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# Paget Disease of the Vulva: One Case Report

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## Abstract

Extramammary Paget's disease is a rare neoplasm that usually develops in apocrine gland-bearing areas. Paget's disease of the vulva is a rare form of extramammary Paget's disease affecting postmenopausal women. We report the case of a 39-year-old woman who presented with a vulvar lesion suggestive of Paget's disease involving only the labia majora and minora. Biopsy confirmed the diagnosis of non-invasive Paget's disease. Extension examination was negative. The patient underwent a superficial vulvectomy with healthy margins on histopathology. Postoperative management was uncomplicated.

**Keywords:** Paget Disease of the Vulva; Extramammary Paget's Disease; Vulvar Lesion

**Abbreviations:** PDV: Paget's Disease of the Vulva, WHO: World Health Organization, EMPD: Extramammary Paget's Disease, EMPD-V: Extramammary Paget's Disease of the Vulva

## Introduction

Paget's disease of the vulva (PDV) is a rare pathological condition accounting for 1% to 2% of all female genital tumors **[1,2]**. It was originally described by William Dubreuil, a French dermatologist, in 1901 **[3]** and is classified by the World Health Organization (WHO) as an epithelial cancer arising from epidermal pluripotent stem cells of the follicular-apocrine and sebaceous cell units.

The standard treatment for Paget's disease is surgical excision. However, the recurrence rate is high (between 30 and 60%), regardless of the surgical technique used (focal resection, simple or even radical vulvectomy) **[4]**. To limit the functional and aesthetic sequelae, other more conservative treatments have been proposed, such as radiotherapy, CO2 laser, topical dynamic phototherapy (PDT), topical application of imiquimod or 5-fluorouracil (5-FU), as first or second intention, with variable efficacy **[5]**.

We report a case of a 39-year-old woman with a vulvar lesion diagnosed as non-invasive Paget's disease who was treated in our structure.

## Case report

A 39-year-old single woman with no specific pathological history who presented with itchy heterogeneous erythematous vulvar lesions suggestive of Paget's disease.

Lesions, affecting the labia majora and labia minora, without involvement of the clitoral hood or the urethral meatus **(Fig. 1 A and Fig. 1 B).** A biopsy confirmed the diagnosis of non-invasive Paget's disease. The gynecological and digestive extension workup was negative. The patient underwent a superficial vulvectomy. The anatomopathological examination revealed in the:

- **Right vulvectomy:** the large and small lip: There is a poorly limited reddish erythematous plaque at the large and small lip junction (intraepidermal tumor proliferation arranged in isolated cells sometimes organized in clusters. Tumor cells have an abundant pale cytoplasm and atypical irregular large nuclei. Immunohistochemical study shows that tumor cells express cytokeratin 7 and do not express PS 100), measuring 2x1x0.1 cm, it remains 3 cm from the upper limit, 2.2 cm from the lower limit, 2 cm from the lateral limit and 0.4 cm from the vaginal limit, 0.6 cm from the lesser lip and 0.2 cm from the deep limit.
- **Small left lip:** Skin flap site of tumor proliferation of the same appearance described above.

The development was marked postoperatively by good healing **(Fig.2A and Fig.2B)**.

