

Salientamos ainda o facto do *Clostridium* ser um microorganismo ubiqüitário, com diversas espécies potencialmente causadoras de doença grave, particularmente pela produção de toxina como é o caso de *Clostridium botulinum* e *Clostridium tetani* - este último, um agente com elevada capacidade de sobrevivência e causador de doença rara mas quase sempre invariavelmente fatal.

#### AGRADECIMENTOS

Os autores agradecem a Luís Paulo Trindade, da Unidade de Cuidados Intensivos Polivalente do Hospital Sousa Martins, na Guarda.

#### PROTEÇÃO DE PESSOAS E ANIMAIS

Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica

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e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial.

#### CONFIDENCIALIDADE DOS DADOS

Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação de dados.

#### CONSENTIMENTO DO DOENTE

Obtido.

#### CONFLITOS DE INTERESSE

Os autores declaram não terem qualquer conflito de interesse relativamente ao presente artigo.

#### FONTES DE FINANCIAMENTO

Não existiram subsídios ou bolsas que tenham contribuído para a realização do trabalho.

## Aggressive Angiomyxoma: A Rare Tumor of Male Pelvic Cavity

## Angiomixoma Agressivo: Um Tumor Raro na Pelve Masculina



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Acta Med Port 2018 Nov;31(11):693-696 • <https://doi.org/10.20344/amp.9062>

#### ABSTRACT

Aggressive angiomyxoma is an uncommon, benign, slow-growing, and locally infiltrative soft tissue neoplasm which is located primarily in the genital region and perineal interstitial tissue of female patient in the fourth decade of life. Its occurrence in male patients is even more unusual and commonly appears at a later age. The mainstay of treatment typically involves surgical excision with tumor-free margins, and despite complete resection, local recurrences are common. Here, an unusual case of aggressive angiomyxoma occurring in the pelvic region of a 55-year-old man and its treatment is discussed due to its rarity.

**Keywords:** Myxoma; Neoplasm Invasiveness; Pelvic Neoplasms

#### RESUMO

O angiomixoma agressivo é uma neoplasia benigna rara, de crescimento lento, localmente infiltrativa que se localiza primariamente no tecido intersticial da região genital, pélvica e perineal de doentes do sexo feminino na quarta década de vida. O aparecimento em doentes do sexo masculino é ainda mais incomum e habitualmente aparece em idade mais avançada. O tratamento tipicamente envolve a excisão cirúrgica com margens livres e, apesar da ressecção completa, a recidiva local é comum. Apresentamos o caso clínico raro de angiomixoma agressivo na região pélvica de um homem de 55 anos de idade bem como o seu tratamento.

**Palavras-chave:** Invasividade Neoplásica; Mixoma; Neoplasias Pélvicas

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Recebido: 09 de abril de 2017 - Aceite: 07 de maio de 2018 | Copyright © Ordem dos Médicos 2018



## INTRODUCTION

Aggressive angiomyxomas (AAMs) are benign mesenchymal neoplasms of the soft tissue comprised of spindle-shaped or stellate neoplastic cells and thickened blood vessels within a hypocellular myxoid stroma.<sup>1</sup> The term 'aggressive' has been used to describe the infiltrative pattern of the blood vessels and high probability of local recurrence (in up to 72% of cases), not to indicate its malignant potential.<sup>2,3</sup> It always involves the pelvic, genital, and perineal region. AAMs occur in ages ranging from 15 to 63 years, with a female predominance (male/female ratio of 1:6.6).<sup>4</sup>

