Journal of Obstetrics, Gynecology and Cancer Research | ISSN: 2476-5848

Post-menopausal Vulvar Aggressive Angiomyxoma. Can Diabetes Mellitus and Overweight be a Risk Factor? A Rare Case Report

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Article Info

10.30699/jogcr.9.1.102

Received: 2023/01/06;
Accepted: 2023/03/26;
Published Online: 22 Jan 2024;

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ABSTRACT

Invasive angiomyxoma as a mesenchymal tumor with a high recurrence rate has been reported mainly in reproductive age according to its association with the estrogenic level of plasma. Above that, it seems there is a need for further treatment despite complete resection of the tumor, to eliminate the hormonal state. In the present study, we sought to introduce a rare case of invasive angiomyxoma in a post-menopausal but high-risk woman, discuss the relativity of risk factors in all hormonal-dependent gynecological malignancy, and intend to seek help from colleagues' opinions and experiences about treatment. It is clearly of great importance to emphasize the role of individualized medicine in such a rare case, in conclusion, there is not any debate on the role of surgical resection but the necessity of changing in lifestyle or adjuvant systemic or local therapy, and the needed duration is doubtful.

Keywords: Angiomyxoma, Vulva Neoplasms, Malignant Mesenchymal Tumor, Diabetes Mellitus, Body Mass Index, Menopause, Gonadotropin-releasing Hormone, Tamoxifen

Introduction

Gynecological malignancies are mostly well known as a hormonal-dependent neoplasm, including endometrial hyperplasia and cancers, epithelial ovarian cancers, breast cancer, and even some type of mesenchymal tumors. One of the rarest estrogen-receptor-positive cancers is angiomyxoma of the vulva, a mesenchymal tissue tumor that usually presents with a painless mass in a young reproductive age woman. Fortunately, this tumor behaves benignly with a low growth rate, but the nature of local invasion concerns the recurrence potency. Focusing on the rarity of this tumor and limited case reports in this regard, there is a controversial opinion about the best way of management and surveillance in this case **(1-3)**.

Here we intended to present a rare case of vulvar angiomyxoma in a postmenopausal aged female, in order to discuss the outcome.

Case presentation

A 52-year-old gravida2 para2 woman had been referred to the gynecological-oncology department with a complaint of growing a painless mass over the past year on the left side of labia major. We obtained the patient's informed consent and license of the ethical committee in Isfahan University of Medical Sciences, with the ethics number of IR.MUI.MED.REC.1399.577. Her medical history consisted of menopausal status from 2 years ago, high blood pressure under control by Enalapril with a dose of 30 mg at 12-hour intervals, and an insulin-dependent diabetes mellitus. There was no worrisome finding in the general examination, except for her obvious overweight condition; the body mass index (BMI) was 37 kg/m2. Assessment of external genitalia confirmed her

complaint; there was a large firm mass of about 5×6 cm with no sign of erythema, or tenderness, exactly in her left labia majora.

The patient was planned for surgical intervention. The initial pre-operative workup was not significant and the ultrasound examination of the vulvar was in favor of soft-tissue overgrowth in differentiating lipoma (Figure 1), but incising the mass in the operating room revealed a soft gelatinous yellow tissue that was excised completely (Figure 2).

Lastly, the histology demonstrated the nature of the tumor, the presence of spindle cells in a myxoid type stroma with separate vessels, spare mitosis, and atypia confirmed the diagnosis of invasive angiomyxoma (Figure 3). According to multidisciplinary consultation, due to patient comorbidities and no tumoral remnant in surgical margins, close surveillance but no additional therapy was recommended. Now after two-year of follow-up, the patient is in a healthy situation and there is no sign of recurrence in her physical and imaging studies (Figure 4).

