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Aggressive Angiomyxoma of the Pelvis and Perineum: A Case Report

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Abstract **Case Report**

Aggressive angiomyxoma is a rare mesenchymal tumor that typically manifests as a slowly growing soft tissue mass in the perineal area, specifically in the paravulvar and pararectal regions, affecting young adult women. MRI is considered the best imaging tool for depiction of this lesion. The confirmation diagnosis is histological and the treatment is surgical. In this report, we present a case of aggressive angiomyxoma and provide a correlation between clinical and pathological findings. We offer a detailed analysis of its imaging features, with a particular focus on magnetic resonance imaging (MRI). Additionally, we include a comprehensive review of the current literature on this subject.

Keywords: Aggressive angiomyxoma, pelvis, perineum, Woman; MRI.

INTRODUCTION

Aggressive angiomyxoma is a rare mesenchymal tumor that exhibits slow growth and is predominantly found in the vulvoperineal and pelvic regions, typically affecting women of reproductive age [2]. The clinical diagnosis is often overlooked and MRI plays an important role in making an accurate diagnosis and determining the surgical approach of the tumor [5]. Surgical excision is the primary treatment for aggressive angiomyxoma (AA) with a high rate of local recurrence, typically occurring in the first years after the initial surgery [3].

CASE PRESENTATION

Mme EL F, 55 ans, nulliparous, postmenopausal, with the following medical history; excision of a vulvar mass with histological findings of atypical lipoma in 2003, a local recurrence, excision performed with histological findings of cellular angiofibroma in 2008, a second local recurrence, excision performed with histological findings of aggressive angiomyxoma. Actually, presents to our institution with a vulvar mass witch examination finding a mass measuring 15 cm in length occupying the pubic mound and labia majora, firm in consistency, adherent to the skin, and mobile deep within. An MRI of the pelvis reveals a large tumor arising from the vulvar region, locally infiltrating, showing hypo intensity on T1-weighted imaging, discrete heterogeneous hyperintensity on T2-weighted imaging, and enhancement after gadolinium injection. Histopathological examination of the surgical biopsy revealed a spindle cell tumor proliferation, with histological and immunohistochemical features suggestive of an angiomyxoma. No secondary localization was detected on the staging evaluation. The case was discussed at the multidisciplinary team meeting: recommendation for total vulvectomy.

