Case Report

Aggressive Angiomyxoma: An Uncommon Entity with Literature Review

Abstract

Aggressive angiomyxoma (AA) is a soft-tissue tumor of mesenchymal origin, not so common, but it occurs and exhibits marked tendency for local recurrence with extremely low risk of distant metastasis. The most common sites of origin include vulvovaginal region, perineum, and pelvis of reproductive age females. We report a case of AA in a 36-year-old female presenting with polypoidal vulval growth on the right side with a previous history of surgical resection at the same site 4 years before. Fine needle aspiration cytology of the growth revealed only blood and no cellular component. The tumor was excised and submitted for histopathological examination. A diagnosis of aggressive angiomyxoma was made based on characteristic histological features.

Keywords: *Aggressive angiomyxoma, mesenchymal tumors, vulva*

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Introduction

Aggressive angiomyxoma (AA) is a rare slow-growing tumor of mesenchymal origin, which affects women of childbearing age. These tumors are famous for having marked propensity for local recurrence, hence the term aggressive designated to them. Destructive recurrence may occur, but it is now appreciated that more than one recurrence is very uncommon. Aggressive angiomyxoma was first described by Steeper and Rosai in 1983.[1] Clinically, these tumors are mistaken for a number of benign and malignant lesions chiefly including fibroepithelial polyp, bartholin gland cyst, leiomyoma, myxoid lipoma, and liposarcomas[1,2] Recently, the terminology has been changed to deep angiomyxoma instead of aggressive as the recurrences occur many years after the initial excision.[3] In fact, it has become evident that these neoplasms have a favorable, less aggressive course if initially completely excised with negative margins.[4] Wide local excision with 1-cm margin is considered optimal.

Case Report

A 36-year-old female presented with a slow-growing polypoidal growth on the right of the vulva in Obstetrics and gynecology Department of NC Medical College of Israna, Panipat. A clinical diagnosis of vulval carcinoma/rhabdomyosarcoma was made. Fine needle aspiration cytology of the lesion was nondiagnostic. The tumor was excised with wide margin and submitted for histopathological examination in Department of Pathology.

Grossly, the specimen comprised of a polypoidal growth measuring 8.5 cm × 8.0 cm × 6.5 cm [Figure 1]. Cut surface was glistening and homogeneous with focal areas of hemorrhage. Multiples microsections examined from tumor mass revealed monotonous and hypocellular areas composed of small spindle and stellate fibroblasts. Stroma was myxoid with collagen fibers and prominent dilated, thick-walled blood vessels [Figures 2 and 3]. Mitosis was not appreciated, and no evidence of tumor necrosis was seen. Surgical margins were free from tumor infiltration. A histopatological diagnosis of AA was made.

Discussion

The diverse mesenchymal tumors and tumor-like lesions that occur within female genital tract include a number of entities that have only been recently characterized. Among these, one entity is AA, which typically occurs in adult women with a median age in the fourth decade and is a nonmetastasizing, locally infiltrative neoplasm that commonly involves the perineum.[5,6] This lesion is occasionally reported in man, male to female ratio 1:6.[7,8] Histologically, AA is sparsely cellular tumor with prominent stroma

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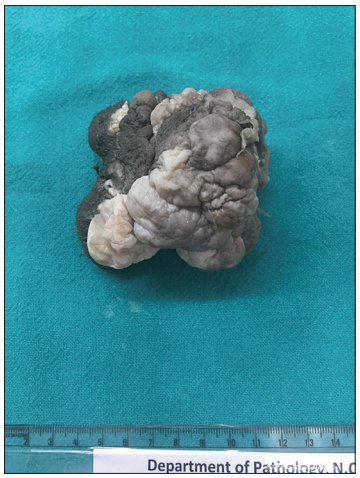

Figure 1: Gross image of aggressive angiomyxoma showing polypoidal growth

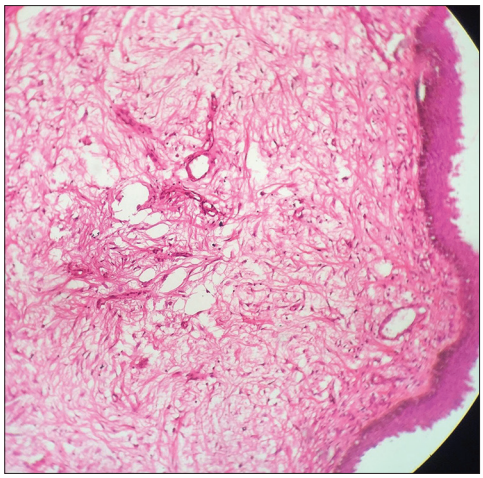

Figure 2: Photomicrograph showing stellate cells lying in abundant myxoid stroma (H and E, ×100)

