CASE REPORT

Squamous Cell Carcinoma of the vulva: Case report.

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ABSTRACT

Squamous cell carcinoma (SCC) is the most common malignant tumor of the vulva, affecting mainly postmenopausal women. It is classified according to its etiology as human papillomavirus (HPV)-dependent and HPV-independent, both of which originate in the context of a chronic dermatosis. Diagnosis is made by histopathological study.

The case of a 56-year-old patient with a diagnosis of well-differentiated squamous cell carcinoma of the vulva is presented and a review of the subject is made.

INTRODUCTION

Squamous cell carcinoma of the vulva constitutes 90% of all vulvar malignancies and arises from pre-invasive epithelial lesions that present cellular atypia with the possibility of progressing to invasive disease, unified within the concept of vulvar intraepithelial neoplasia (VIN).[1]

About 1/3 of vulvar squamous cell carcinomas are caused by human papillomavirus (HPV), and the usual precursor lesion of this group is the high-grade squamous intraepithelial lesion. However, most are HPV-independent and arise in the context of chronic dermatoses.[2]

CASE PRESENTATION

Female patient of 56 years old, with personal pathological history of arterial hypertension and within the gynecological history she had 7 gestations of which she presented 2 cesarean sections, 1 delivery and 4 abortions, her menopause was at the age of 50 years old. Pap smear and mammography showed normal results. She had no relevant pathological family history.

She went to the Centro Dermatológico Úraga, presenting a lesion in the vulvar area of 1 year and 6 months of evolution, which began as a papule of 0.5 cm in diameter located on the right labium majus of gradual growth, accompanied by pruritus and mild pain, during this period of time the patient was treated with topical ointments without improvement in other medical centers.

Physical examination of the right upper lip showed an erythematous-violaceous infiltrated plaque 5 cm in diameter, an ulcer with irregular borders on the lateral border and another smaller ulcer on the proximal border, accompanied by serohaematic and fetid secretion (Figure 1). Dermoscopy showed an erythematous plaque with scaly areas, pink background, punctate vessels and whitish striae on the surface (Figure 2).

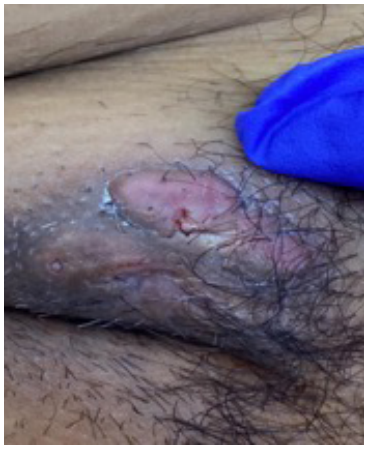

Figure 1. Physical examination revealed an infiltrated erythematous-violaceous plaque, 5 cm in diameter, with a lateral ulcer of irregular borders and another smaller ulcer on the proximal border of the lesion.

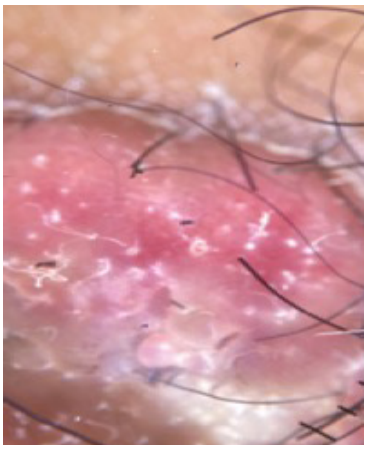

Figure 2. Dermoscopy shows an erythematous plaque with scaly areas, pinkish background, punctate vessels and whitish striae on the surface.