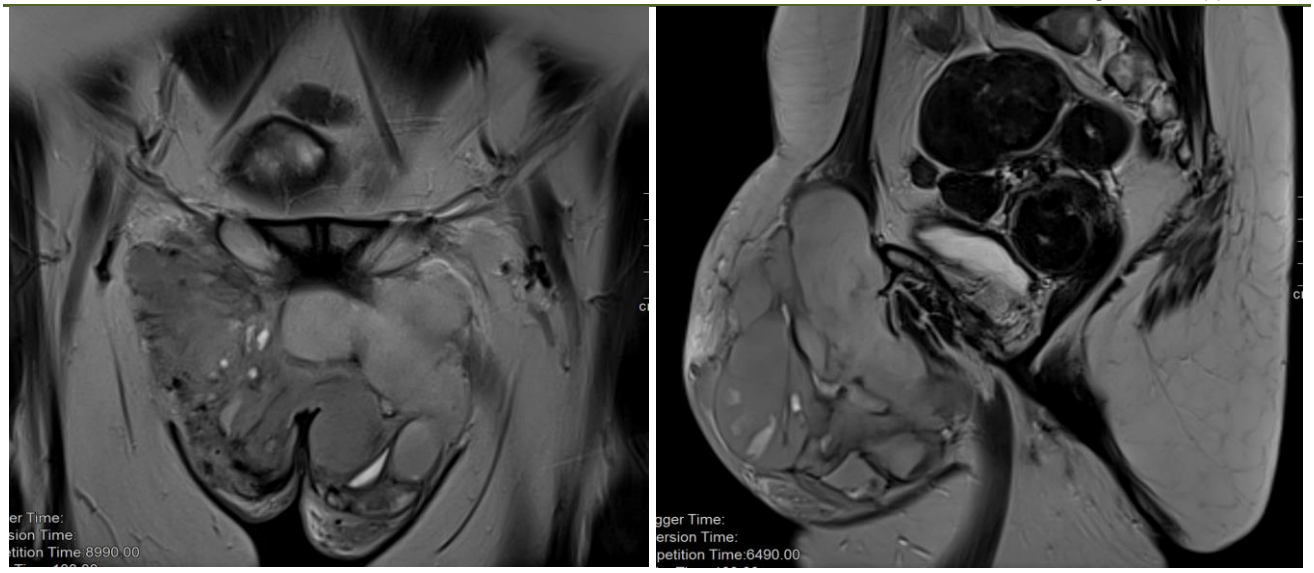

Figure 1: Coronal and sagittal section T2 showed a large heterogeneous mass at the perineal region with discrete high signal

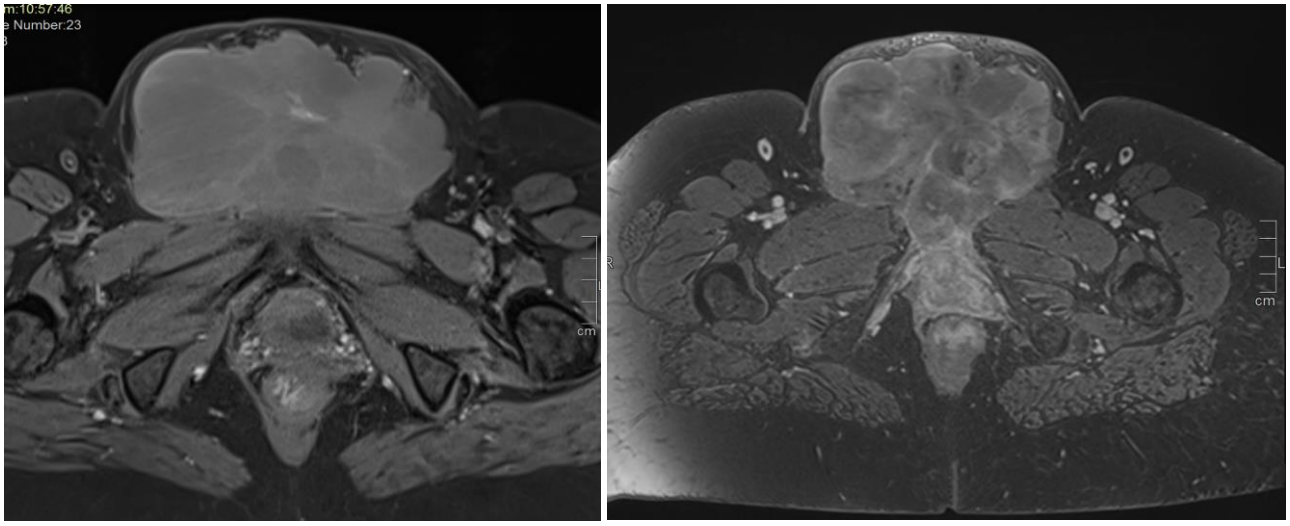

Figure 2: Axial section T with and without gadolinium injection showed an avidly heterogenous enhancing of the mass at the perineal region

DISCUSSION

Aggressive angiomyxoma is a rare mesenchymal tumor that arises from the connective tissues of the perineum or lesser pelvis, typically affects young women in their fourth decade in 90% of cases [1, 3]. It has a distinct imaging finding, and the radiologist plays an essential role in its diagnosis and his management.

Clinical presentation is non specific, characterized by the slow growth of a soft and mobile mass in the paravaginal or pararectal space (without anatomical constraints on its expansion), often asymptomatic [1]. The lesion is typically discovered once it has become sizable [2].

When the tumor is symptomatic, clinical manifestations include pelvic heaviness, vulvovaginal pain, dysmenorrhea, dyspareunia, and digestive and urinary disturbances. [2].

Macroscopically, the lesion appears as a soft, gelatinous, non-encapsulated mass with indistinct borders, having a multilocular contour and infiltrating adjacent soft tissues [1].