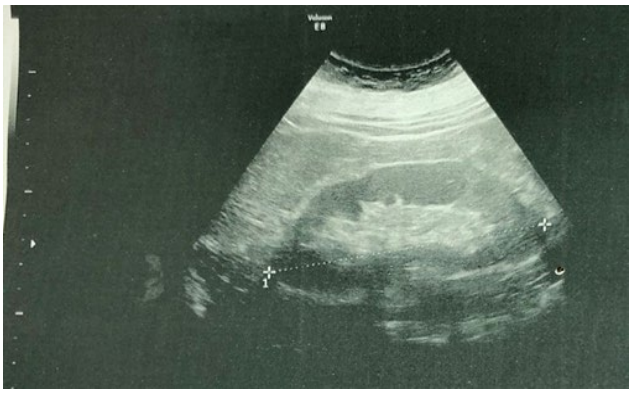

Figure 1. Ultrasound imaging of the vulva (labia major) shows a mass of 5.5 by 6.3 by 3 cm with a hyper and hypo internal echo relative to the adjacent tissue.

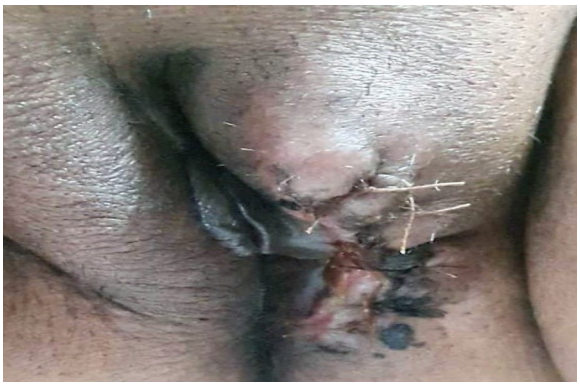

Figure 2. Clinical view of the site of tumor resection in the left side of labia major, just after surgery

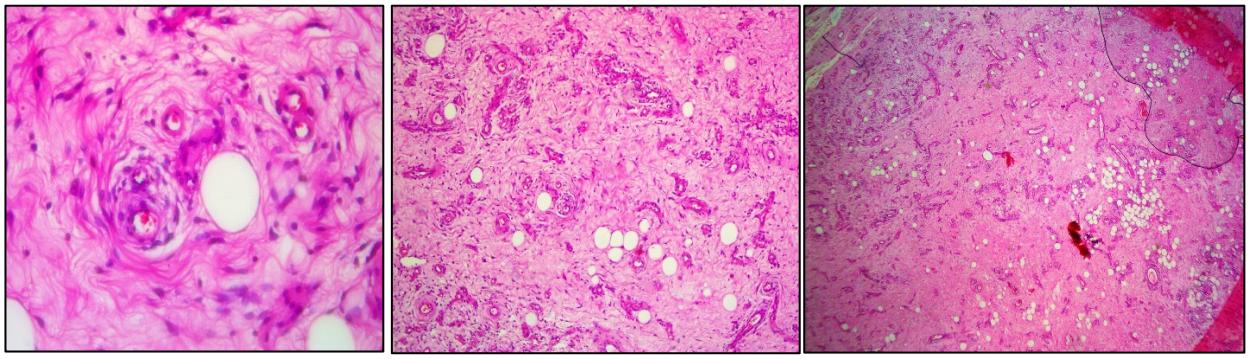

Figure 3. Microscopic finding revealed spindle-shaped cells in the myxoid stromal space with blood vessels, and adipose tissue. No pleomorphism or nuclear mitosis (magnification of 10, 40, and 100HPF)