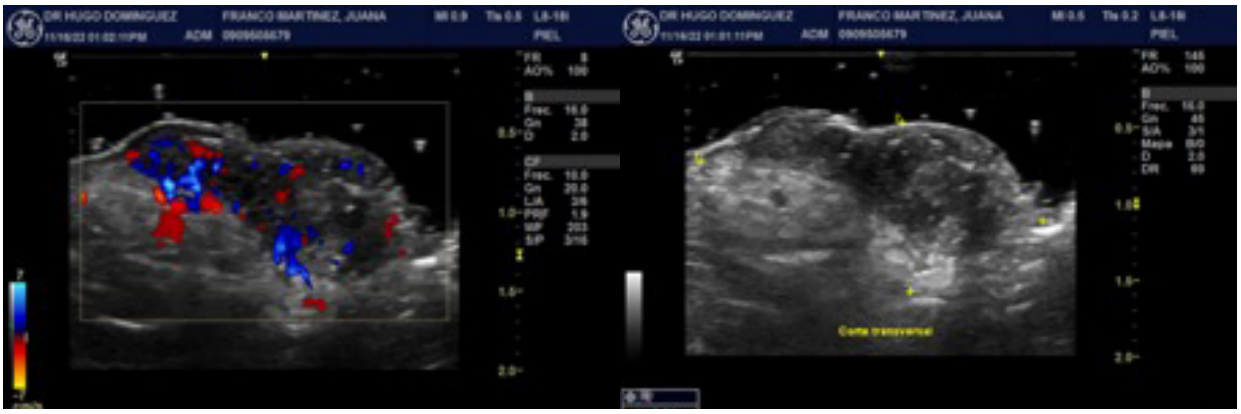

Figure 3A. On ultrasound, a solid, dermal and hypodermal, heterogeneous focal area (hypoechoic in its upper zone, hyperechoic in its lower zone), with slightly defined contours is visualized. 3B. Doppler ultrasound shows a prominent vascularization in its interior.

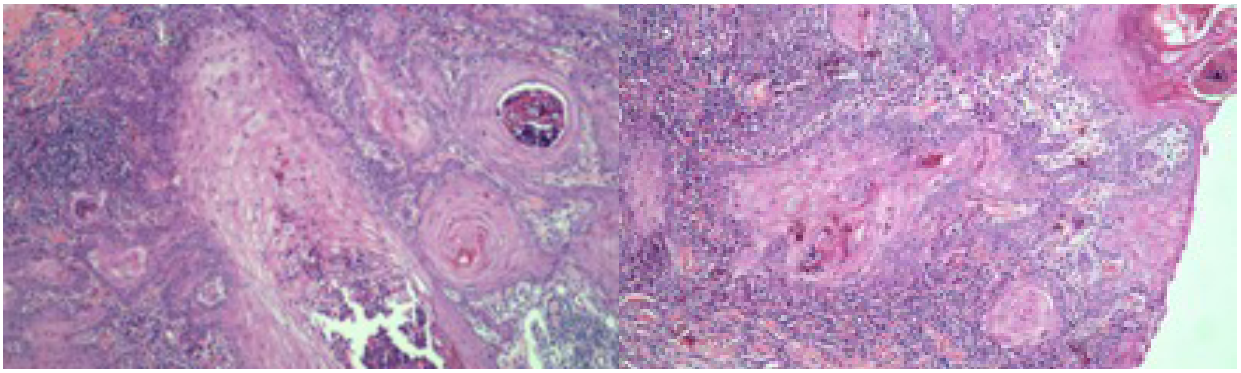

Figure 4. Histopathologic findings demonstrated a dense lymphocytic infiltrate. 4B Nests of keratinocytes with broad eosinophilic cytoplasm, loss of polarity, pleomorphism, and nuclear atypia are observed. Proliferation extends to the deep dermis. Surgical edges involved, compatible with squamous cell carcinoma.

Ultrasound was performed, which reported a solid focal area, dermal and hypodermic, heterogeneous (hypoechoic in its upper zone, hyperechoic in its lower zone), with slightly defined contours. Color Doppler showed a prominent vascularization in its interior (Figure 3). Within the differential diagnosis the following pathologies were considered: Dermatofibrosarcoma Protuberans, Secondary Fistula, Spinocellular Carcinoma (Figure 3).

