

A CASE REPORT ON ANGIOMYXOMA OF VULVA - A RARE MESENCHYMAL TUMOR IN REPRODUCTIVE AGE GROUP WOMEN

ASHISHJOT KAUR¹, PRABH SIMRANPAL¹, ARSHDEEP KAUR*¹, ANJU GUPTA¹

Department of Obstetrics and Gynaecology, Government Medical College, Patiala, Punjab, India.

*Corresponding author: Arshdeep Kaur; Email: arshdeepkaur658@gmail.com

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ABSTRACT

Aggressive angiomyxoma (AAM) is a mesenchymal tumor that arises from perineal or lower pelvic connective tissue. This tumor is locally aggressive. It has mostly been found in reproductive age group females. The correct diagnosis and follow-up of this tumor are very important, as it has similar presentation to many other vulvar masses and it tends to recur due to its locally aggressive nature. We report here a case of 40-year-old female P2L2, who presented with a soft, non-tender swelling on the left labia majora.

Keywords: Aggressive angiomyxoma, Mesenchymal tumor, Vulvar mass, Histopathology, Recurrence.

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INTRODUCTION

Aggressive angiomyxoma (AAM) is a mesenchymal tumor that arises from perineal or lower pelvic connective tissue. This tumor is locally aggressive. This tumor is much more common in women as compared to men, with a female-to-male ratio of 6.6/1. It has mostly been found in reproductive age group females [1]. It mainly involves anatomic sites including pelvis, vulva, vagina, perineum, and bladder. It is often misdiagnosed as Bartholin duct cyst, Gartner duct cyst, vaginal prolapse, lipoma, vulvar mass or vulvar abscess, or vaginal prolapse, as the presentation of AAM is very similar to these conditions [2]. Term "aggressive" is used for this tumor due to its infiltrative nature and high rates of local recurrences. AAM has been defined as "tumor of uncertain differentiation" by the World Health Organization. Approximately 350 cases of AAM have been reported to date [3].

CASE REPORT

A 40-year-old female presented to Gynecology outpatient department, Government Medical College, Patiala, Punjab, India, on March 2nd, 2022, with the complaint of swelling on the perineal area for 1 year. On local examination, there was a 4 × 2 cm soft, non-tender swelling in subcutaneous soft tissue lateral to left labia majora. Ultrasonography showed a hypoechoic lesion of size 38 × 21 mm in subcutaneous tissue, lateral to labia majora on left side, suggestive of lipoma. FNAC was done and only blood was aspirated. The swelling was excised on March 7th, 2022 and sent for histopathological examination. Fig. 1 shows gross image of vulvar mass after excision and Fig. 2 shows cut section of vulvar mass. On microscopic examination, H&E-stained sections show well-circumscribed mass comprising of variable-sized blood vessels with medial hypertrophy. There was proliferation of perivascular spindle-shaped stromal cells embedded in myxoid stroma. The cells were small without nuclear atypia. There was absence of necrosis and mitotic figure. Findings were suggestive of angiomyxoma. Immunohistochemistry was positive for desmin and vimentin. No recurrence has been reported in this patient so far, on follow-up visits for 17 months.

DISCUSSION

AAM has high rates of recurrences, rapid growth rate, and potential for morbidity and these features make AAM clinically very important.

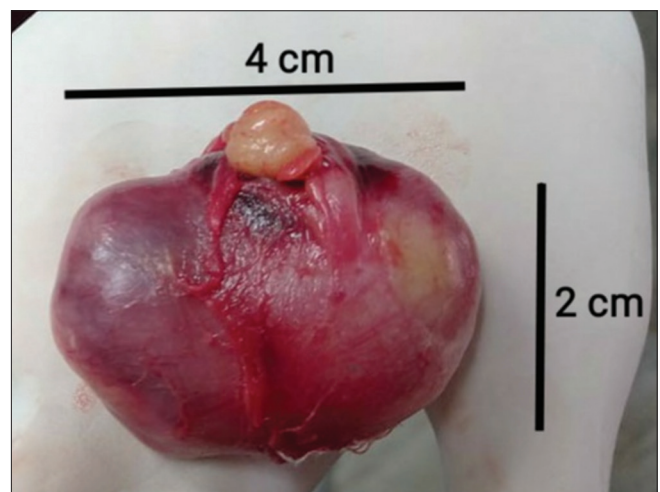


Fig. 1: Gross image of vulvar mass after excision



Fig. 2: Cut section of vulvar mass

Diagnosis of this tumor is made using imaging and biopsy. Treatment of choice is surgery of primary disease. Surgery is also the main treatment modality for recurrences and metastasis. Neoadjuvant and adjuvant treatment can be given using medical methods such as GnRH agonists and SERM and radiotherapy. As this tumor has high rates of local relapse, so long-term follow-up is indicated in treated cases [4].

CONCLUSION

Whenever a female of reproductive age, presents with a vulval or perineal mass, angiomyxoma should be considered as a differential diagnosis. Patients diagnosed with angiomyxomas should be followed up as these tumors tend to recur.

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AUTHORS CONTRIBUTION

Dr Ashishjot Kaur, Dr Prabh Simranpal, and Dr Arshdeep Kaur were involved in writing of case report. Dr Anju Gupta was involved in history taking and examination of the patient in the OPD and performed the excision of vulvar swelling in operation theater. Dr Anju Gupta and Dr Arshdeep Kaur were involved in follow-up of the patient postoperatively.

CONFLICT OF INTEREST

All the authors declare that they have no conflict of interest.

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INFORMED CONSENT

Written informed consent was obtained from the patient.

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