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Case Report

Surgical Treatment of a Vulvar Paget Disease: A Case Report

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Abstract

Paget disease of the vulva is a chronic, rare type of vulvar malignancies that occurs in postmenopausal women. Vulvar Paget's disease is a rare neoplasm of the vulva that presents as a dermatological benign disease. Recurrences are common and the selection of the accurate management is very challenging. We hereby, are reporting the case of a 54 years old female patient diagnosed with a vulvar Paget disease that was only treated via a surgical resection, without any adjuvant therapy and that resolved with no signs of recurrence.

Introduction

Paget disease of the vulva is a chronic, rare type of vulvar malignancies that occurs in postmenopausal women, usually in their sixth to seventh decades, with a small percentage occurring in young patients.

It is known to be a very confusing disease because of its uncertain etiology and physiopathology that could mimic many types of dermatoses. In addition, there is no standardized established treatment since its first description hundred years ago.

The main clinical manifestation is pruritis associated with tender lesions on the vulva. The patient may experience the symptoms for several years before seeking a medical advice.

We hereby, are reporting the case of a 54 years old female patient diagnosed with a vulvar Paget disease that was only treated via a surgical resection, without any adjuvant therapy and that resolved with no signs of recurrence.

Case Presentation

A 54 years old menopausal woman, known to have a past history of breast adenocarcinoma treated with mastectomy, ipsilateral axillary lymphadenectomy with adjuvant chemotherapy, was admitted in the department of obstetrics and gynecology, for painful vulvar lesions that appeared on the right side of her vulva. Based on the patient's story, the lesions began to appear 6 months prior to her consultation, associated with pruritus, inflammation, redness and sometimes, pain. She was repetitively treated by topical corticosteroids with minimal improvement and recurrences.

On physical examination, the patient had on the right vulva, an erythematous, papular, pink lesions, with hyperkeratosis taking over the upper part of the labia majora. Cervical and rectal exams were normal, without any apparent lesion on the left vulva.

She was counseled to get a vulvar biopsy for further investigations. The patient was cooperative and had her biopsy done. The pathology report of the biopsy was compatible with vulvar Paget disease. In fact, microscopic examination showed an acanthotic epidermis colonized by large cells with an irregular nucleus and moderately abundant cytoplasm. Glandular lobules of mammary type were detected in the dermis, along with vacuoles that were seen after coloration with Alcian Blue.

After the patient's consent, a frozen-section guided wide local resection of the lesion was performed. The patient didn't show any recurrence since the surgery with improvement of her symptoms and disappearance of the pain over one year follow-up. No adjuvant treatment was used in this case.

Discussion

Sir James Paget was the first to discover the mammary Paget disease and described it in 1874 [1], as a lesion around the areola and the nipple. It was followed by another case reported by Crocker in 1889 [2], where it was described as lesions appearing on the scrotal area. On 1901, the vulvar Paget's disease was illustrated by Dubreuilh [3], who defined the extra mammary Paget disease.

Vulvar Paget's disease is an extremely rare, recurrent vulvar malignancy that could be misdiagnosed with other various forms of dermatoses. It arises either from the apocrine gland or from an adjacent form of adenocarcinoma including the bladder, the rectum or the anal region. The vulva remains the most affected area, as it consists 65% of the extramammary disease [4].

Vulvar Paget's disease represents less than 1.5% of the vulvar malignancies [5], and it is mostly seen in white and Caucasian postmenopausal females. The pathophysiology of the disease is not very well known. Some theories state the fact that the cells seen in Paget's disease are usually found in the basal layer of the epidermis of the genital area, as malignant keratinocytes, transformed in situ [6] via the angiogenesis mechanism. Consequently, the Paget's cells are larger than keratinocytes, having clear chromatin, prominent nucleolus and a gray–blue cytoplasm, with vacuoles after hematoxylin and eosin staining.

The clinical presentation begins with the development of superficial pink-red pruriginous lesions distributed in an island shape, often seen in an eczematous form, and can sometimes be painful [7]. The vulvar skin may be thick giving the false impression of leukoplakia. Generally, Any palpable mass should suspect an underlying invasive disease.

Wilkinson and Brown classified the vulvar Paget's disease into two forms:

- The primary (cutaneous) Paget's disease which is an intraepithelial adenocarcinoma arising within the epidermis.

- The secondary form, also called noncutaneous Paget's disease, and arises from anorectal and bladder adenocarcinoma mainly, or from other origins such as the cervix, the endometrium and the ovary [8].

The definitive diagnosis is made via a direct biopsy of the lesion, followed by immunohistochemical studies in order to distinguish between the two forms.

