

Basal Cell Carcinoma of the Vulva

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ABSTRACT

"The basal cell carcinoma (BCC), a malignant tumor representing 75% of all skin cancers other than melanoma [1], is rare on non-exposed skin such as the perianal and genital regions [2]. We describe a woman with BCC of the vulva treated by partial hemivulvectomy. All excision margins were disease-free. The patient recovered without complications. Approximately 200 cases of BCC on perianal and genital skin have been reported in the literature [3,4]. Although the etiology of vulvar BCC is unknown, early diagnosis is important. As BCC on these sites can sometimes appear innocuous, biopsy of all suspicious lesions is recommended [5,6]."

Keywords

Basal cell carcinoma, Non-exposed skin, Vulvar basal cell carcinoma, Vulvar cancers.

Introduction

Basal cell carcinoma (BCC) is a malignant tumor that represents 75% of all skin cancers other than melanoma. The most frequently involved factor in the pathogenesis of BCC is exposure to ultraviolet rays; BCC has a frequency of over 80% in elderly patients, appearing on sun-exposed areas of the head and neck. BCC is rare on non-exposed skin such as perianal and genital regions, and other etiological factors should be considered in these cases. Approximately 200 cases of BCC on perianal and genital skin have been reported in the literature, and BCC on these sites is estimated to represent less than 1% of all vulvar BCC and less than 5% of vulvar cancers.

A Case Report

A 50-year-old single nulligravida woman was referred to our department for a pigmented nodular, budding lesion on the right labium majus (Figure 1). She reported one year of vulvar discomfort and itching, unsuccessfully treated with a corticosteroid ointment. Her general condition was good, with no history of sexually transmitted diseases, radiation or smoking, and no personal or family history of skin cancer.



Figure 1: Pigmented nodular budding lesion of the right labium majus.

The lesion measured 1.5 x 2 cm (figure 1) and no lymphadenopathy was noted in the inguinal region. The patient underwent a biopsy of the lesion, which showed infiltrating and pigmented basal cell carcinoma on histopathological examination. Subsequently, the patient underwent partial hemivulvectomy (Figures 2 and 3) under spinal anesthesia and she recovered without any complications.



Figure 2: Operative specimen of right hemivulvectomy.



Figure 3: Final result after hemivulvectomy.

Definitive Anatomopathology

The examination showed a downward growth of basaloïd cells with peripheral palisading of tumor islets (figure 4). In situ hybridization did not reveal DNA from human papillomavirus (HPV) types 6, 11, 16, 18, 30, 31, 33, 35, 45, 51 and 52. All excision margins were free of disease. A vulvar BCC was diagnosed.

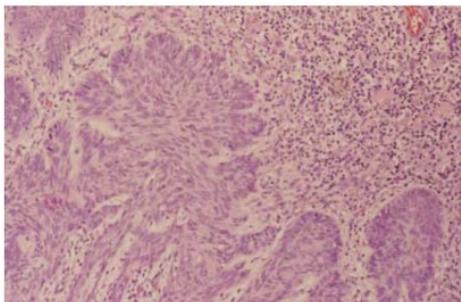


Figure 4: Examen anatopathologique montrant une croissance basaloïde avec une palissade périphérique des îlots tumoraux.

Discussion

Over 250 cases of vulvar BCC have been reported in series [7-19] or case reports [20-25]. BCC represents on average 2 to 3/100 of vulvar cancers. It typically affects white women over the age of 60 [16]. Proposed risk factors include those common to cutaneous

BCC, including chronic exposure to arsenic, basal cell nevus syndrome, and chronic irritation. Ultraviolet (UV) radiation does not play a direct role in the development of vulvar BCC, but UV-induced suppression of immune surveillance at a distance may be involved in the pathogenesis of BCC in non-sun-exposed sites [26]. In these regions, relative systemic immunosuppression induced by UV radiation may act in concert with other factors, such as advanced age and local trauma, to precipitate tumor development. The most common predisposing factor for vulvar BCC is prior radiotherapy; the literature also mentions a delay of several years before vulvar BCC manifests following radiotherapy [16,19].

Although BCCs are generally slow-growing tumors, they are locally invasive and destructive and may recur or metastasize. They tend to grow along the anatomical pathways of least resistance, and invasion of bone, cartilage, and muscle is a late event. Vulvar BCCs are nonspecific and do not suggest BCC: exophytic, ulcerated, pedunculated, infiltrating, nodular, and pigmented lesions have been described. They were located on the non-mucosal surface of the labia majora. Bilateral or multifocal vulvar BCCs are rare. The aggressiveness and recurrence of BCCs vary depending on the histological profile. Morpheiform, metatypical (basosquamous), adenocystic, and infiltrating tumors are more aggressive and tend to recur [27], associated with deep local infiltration and occasional perineural extension. Metastatic BCC is rare, with an incidence of 0.0028 to 0.1%. The average time from initial presentation is 9 years [28].

Aggressive histological patterns appear to be a common denominator in cases of metastatic vulvar BCC. Perineural invasion was noted in one case. The most common site of metastases in these cases was regional lymph nodes. Metastatic lymphadenopathy (subclavian) was reported in only one case. One case presented with cutaneous metastases [29]. The survival of patients with metastatic vulvar BCC cannot be determined from the current literature because long-term follow-up has not been evaluated. A short survival period of 8 to 10 months is expected after diagnosis of metastases in patients with metastatic BCC from other cutaneous sites [30]. Most authors agree that vulvar BCC should be treated surgically. Complete excision with histologically clear margins is sufficient. When surgery is contraindicated or excision is incomplete, radiotherapy is an alternative but often results in local complications. Mohs micrographic surgery has been recommended by some as the method of choice for a large or histologically aggressive BCC [31,32]. Local recurrence, likely due to inadequate margins, is observed in approximately 10 to 20% of cases [6-7]. Although the etiology of vulvar BCC is not known, early diagnosis is important. As BCCs in these sites sometimes appear indolent, a high index of suspicion is necessary to make the diagnosis.

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