AGGRESSIVE ANGIOMYXOMA OF VULVA

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ARTICLE INFO

Received : 09 July, 2021

Accepted : 28 December, 2021

Published : 21 February, 2022

CR 44

DOI: https://doi.org/10.3126/bjhs.v6i3.43246

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Citation

Aggressive Angiomyxoma of Vulva. Madhav Panthi, Neeta Kafle, Tarun Pradhan. BJHS 2021;6(3)16. 1661-1664.

ABSTRACT

Aggressive angiomyxoma is a rare, slowly growing, and benign tumor of mesenchymal origin that usually occurs in the pelvis and perineum of young females. Steeper and Rosai in 1983, first described angiomyxoma. The term “aggressive” underlines the tumour's locally infiltrative nature and high risk for local recurrence. The rate of relapse varies from 35% to 72%, however, it rarely metastasizes so it has an overall good prognosis. Diagnosis is mainly made on histopathology after surgical resection. The optimal treatment for aggressive angiomyxoma is wide local excision with tumor free margin. We here report a case of aggressive angiomyxoma in 22 years female who presented with mass on left labia majora.

KEYWORDS

Aggressive angiomyxoma, local recurrence, mesenchymal tumor

INTRODUCTION

Angiomyxoma is soft tissue neoplasm with a predilection for pelvic and perineal regions and has a tendency for local recurrence. It is composed of small stellate and spindle cells in a myxoedematousstroma with entrapped regional structures.[1] Aggressive angiomyxoma is rare. Steeper and Rosai in 1983, first described angiomyxoma and reported a case series of 9 female patients.[2] There is predilection for the perineum of reproductive-age women. Very few cases have also been described in men, usually involving the scrotum. Female to male ratio is 6.6/1 and the term "aggressive" underlines the tumour's locally infiltrative nature, but mostly the high rate of local recurrence.[3]It usually presents as a vulval polyp and is diagnosed on histopathology. Estrogen and progesterone receptors are commonly found in aggressive angiomyxoma.[4] There is lack of agreement among pathologists regarding the tumour pathogenesis; however, a fibroblastic/myofibroblastic origin seems most likely.[5] We here report first diagnosed case of aggressive angiomyxoma in our institution in 22 year old female.

CASE PRESENTATION

A 22 years old female presented to our gynecology out-patient department with mass on the left labia majora for one year. Initially the mass was small and increased in size for last six months. There was no history of any vulvar or vaginal discharge, bleeding or pain except a hanging sensation while standing. Menstrual cycles were irregular ranging from 35-70 days but with a normal flow. There was no history of prior perineal surgery. There was no significant past history, no significant family history. Examination of perineum revealed a pedunculated mass from left labia majora measuring approximately 10cm x 8cm x 4cm with stalk 4cm (Figure1). There was no sensation of pain and touch over the mass. Overlying skin showed some area of excoriation and ulcer. There was no associated regional or distant lymphadenopathy. Other systemic examinations were found to be normal. Preoperative investigations were within normal limits. Ultrasonography of abdomen and pelvic had nothing to add. Patient was planned for wide local excision of the mass. Mass was excised under spinal anesthesia and was sent to histopathological examination. Mass was pedunculated and measured 9cm x 6cm x 3cm with 5 cm stalk. Perineal area was healthy after excision (Figure 2). There was no reactionary hemorrhage on postoperative period. On gross examination the mass was soft in consistency with glistening, gelatinous cut surface (Figure 3). Histopathogical examination revealed proliferation of many variable sized thick and thin walled blood vessels. There was proliferation of spindle shaped cells and stellate shaped cells, stroma was edematous and showed focal myxoid areas intermingled with collagen fibers (Figure 4,5). On the background of clinical presentation and histopathologicalreport diagnosis of aggressive angiomyxoma was made out. The patient had complete resolution of the swelling and remains asymptomatic during our first six months of follow up.