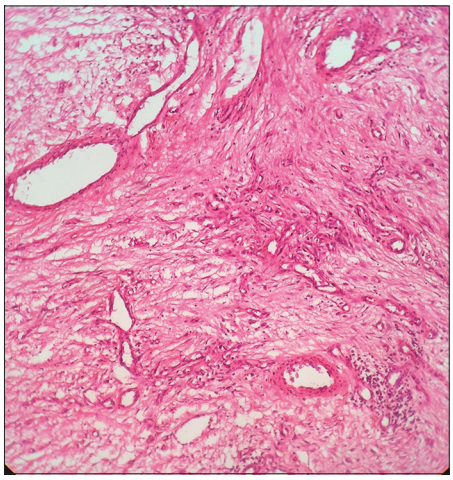

Figure 3: Photomicrograph showing blood vessels of varying caliber (H and E, ×400)

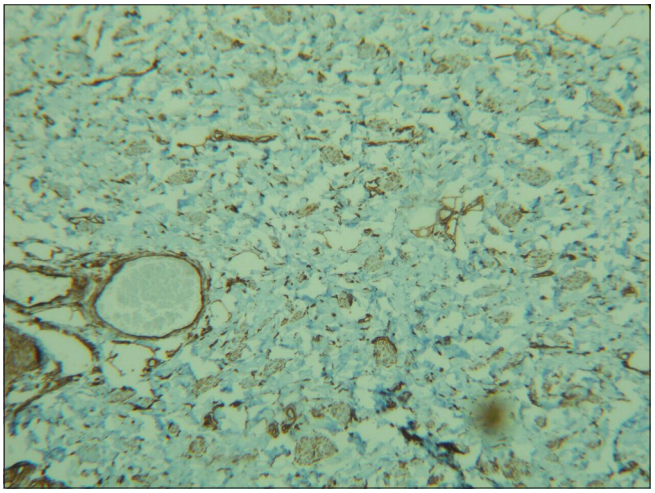

Figure 4: Photomicrograph showing diffuse vimentin positivity in tumor cells (H and E, ×100)

studded with numerous haphazardly arranged blood vessels that range in size from thin-walled capillaries and venules to larger muscular arteries.[9,10]

Immunohistochemically, the tumor cells show diffuse reactivity for vimentin [Figure 4] and desmin. Perhaps, the most characteristic feature is estrogen and progesterone positivity.[5]

Cytogenetic analysis and fluorescent *in situ* hybridization have confirmed the presence of HMGA2 gene rearrangement in majority of AAs thus depicting the same molecular genetic background as other common mesenchymal tumors.[11]

Important differentials of AA include various benign and malignant soft-tissue lesions chiefly including fibroepithelial polyp, angiomyofibroblastoma, superficial angiomyxoma, and myxoid lipomatous tumors.[12] Fibroepithelial stromal polyp is relatively common benign lesion of the vulvovaginal region, close clinical mimic of AA. Histologically, fibroepithelial stromal polyps are lined by unremarkable squamous epithelium with underlying edematous or myxoid stroma. Local excision is usually curative.

Angiomyofibroblastoma is a recently described benign and myxoid vascular soft-tissue tumor of the vulva, relatively well circumscribed with much lower rate of recurrence.[12] Histologically multinucleated giant cells with linearly arranged nuclei are key histological features along with hypocellular and hypercellular zones composed of cytologically bland cells.

Superficial angiomyxomas are commonly located in extragenital sites but may arise in vulvovaginal region.[10] Histologically, tumor lacks large caliber vessels, as seen in AA.

Another important differentials include myxoid lipoma and myxoid liposarcoma. Myxoid lipomas contain abundant mature adipose tissue in myxoid stroma with rare stellate cells. Myxoid liposarcoma contain characteristic lipoblast and arborizing network of capillaries as opposed to disorganized thin- and thick-walled blood vessels of AA.[5]

The treatment of choice for AA is wide local excision with 1-cm margin. As this tumor is locally invasive and tends to infiltrate deep into pelvic soft tissues, preoperative knowledge of the tumor extent is important in determining surgical approach MRI is investigation of choice to determine the extent of deep invasion of AA because of characterstic imaging features.[13]

Conclusion

Our case report concluded that AA is a rare, benign mesenchymal tumor occurring in women of reproductive age group. Despite the morbidity associated with tumor recurrence and repeated surgeries, the prognosis is generally considered good. Close clinical follow-up is strongly recommended to detect early recurrence for appropriate patient management, either with wide local excision or more recent treatment modalities including hormonal therapy.

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Conflicts of interest

There are no conflicts of interest.

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