Case Report

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Aggressive angiomyxoma of vulva in a young female: a rare entity with an unusual presentation

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ABSTRACT

Aggressive angiomyxoma is a rare benign neoplasm usually involving the pelvic region. It is six times more common in females than males. It occurs almost exclusively in adult females. The term aggressive signifies the neoplastic character of the blood vessels, its locally invasive nature, and the high risk of local recurrence. Here we report a rare case of an eighteen year old female, who had a vulvar swelling for over two years, which turned out to be aggressive angiomyxoma after wide local excision on histopathological examination.

Keywords: Aggressive, Angiomyxoma, Benign, Invasive, Neoplasm, Vulvar

INTRODUCTION

The main purpose of this case report is to enhance our ability to suspect a case of aggressive angiomyxoma on clinical basis and thereby enhancing our knowledge on the nature of this tumor and its follow up after surgery.

Aggressive angiomyxoma is a rare mesenchymal tumor of the pelvis and perineum that occurs almost exclusively in adult women. It was first described by Steeper and Rosai.² Although categorized as benign, it has infiltrative potential into skeletal muscle and fat. This tumor most frequently occurs in sites within lower pelvis including vulva, vagina or inguinal regions. Rarely, this tumor appears in men, simulating inguinal hernia, testicular neoplasm, spermatic cord neoplasm, hydrocele, or spermatocele.³ Due to its rarity the misdiagnosis rate was reported to be 80%.4

Considering the rare occurrence of this tumor it is important to closely observe its presentation and prognosis for better understanding of its progression.

CASE REPORT

An eighteen-year-old female presented with a right sided vulvar mass of more than two years duration (Figure 1). Initially patient noticed a small pea sized swelling in vulva region without any significant symptoms. She did not take any treatment for the same as it was not interfering with her day to day activities. Six months back she experienced exponential growth in the swelling with mild pain. There was no history of urinary or menstrual disturbance. Patients denied any bleeding or discharge from the local site but in the last few weeks, she experienced difficulty in sitting due to the swelling. Local examination revealed a firm to hard swelling arising from right labia majora mimicking the penis and scrotum of males. The swelling was freely mobile and there was no tenderness or discharge associated with it. There was no regional lymphadenopathy. Wide local excision of the swelling was done. Histopathological examination revealed features suggestive of aggressive angiomyxoma (Figure 2). The patient is under follow up till date (one year after the surgery) and there is no further recurrence.



Figure 1: Preoperative right sided vulva swelling in an eighteen year old patient.

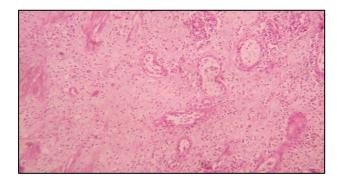


Figure 2: Cells with a spindled and stellate morphology are seen, embedded in a loose collagenous/ myxomatous background containing scattered vessels of different calibre.

DISCUSSION

Aggressive angiomyxoma is an uncommon benign tumor that occurs predominantly in females involving pelvic and perineal areas. On clinical examination it is usually mistaken for vulvar abcess, Bartholyn's cyst, Gartner's duct cyst, vaginal prolapse, pelvic floor hernia, vaginal mass or polyp and obturatory or levator hernia.⁴ Although benign, the term "aggressive" emphasizes the frequent local recurrence and its infiltrative nature.⁵ To our knowledge, there are only three cases of metastasised aggressive angiomyxoma described in literature. Usually the patients present with a slow growing mass which is otherwise asymptomatic in nature. Other associated symptoms may include dull aching pain, or urinary symptoms like dysuria, urinary retention dyspareunia. The size of this tumor may range from 10cms to as large as 40cms in the largest diameter. The pathogenesis of this tumor is unclear. Recently an association between chromosome 12 translocation (12q13-15) and consequent aberrant expression of the high-mobility group protein isoform 1-C (HMG1-C) has been suggested.⁷

Wide surgical excision is by far the most accepted treatment modality. The use of hormonal treatments with

raloxifene, tamoxifene and GnRH agonists for residual and recurrent disease has also been suggested. The local recurrence rate for aggressive angiomyxoma is about 30-40% and can occur after 10-15 years of primary excision (2 months to 15 years). In suspicious cases long term follow up with CT scan or MRI has been recommended.

CONCLUSION

This particular case demonstrates that aggressive angiomyxoma can occur in different age groups with unusual tumor shapes and sizes. Aggressive angiomyxoma should be considered as differential diagnosis when patient presents with painless vulva mass. Wide surgical excision with tumor free margins is the basis of curative treatment. Along with it adjunctive therapy may be necessary for residual or recurrent tumors. Long-term follow-up with MRI or CT scan is recommended in some cases.

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