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Deep angiomyxoma of the vulva in south India: A Case Report

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Abstract

Deep Angiomyxoma is very rare, slowly growing, and benign tumor of mesenchymal origin which affects women in perimenopausal age group and is associated with a high risk of local recurrence. A case of a 42-year-old female is presented, with a large firm gelatinous mass on the right labia majora, measuring 5.5 × 6 cm. The patient underwent wide local excision of the tumor. Histopathological examination showed a hypocellular neoplasm composed of uniformly distributed stellate cells in an abundant myxoid matrix and variably sized vessels suggestive of deep Angiomyxoma. 1 year followup showed no recurrance. Deep angiomyxoma of the vulva has to be distinguished from benign myxoid and malignant myxoid neoplasm. Wide local excision of the tumour is the treatment of choice. Long term follow up is indicated in view of local recurrence.

Keywords: Angiomyxoma, India, Rare, Vulva

Introduction

Deep angiomyxoma is a very rare, slowly growing, and benign tumor of mesenchymal origin, presenting in women of reproductive age, which has high risk for local relapse[1]. Deep Angiomyxoma was first described by Steeper and Rosai in 1983. It is a very rare local mesenchymal tumor of unknown etiology usually affecting vulva, labia major, buttocks, or pelvis of women in reproductive age[3]. Very few cases reports have also been described in men, which affect the scrotum [2]. Pathophysiology is unclear, but estrogen and progesterone influence may also play a role [4]. Women are affected predominantly in early reproductive years Adult men are less commonly affected. Female to male ratio is 6:1. It is Rare in children [4]. Controversy exists regarding management in view of local recurrence. It is a mesenchymal lesion, angiomyxoma mimics disorders which range from non neoplastic vulval hypertrophy and lymphedema to malignant tumours like myxoid liposarcoma.

Presentation

A 42-year-old female is presented, with a large firm gelatinous mass on the right labia majora, measuring about 5.5 × 6 cm with duration of 2 years and increased in size for 6 months.(Fig.1a) It was painless, soft, spongy in consistency, and non-tender. Menstrual cycles were regular with a normal flow. Examination of the cervix, external urethral meatus and vagina were unremarkable. Bilateral inguinal lymph nodes were not enlarged. Ultrasound of the abdomen and pelvis was normal. Cystoscopy was done in view of mass close to

the external urethral meatus, which showed normal urethral meatus, urethra and bladder. She underwent wide local excision of the lesion (Fig.1b). There was moderate bleeding during the procedure but the postoperative course was uneventful. Histopathologically, lesion showed a neoplasm covered by stratified squamous epithelium. The neoplasm was paucicellular (Fig.2a) composed of uniformly distributed spindle to stellate cells with ill-defined cell borders, moderate amount of eosinophilic cytoplasm and an elongated bland nucleus. Interspersed were thin fascicles of smooth muscle bundles and variably sized numerous thin and thick walled blood vessels (Fig.2b, 2c). These cells were set in an abundant myxoid and collagenous stroma(Fig.2d). Differential diagnosis at this stage included fibroepithelial polyp and, angiomyofibroblastoma both being commonly reported in Vulva. Because of the larger size, ill defined margins and absence of predominantly plump cells with perivascular accentuation, Angiomyofibroblastoma was ruled out. Angiomyofibroblastoma is usually a well circumscribed, smaller neoplasm with hypercellular and hypocellular areas with more plump cells with perivascular accentuation. Lack of bizarre stomal cells ruled out fibroepithelial polyp. There were no atypical cells or increased mitosis. With the above morphological features, it was diagnosed as Deep Angiomyxoma. 1 year follow-up revealed no recurrance.

Discussion

Deep angiomyxoma is the benign tumor commonly presents as an asymptomatic mass in the genital area of women in their reproductive life. The term “deep” denotes its propensity for local aggression and recurrence after excision. Approximately 70% - 80 % of the cases have recurrence after a period of 5 years postoperatively and it has been reported even 20 years after surgery. Local recurrences are treated with reoperation with wide local excision [1]. Some controversy exists with regard to need chemotheraphy. Some studies advocate that tumour cells are positive for estrogen and progesterone receptors[2,3]; hence hormonal therapy like tamoxifen and gonadotropin-releasing hormone analogue will reduce the tumor size and helps to make complete excision in large tumors and in the treatment of recurrence [5, 6]. Some studies also conclude that there is No role in Chemotherapy and radiotherapy in view of low grade mitotic figures. However most studies have consensus for Long term follow up with local examination and MRI pelvis for local recurrence [1]. Here we observed that wide local excision of the tumour even without chemotheraphy, with good follow- up can prevent local recurrence as observed in some of the studies. Angiomyxoma is a mesenchymal lesion. Immunohistochemically, these tumors stain for vimentin and desmin. The tumor cells also show some expression for muscle actin. Superficial angiomyxoma, angiomyofibroblastoma, and smooth muscle tumors like leiomyoma, hamartoma, liposarcoma also need to be considered in the differential diagnosis of a solid smooth mass in the perineum [2]. We observed that a wide range of vulvar mesenchymal lesions exist and angiomyxoma has to be differentiated from the mimics which range from non neoplastic vulval hypertrophy and lymphedema to malignant tumours like myxoid liposarcoma.

Conclusion

Deep Angiomyxoma is rare entity in the Vulva, especially when it is painless lesion, particularly in premenopausal women in their third to fourth decades of life. CT/MRI pelvis is advocated in this situation for the extent of the mass. A wide range of vulvar mesenchymal lesions exist and angiomyxoma has to be differentiated from the mimics which range from non neoplastic vulval hypertrophy and lymphedema to malignant tumours like myxoid liposarcoma. Wide local excision is the treatment of choice for deep angiomyxoma of vulva with low recurrence rate. Long term follow up is required with genital examination and imaging studies.

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