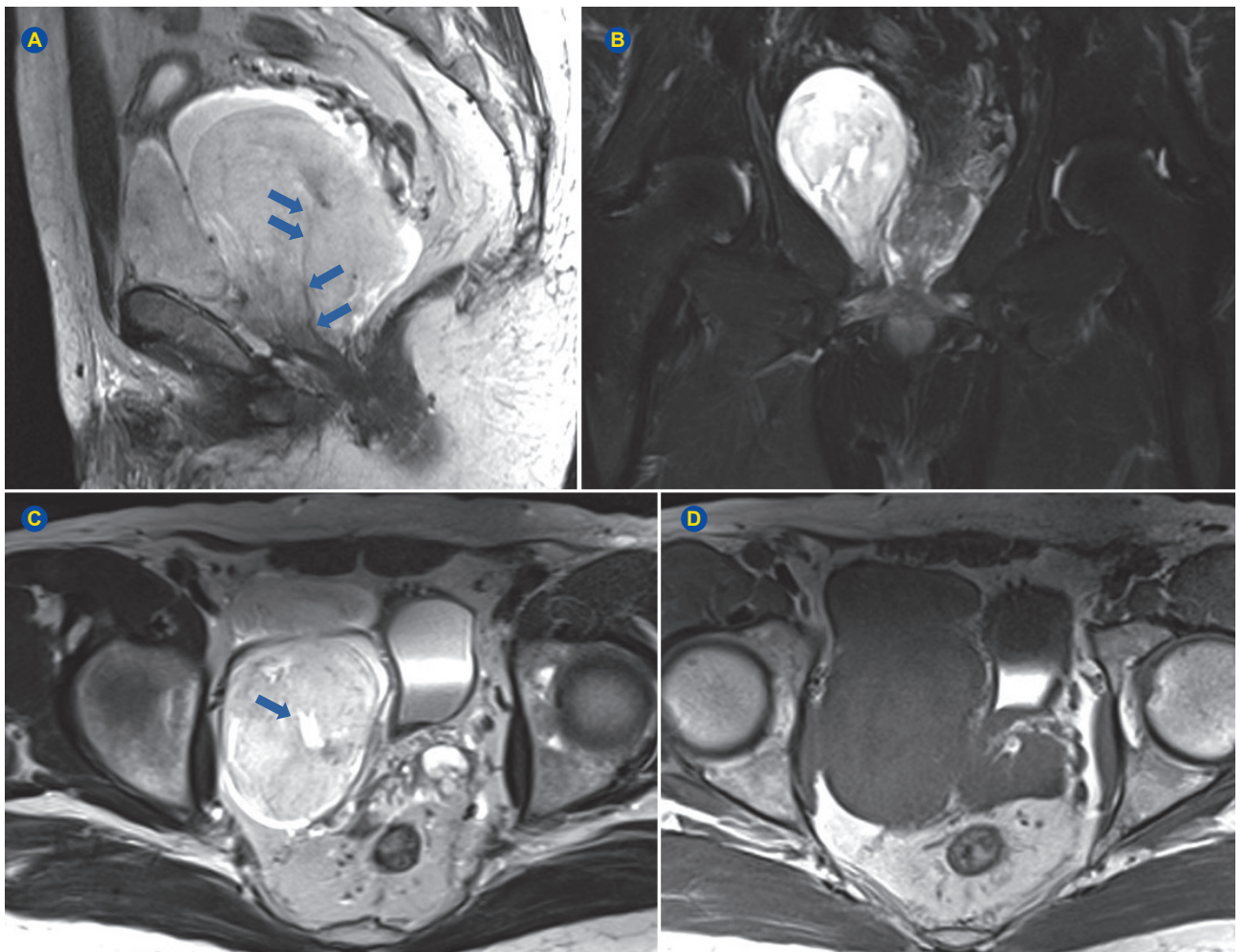
Since its first description by Steeper and Rosai in 1983, less than 50 cases in male patients have been reported in the literature.<sup>5,6</sup> AAMs usually arise mainly in the perineal region, scrotum or inguinal area in males. The most common manifestation is a slowly growing soft tissue mass with diameters ranging from 3 to 60 cm. In men, the mean age is around 45 years and ranges from 1 to 82 years.<sup>5,7</sup> Herein, we discuss a case of AAM arising from the pelvic region in a male patient.

## CASE REPORT

A 55-year-old man with a one-year history of constipation,

abdominal swelling, and symptoms of prostatism such as nocturia, urgency, hesitation, and post-micturition dribbling presented to our clinic. Physical examination revealed a palpable suprapubic mass; on rectal digital examination, there was an immobile, rigid mass in the anterior part of the rectum that was not related to the rectum. Moreover, there was no regional lymphadenopathy. His past medical history was only notable for hypertension and diabetes mellitus and surgical history was significant only for hemorrhoidectomy. Laboratory findings including tumor markers were unremarkable. The colonoscopy was normal except for internal hemorrhoids. Ultrasonography demonstrated a homogeneous and mixed echogenic mass in the pelvic area. In addition, magnetic resonance imaging (MRI) showed a well-defined and lobulated pelvic mass measuring 10 x 9 x 5 cm between the obturator internus muscle, the prostate, and the bladder and extending to the anterior wall of the abdomen. The mass had similar signal intensity to muscle on T1 and shows heterogeneous high signal intensity on T2-weighted sequences (Fig. 1).

