

Extramammary Paget's disease found by abnormal vulvar brush sampling

B. K. J. Yu¹, C. R. Lai², M. S. Yen¹, N. F. Tou¹, K. C. Chao¹, C. C. Yuan¹

Department of Obstetrics and Gynecology¹, Pathology², Taipei Veterans General Hospital, National Yang-Ming University, National Defense Medical Center, Taipei (Taiwan)

Summary

Introduction: Doctors are usually reluctant to perform a vulvar biopsy on a patient with non-specific chronic vulvitis – especially because of the rarity of vulvar malignancy in young women – until the lesion is suspected of being malignant. Therefore, most cases of extramammary Paget's disease (EMPD) were originally misdiagnosed as chronic and recurrent vulvar lesions. Late diagnosis of invasive lesions occurring in elderly females have resulted in cases of death.

Case: A 37-year-old patient showed an extended lesion on the vulva and perineum. In addition, abnormal cells were found from a vulvar scrape smear, and a following punch biopsy was used to diagnose and determine the extension of the disease.

Conclusion: Diagnosis and demarcation of EMPD remain difficult due to the multifocal lesions and subtle nature of the disease. Brush sampling taken from suspicious areas can be a guide for multiple biopsies to demarcate the lesion before major surgery. A brush biopsy is presented as a first-step method to detect vulvar malignancy.

Key words: Extramammary Paget's disease; Tissue sampling; Cancer screening.

Introduction

Extramammary Paget's disease (EMPD) is classified as a non-squamous intraepithelial neoplasia of the vulva [1] and is a very rare disease that occurs mostly in females over 50 years of age [2]. There is no significant characteristic of EMPD; therefore, late diagnosis is often from punch biopsies. Some cases were first diagnosed as invasive adenocarcinoma [1]. Doctors are usually reluctant to perform a vulvar biopsy on a patient with non-specific chronic vulvitis, especially in young women, until the lesion is suspected to be malignant. On the other hand, the rarity of vulvar malignant cases may hinder the doctor from performing an invasive biopsy due to chronic vulvitis. We present a case of EMPD in a 37-year-old patient with an extended lesion on the vulvar and perineal area where diseased cells were found in a vulvar smear taken by a sampling brush.

Case Report

A 37-year-old gravida 2, para 2, Asian female presented with a two-year history of recurrent itching in the vulva that had been previously diagnosed as chronic vulvitis. Physical examination showed an edematous lesion about 28 mm in width and 77 mm in length bilaterally on the vulva and perineum, including the clitoris, labia minor and major. The base of the lesion was erythematous, and the lesion itself was moist with exudate and partially covered with a dull white film, consistent with descriptions of the "cake-icing effect" seen in some cases of EMPD (Figure 1). Pelvic and breast examinations revealed no abnormalities and inguinal lymph nodes were non-palpable. The patient's physical symptoms were consistent with a diagnosis of chronic vulvitis. Following puffs with 10% xylocaine spray (Astra Sodertalje, Sweden), a brush smear was taken from a catarrh area of the vulva using a SpiraBrush (Trylon Corp.,

Torrance, CA) whereby glandular cells were observed (Figure 2). A colposcopy examination was performed on the perineum after applying 5% acetic acid solution and showed a distinct lesion with a mild acetowhite appearance and blurred margins.

The scrape smear showed positive Paget's cells. Biopsies were then performed using a 2-mm Keyes punch over the extended lesion as well as the entire vulva and perineum to define the margins of the lesion before performing major surgery. Eleven out of 19 specimens tested positive for nonin-

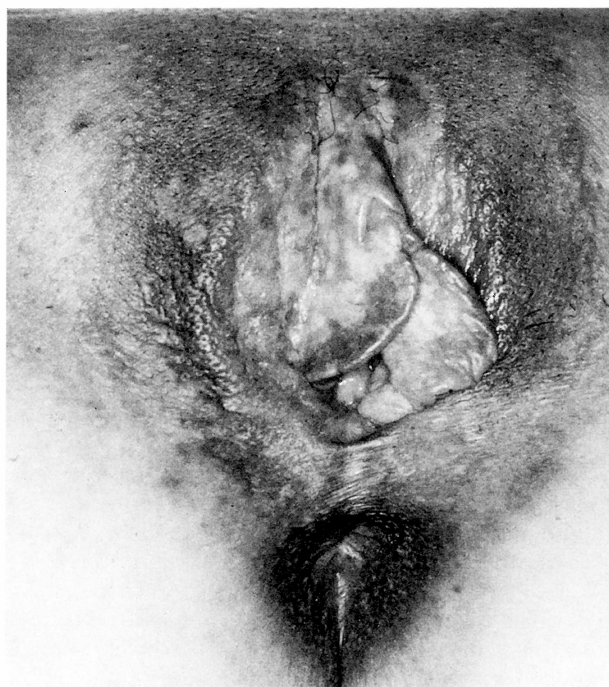


Figure 1. — Appearance of EMPD on the vulva. An edematous lesion with "cake-icing appearance" on the vulva and perineum, including the clitoris, labia minor and major, and the perineal region.



