GIANT AGGRESSIVE ANGIOMYXOMA OF THE VULVA: CASE REPORT

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SUMMARY – A 57-year-old multiparous housewife was hospitalized at University Clinical Center with painless, ulcerated, huge tumor of the vulva, which had progressively increased in size during the last five years. It was a firm, ulcerated mass involving the left vulva and measuring 35 cm in diameter. The vaginal orifice was deviated to the right by the tumor. The adnexa and the uterus were normal. The patient underwent total excision of the tumor in general anesthesia, and histology confirmed aggressive angiomyxoma. She had an uneventful postoperative period with satisfactory healing of the wound.

Key words: *Myxoma – surgery; Myxoma – pathology; Vulvar neoplasms – pathology; Vulvar neoplasms – surgery; Case reports*

Introduction

Vulva is the site where a wide variety of tumors may occur, many of them benign. Aggressive angiomyxoma is an uncommon locally infiltrating mesenchymal tumor that commonly occurs in the vulvovaginal region, perineum and pelvis of women[1,2], arising from connective tissue of the lower pelvis or perineum and having a locally aggressive course[3]. Aggressive angiomyxoma is described histologically as a mesenchymal tumor composed of fibroblasts within a strong myxoid background. Vascular proliferation is also prominent and virtually no mitoses are present. Patients often present with nonspecific symptoms that are frequently misdiagnosed as some more common entities such as a Bartholin cyst, vulvar fibroma, lipoma and hernia.

Tumor cells are characteristically positive for estrogen and progesterone receptors, suggesting a hormonal role in the development of the tumor. Chromosomal translocation of the 12q13-15 band involving the HMGA2 gene has been described[4]. Standard-of-care treatment for angiomyxoma is surgery.

In this case, a huge aggressive angiomyxoma of the vulva on the left labium majus had developed for more than five years.

Case Report

A 57-year-old married woman, housewife, was admitted to the hospital with a giant vulvar tumor that had been progressively developing for more than five years. The size of the tumor was 35 cm in diameter. The patient's medical history was not significant. She had five children, did not smoke or drink alcohol. Initially, she had no symptoms, but later the tumor caused difficulty on walking due to tumor weight and size. The patient suffered from depression and emotional stress too. There was a broad area of ulceration, necrosis and bleeding.

The pedicle of the tumor was located in the upper left labium majus, extending downward to the knees. Internal genital organs showed no abnormalities. Dif-

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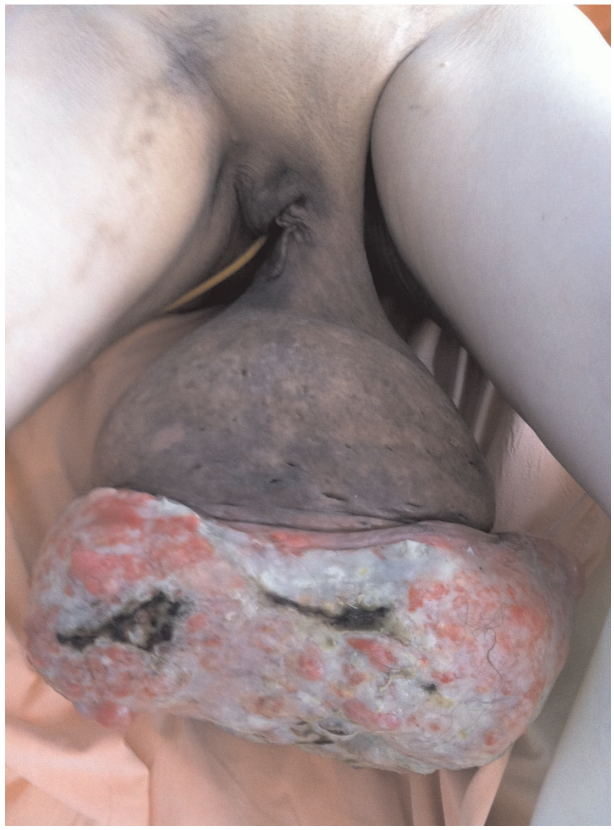