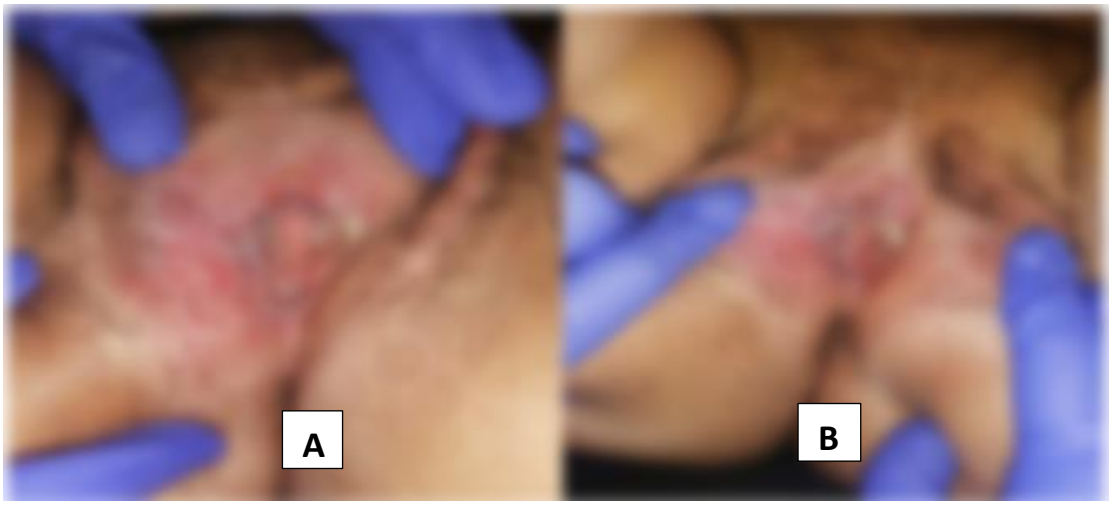


**Figure 1 (A & B):** Appearance of vulvar lesions


**Figure 2 (A & B):** the result after surgery

## Discussion

Extramammary Paget's disease (EMPD) is a distinct form of relatively rare cutaneous neoplasm with unclear histogenesis. Importantly, it may be associated with regional or distant malignancy, necessitating a thorough work up and investigation **[6]**. Extramammary Paget's disease of the vulva

(EMPD-V) predominantly affects postmenopausal Caucasian women over the age of 50, with a median age of 65 years **[7-8]**. The disease is rare in black patients **[9]**; interestingly, an increasing number of patients have been observed in Asian populations over the past 4-5 years. Seven familial cases (six Japanese, one British) have been published, but the genetic basis is unknown **[10]**.

In most patients, PDV causes symptoms of irritation, itching and burning. PDV can be asymptomatic in some patients: approximately 5-15% of patients have no symptoms at the time of diagnosis (Perez et al, 2014; De Magnis et al, 2013). In our patient it manifested as pruritus.

When PDV is suspected, a detailed history should be taken, including a history of vulvovaginal, gastrointestinal and urological complaints. In addition, a full gynecological examination should be carried out, including vulvar, vaginal and rectal examinations. Any raised, pigmented or otherwise suspicious lesions should be followed up with a thorough report, digital photography and histological examination. Digital photography can help to monitor the course of the disease. Invasive disease should be excluded, preferably by vulvar mapping, including multiple biopsies of the involved and surrounding uninvolved skin. In the case of a small unifocal lesion, a single biopsy may be considered, with complete excision of the visible lesion. The diagnosis is confirmed by the histological presence of Paget's cells **[11]**.

Differential diagnoses may include cutaneous candidiasis, tinea cruris, seborrhoeic dermatitis and psoriasis, Bowen's disease or melanoma **[12]**, which explains the multiple topical treatments prior to biopsy and for this reason PDV should only be diagnosed by vulvar biopsy. The importance of immunohistochemistry in the diagnosis of PDV has now been mentioned in all studies. It is important for differential diagnosis with other vulvar diseases such as melanoma and squamous cell carcinoma. Immunohistochemistry can be used to highlight secondary forms of Paget's disease of the urinary tract and gastrointestinal cancers **[12]**.

Surgery is still considered the gold standard for the treatment of PDV **[13]** and is a factor that increases overall survival **[12]**. Due to the local aggressiveness of these lesions, it is an extensive vulvar surgery with removal of the clitoris. Clinically healthy margins of 2 cm are required as the boundaries of the lesion, which is very poorly defined, are difficult to assess preoperatively. In the event of invasion, inguinofemoral dissection is associated with a sentinel node technique which is currently being evaluated **[12]**.

In addition, recurrence rates after surgical treatment are high and morbidity is impressive (Perez et al, 2014; Fanning et al, 1999). There is therefore an urgent need for alternative treatment options for PDV: topical imiquimod cream, chemotherapy for metastatic EMPD, radiotherapy and other topical treatments **[11]**.

## Conclusion

PDV preferentially affects postmenopausal women. Clinical diagnosis is often delayed after onset because of the atypical symptoms and the small number of cases, if any, seen by clinicians. PDV may be invasive or non-invasive from the outset and the final diagnosis is based on anatomopathological features. Surgery is the treatment of choice for women with invasive PDV who are in good general health, although it is often perceived as mutilating. Topical treatment with imiquimod seems promising for non-invasive cases and is less likely to cause serious side effects. Regardless of treatment, recurrence is common and patients require close monitoring.

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