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Case Report

Paget Disease of the Vulva: A Diagnosis not to be Forgotten

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ABSTRACT

Background: Paget disease of the vulva is a rare intraepithelial adenocarcinoma. Diagnosis is based on the histopathology results. It is associated with invasive adenocarcinomas, but other synchronous neoplasms can also occur. Treatment typically consists of wide local excision or vulvectomy. Local recurrence is frequent, and it may occur several years after initial treatment.

Materials & Methods: We present a case of vulvar Paget's disease treated with wide local excision.

Results & Discussion: An 85-year-old patient was referred to a gynecology consultation due to complaints of persistent vulvar itching. When she was first evaluated by a gynecologist, she had mild vulvovaginal atrophy, with a whitish color along both labia majora. At this time, a course of corticosteroid therapy was attempted. When she returned about four months after the initial assessment, she presented with an eczematoid and ulcerated appearance lesion with about 7 cm on the transition between the right labia majora with labia minora. At this time, a punch biopsy was done, which confirmed Paget disease of the vulva. All additional exams came back negative. She was further submitted to wide local excision of the lesion. Currently, the patient remains under follow-up at a gynecology consultation and is stable with no recurrences of the disease.

Conclusion: With this case presentation, the authors intend to emphasize the importance of considering all differential diagnosis of vulvar lesions, even those that are less frequent. Long-term follow-up is indicated in these cases because of the high risk of recurrence and the increased risk of synchronous carcinomas.

Keywords: Paget disease of the vulva, Synchronous neoplasms, Recurrence

1. Introduction

Paget disease of the vulva is a rare intraepithelial adenocarcinoma that accounts for less than 1% of all vulvar malignancies. It usually affects white, postmenopausal women in their 60s and 70s; its most common presentation symptoms are pruritus (present in 70% of patients) and vulvar discomfort[1-4]. Lesions usually have an eczematoid, ulcerated or crusty appearance which typically begins in the vulvar areas with hair, most frequently on the labia majora[5]. They are well demarcated and have slightly elevated edges and a red background, often dotted with pale islands. This condition is usually multifocal and may occur anywhere on the vulva, perianal area, or inner thigh; extension into the vagina is rarer but has also been reported[6].

Diagnosis is based on the histopathology results of a vulvar biopsy. This is crucial to make a correct diagnosis and it should be performed in all patients with suspicious lesions that persist even after adequate antieczema treatment.

This disorder is associated with invasive adenocarcinomas in up to 25% of patients[1,7-13]. In addition to these, the association with other synchronous neoplasms has been described, namely involving the breast, bladder, urethra, cervix, ovary, and rectum (in approximately 20-30% of patients)[3,13,14]. As so, evaluation may include mammography, urine cytology, colonoscopy and/or transvaginal ultrasound[15].

Treatment typically consists of wide local excision (a 2 cm

margin is preferred) or vulvectomy with or without inguinal lymphadenectomy, depending on the histological findings and extent of disease; however, more conservative surgery can be done in selected patients (eg, those with poor performance status and older age)[16].

Other treatments for localized, inoperable, or recurrent disease may include laser therapy, photodynamic therapy, radiation or even chemotherapy[17-19]. Management with topical 5% imiquimod cream has also been shown to be a safe conservative treatment option in these cases, with complete response rates of 22-90% of cases[20-23].

The occurrence of lymphovascular involvement and greater depth of invasion are typically poor prognostic markers[24].

Local recurrence is frequent (12-58%); it may occur several years after initial treatment and even in patients with negative surgical margins, presumably because of multicentricity and microscopic extension of disease beyond clinically visible margins[7-9,25,26].

2. Materials and Methods

We present a case of vulvar Paget´s disease treated with wide local excision.

3. Results and Discussion

An 85-year-old patient was referred to a gynecology consultation due to complaints of persistent vulvar itching, burning and discomfort with at least eight months of evolution. The patient denied dysuria or any vaginal discharge. When symptoms first began, she was evaluated by her family doctor, who prescribed topical and oral antifungal; still the patient did not notice any improvements.

