

Case report:

Aggressive Angiomyxoma: A Rare Perineal Mass

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Abstract

Aggressive angiomyxoma (AAM) is a rare tumour that usually occurs in females at reproductive ages and affects pelvic region. We herein report a case of perineal AAM to contribute to the literature about pathological features and clinical outcomes of this tumour. A 36 year old female with no history of chronic diseases presented to our hospital with a nontender perineal mass. The mass was present for two years and it first appeared during pregnancy. She underwent surgery for local resection. The histology of the mass was consistent with AAM and multifocal extension into surgical margins was observed. With immunohistochemical staining the tumor was positive for desmin, CD31, CD34, ER and PR; poorly focal positive for SMA and negative for S-100. Ki67 was less than 1%. Due to surgical margin positivity she had a second operation. After the resection with clear margins, patient showed no signs of recurrence for 7 months. Resections with positive surgical margins were mostly concluded as recurrent with wide time range and recurrence rates, extended surgical resection is gold standard for management of this tumour.

Keywords: Perineal tumour, Vulvar mass, Soft Tissue Lesions, Mesenchymal Tumour

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Introduction

Aggressive angiomyxoma (AAM) is a locally infiltrative soft tissue lesion which is described as a tumour with undifferentiated by World Health Organization in Classification of Bone and Soft Tissue. Female/male incidence ratio is 6:1¹. Generally, this tumour is found in the pelvic and perineal region of female but there are cases with intrabdominal AAM located in liver and pelvic ureter^{2,3}. Patients with perineal AAM usually present with a nontender, edematous lesion which is found as a reducible mass in physical examination^{4,5,6}. As a rare entity AAM is often misdiagnosed with Bartholin cyst, vulvar lesions, condyloma acuminatum, lipoma or pelvic floor hernia. The lesions are usually much more bigger than they seem on inspection and palpation due to their tendency to grow towards the deeper soft tissues⁷. The main treatment approach is surgical resection with negative margins since there is only

a few data about recurrence rates^{6,7}

We herein report a case of perineal AAM to contribute to the literature about pathological features and clinical outcomes of this tumour.

Case Report

A 36-years old female with no history of chronic diseases presented to our hospital with a nontender perineal mass. The mass was present for two years and it first appeared during pregnancy. In lithotomy position on right gluteal region, 8 cm from the anus; soft, nontender mass with fluctuation was palpated. Laboratory tests were within normal limits. Ultrasound showed a mass measuring 73 x 68 x 48 mm and consistent with abscess containing air-dense fluid level. With Magnetic resonance imaging (MRI), the mass was isointense on T1-weighted and hyperintense on T2-weighted images, starting from adjacent of right vaginal wall to intergluteal cleft (Figure 1,2).

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No diffusion restriction on diffusion-weighted imaging was noted. She underwent surgery for local resection. Macroscopic examination showed a solid tumour that was homogeneous, dark grey colored and glistening on the surface (Figure 3). The histology of the mass was consistent with AAM and multifocal extension into surgical margins was observed. With immunohistochemical staining the tumor was positive for desmin, CD31, CD34, ER and PR; poorly focal positive for SMA and negative for S-100. Ki67 was less than %1. Due to surgical margin positivity she had a second operation. After the resection with clear margins, patient showed no signs of recurrence for 7 months.