In light of the preoperative clinical and imaging findings, surgical excision of the mass was planned after informed



**Figure 1** – (A) sagittal, (B) coronal, (C) axial T2-weighted MR scans show hyperintense and heterogeneous mass involving pelvic cavity with swirled appearance (arrows). (D) axial T1-weighted MR image identifies a homogeneous and isointense lesion.

consent of the patient. On exploration, a large encapsulated mass measuring approximately 12 cm in diameter was found and was completely excised; there was no invasion into the adjacent structure. There were no intraoperative complications, and blood loss was estimated at 150 mL.

Macroscopically, the resected specimen (12.5 x 8.5 x 5 cm) had a smooth grayish external surface and was partially encapsulated with no invasion. The surface of the tumor showed a glistening, grossly gelatinous, and homogeneous appearance with no necrosis or hemorrhage. Microscopically, the tumors showed stellate and uniform spindle-shaped cells in a loose hypocellular myxoedematous matrix with some collagen formation and thickened vessels of varying size. Vascular features were characterized by perivascular hyalinization. Also, there was nearly no mitotic activity or cytologic atypia (Figs. 2A and 2B).

Immunohistochemical studies revealed positive reactivity for desmin, but negative reactivity for CD31, CD34, CD117, S-100, SMA, myogenin, CK, and EMA (Fig. 2C). The tumor demonstrated a very low Ki-67 proliferation (1%). The clinical and pathological findings were compatible with

aggressive angiomyxoma. The patient has been in regular follow-up for sixteen months and is currently disease-free.

## DISCUSSION

Aggressive angiomyxoma is a relatively rare infiltrative mesenchymal neoplasm of the soft connective tissue that contains myxoid and vascular components.<sup>1,2</sup> In contrast to its locally aggressive and infiltrative nature, AAM is a slowly growing benign tumor. Approximately 70% of the cases locally recurred within the first three years.<sup>3,5,8,9</sup> Although most of the cases predominantly occur in female patients, occurrence of AAM in male patients is extremely rare and less than 50 cases have been reported up to now.<sup>5</sup> In male patients, AAMs usually arise from the scrotum (38%), spermatic cord (33%), perineum (13%), and pelvic cavity (8%) with proximity to anorectal structures. The average age of male patients is 46 years and ranges from 1 to 82 years.<sup>5,7</sup> There are no characteristic symptoms associated with AAM. Patients usually present with a painless, slowly growing mass and a feeling of local pressure.<sup>6,10</sup>

Accurate diagnosis of AAMs often remains difficult because of its unique clinicopathological characteristics and rarity. However, it has a typical appearance on radiological examinations. Ultrasonography usually shows a homogeneous and hypoechoic mass, with tubular or fusiform morphology, lipoma-like consistency, and internal vascularization. Characteristic computed tomography (CT) features are a mass hypodense or isodense to muscle with well-defined margins that shows variable enhancement after contrast infusion. MRI is the best imaging technique for characterization of this kind of tumors and identifies AAM as an isointense or hypointense lesion on T1-weighted and high signal intensity on T2-weighted images.<sup>11</sup> Both CT and MRI show a typical swirling or layering appearance as definitive diagnostic feature which is found in the majority of patients (83%).<sup>2,7,11</sup> Additionally, angiography typically demonstrates a hypervascular mass and increased vascularity in the area of the tumor.<sup>7</sup>

On gross examination, AAM is a large, lobulated, gelatinous, and partially or completely encapsulated mass. The tumor size can range from 3 to 60 cm, and fingerlike projections are commonly reported.<sup>5,11</sup> Histologically, AAMs are infiltrative, poorly circumscribed, and consist of stellate or scattered spindle cells in a myxoid matrix with large hyalinized vessels of varying size. There is no evidence of necrosis, significant mitotic activity or nuclear atypia.<sup>5-7</sup> In immunohistochemical staining, the tumor cells are typically positive for desmin, vimentin, smooth muscle actin, muscle-specific actin, and CD34; negative for Ki-67 (MIB) and S-100.<sup>4,7</sup> Furthermore, most of AAMs have positive estrogen and progesterone receptor status, which may expand the available therapeutic options.<sup>3</sup> More recently, a translocation on chromosome 12 (12q13-15) with aberrant expression of DNA architectural factor and proto-oncogene *high mobility group A protein 2 (HMGA2)* involved in DNA transcription has been revealed in AAM and may be potentially helpful in the diagnosis of difficult cases.<sup>7</sup> The differential diagnosis

