

Vulvar basal cell carcinoma: report of a case involving the mucosa and review of the literature

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Abstract

We report the case of woman who presented a vulvar basal cell carcinoma (BCC) on the inner part of the labium majus, treated with local resection. Vulvar BCC is a rare cancer but can be long misdiagnosed due to a non-specific presentation. Though even rarer, BCC involving the mucosal side of the labium majus has to be considered in the differential diagnosis of the vulvar tumors. A complete excision with free margins is the treatment most recommended. Other recommendations include the early identification of aggressive subtypes, which carry a greater risk of recurrence and spreading potential as well as a long-term follow-up with exhaustive muco-cutaneous examination.

Case Report

A 57-year-old woman presented with an asymptomatic genital eroded mucosal lesion that she had noticed two years earlier (Figure 1). Physical examination showed a papillomatous lesion of 2.2X1.5 cm, located on the mucosal surface of the left labium majus. There was visible pigmentation and bleeding. Lymph nodes areas remained free. The patient's medical history was unremarkable. Differential diagnosis was to be made between pemphigus vegetans and tumors of the vulvar area. A resection-biopsy was performed under local anesthesia and revealed a basal cell carcinoma of compact, nodular and pigmented type, with erosion on surface. All margins of excision were free of disease. Follow-up to date showed no evidence of recurrent or metastatic carcinoma.

Discussion

Basal cell carcinoma (BCC) of the vulva represents 2-3 % of the vulvar cancers and less than 1% of all BCC. Out of around 250 cases

published so far, only twenty cases of BCC of the clitoris, the labia minora or the medial non-hair bearing aspect of the labia majora have been described.¹ Review of the literature shows that, considering other mucosae, four cases related to the buccal mucosa have been published.²⁻⁵ To our knowledge, no case involving the glans is reported. In the literature, vulvar BCC is commonly reported to affect white women, mostly in the post-menopausal period⁶ though some cases affecting younger women have been described.¹ Diagnosis is difficult as presentation and clinical manifestations are unspecific. As a result, the delay for diagnosis reaches 5 to 6 years on average.¹ Complaints mostly range from the discovery of an asymptomatic labial lesion to the evaluation of pruritus, pain or bleeding. Clinical manifestations are diverse and do not usually suggest BCC, lacking characteristic pearly and telangiectatic aspects.¹ They are showing an exophytic, ulcerated, pedunculate, infiltrating, nodular or pigmented lesion, mostly located on the non-mucosal surface of the labia majora.⁷ Differential diagnosis has to be made between pemphigus vegetans and tumors of the vulva (Table 1).^{1,6,8-15} BCC of the vulva is not located on a surface exposed to UV and therefore other risk factors need to be determined. To date, there hasn't been any evidence of clearly identified risk factors, especially for BCC involving mucosa. Possible associations remain prior radiation therapy^{1,16} and previous trauma such as a burn or a scar.¹⁶ Common risk factors to cutaneous BCC have to be considered: Gorlin syndrome,^{17,18} chronic radiation,^{1,16,19} chronic arsenic exposure²⁰, mutations in p53,²¹ xeroderma pigmentosum.¹¹ Immunosuppressive medication has been suggested.¹⁶ A few cases have been reported in association with pre-existing lesion: lichen sclerosus et atrophicus,²² Paget's disease,²³ multiple tumors of the follicular infundibulum.¹

Biopsy of any suspect lesion is widely recommended.^{7,16,24} Once diagnosis is confirmed, conservative surgery is most indicated, with free margins resection.^{1,25-27} Some cases of relapses have been reported and are possibly due to inadequate margins. Local recurrence varies from 0-25% in published reports¹ with an average of 10-20%.⁷ The aggressiveness and recurrence of BCC vary according to histological pattern.⁷ Tumors of the morphea-like (nodular, sclerosing), metatypical (basosquamous), adenocystic or infiltrative types are more aggressive, leading to a higher rate of recurrences. An aggressive BCC is associated with often deep local infiltration and occasional perineural extension.⁷ Several cases of metastasizing BCC have been reported with an incidence of 0.0028-0.1% and with a mean time from initial presentation reaching 9 years.⁷ These cases raise a challenging differential diagnosis in which it can be difficult to distin-

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Key words: vulvar basal cell carcinoma.

Received for publication: 3 August 2011.
Accepted for publication: 31 August 2011

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Dermatology Reports 2011; 3:e37
doi:10.4081/dr.2011.e37

guish BCC from adnexal tumors. Eventually, BCC may be associated with another vulvar tumor, such as melanoma or epidermoid carcinoma.¹ Most reported associated cancer with BCC of the vulva is uterine neck cancer (UNC).¹ Considering the role of HPV in the genesis of UNC, a few studies have assessed the presence of HPV in genital BCC on small



Figure 1. Eroded and pigmented papillomatous lesion located on the mucosal surface of the left labium majus.

Table 1. Vulvar cancers.

Squamous-cell cancers	90%
Non-squamous tumors	10%
Malignant melanoma	
Bartholin's gland carcinoma	
Verrucous carcinoma	
Paget's disease	
Adenosquamous carcinoma	
Basal-cell carcinoma	
Sarcoma	
Leiomyosarcoma	
Malignant fibrous histiocytoma	
Dermatofibrosarcoma protuberans	
Kaposi's sarcoma	
Metastatic malignant disease	
Lymphoma of the vulva	
Merkel-cell cancer	

series of patients and with uneven results.^{1,28,29} Yet the implication of HPV in the BCC pathogenesis has not been proved. Most authors underline the fact that there is a possible significant morbidity and occasional mortality if a lesion such as BCC is neglected or improperly treated.¹⁹

Considering the rates of relapse and possible aggressiveness of BCC, a close long-term follow-up is essential in order to decrease skin cancer morbidity through early detection and treatment.¹⁷

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