Aggressive Angiomyxoa of Vulva: Wide local excision case report

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Abstract

Aims and Background: Aggressive angiomyxoma is rare tumor of pelvic and perineal organs, occurring usually in women of reproductive age, and carrying a high tendency to local infiltration and relapse. It is non-malignant but can be locally aggressive and frequently recurs. Misdiagnosis is frequent. Wide excision is the preferred method of treatment but the challenge is to perform the degree of extirpation necessary to prevent recurrence. Most literature about Aggressive angiomyxoma consists of isolated case reports.

Presentation of Case: We reported a case of 35-year-old Indian woman complaining of a slow and progressive growth of a left vulvar labia majora pedunculated tumor with long a stalk was detected second time after first resection 2yrs back, measuring of 15.0x8.0 cm. Wide local surgical resection of the tumor were performed. Histopathology diagnosed a large aggressive angiomyxoma with uninvolved resection margins. Patient remains without recurrence at 8.0-months follow up.

Discussion and Conclusion: Non Aggressive angiomyxomais rare but should be excluded in any large vulval mass. We expect that awareness accompanied with wide free safe margin excision has important role to prevent the recurrence of tumor.

Keywords: Aggressive angiomyxoma, vulva.

1. Introduction

Aggressive angiomyxoma (AA) is a rare, locally aggressive myxoidmesenchymal neoplasm arising in the pelvis and perineal regions. The name angiomyxoma was chosen because of the similarity to myxoma and the notable vascular component. The term Aggressive angiomyxoma was not coined until 1983 but similar tumors were described as early as in the 1860s. AA was first described in 1983 by Steeper and Rosai.[1] They presented AA as a vulgar polyp clinically and diagnosed it histologically. It usually occurs in women, especially middle-aged; 95% of total cases are found in females.[2] However, it has later been reported in males. In men, the tumour involves analogous sites including the scrotum and inguinal area and usually appears at an older age.[3] It’s suspected there might be a relation with hormonal status that might explain a female to male ratio of slightly more than 6:1. Have been reported about 150 cases in the world medical literature.[4] This benign tumor usually presents in middle aged adults as single nodule or polypoid lesion that may be clinically confused with skin tag or neurofibroma.[5]

2. Case report

35-year-old Indian woman para four, presented with polypoidal, slow-growing painless mass for two year duration, arising from the left labia majora which was causing her difficulty in walking and restricting her free movement. There were no associated symptoms like pain and no urinary or bowel complaints. Past and menstrual history was unremarkable. Patient was admitted in August 2016, and clinical examination showed a large skin covered non-tender, pedunculated polypoidal mass involving the middle part of left labia majora. The soft doughy tumor mass attached arising from vulva of size 15.0x8.0 cm (Figure 1 and 2).
Speculum examination for vagina and cervix were also normal. The inguinal lymph nodes were not enlarged. An ultrasound revealed a subcutaneous hypo-echoic cyst in the left labium major. The uterus, adnexa, and other pelvic organs were normal. CT scans showed a mass of soft tissue originating from the left side of the vulva, without infiltration. Laboratory tests for tumour markers were negative.

The patient underwent wide local excision of the tumor remnants and partial vulvectomy surgery. The pedicle was ligated at the base, excised and labial reconstruction was done. (Figure 3 and 4)

On slicing, cut section was homogenous, white fleshy, soft to feel. No areas of haemorrhage or necrosis were identified.

Microscopically, the tumor was composed of spindle and stellate-shaped cells embedded in a loose myxoid matrix. These cells showed low to moderated cellularity and had eosinophilic cytoplasm with no significant nuclear pleomorphism and mitosis. Variable-sized thin-walled capillaries and thick-walled vascular channels were haphazardly arranged in the stroma. Some of these vessels showed perivascular hyalinization in the vascular walls. (Figure 5)

3. Discussion

At least 100 cases of aggressive angiomyxoma involving the female pelvis or perineum, or both, have been described. At least 14 of these primarily involved the vulva. It is an unusual tumor derived from fibroblasts or myofibroblasts with nuclei that have no atypical features or
It has a strong propensity for local recurrence, and can be aggressive. The preferred method for detecting recurrences is good. Long-term follow-up is necessary and MRI is the preferred method for detecting recurrences.

4. Conclusion

Although a rare diagnosis, AAM can present with unusual features. Detailed radiological examination is helpful in detecting the problem, but histology is the gold standard for diagnosis. Wide excision is curative and prognosis of such tumors is good. Long-term follow-up is necessary and MRI is the preferred method for detecting recurrences.

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