Microscopically, aggressive angiomyxoma is characterized by scattered stellate and spindle cells embedded in a myxoid matrix, accompanied by abundant fibroblasts, myofibroblasts, and vessels of variable sizes. It exhibits low mitotic activity and the absence of nuclear atypia [3]. Immunohistochemistry reveals positive staining for vimentin, desmin, actin, CD34, and factor VIIIa, and negative staining for S-100 protein. Additionally, estrogen and progesterone receptors are typically positive [3].

The definitive diagnosis is anatomopathological, performed on perineal biopsies guided by ultrasound. On ultrasound, aggressive angiomyxoma appears hypoechoic, even pseudo-cystic. Its tissue nature is confirmed by the presence of vascularity, highlighted in Doppler mode [2]. However, recent articles describe a hyperechoic pattern and also note a mixed appearance with alternating hypoechoic and hyperechoic layers [3].

On a CT scan, the tumor appears spontaneously hypodense compared to the muscle, sometimes even pseudo-cystic. There is not a specific feature on ultrasound and CT scan and a pelvic MRI is the optimal imaging technique, for characterization of this kind of lesions [2].

In MRI, the lesion appears well-defined with hyperintensity on T2-weighted images, hypo intensity on T1-weighted images, and shows heterogeneous contrast enhancement: it exhibits a swirled or striated appearance, characterized by lines of hypo intensity within the enhanced tumor [1, 2]. This appearance indicates the stretching of the fibrovascular stroma on either side of the pelvic diaphragm [2] and it's the main imaging feature that supports the diagnosis 3. The behavior of these lesions on dynamic contrast-enhanced (DCE) sequences has been recently reported in radiologic literature, demonstrating progressive and heterogeneous enhancement with a layered pattern that becomes more pronounced in late venous phases [3]. This type of tumor also exhibits mild restriction on Diffusion-Weighted Imaging (DWI) with characteristic high mean values in Apparent Diffusion Coefficient (ADC) maps. These values are in line with recent literature and are consistent with the low mitotic activity observed in these tumors [3].

In both CT and MRI, the tumor typically manifests as a well-defined mass, in paravaginal or pararectal region, pushing aside neighboring organs without infiltrating them (urethra, vagina, anal sphincter, and rectum). Those elements support this diagnosis and allow differentiation from retrorectal tumors [1, 2].

These two techniques enable the evaluation of the tumor's extent by determining its expansion on either side of the pelvic diaphragm, influencing the surgical planning.

However, MRI remains the optimal tool due to its superior contrast resolution in the pelvic region. After surgery, it facilitates the detection of any remaining macroscopic residue and provides ongoing surveillance for recurrences with similar characteristics to the initial tumor.

In pathology, the differential diagnoses include benign lesions such as superficial angiomyxoma, cellular angiofibroma, myxoid leiomyoma, and deep abdominal fibromatosis (desmoid tumor), as well as malignant conditions, notably myxoid liposarcoma [2].

The treatment is surgical, involving complete excision in a single block. Incomplete excision often leads to frequent recurrences [2]. To reduce the likelihood of local recurrence, adjuvant therapies such as chemotherapy and external radiotherapy have been attempted, but results have been disappointing due to the low mitotic activity of these tumors [3].

During the last years, favorable outcomes have been achieved with preoperative angiographic embolization of the tumor. Additionally, gonadotropin-releasing hormone agonist therapy has shown significant tumor volume reduction, potentially avoiding the need for mutilating surgery [3].

Despite the morbidity related to recurrences and occasionally repetitive surgical interventions, the overall prognosis for aggressive angiomyxoma remains favorable [2]. Clinical follow-up and periodic MRI are recommended [2].

CONCLUSION

Aggressive angiomyxoma is a rare tumor characterized by a typical presentation as a locally infiltrative and slowly growing perineal soft tissue mass, primarily affecting young or middle-aged women with very low malignant potential. Distinctive imaging features play a crucial role in achieving a correct preoperative diagnosis and determining the optimal surgical approach. While ultrasound and CT appearances are not specific, MRI is considered the most effective imaging tool for depicting this lesion. This tumor should be managed in a specialized setting for soft tissue tumors to prevent local recurrence.

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