The patient had a personal history of hypertension, dyslipidemia, cardiac arrythmia and depression and she was taking medication for all those disorders. As relevant surgical history, the patient had only undergone cholecystectomy. She had no family or personal history of skin disease and no known drug allergies. Menarche at the age of 14; gravida 6 para 6 (6 vaginal births); menopause at 47-years old, with no use of hormonal therapy.

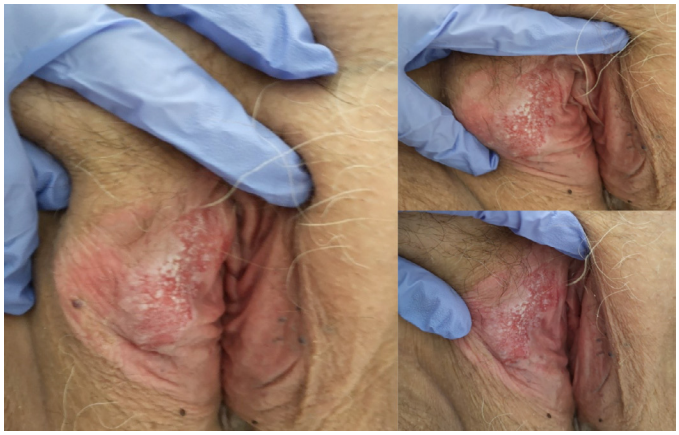
About six months later, when she was first evaluated by a gynecologist, she had mild vulvovaginal atrophy, with a whitish color along both labia majora, which also had a rough surface. At this time, a course of corticosteroid therapy was attempted, and the patient also began an emollient cream.

She missed the evaluation appointment six weeks later and was only re-examined again about four months after the initial assessment. When she returned, she had already abandoned corticosteroid therapy. She referred a partial initial improvement, however pruritus persisted, and it got worse when she stopped applying corticosteroid therapy.

On examination, she presented erythema on labia minora and labia majora bilaterally; on the transition between the right labia majora with labia minora, it was notable an eczematoid and ulcerated appearance lesion with irregular borders with about 7 cm of maximum diameter (**Figures 1**). At this time, a 5 mm length cylindrical punch biopsy of this region was done, which confirmed Paget disease of the vulva.

Posteriorly, faced with this diagnosis, mammography and breast ultrasound, cystoscopy, colonoscopy, and transvaginal ultrasound were requested to exclude malignant extra-vulvar disease. All of them were negative.

Figure 1: Clinical aspect of the lesion.



Then, she was submitted to wide local excision of the previously mentioned lesion. The post-operative period was uneventful.

The histological study of the surgical specimen confirmed the existence of vulvar Paget's disease; on immunohistochemistry evaluation, there were cells containing cytokeratin 7 (CK 7) and GATA3 and lack of expression of CDX-2, cytokeratin 20 (CK 20), estrogen and progesterone receptors and Melan-A. The surgical excision planes were free of the lesion, but with minimum margins of 8mm on both sides. The case was discussed in a multidisciplinary group meeting, and surveillance was decided.

Currently, the patient remains under follow-up at a gynecology consultation and is stable with no recurrences of the disease.

4. Conclusion

With this case presentation, the authors intend to emphasize the importance of considering all differential diagnosis of vulvar lesions, even those that are less frequent.

Pruritus and vulvovaginal erythema may be caused by a range of conditions that varies from immunological, infectious, or even malignant causes; it often represents a frequent cause of misdiagnosis that can lead to delays in effective treatment.

Our patient fitted the typical characteristics of being white, postmenopausal women; the fact that the symptoms persisted despite initial corticosteroid treatment was a red flag.

Long-term follow-up is indicated in these cases because of the high risk of recurrence and the increased risk of synchronous carcinomas[1,7-11]. Therefore, the vulva should be inspected at least once a year with a low threshold for biopsy and screening and surveillance for tumors at other sites must be considered.

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