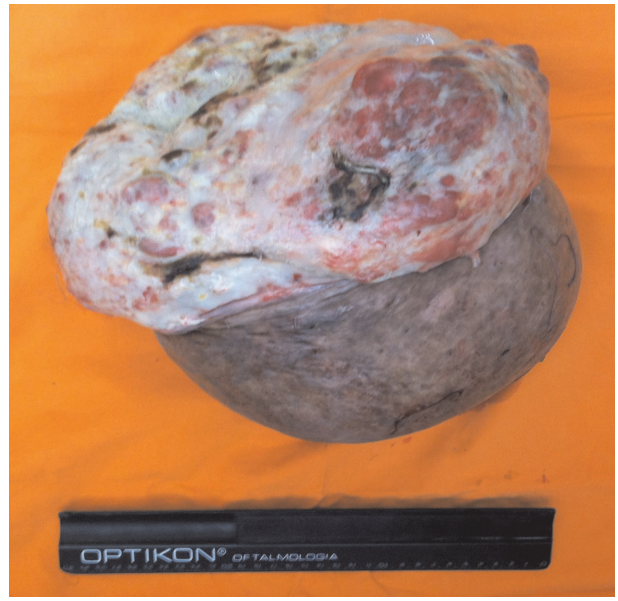

Fig. 2. Aggressive angiomyxoma (after removal).

Fig. 1. Giant aggressive angiomyxoma of the vulva.

ferential diagnosis included lipoma, fibroma, inguinal hernia, and other benign tumors of the vulva.

The patient underwent operative removal of the tumor (Fig. 1). Histologic examination showed it to be aggressive angiomyxoma, 35x24x16 cm in size, weighing 8.1 kilograms (Fig. 2). Complete excision of the tumor resulted in cure, relieved the patient's great discomfort, and improved her social and emotional status.

Histologic examination of the specimen revealed a hypocellular mesenchymal tissue with spindled and stellate cells with ill-defined cytoplasm, loosely scattered in myxoid stroma, without evidence of nuclear atypia and mitosis. Histopathology of the specimen showed capillary and cavernous vascular spaces stuffed with blood and separated by edematous fibrous and myxomatous tissue. Immunohistochemistry was positive for desmin, estrogen and progesterone receptors, and negative for S-100 protein. On histopathology, a huge area of necrotizing tissue, fibrin and degenerated polymorphonuclear leukocytes was found at the edge of resection.

Discussion

Aggressive angiomyxoma is an uncommon mesenchymal neoplasm occurring predominantly in the pelvic-perineal region, first described in 1983 by Steeper and Rosai[4]. It presents as a painless, poorly circumscribed vulvar mass and clinically simulates fibroma, lipoma, Bartholin gland cyst and hernia. The majority of these tumors occur in premenopausal women, predominantly in the 30-50 age group.

Aggressive angiomyxoma is considered as an aggressive neoplasm because of its propensity for local recurrence. The recurrence rate has been reported to be as high as 30%-70%, and most of these arise within two years after surgery, but it may occur as late as 20 years later[5-7]. In spite of the benign nature of this neoplasm suggested by histology, two cases of distant metastases were documented in previous reports, i.e. massive bilateral pulmonary, mediastinal, iliac and aortic lymph node and peritoneal mestastases[8], and multiple local recurrences and metastases in the lungs[1] in one case each.

Surgical excision is the treatment of choice, although treatment with gonadotropin-releasing hormone agonists is an emerging therapy. Metastases are exceedingly rare, and overall, the prognosis is good.

In conclusion, aggressive angiomyxoma is an uncommon mesenchymal myxoid tumor characterized by frequent local recurrence, currently considered as a non metastasizing tumor. Some unusual cases documented in some reports may change the current concept of aggressive angiomyxoma as a non metastasizing tumor. This suggests that aggressive angiomyxoma can no longer be considered a purely localized disease, since it can be, in a small percentage of cases though, regarded as a tumor of intermediate malignancy having unpredictable and sometimes even unfavorable outcome.

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Sažetak

VELIKI AGRESIVNI ANGIOMIKSOM VULVE: PRIKAZ SLUČAJA

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Višerotkinja u dobi od 57 godina, domaćica, hospitalizirana je na Univerzitetskom kliničkom centru zbog bezbolnog, ulceriranog, velikog tumora vulve koji je progresivno narastao u posljednjih pet godina. Tumor je bio čvrsta, ulcerirana tvorba na lijevom dijelu vulve, promjera 35 cm. Otvor vagine je povučen prema desno pod djelovanjem tumora. Adneksa i maternica bili su normalni. Izvedena je potpuna ekscizija tumora u totalnoj anesteziji, a histološka dijagnoza je potvrdila agresivni angiomiksom. Rani poslijeoperacijski tijek protekao je bez komplikacija sa zadovoljavajućim zaliječenjem rane.

Ključne riječi: *Miksom – patologija; Miksom – kirurgija; Vulva, tumori – patologija; Vulva, tumori – kirurgija; Prikazi slučaja*