The treatment of vulvar Paget's disease was mainly surgical, through a wide, deep local excision of the skin (including the appendages), for intraepithelial Paget's disease. As for the invasive form, either a partial or a total vulvectomy is made. However, high rate of recurrences (20-65%) were found with positive margins after the frozen section assessment. In fact, positive margins are associated with a higher local recurrence rate than the negative ones. If an underlying adenocarcinoma is present, the treatment of the lesion would be similar to that of the squamous cell carcinoma of the vulva. Most of the recurrences were reported at ten years after the surgical excision. This makes the long term follow-up mandatory for the patient's safety. The disappointing outcomes of the surgical treatment consist of the high morbidity/side-effects ratio which has encouraged many doctors to try alternative treatments such as topical applicable solutions like 5FU and imiquimod 5% cream, laser ablation, or the radiotherapy that was used as a postoperative treatment to prevent local recurrences 25. However, the exclusion of the surgical treatment and its replacement by the latter alternatives, is still debatable and subjected for too many controversies. Hatch and Davis reported two cases of patients with vulvar Paget's disease achieving a biopsy-confirmed resolution of their disease after topical application of imiquimod 5% cream [9]. Sendagorta *et al* reported a complete histological and clinical remission of the vulvar Paget's disease, in three patients treated by a daily application of imiquimod 5% cream for 3 weeks. Only mild irritation and tenderness were observed as side effects [10].

On the other hand, the topical photodynamic therapy (PDT) has shown equivalent results in comparison with the surgical treatment in the case of extramammary Paget's disease. In fact, there is a complete clinical response at six months following two cycles of phototherapy [11]. Even if the recurrence rate was high,

the PDT treatment is optimal to be repeated without any functional or physical consequences. PDT resulted in the disappearance of pain and the improvement of the quality of life.

Conclusion

Vulvar Paget's disease is a rare neoplasm of the vulva that presents as a dermatological benign disease. Consequently, its diagnosis is delayed with bad results on the prognosis.

Recurrences are common. and the selection of the type of management is very challenging. In fact, until nowadays there is no standard treatment that exists. So every decision will be taken according to many factors (the size, the location, the symptoms, etc.).

The most important issue is that the biopsy should never be delayed facing any recurrent pruritic vulvar lesion that didn't respond to the usual therapy.

Conflict of Interest

No conflict of interest to declare

Consent and Ethical Approval

Obtained from the patient to publish the case.

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Bibliography

1. Paget, J. (1874). On disease of the mammary areola preceding cancer of the mammary gland. *St Bartholomew's Hospital Research, 10*(87).

2. Crocker, H. R. (1889). Paget's disease affecting the scrotum and penis. *Transactions of the Pathological Society of London, 40,* 187-191.

3. Dubreuilh, W. (1901). Paget's disease of the vulva. *British Journal of Dermatology, 13*(11), 403-413.

4. Chanda, J. J. (1985). Extramammary Paget's disease: prognosis and relationship to internal malignancy. *Journal of the American Academy of Dermatology, 13*(6), 1009-1014.

5. Jones, I. S. C., Crandon, A. & Sanday, K. (2011). Paget's disease of the vulva: diagnosis and follow-up key to management; a retrospective study of 50 cases from Queensland. *Gynecologic Oncology, 122*(1), 42-44.

6. Lagios, M. D., Westdahl, P. R., Rose, M. R. & Concannon, S. (1984). Paget's disease of the nipple. Alternative management in cases without or with minimal extent of underlying breast carcinoma. *Cancer, 54*(3), 545-551.

7. Shaco-Levy, R., Bean, S. M., Vollmer, R. T., *et al*. (2010). Paget disease of the vulva: a histologic study of 56 cases correlating pathologic features and disease course. *International Journal of Gynecological Pathology, 29*(1), 69-78.

8. Wilkinson, E. J. & Brown, H. M. (2002). Vulvar Paget disease of urothelial origin: a report of three cases and a proposed classification of vulvar Paget disease. *Human Pathology, 33*(5), 549-554.

9. Hatch, K. D. & Davis, J. R. (2008). Complete resolution of Paget disease of the vulva with imiquimod cream. *J Low Genit Tract Dis., 12,* 90-94.

10. Sendagorta, E., Herranz, P., Feito, M., Ramirez, P., Floristan, U., Feltes, R., *et al.* (2010). Successful treatment of three cases of primary extramammary Paget's disease of the vulva with Imiquimod-proposal of therapeutic schedule. *J Eur Acad Dermatol Venereol., 24*(4), 490-492.

11. Clément, E., Sparsa, A., Doffoel-Hantz, V., *et al.* (2012). Traitement de la maldie de paget extramammaire par photothérapie dynamique topique. *Annales de Dermatologie et de Vénéréologie, 139*(2), 103-108.