International Journal for Multidisciplinary Research (IJFMR)
E-ISSN: 2582-2160 ● Website: www.ijfmr.com ● Email: editor@ijfmr.com

Aggressive Angiomyxoma of Vulva in Young Female: A Rare Case Report

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Abstract

Aim/Background: Aggressive angiomyxoma is a rare and benign mesenchymal stromal tumour predominantly occurring in vulvovaginal, pelvis and perineal regions in young females. The tumour has very high risk of local recurrences with a low likelihood of metastasis. Because of its rarity It is usually misdiagnosed.

Materials & Methods: We report here a rare case of 32-years-old female patient who presented with slow growing swelling on the right sided labia majora for the last 1 year. USG revealed hypoechoic lesion in vulvar region reaching upto the right pararectal region. Patient had undergone surgery for the tumour. Histopathology report was indicative of Aggressive Angiomyxoma of Vulva.

Results: After taking informed and written consent. Wide local excision of the mass and surrounding margins was done under anaesthesia. Patient had undergone Hormonal therapy with GnRH injections (Leuprolide) post-surgery. Her postoperative recovery was uneventful. No complications and recurrence was noted on regular visit to Outdoor patient department uptill now.

Conclusion: Aggressive angiomyxoma is a benign and aggressive mesenchymal stromal growth affecting Pelvic and External genital region of young females. Surgical excision is the treatment of choice, but the tumour carries a high risk of recurrence after complete excision. Hormonal Therapy plays a key role in preventing recurrence. A long-term surveillance should be done for a period of at least 12 months post-surgery for the detection of any recurrence.

1. INTRODUCTION

Aggressive angiomyxoma is a rare and benign mesenchymal stromal tumour most commonly involves pelvis and perineum of young females of reproductive age group. It was initially described in 1983. The tumour has significantly increased chances of local recurrences with reduced metastatic tendency. It can be misinterpreted due to its infrequency. So far, the underlying causes for Aggressive Angiomyxoma remains unclear. Here we report a case with giant vulvar aggressive angiomyxoma and describe the clinical presentation, diagnostic modalities and treatment procedures related with the disease.

2. CASE REPORT

We report a case of 32-years-old female (gravida 2, para 2)patient who presented with slow growing swelling on the right sided labia majora with mild pruritis for the last 1 year. Patient has previous history of same 3 years back for which surgical management was done.

Local examination was indicative of well circumscribed, non-tender, pedunculated, spongy mass of size 15cm x 5 cm.

Ultrasonography of pelvis was suggestive of hypoechoic lesion (126x45)mm seen in vulva extending up to right pararectal region.

On Histopathology, the tumour was composed of spindle and stellate-shaped cells in a myxoid matrix suggestive of aggressive angiomyxoma with tumour free margins. Examination of tumour biomarkers like carcinoembryonic antigen, CA125 and CA199 or alpha-fetoprotein were within normal range. No lymph nodes seen with normal adnexa, urinary bladder and rectum.

Tumour was completely resected including 2 cm of surrounding normal skin margins. Patient had undergone Hormonal therapy with GnRH injections (Leuprolide) post-surgery. Her postoperative recovery was uneventful. Close follow-up of the patient is being done without recurrence, 4 months post-surgery. A recent Ultrasound of her abdomen and pelvis was normal.