A biopsy of the lesion was performed and histopathological study revealed keratinocyte nests with large eosinophilic cytoplasm, loss of polarity, pleomorphism, and nuclear atypia, with proliferation extending to the deep dermis. Compromised surgical edges (Figure 4). The histopathologic diagnosis was well-differentiated squamous cell carcinoma invading deep dermis.

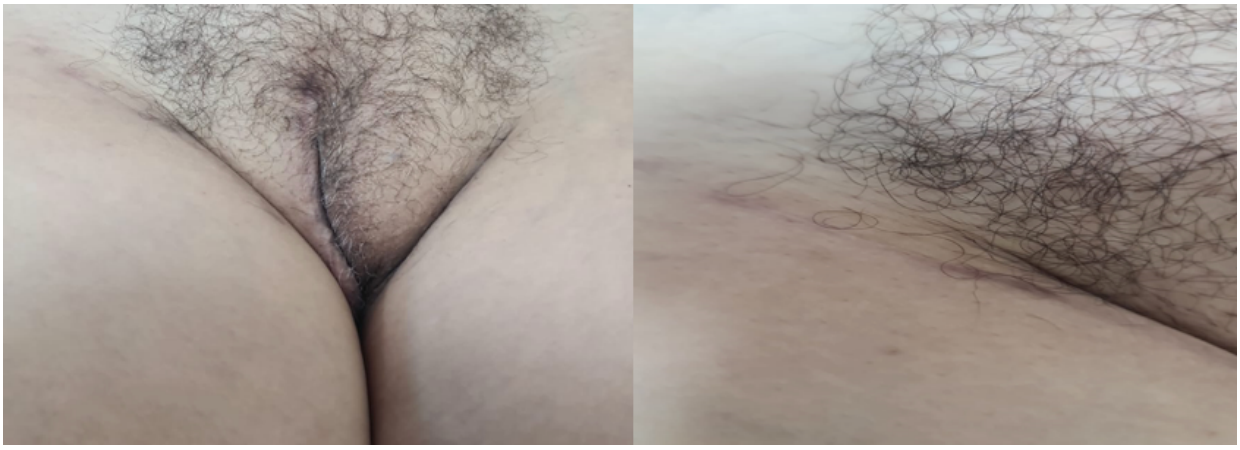

Figura 5A. Postoperative 6-month postoperative right partial vulvectomy with optimal results. 5B. Negative sentinel lymph node postoperative picture.

The patient was referred to the Oncology service for surgical treatment where she underwent right partial vulvectomy, with resection limits free of neoplastic lesion, sentinel lymph node was studied during surgery, marked by negative radioisotopes, for which reason lymphadenectomy was not indicated (Figure 5). The patient was evaluated with the Karnofsky scale being 100% asymptomatic, with no evidence of disease in the first control 6 months after surgical treatment. Follow-up every 6 months for 5 years is proposed.

DISCUSSION

Vulvar cancer is a very rare neoplasm, constituting 5% of all gynecologic cancers.[9] Squamous cell carcinoma (SCC) is the most common malignant tumor of the vulva, mainly affecting postmenopausal women.[3] Of all vulvar malignancies 90% consist of squamous cell carcinoma and arise from the precursor lesion, vulvar intraepithelial neoplasia (VIN).[4] About 1/3 of SCCs are caused by human papillomavirus (HPV), and the precursor lesion of this group is the usual VIN/high-grade squamous intraepithelial lesion (uVIN/HSIL). However, most SCCs are HPV-independent and arise in the context of chronic dermatoses.[2,5] An estimated 44,235 new cases are diagnosed annually worldwide.[6]

