy are not useful because of low mitotic activity of tumor cells.[8] Embolization of feeding vessel have also been tried but not successful because of multiple arterial sources.[3] These treatment modalities can be tried preoperatively in bulky tumors to decrease the extent of surgery and postoperatively to prevent recurrence. Recurrence is common after surgical resection, and the reported rate is almost 50–70%.[9] Most of them recur within 5 years of resection but late recurrence up to 14 years have also been reported. It was an assumption that recurrence occurs when resection margin is not tumor free, but this is not the case. Chan et al. did review of 100 cases, after 10 years of follow-up recurrence rate in patients with and without clear resection margin was 50% *vs.* 60%.[3] These tumors occur in reproductive age females, so fertility preservation is an issue while doing a resection. Further, these tumors are benign and not life-threatening. These are locally aggressive and do not metastasize, but three cases of metastasis have been reported in literature till date. The first case reported by Siassi et al. in 1999, was 63-year-old female with pulmonary, mediastinal, lymph node and peritoneal metastasis.[10] Second reported by Blandamura et al. in 2002, was a young woman with lung metastasis.[11] And the third one reported by Junzu in 2012, was 37-year-old with recurrence and invasion into the veins including the inferior vena cava and the right atrium and with pulmonary metastases.[12] Keeping these

facts in consideration, though one should try for complete resection, incomplete resection can be done when extensive surgery is anticipated in a large tumor and fertility is being compromised in women desirous of it.

CONCLUSION

In spite of wide excision with a tumor-free margin, the recurrence rate is high, but metastasis is rare. In view of this high recurrence rate, long-term follow-up is required. Follow up is done with examination and imaging in case of clinical suspicion.

CLINICAL SIGNIFICANCE

Correct diagnosis of aggressive angiomyxoma is important as these tumors are benign and does not need radical surgery but recurrence is common, so long-term follow-up of the patient is needed. So, we reported this case to sensitize clinician about this unusual entity.

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