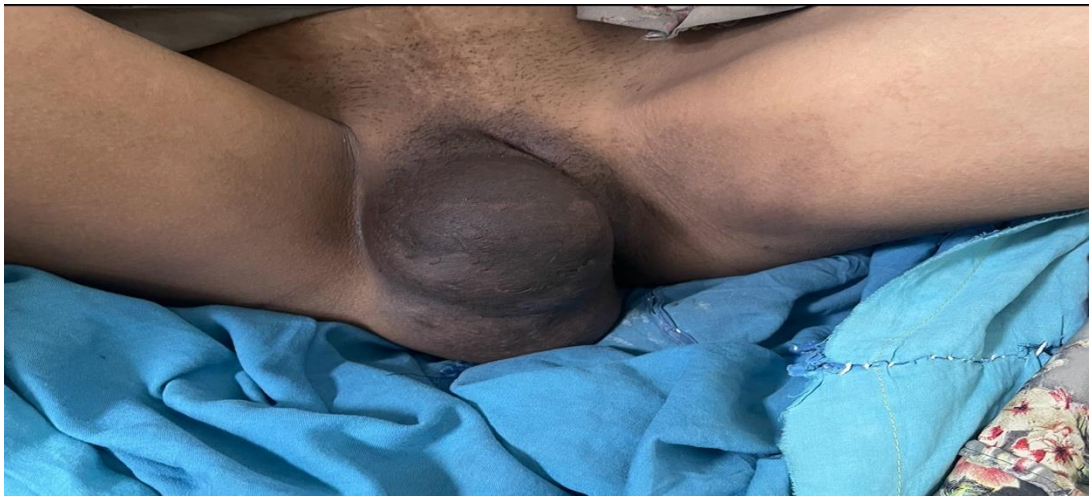

Figure 1. 15 cm x 5 cm mass at right sided labia majora

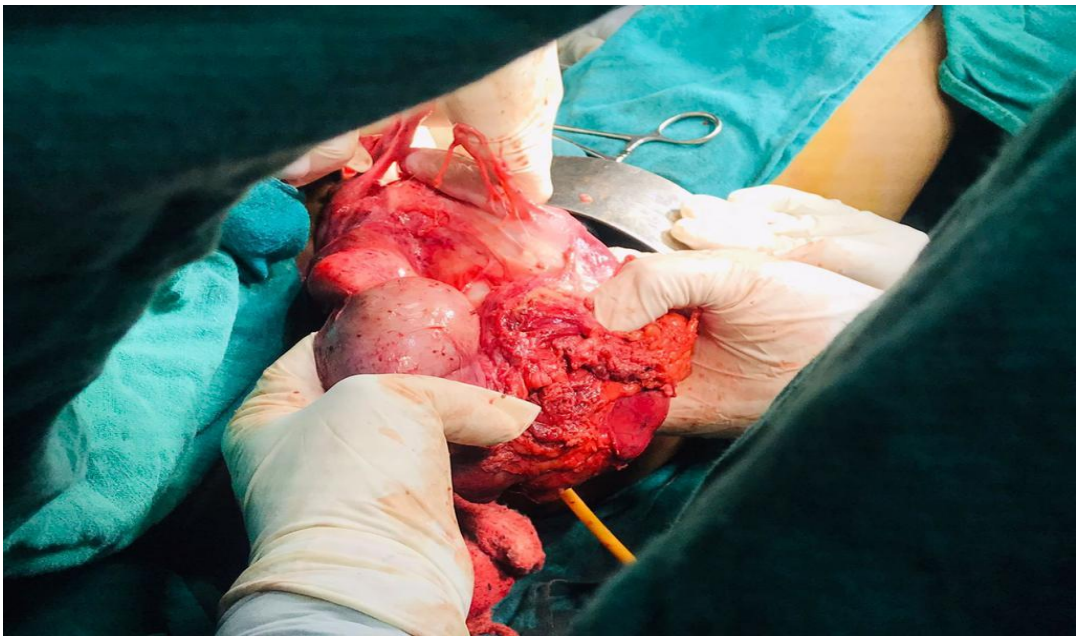

Figure 2. Surgical resection of tumor

3. DISCUSSION

Steeper and Rosai initially described nine cases of Aggressive angiomyxoma in 1983.It arises from mesenchymal cells most commonly occurring in pelvic and perianal region, among females of reproductive age group (30-40 yrs.).

Differential Diagnosis includes angiomyolipoma, histiocytoma myxoid neurofibroma, leiomyosarcoma, rhabdomyosarcoma, lipoma. It is usually misdiagnosed as Vaginal polyp, Vaginal prolapse, Vaginal cyst, Bartholin cyst, Gartner`s duct cyst, pelvic floor hernia.

Surgical excision is the mainstay of treatment with high chances of recurrence (>50%). On histopathology, the tumour is constituted of small spindle and stellate fibroblasts with collagen fibres. The tumour markers CA125, CEA, CA199 usually remains within normal range.

Radiotherapy and chemotherapy has no role in the management, but hormonal therapy with GnRH, Tamoxifen and raloxifene possesses therapeutic effect such as shrinking AAM or retarding the tumour growth.

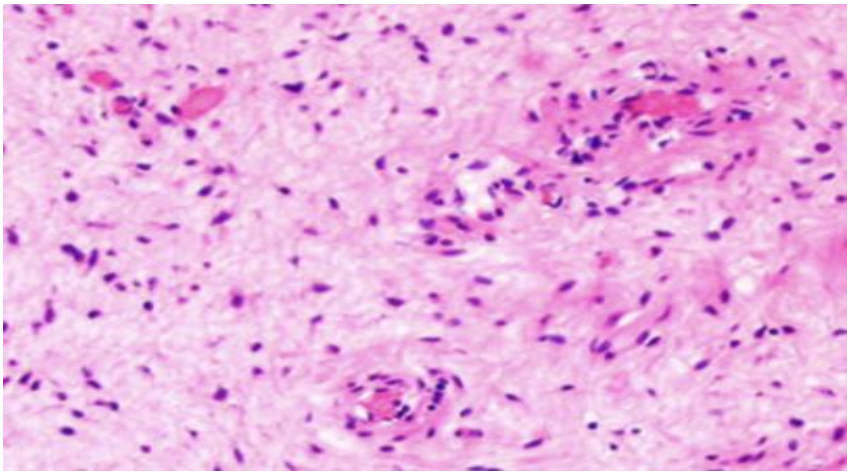

Figure 3. Histopathology image showing spindle and stellate fibroblasts with collagen fibers.

4. CONCLUSION

Aggressive angiomyxoma is one the differential diagnosis for vulvovaginal, pelvic and perianal swelling. Pre-operative diagnosis requires imaging to determine complete extent and location of tumour. Surgical excision is the mainstay of treatment. Close follow-up of patient plays crucial rule in detecting any recurrences. Usually Hormonal therapy is given post-surgery in order to control recurrence. Correct diagnosis and appropriate management using a multidisciplinary approach are very important in management of such patients.

The authors have no Financial disclosures to report. The authors mentions that they have no conflict of interests with the material presented in the case report.

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