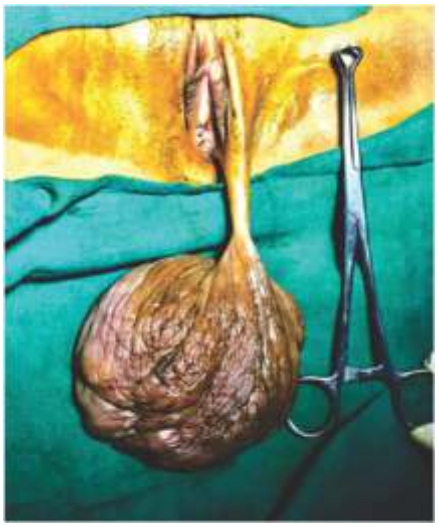

Figure 1: Gross pedunculatedmass with stalk.

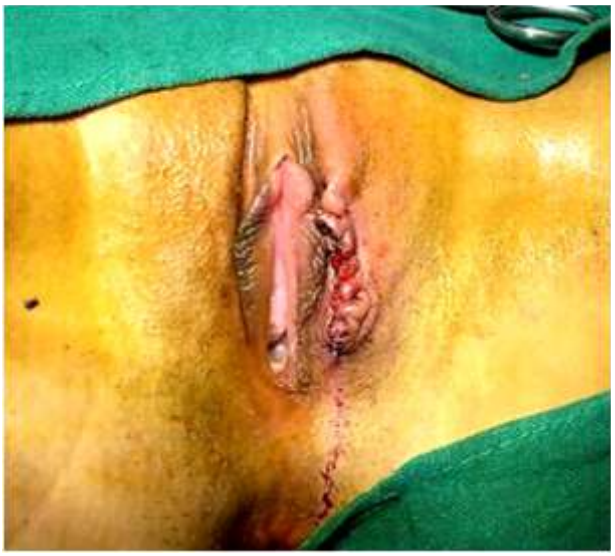

Figure 2: After excision of the mass

Figure 3: Shows glistening, gelatinous cut surface.

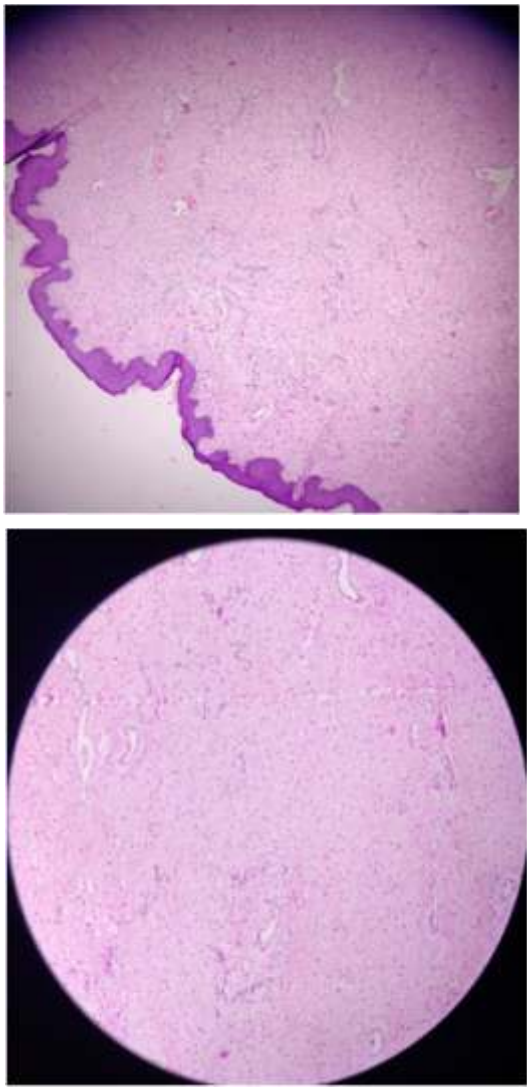

Figure 4 and 5: Histopathological pictures of Aggressive Angiomyxoma under low and high power respectively.

