Case Report

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AGGRESSIVE VAGINAL ANGIOMYXOMA MIMICKING URETHRAL TUMOR

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ABSTRACT

This is a case report of a 32-year-old female patient with a neoplasia mimicking a urethral tumor. Following anterior pelvic exanteration, vulvectomy, bilateral inguinal lymphadenectomy, the pathological study established the diagnosis of aggressive vaginal angiomyxoma, CD-34 labeled.

Key words: urethra; vagina; vulva; angiomyxoma
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INTRODUCTION

Aggressive angiomyxoma is a rare neoplasm of the soft tissues affecting mostly female patients (6:1) in the third decade of life. It affects the genital and pelvic area and has a propensity for local recurrences. This tumor was first described and named aggressive angiomyxoma by Steeper & Rosai in 1983 (1). The characteristics of the tumor are thick-walled, medium sized vessels scattered amongst neoplastic stromal cells in a myxoid-apperaring background. It is sometimes classified as low-grade sarcoma, and metastasis, generally, does not occur (2).

CASE REPORT

A 32-year-old female patient underwent surgery for a urethral tumor in March 2001. On that occasion, the pathological study established the diagnosis of urethral caruncle. Between 2001 and 2002, she had a pregnancy which went to term. On September 2002, she observed a local recurrence. The biopsy was rhabdomyosarcoma. After (30 days), the urethral disease enlarged considerably and large tumors appeared in the inguinal regions. The CT confirmed the extension of the lesions showing a large tumor comprising the urethra, vagina and inguinal regions bilaterally.

Neoadjuvant chemotherapy (QT) was performed between November 2002 and March 2003 with the following schedule: ifosfamide, actinomycin D, vincristine, cyclophosphamide, doxorubicin and VP16. In spite of CT, the tumor increased and tumor exteriorization to the vulva and great enhancement of the inguinal mass occurred (Figure-1).

In March 2003, the patient underwent anterior pelvic exanteration, bilateral inguinal lymphadenectomy, vulvectomy (Figure-2) with autologous myocutaneuos flaps reconstruction and urinary diversion (sigmoid) and colostomy (Hartman's procedure). The surgical procedure evolved without intercurrences. The patient is well and disease-free after a 2 year follow-up.

The pathological examination of the inguinal lesions showed a poorly differentiated neoplasm with necrosis and profuse hemorrhage, suggesting high-grade sarcoma. The vaginal lesions had the characteristics of a fibromyxoid and vascular tumor without atypias or special features, suggesting angiofibroma myxoid. A immunohistochemistry examina-

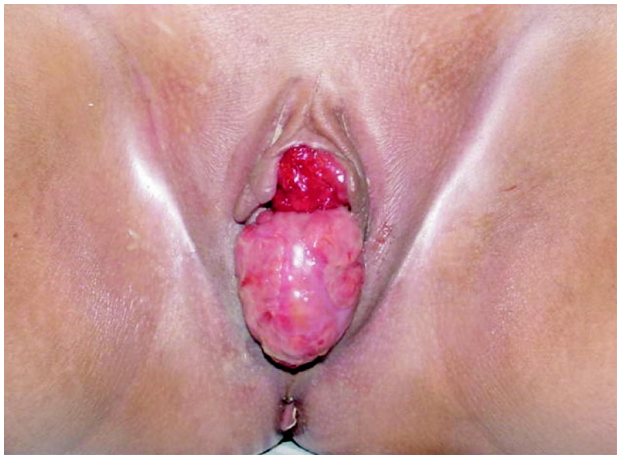

Figure 1 – *Tumor exteriorization through the vulva.*

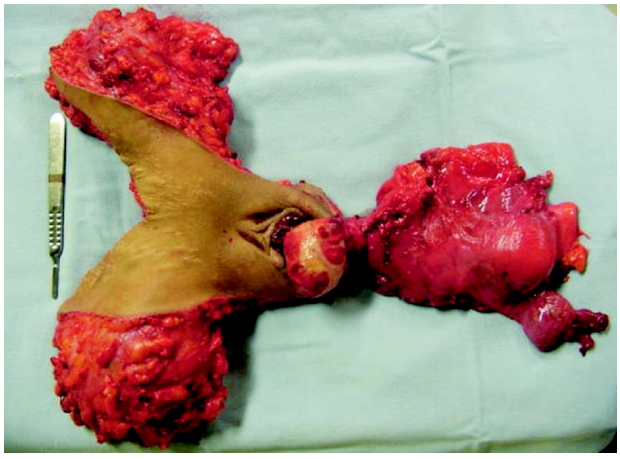

Figure 2 – *Surgical specimen with in bloc bilateral inguinal lymph nodes, vulvectomy and anterior pelvic exenteration.*

tion showed tumor positivity for CD-34 appointed strongly for angiomyxoma.

COMMENTS

Aggressive angiomyxoma is a rare mesenchymal tumor arising from soft tissue of the pelvis and perineum. It almost exclusively involves the genital, perineal and pelvic regions of women, with great incidence occurring in the fourth decade (3).

About 150 cases have been published in the literature since 1983 when it was first described by Steeper & Rosai (1). The tumor has a high recurrence rate with 50-70% of patients exhibiting relapse after surgical resection, often appearing many years after the first excision (2).

The diagnosis of aggressive angiomyxoma is usually made by the pathologist. Its differential diagnosis includes myxoma, myxoid liposarcoma, sarcoma botryoides, myxoid variant of malignant fibrous histiocytoma, nerve sheath myxoma and other soft tissue tumors with secondary myxoid changes (1).

Given the topographical variability of this genital tumor, no standardized surgical procedure has been described in the literature, but its complete excision is crucial for disease eradication (3).

Among the 12 cases of aggressive angiomyxoma pathologic diagnosis of the Amezcua et al. series, with a follow-up ranging from 2 to 60 months (mean of 19 months), 11 patients were still alive and 10 patients were disease-free without any incidence of recurrence (2).

This present case (disease-free after a 2 year follow-up) demands attention because of the presence of complex lesions arising from the urethral meatus mimicking caruncles.

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