

Vulvar basal cell carcinoma in a patient with long-standing lichen sclerosis



Jeanette R. Comstock, BA,^a Carina M. Woodruff, MD,^b Siegrid S. Yu, MD,^b and Rachel I. Kornik, MD^c
Madison, Wisconsin and San Francisco, California

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INTRODUCTION

Lichen sclerosis (LS) is a chronic, inflammatory dermatosis most commonly affecting the vulva. It is estimated that LS is associated with up to a 5% risk of vulvar squamous cell carcinoma (SCC) development.¹ We present an unusual case of basal cell carcinoma (BCC) of the vulva arising in a foci of vulvar LS after treatment with topical steroids and tacrolimus ointment.

REPORT OF A CASE

A 44-year-old woman with a medical history of more than 10 years' of LS and morphea presented with a 6-week history of persistent pain on the left labium majus and generalized vulvar irritation and erythema. The patient denied a history of bleeding, trauma, or radiation to the affected area. Prior treatment for her LS included high-potency topical steroids, tacrolimus ointment (applied to the affected area for less than 1 year), vaginal dilator, and coconut oil. A skin culture was obtained and grew group B *Streptococcus*. The patient was treated with cephalixin and bleach baths. The generalized vulvar irritation resolved, but the focal tenderness on the left labium majus remained. The patient was reluctant to undergo biopsy. Subsequent therapies included courses of mupirocin and increasing frequency of desoximetasone with tacrolimus.

Examination was significant for a subtle 0.5 × 0.7-cm pink macule on the medial left labium majus in addition to previously noted loss of the labia minora and fusion of the clitoral hood consistent

Abbreviations used:

BCC: basal cell carcinoma
LS: lichen sclerosis
SCC: squamous cell carcinoma

with LS (Fig 1). A shave biopsy found collections of basaloid cells arising from the undersurface of the epithelium and separated by clefts from adjacent fibromyxoid stroma, consistent with a diagnosis of superficial basal cell carcinoma. Staining with cytokeratin 20 and T-cell death-associated gene 51 immunoperoxidase stains was negative. The patient underwent Mohs micrographic surgery. Notably, the tumor had marked microscopic extension and required 5 stages for clearance, with the final surgical defect measuring 1.7 × 2.5 cm (Fig 2). The wound was left to heal by second intention. Surveillance colposcopy and biopsies 6 months later found no recurrence.

DISCUSSION

SCC is by far the most common cancer affecting the vulva, accounting for 90% of all vulvar malignancies.¹ The association between LS and SCC is well established.² The mechanism of the carcinogenic progression from LS to SCC is not clearly elucidated but is thought to be related to high cell turnover in the setting of chronic irritation.¹

In contrast, BCC is extremely rare in the perianal and genital regions with an estimated prevalence of less than 1%.³ Exposure to ultraviolet radiation, fair

From the Department of Dermatology,^c The University of Wisconsin School of Medicine and Public Health^a and the Department of Dermatology, University of California, San Francisco.^b

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Correspondence to: Rachel Kornik, MD, University of Wisconsin School of Medicine and Public Health, 1 South Park Street, 7th Floor, Madison, WI 53715. E-mail: rkornik@dermatology.wisc.edu.

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Fig 1. BCC before biopsy. **A**, Single 0.5- × 0.7-cm pink macule on the medial left labium majus in the setting of pre-existing loss of the labia minora and fusion of the clitoral hood before biopsy. **B**, Pre-Mohs micrographic surgery. An ill-defined 0.5- × 0.7-cm pink macule coinciding with the initial biopsy site was demarcated for the first stage of Mohs micrographic surgery.



Fig 2. After Mohs micrographic surgery. After surgery, the defect measured 1.7 × 2.5 cm.

skin, and advancing age are known risk factors for the development of BCC.⁴ However, the pathophysiology of BCC development in non-sun-exposed regions remains unknown. Vulvar BCC develops

most commonly in middle-age to elderly women (average age, 73 years) and tends to present with a large, often ulcerated lesion (average size, 1.95 cm).⁵ Associations between genital BCC and preceding immunosuppressive therapy, chronic irritation, pelvic radiation, and prior trauma have been reported.^{4,5} To our knowledge, there are very few cases of BCC arising in a foci of LS.³

The potential pathogenic role of tacrolimus in this case remains unclear. LS compromises the skin barrier, potentially allowing tacrolimus to penetrate and accumulate in the epidermis and dermis, where it is theorized to impede DNA repair and decrease apoptosis of keratinocytes leading to abnormal cell differentiation. Oral calcineurin inhibitors confer an increased risk of nonmelanoma skin cancer in organ transplant recipients.⁶ However, the literature has not definitively correlated topical calcineurin inhibitors with an increased risk of skin cancer in atopic dermatitis patients, and there are no data to suggest an elevated cancer risk in other inflammatory dermatoses.⁷

The presentation of vulvar BCC can be very subtle and nonspecific, with symptoms that can closely mimic LS. As such, this case highlights the importance of clinician vigilance in patients with existing LS for the subtle symptoms of vulvar BCC that should prompt timely biopsy and treatment. In addition,

patients with vulvar BCC are generally older, making this case of a relatively young woman presenting with a BCC very unusual.

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