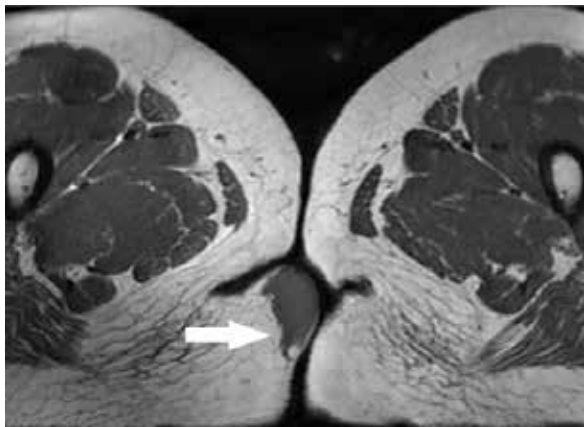


Figure 1. T1-weighted axial view of MRI scan shows soft tissue mass

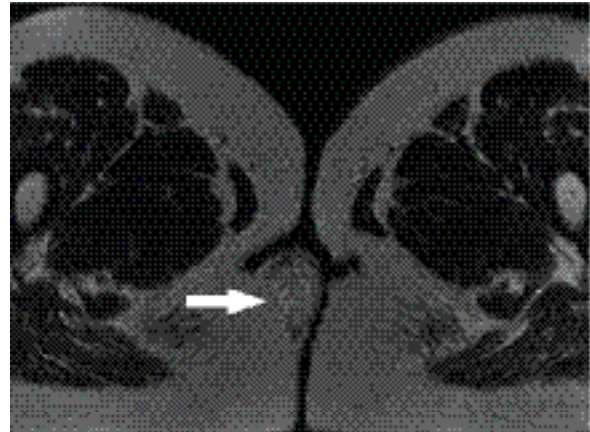


Figure 2. T2-weighted axial view of MRI scan shows soft tissue mass

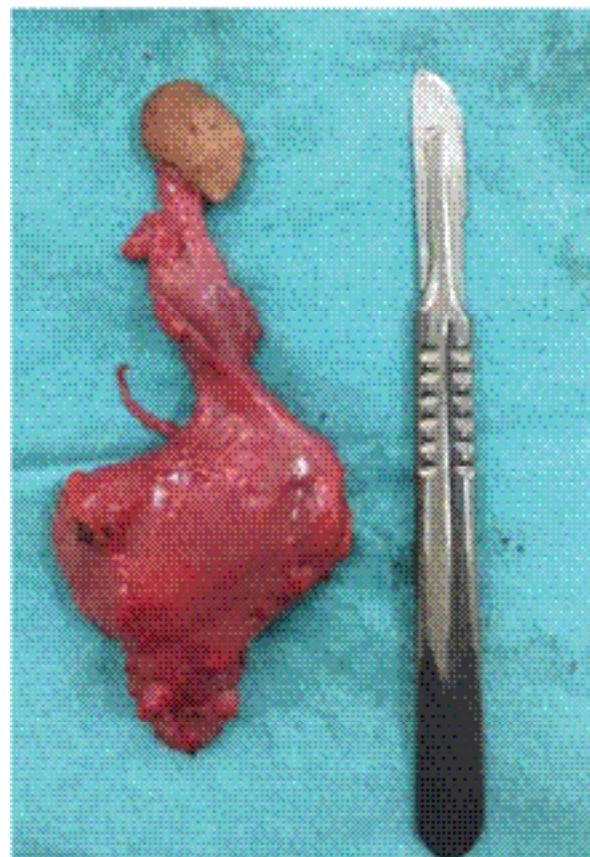


Figure 3 Gross pathological examination of specimen

Discussion

AAM is first described by Steeper and Rosai in 1983 as an infiltrative and recurrent neoplastic tumour of the blood vessels⁸. The etiology remains unclear, but significantly increased female dominance, incidence peak at reproductive ages and reported cases during pregnancy suggest an hormonal involvement to pathogenesis^{4,9}.

Most cases locates in pelvic and perineal region but in a review study, Sato et. al have reported a patient which was an AAM, arising from the liver and other cases from the literature located in larynx, oral floor, spraclavicular fossa and lungs were encountered². Clinical characteristics are similar to perineal lesions, therefore there is no specific findings on physical examination. Most cases are misdiagnosed with other vulvar pathology or levator hernia⁶.

Ultrasonographic examination AAM appear as homogeneous and hypoechoic lesions and on colored Doppler blood flow is usually observed^{6,10}. On MRI, due to increased water content and loose

matrix of these lesions, they appear hyperintense on T2-weighted imaging and masses demonstrate a 'swirled' pattern^{3,9,10}. Gilardi et al. reported moderate FDG uptake of an AAM with 3.75 SUV max¹¹.

The tumor cells are bounded by fibrofatty tissues without a well shaped border and surfaces are myxedematous or gelatinous, and gray reddish-brown. They appear as spindle-shaped cells with lightly stained or eosinophilic cytoplasm^{6,13}. The nuclei are oval-shaped, bland, and lightly stained with a single, small, centrally located

nucleolus. Mitotic figures are usually absent. By immunohistochemistry, tumour cells show strong expression of vimentin, desmin, ER, and PR. On the other hand, partial or weak expression was observed for SMA, actin, CD34, and S-100, whereas the Ki-67 index was 1% to 3%^{4,6}.

Most of the patients were treated surgically but there are studies that investigated pharmacological approaches. In a case series with 7 patients, Magtibay et al reported a patient that was administered tamoxifen and during treatment tumour progression was observed. Another patient in this series, a 59 year old woman, received preoperative radiation and angiographic embolization (after an unsuccessful resection), followed by intraoperative radiotherapy and definitive surgery. Tumour margins were clear, yet after 42 months patient had evidence of recurrence on CT¹². Im et. al. reported a case treated with both surgery and gonadotropin-releasing hormone agonist. The tumor showed reduction after the treatment but recurred after 10 months¹⁴.

Conclusion

Aggressive angiomyxoma is a rare entity, but with increased number of reported cases diagnostic

and pathological features of this tumor was enlightened. Resections with positive surgical margins were mostly concluded with recurrence in varied time intervals. In summary due to high recurrence rates, extended surgical resection is gold standard for management of this tumor.

Conflict of Interest

No conflict of interest has been disclosed by the authors.

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