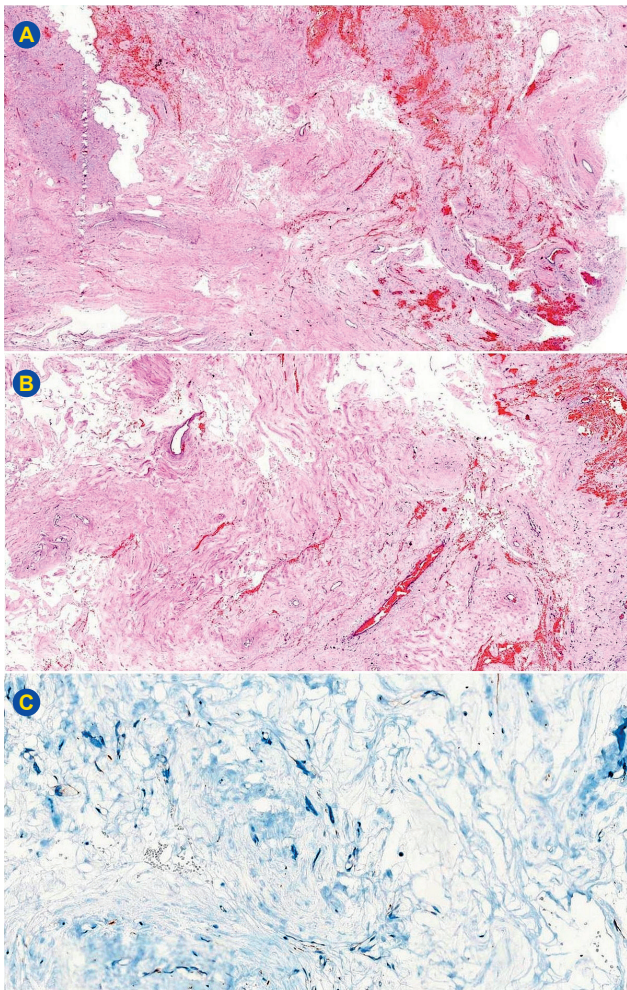


Figure 2 – (A, B) hypocellular hyalinized lesion consists of scattered spindle cells in myxoid matrix (H&E, x 14 and x 50); (C) diffuse cytoplasmic desmin expression in tumor cells (Desmin, x 240).

should be considered with various benign and malignant soft tissue tumors with myxoid pattern such as superficial angiomyxoma, myxoid liposarcoma, cellular angiofibroma, angiofibrosarcoma, and myxoid malignant fibrous histiocytoma.<sup>3</sup>

Complete surgical resection (R0 resection) is the primary treatment of choice for AAM, but local recurrence rates are high at approximately 70%.<sup>2</sup> An additional resection for recurrence is possible in many cases with technical difficulty. Moreover, the use of hormonal therapy may be an effective therapeutic option in patients with positive estrogen and progesterone status for recurrent cases or incompletely resected tumors.<sup>3,4</sup> Less radical surgery with an adjuvant therapy including radiotherapy, chemotherapy, and preoperative angiographic embolization of the tumor has also been discussed in the literature.<sup>5,11,12</sup> Close clinical follow-up with imaging studies especially MRI scans is necessary, because recurrences may occur several years after the initial surgical excision.

In conclusion, aggressive angiomyxoma is a locally infiltrative tumor and has a strong tendency for local recurrence, although it is classified as a benign tumor. Complete surgical excision with tumor-free margins is important and close postoperative follow-up is needed because of the high risk of local recurrence. Alternatively, hormonal therapy may be effective for recurrent cases.

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Lastly, AAM should be distinguished from other, more common neoplasms of the pelvis and perineum, regardless of gender.

## ACKNOWLEDGEMENTS

The authors would like to sincerely thank Akin Firat Kocaay and Gulsah Kaygusuz for their contribution. We would also like to thank Stéfano Fiúza and André Goulart for assistance in translating the abstract into Portuguese.

## OBSERVATIONS

The manuscript's abstract has been presented in the 20<sup>th</sup> National Surgical Congress'16 as a poster, 13-17 April 2016 - Antalya, Turkey.

## PATIENT CONSENT

Written informed consent was obtained from the patient for publication of this case report.

## CONFLICTS OF INTEREST

The authors declare that there are no potential conflicts of interest.

## FUNDING SOURCES

The authors received no financial support for the research, authorship, and publication of this article.