HPV-associated SCC usually affects younger women, commonly shows strong block-type staining for p16 on immunohistochemistry (IHC), usually develops from an HPV-induced intraepithelial precursor called a high-grade squamous cell lesion (HSIL), characterized by basal-like immature cells affecting the entire thickness of the epithelium, also known as vulvar intraepithelial neoplasia (VIN) of the usual type.[5] Low-grade squamous intraepithelial lesions represent benign warts or mild atypia with coilocytic change and are often due to low-risk HPV types 6 and 11. High-grade squamous intraepithelial lesions are usually derived from high-risk HPV types 16, 18 and 33 and represent moderate to severe atypia.[7] Clinically, they present near the labia minora as a warty, erythematous or white macule or papule.[8]

In contrast, HPV-independent SCC affects older women, aged 60-80 years, commonly shows mature, well-differentiated keratinizing features, does not overexpress p16, frequently shows an abnormal p53 IHC pattern, and usually arises in the context of chronic inflammatory lesions of the vulvar skin, such as lichen simplex chronicus and lichen sclerosus.[5] Clinically, lesions may appear as focal grayish-white discolorations with a rough surface, vaguely defined thick white plaques, or raised nodules.[8]

Most studies report a progression rate of vulvar intraepithelial neoplasia to carcinoma of around 7%.[1]

The final diagnosis of vulvar squamous cell carcinoma is made by histopathology, distinguishing between well-differentiated forms and poorly differentiated tumors. In HPV-independent well-differentiated SCC histopathology shows acanthosis, parakeratosis, elongation and anastomosis of the intercellular ridges with prominent

intercellular bridges. Cytologic atypia is limited to the basal and parabasal layers of the epithelium and consists of a slightly increased nuclear-to-cytoplasmic ratio, retained eosinophilic cytoplasm and nuclear enlargement with prominent nucleoli. One of the most useful features is related to the phenomenon of "premature differentiation or keratinization".[7,8] Whereas in HPV-dependent poorly differentiated SCC one finds nests of immature basal-type squamous cells with scant cytoplasm. The cells show a basophilic cytoplasm and a high cytoplasmic nuclear ratio, absence of keratin pearls and presence of geographic necrosis or comedo-like necrosis.[3]

The dermoscopic findings in well-differentiated SCC are: presence of multiple lobules, rounded, separated by erythematous septa, few linear irregular vessels. While in poorly differentiated SCC there is altered and 'chaotic' dermoscopic architecture, absence of lobules and septa, linear irregular vessels surrounded by thinner, contoured vessels, severe vascular polymorphism and great variability in vessel diameter.[3]

Treatment options depending on the stage of the disease include local excision, topical imiquimod, cidofovir, 5-fluorouracil, photodynamic therapy, and laser ablation.[8]

The treatment of women with advanced vulvar cancer is complex and should be carried out by a multidisciplinary team.[10] Ideally, the status of the patient's inguinal lymph nodes should be determined first, as this will help to plan her treatment.[11] Radical wide local excision of the vulvar tumor, whenever possible, is the gold standard treatment.[10] The type of surgery is individualized for each patient, depending on the size and location of the tumor.[12] Chemoradiotherapy is recommended when there is anorectal, vaginal or urethral involvement; if the tumor is fixed to the bone; or if macroscopic lymph node involvement is observed.[13]

The prognosis of vulvar SCC is comparatively poor, and treatment is sometimes difficult because approximately 40% of patients with vulvar carcinoma are not diagnosed until their cancer has reached an advanced stage.[14]

The Karnofsky scale measures the functional capacity of the patient with a score ranging from 0 to 100, with 100 being normal without complaints or evidence of the disease and 0 being death, taking into account the patient's ability to perform daily tasks and dependence on medical assistance.[15]

Recurrences of vulvar cancer usually occur in the first 1 to 2 years from initial diagnosis. However, there are several cases of recurrent vulvar SCC occurring 5 years after initial diagnosis, making routine surveillance imperative.[7]

CONCLUSION

Squamous cell carcinoma of the vulva is a rare malignant neoplasm, often underdiagnosed, so a physical examination complemented by dermoscopic and histopathologic findings are important to perform the correct approach and timely treatment, thus improving the quality of life of patients and increasing the survival of those who suffer from it.

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