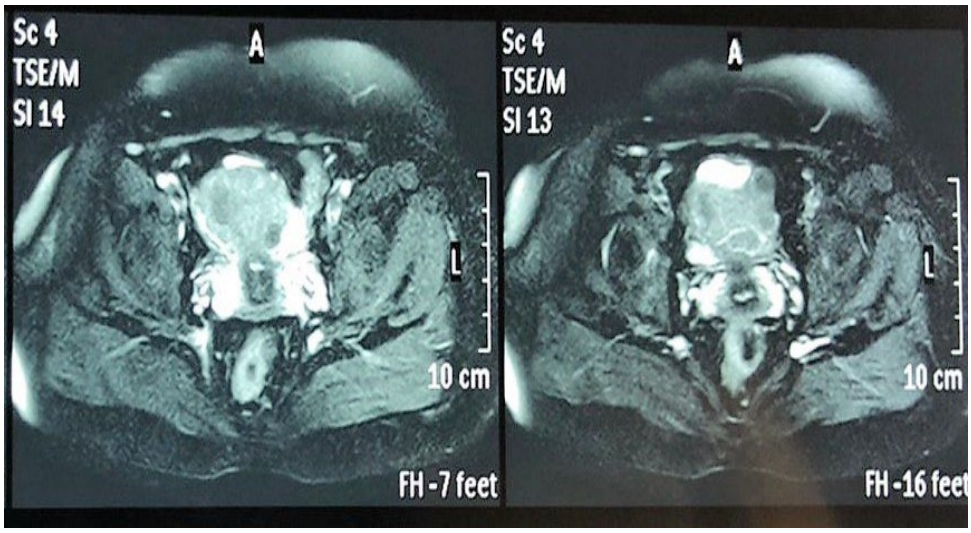

Figure 4. **MRI image of the left large labia after surgery with no evidence of tumor remnants**

Discussion

Invasive angiomyxoma was first described in 1983, as a painless mass in the external genitalia of childbearing age women, and rarely in the near menopausal age range **(3, 4)**. It's vascular, and infiltrate traits make it hard to respect the tumor completely in one session, especially in a young patient with cosmetic concerns, and the remaining surgical margin might be the cause of prompt recurrences **(5, 6)**. But its rarity makes it hard to assess the exact underlying factor for tumoral progression, and besides that, the standard management to decline the high chance of relapse, that is in about 80% of cases based on literature **(2)**. Referring to limited accessible studies, Sermpetzoglou et al., Believed in the overgrowth of mesenchymal cells based on the detection of desmin and actin elements in the immunohistochemically staining of these tumors, but Sozutek et al. emphasized the molecular substructure, they demonstrated a few changes in chromosome 12, the HMGIG gene that supposed to be the reason of abnormal mesenchymal cell development **(1, 2)**. Although neither of these hypotheses could explain the reproductive age limitation of this disease, the mystery was solved by Fine et al. they proposed angiomyxoma susceptibility to estrogen **(4)**. So, it seems that the lone reasonable explanation for angiomyxoma occurrence in a post-menopausal woman similar to the present case should be the presence of other well-known risk factors for a high level of plasma estrogen, like excess of fat tissue, or insulin-dependent diabetes mellitus as a lipogenic factor in increasing the body fat by itself. As a result, similar to the present case, the excessive adipose tissue contributes to converting the adrenal-provided androgen hormone to estrone, as a source of abnormally high estrogen levels even in post-menopausal age, needless to remind the importance of these factors in recurrences. There is remaining debate on the role of oophorectomy in reproductive ages, GnRH agonist, or other hormonal therapy like tamoxifen in this case **(1, 5)**. Focusing in particular on the vascular and myxoid nature of this tumor, imaging evaluation of tumor, especially computerized tomography (CT scan) or MRI is supposed to be diagnostic in initial and surveillance assessment **(5)**. But considering the present case, the ultrasonography is said to be non-characteristic, so MRI is recommended to be used in the first three years of postoperative follow-up, although there is a need for long-term surveillance based on a rare report of recurrences even after 14 years **(2)**. The clinical implication of this study is the certain role of completer resection with at least a one-centimeter free margin, in increasing the years of free disease survival **(6, 7)**. Adjuvant radiation and/or chemotherapy seem not to be efficient due to the rapid cell division of these tumors, except in rare cases with residual tumors after surgery **(1, 6)**. Furthermore, there is a rare report on unexpected distant metastasis **(2, 5)**, considering all of the above, individualized evaluation and management are highly recommended. Although the possibility of distant metastasis is very low, it should be considered **(2, 5)**.

Conclusion

It is clearly of great importance to emphasize the role of individualized medicine in such a rare case, in conclusion, there is not any debate on the role of surgical resection but the necessity of changing in lifestyle or adjuvant systemic or local therapy, and the needed duration is doubtful.

Acknowledgments

We want to admire all who have participated in this horrible situation of the COVID-19 pandemic, especially the health care providers.

Conflict of Interest

There are no conflicts of interests.

Funding

None.

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How to Cite This Article:

Mousavi Seresht, L., Farhadi Dehkordi, A. R., Danesh Shahraki, A., Hedaiat, P., Haghollahi, F. Post-menopausal Vulvar Aggressive Angiomyxoma. Can Diabetes Mellitus and Overweight be a Risk Factor? A Rare Case Report. J Obstet Gynecol Cancer Res. 2024;9(1):102-5.

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