DISCUSSION

In general, angiomyxomas are classified either as superficial (also known as cutaneous myxoma) or aggressive angiomyxoma. Superficial angiomyxoma may occur in the setting of Carney complex.[5,6] This lesions is observed predominantly in male middle aged adults and can arise anywhere in the superficial tissues, but mostly it involves the trunk, lower extremities, and head and neck. Clinically, most lesions appear as slowly growing polypoid cutaneous lesions and are easily confused with a cyst, skin tag, or neurofibroma.[5] But aggressive angiomyxomasmainly occurs on the vagina, vulva pelvic cavity, perineum, hips and crissum in reproductive female. The rate of relapse varies from 35% to 72%.[7] However, it rarely metastasizes so it has an overall good prognosis.[8,9] Tumor size is highly variable and ranges from 1 cm to 60 cm. Most patients with deep aggressive angiomyxoma present with a slow-growing mass in the pelvicoperineal region that is either asymptomatic or associated with regional pain, dyspareunia, or a pressure-like sensation. But in our case the mass was rapidly growing over six months and was asymptomatic otherwise except the sensation of hanging mass. The lesions frequently have a lobular contour with adherence to fat, muscle, and other regional structures. A soft, firm, or rubbery consistency may be present, and a glistening, myxooedematous, pink or reddish-tan cut surface is usually evident. Cystic change has occasionally been noted.[1]Differential diagnosis of aggressive angiomyxoma includes angiomyofibroblastoma, bartholin gland cyst, vaginal polyp , leiomyoma , leiomyosarcoma, lymphangioma , malignant fibrohistiocytoma, myxolipoma, myxoid leiomyoma, myxoidneurofibroma, slerosing hemagioma.[10] Sometimes it may misdiagnosed with an obturator hernia.[11] The optimal treatment for aggressive angiomyxomais wide local excision with tumor free margin, as this tumor is locally invasive and tends to infiltrate deep into pelvic soft tissues. There are no guidelines in the postoperative management of vulvar aggressive angiomyxoma; however due to the high recurrence rate and potential morbidity associated with undiagnosed recurrences, several authors have recommended periodic evaluations with physical examination and MR imaging up to fifteen years after treatment.[12] We advised our patient for regular follow up to evaluate any signs and symptoms of local recurrence. Aggressive angiomyxomashows some immunohistochemical markers. Studied carried out by Chen et al in 5 patients concluded that tumor cells showed strong expression of vimentin, desmin ,estrogen receptor(ER) and progesterone receptor(PR) on the other hand, partial or weak expression was observed for smooth muscle actin(SMA) , actin , CD34 , and S-100.[13]Similarly study carried out by Jingping and Chunfu concluded that aggressive angiomyxoma tended to be strongly positive for vimentin, smooth-muscle actin, and CD34 but mostly negative for S-100.[14] Although the first line of treatment of aggressive angiomyxoma is surgery but when organs, such as the rectum and bladder to which the tumor may be attached, are spared as the morbidity of extensive surgery may not be justified due to its high recurrence rate even after complete resection. So hormonal therapy with GnRH agonists may be of value in managing cases of aggressive angiomyxoma, either primary or recurrent, which are not amenable to surgical excision.[15]

CONCLUSION

We conclude that angiomyxoma is a rare, locally aggressive tumor, which originates from mesenchymal tissues with the tendency of local recurrence. It usually presents as a vulval mass and its definitive diagnosis is made on histopathology. The first line of management of aggressive angiomyxoma is wide local excision with tumor free margin. As there is risk of recurrence of disease regular follow up is mandatory. We resected the tumor with no visible or palpable residual tumor and there are no signs of recurrence after six months without any hormonal therapy.

CONFLICT OF INTEREST

No any conflict of interest.

FINANCIAL DISCLOSURE

None

PATIENT CONSENT

Written informed consent was obtained from the patient for the publication of this case report and the images.

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