Case Report

A rare case of angiomyxoma vulva

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ABSTRACT

Aggressive angiomyxoma is a rare, locally aggressive soft-tissue tumor that has a high propensity for local recurrence. It involves mainly the pelvis, vulva, perineum, vagina and urinary bladder in adult women in the reproductive age. Considering its locally aggressive nature, appropriate management and long-term follow-up is necessary. Though rare, this tumor needs to be considered in the differential diagnosis of vulval tumor. We describe a case of angiomyxoma in a 32-year-old young patient presenting with a large recurrent swelling of the left labia majora.

Key words: Aggressive angiomyxoma, vulval cancer, vulval growth

INTRODUCTION

Aggressive angiomyxoma (AA) is a rare, mesenchymal, locally aggressive soft-tissue tumor.[1] It arises as cystic swelling from pelviperineal region of adult females of reproductive age group, often confused with bartholin cyst. It is derived from fibroblasts or myofibroblasts with nuclei that have no atypical features or mitotic activity. It is a slowly growing tumor with a high propensity for local recurrence. Although it does not metastasize, it requires long-term follow-up because of its nature. About 100 cases have been reported in the literature.

CASE REPORT

We describe a case of 32-year-old patient who presented with slowly growing mass in the left labia majora. She had a history of similar swelling in the past, which was excised 1-year ago. On examination, a spherical mass of 10 by 10 in left labium majora extending from mons pubis to fourchette was seen. It was non-tender, had gelatinous consistency. The overlying skin was normal. A probable diagnosis of recurrent bartholin cyst was made and patient was planned for excision of the mass. Per operatively, exact extent of the tumor was appreciated. Due to the close proximity of the mass to urethra anteriorly and rectum posteriorly, complete excision was not done. The post-operative period was uneventful and the wound healed well. The histopathology showed a poorly demarcated hypocellular neoplasm composed of bland spindle cells with round to ovoid nuclei and patchy eosinophilic cytoplasmic process set in a myxoid matrix also containing variable small to medium sized blood vessels, with some having thickened wall showing hyalinization, fibrocollagenous tissue and at places medial hypertrophy. Other sections showed epidermis with underlying dermis and dermal appendages confirming it to be deep AA The patient is under follow-up. She will be evaluated for the unexcised tumor mass and will be considered for gonadotrophin agonist therapy accordingly.

DISCUSSION

AA is an uncommon mesenchymal neoplasm occurring predominantly in the pelvi-perineal region of adults, first described in 1983 by Steeper and Rosai.[1] In the latest World Health Organization-classification AA is classified under "tumors of uncertain differentiation."[2]

It is seen in females, commonly in reproductive age with peak incidence during the 3rd decade of life, suggesting that estrogen may stimulate its growth. It generally involves the genital, perineal and pelvic region, with vulvar region being the most common site. It presents as a painless, poorly circumscribed gelatinous vulvar mass and clinically simulates a bartholin gland cyst or an inguinal hernia. The

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tumor grows slowly and does not metastasize. However, it is locally aggressive and tends to recur (36-72%) after resection.[3] Our patient also presented with recurrent swelling within 1 year of resection.

Imaging of these tumors is important to determine extent and the optimal surgical approach. In sonography, angiomyxoma is seen as a hypoechoic mass. Angiography usually shows a generally hypervascular mass. It has a characteristic appearance on computed tomography and magnetic resonance imaging and these techniques reveal the extent of the tumor as well.[4]

Treatment is usually surgical in the form of wide local excision. Pre-operative angiographic embolization, pre-operative external beam irradiation and intraoperative electron beam radiotherapy have been mentioned in the literature to decrease the chances of local recurrence.[5] Hormonal treatment with a gonadotropin-releasing hormone agonist has been used by some as neoadjuvant therapy for decreasing the size of tumor before surgical excision. It has also been used to take care of residual tumor in cases of incomplete excision and by some for small AA as primary therapy.[6]

On gross examination, the tumor can be of various sizes but are often relatively large (more than 10 cm). They are typically poorly demarcated and characteristically have a soft, gelatinous appearance but occasionally may show a white and rubbery cut surface in a recurrence, presumably due to involvement of surgical scar tissue. In our patient, the gross examination showed soft rubbery mass with attached muscle tissue and solid white areas consistent with recurrence. Histologically angiomyxoma is a mesenchymal tumor, composed of bland spindle cells with round to oval nuclei and palely eosinophilic cytoplasmic processes set within a strong myxoid background.[7] The matrix contains medium sized thick and often hyalinized blood vessels. The presence of bundles of smooth muscle cells and condensation of delicate fibrillary collagen around the blood vessels is characteristic. As the tumor is deceptively bland and poorly marginated, it is often difficult to distinguish between neoplastic cells and non-neoplastic vulvovaginal mesenchyme. Majority of the tumors demonstrate positivity for desmin in the myxoid bundles and/or stromal cells, while actin and CD34 may be variably positive. The estrogen and progesterone receptor positivity suggests that AA might be hormone dependent tumors. Deep AA must be distinguished from other relatively site specific mesenchymal lesions such as fibroepethilial stromal polyp angiomyofibrobastoma as well as other myxoid neoplasms that occur in vulva. Deep location, poor circumscription, lack of superficial polypoidal growth associated with a fibrovascular core and bland cytomorphologic features distinguish deep AA from fibroepithilial stromal polyp. Angiomyo-fibroblastoma are small, well-circumscribed tumors composed of plump epitheloid cells arranged in perivascular area and are not aggressive locally.[8] Superficial angiomyxoma has a distinctive lobulated growth, in contrast to the diffuse growth of deep angiomyxoma. Furthermore, it lacks medium sized vessels seen in deep angiomyxoma and it is frequently associated with acute inflammatory cells.[7]

Recent cytogenetic and molecular studies have identified a variety of genetic alterations, involving the chromosome 12, in the region 12q13-15 in angiomyxoma.[9]

To conclude, whether the treatment is with surgery, hormone therapy or both, it is comprehensible that AA requires close, long-term follow-up to monitor for disease recurrence.[10] It should be considered as differential diagnosis for big vulval tumors with gelatinous consistency.

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