Figure 2. — Vulvar brush smear showing Paget's cells with eccentric nuclei and abundant pale lacy-like mucin containing cytoplasm (Papanicolaou stain, 400x).

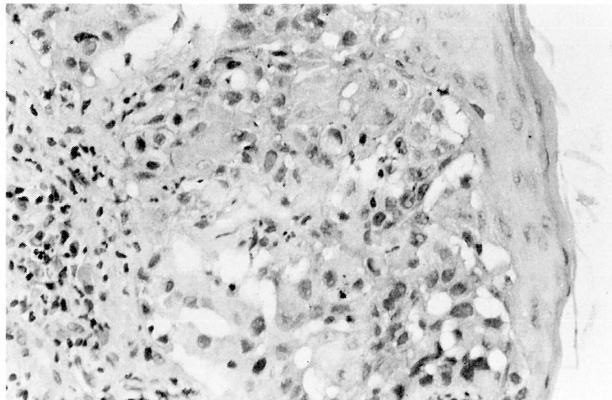


Figure 3. — Paget's cells in the deep dermis, characterized by distinct borders, abundant pale cytoplasm, eccentric nuclei and prominent nucleoli (H&E, 200x).

vasive EMPD. The lesion sites extended over the introitus, perineum, and vulva. A modified radical vulvectomy and superficial inguinal lymph adenectomy were performed on both sides. Due to the large size of the excised portion, a lateral transposition skin flap procedure was performed from both inner thighs for reconstructive surgery. Final reports on the excised lesion showed noninvasive EMPD (Figure 3) and clear margins; samples from all lymph nodes were free from disease.

Discussion

Extramammary Paget's disease (EMPD) is a very rare disease and lacks diagnostic criteria. Most cases are initially diagnosed and treated as cases of chronic vulvitis, resulting in the neglect of a serious preinvasive disease. At 37 years of age, our patient was rather young to be diagnosed with Paget's disease, in that the literature reports the disease as occurring primarily in women between the ages of 50 and 75 [2]. Given EMPD's ambiguous symptoms, the patient may have been misdiagnosed with chronic vulvitis for two years.

So far, a biopsy has been the only method of differentiating Paget's disease from chronic vulvitis. Gynecolo-

gists are reluctant to perform biopsies in patients with non-specific chronic vulvitis, especially in young women, due to the rarity of Paget's disease. Biopsies are performed only when the lesion is suspected of being malignant. Vulvar scrape sampling following puffs of 10% xylocaine spray solution can be a simple and painless method performed in a clinic to screen for vulvar malignancy as well as to examine the extent of the lesion. However, it is rare for Paget's cells to be shed from the lesion except in cases where the lesion has been scraped. As the SpiraBrush is made of hard nylon, the tool provides a useful sampling from the vulvar skin. Thus, liberal application of scrape sampling from the suspected vulvar region can aid in earlier detection of Paget's disease.

A punch biopsy is a conventional method of differentiating between malignant and chronic benign vulvar disease. Diagnosis of EMPD is hampered by its multi-focal nature and lack of typical lesions [3]. An earlier liberal biopsy taken from patients diagnosed with chronic vulvar disease is encouraged as a method of detecting vulvar malignancy [4]. In this case, a biopsy taken 3 mm away from the gross margin of the lesion revealed the Paget's lesion. The size of the excision in an operation is dependent on the extension and severity of the vulvar Paget's. The difficulties of recovery through rehabilitation as well as sexual functional preservation are closely related to the extensiveness of the surgery. Therefore, extended random multiple punch biopsies are mandatory to determine if and where EMPD is present in the vulvar and perineal regions before proper surgery. In this case, 19 punch biopsies were taken from the extended lesion of the patient covering nearly the entire vulvar and perineal region.

The scrape sampling in this case detected Paget's cells. We consider smear or cell block studies from the scrape sampling as a first-step method for cancer screening of patients with chronic vulvar lesions. Following the screening of scrape samplings on any suspicious lesions, intensive multiple punch biopsies can then be taken from target areas of positive cytology detected by the scrape samplings. Therefore, a simple initial procedure based on multiple scrape samplings would allow a thorough examination for malignancy of the external genitalia to result in reduced misdiagnosis, earlier detection, and good accessibility.

References

- [1] Wilkinson E. J.: "Normal histology and nomenclature of the vulva, and malignant neoplasms, including VIN". *Derm. Clinics.*, 1992, 10, 283.
- [2] Kurzl R. G.: "Paget's disease". *Semin. Derm.*, 1996, 15, 60.
- [3] Gunn R. A., Gallager H. S.: "Vulvar Paget's disease- a topographic study". *Cancer*, 1980, 46, 590.
- [4] DiSaia P. J., Creasman W. T.: "Clinical gynecologic oncology. Invasive cancer of the vulva". In: "Paget's Disease" (eds.): DiSaia P. J., Creasman W. T. Missouri: Mosby-Year Book, Inc. 1997, 222.

Address reprint requests to:
 BILL KEN-JEN YU, M.D., Ph.D.
 Department of Obstetrics and Gynecology
 Taipei Veterans General Hospital
 201, Sec. 2, Shih-Pai Rd. Taipei (Taiwan)