

An Unusual Location of Carcinoma: The Clitoris and the Vulva

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Abstract: Vulvar carcinoma is rare, accounting for less than 5% of all vulvar neoplasms and less than 1% of all neoplasms. Vulvar SCCs are usually diagnosed late because they are often asymptomatic and tend to grow at slow rates. They may be invasive and destructive if neglected or improperly treated. Nevertheless, they have a very low propensity for metastatic spread, but frequently recur after simple excision. We report a 45 year-old woman presenting with the complaint of painful vulvar ulceration, small swelling at vulvar region and vaginal bleeding. The physical examination revealed a 3 × 2 cm indurated nodulo-ulcerative lesion involving the clitoris, both labia minora and left labia majora. The histopathology was consistent with the “non keratinizing squamous cell carcinoma” that invaded the subcutaneous tissue without lymph node metastasis on left side and with lymph node metastasis on 2 nodes on the right side. The patient underwent wide local excision with clitoral amputation and remained disease free at post-surgical follow-up after 6 months.

Keywords: squamous cell carcinoma, clitoris, vulva

I. Introduction

Squamous cell carcinoma (SCC) is the common malignancy of the skin, accounting for approximately 70–80% of all cutaneous cancers.¹ The lifetime ultraviolet radiation damage is the most important factor in its pathogenesis, and the vast majority is observed on sun exposed skin, with nearly 85% occurring in the head and neck.² Although SCCs can develop in sun protected areas, genital involvement is very rare, accounting for fewer than 1% of all cases.³ SCC accounts for 90% of all vulvar cancers (Table 1) and occurs most commonly in post-menopausal women.⁴ Since its first description by Temesvary in 1926, 200 cases of vulvar SCC have been listed in the literature.⁵ The etiology of vulvar SCC is unknown. Syphilis, chronic irritation, chronic infection, trauma, arsenicals, and radiotherapy have been implicated as possible precipitating factors.^{5,6} Clinically, vulvar SCC is an indolent and destructive tumor that rarely metastasizes, but the local recurrence rate is as high as 20% in some series.^{5,6}

A.	Squamous-cell cancers (90%)
B.	Nonsquamous cancers (10%)
•	Bartholin's gland carcinoma
•	Malignant melanoma
•	Verrucous carcinoma
•	Paget's disease
•	Basal cell carcinoma
•	Sarcoma
•	Dermatofibrosarcoma protuberans
•	Kaposi's sarcoma
•	Metastatic malignant disease

Table 1 caption a4 Vulvar cancers

In this paper, we report an unusual case of locally invasive BCC located at the clitoris extending to the labia minora and left labium majus of a 45-year-old woman treated with wide local excision.

II. Case Report

A 45-year-old female presented with complaints of painful vulvar ulceration, small swelling at vulvar region and bleeding for at least 4 months. She was a known case of hypertensive since 2 yrs. She did not have a history of an antecedent skin disease or local irritation. She denied any personal or family history of skin cancers or internal malignancies. There was no history of sexually transmitted disease, irradiation or tobacco use. Physical examination revealed indurated ulcerous lesion (3 × 2 cm) with an elevated telangiectatic border on the labium majus (Fig. 1). The left labium minus, the superior portion of the right labium minus, medial part of the left labium majus and clitoris were completely eroded. Inguinal lymph nodes were not palpable and pelvic computed tomography verified the physical examination. The patient underwent wide local excision of the tumor with the amputation of the clitoris. The surgical defect was reconstructed with bilateral advancement skin flaps. The pathologic examination of the excisional biopsy revealed non keratinizing squamous cell carcinoma with the invasion of subcutaneous tissue. All the 12 lymph nodes on left side was free of tumor and out of the 11 lymph nodes on the right side 2 nodes were seen to have tumor cells in it. There has been no tumor recurrence at the post-surgical follow-up after 6 months.

III. Discussion

Vulvar malignancy is a rare malignancy accounting for approximately 2–3% of all neoplasms.^{2,3,5} It usually affects white women over 70 years of age.^{3,6} It usually presents as a nodule or like an ulcer in our case, but it may also have a very nonspecific and indolent clinical appearance. It may mimic other dermatological pathologies such as eczema, psoriasis, seborrheic keratosis or angiokeratoma.² Therefore, it is recommended that all suspicious vulvar lesions should be biopsied for early diagnosis. The previous studies demonstrated that the tumor size ranged between 0.2 and 10 cm and most occurred on the labium majus and less commonly on the labium minus, urethral meatus, prepuce and clitoris.^{5–7} In our case, three parts of the vulva were invaded simultaneously by the tumor. *The etiology of SCC in sun-protected areas remains unknown. The factors other than ultraviolet radiation seem to be involved. The literature suggests that radiotherapy to the pelvic region, chronic pruritus vulvae or ani, chronic vulvovaginitis, previous trauma such as burn or scar, arsenic, certain genetic conditions such as nevoid basal cell carcinoma syndrome and xerodermapigmentosum, immune deficiency, human papillomavirus (HPV) infection (more relevant in squamous cell carcinoma of the genitalia), mutations in the p53 gene and advancing age may all contribute to the development of SCC in these sites.^{2,3,5–7} Although we could not search for the presence of HPV DNA, the advanced age seemed to play a contributor role in our patient. Although local excision is usually curative, recurrence and rare metastases have been reported, particularly in cases of the sclerosing type and those with perineural invasion.^{3,4} Our case had a nodular type of SCC and perineural invasion and lymphatic involvement were not present. Because of the tendency to be locally invasive and recurrent, wide surgical excision or Mons micrographic excision are the recommended therapies for vulvar SCC.^{6–8} One centimeter margins seem to be adequate, particularly at the given age and general condition of the patient.⁴ Selective lymphadenectomy is warranted for the large invasive tumors with lymphatic involvement.^{6,8} In the case of incomplete excision or when surgery is contraindicated, radiation therapy is an alternative but often leads to local complications.^{5,8} Our patient underwent a wide local excision of the tumor with ilioinguinal nodal dissection and reconstruction with advancement skin flaps. At the post-surgical follow-up after 6 months, she is still tumor free. However, because of a high local recurrence rate of vulvar SCCs, close long-term follow-up is necessary.

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