Verrucous carcinoma of the vulva: diagnosis and treatment*

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Abstract: Vulvar cancer accounts for less than 1% of malignancies in women. Verrucous carcinoma of the vulva is a rare histological variation, comprising less than 1% of vulvar cancer cases. Although it is characterized as being locally invasive, the condition is not associated with metastatic spreading. Lesions present in the form of a verrucous, ulcerated, and bleeding tumor that can reach large dimensions. This type of tumor can be mistaken for condylomata, both macroscopically and microscopically. We report the case of an 81-year-old patient with a large vulvar tumor presented for eight years, initially considered as a Buschke-Löwenstein tumor. The patient underwent radical vulvectomy with a V-Y advancement flap technique. This type of tumor should be considered by clinicians dealing with condylomatous ulcerative lesions that do not respond to the usual treatment.

Keywords: Carcinoma, verrucous; Histology; Surgical procedures, elective; Surgical procedures, operative; Therapeutics; Treatment outcome; Vulvar neoplasms

INTRODUCTION

Vulvar cancer is a rare neoplasia, accounting for less than 1% of malignancies in women and for 3-5% of malignancies of the female genital tract. With an estimated incidence is of 1-2 cases per 100,000 women/year worldwide,[1] the condition affects younger women (15% of cases - mean age of 40 years) and women in the 6th and 7th decades of life (85% of cases). In the first scenario, vulvar cancer is usually related to HPV infection (usual-type VIN – vulvar intraepithelial neoplasia). In the latter case, vulvar lesions develop from VIN exhibiting epithelial atypia and are not related to HPV infection. The most common histological types are squamous cell carcinomas (86%), melanomas (4.8%), sarcomas (2.2%), basal cell carcinoma (1.4%), and adenocarcinomas (1.2%).[3]

Verrucous carcinoma of the vulva is a rare histological variation, comprising less than 1% of vulvar cancer cases. Although it is characterized as being locally invasive, the condition is not associated with metastatic spreading.[4] Lesions present in the form of a verrucous, ulcerated, and bleeding tumor that can reach large dimensions. The major concern regarding this type of tumor is that it can be mistaken for condylomata, both macroscopically and microscopically. Therefore, clinicians are expected to include the adjacent stroma when analyzing the tissue in order to differentiate the tumors.[5] We report the case of a patient with a large-volume vulvar tumor, initially believed to be a Buschke-Löwenstein tumor.

CASE REPORT

An 81-year-old patient sought our services at the gynecologic oncology clinic due to a vulvar lesion present for the last eight years. The lesion, approximately 4cm in size, had appeared eight years before and remained stable without any treatment. Over the past eight months, it started to grow progressively, which was accompanied by pain, bleeding, and a foul odor. During these eight years, the patient sought no medical care.

She has Type II diabetes and currently controled with 850mg metformin 3 times a day. She denied other diseases, smoking habits, alcohol consumption, illicit drug use, or other medication use. She reported menopause onset at age 45 and no use of hormone therapy. She also reported five pregnancies, with five home births.

Physical examination revealed normal condition. The vulvar lesion was approximately 15cm in diameter, sessile, fixed at its 7cm base to the left labia majora, and extending to the clitoris. Lesion surface was ulcerated with bleeding and pain to the touch and signals of local infection (Figure 1). The lesion did not infiltrate the vaginal mucosa, fourchette, or perianal region/anus. The lesion was so extensive that we were unable to perform either a speculum examination or material collection for oncologic cytology. We observed an approximately 2-cm, palpable, fibroelastic, and mobile lymph node in the left inguinal region. Biopsy of the vulvar lesion revealed verrucous squamous cell carcinoma with a 1mm area of microinvasion.

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The patient underwent radical vulvectomy with 2cm margins. We used the V-Y advancement flap technique. Due to the patient's advanced age, only the palpable lymph node in the left inguinal region was excised (Figures 2 and 3). Anatomopathological examination of the surgical specimen confirmed a 15.5x8.0cm grade I verrucous squamous cell carcinoma with a 1.0cm invasion area. Surgical margins were not affected and no vascular or perineural invasion was observed (Figure 4). There was an extensive adjacent skin area affected by lichen sclerosus. There was no sign of metastasis in the removed lymph node. Following FIGO 2009 staging criteria, the tumor was classified as IB (size > 2.0 cm). The patient had no dehiscence or surgical wound infection during the post-operational course. After 4 months, she is disease-free (Figure 5).

DISCUSSION

Verrucous carcinoma was first described in 1945 by Ackerman, who reported an uncommon variant of squamous cell carcinoma. Cases involving not only the oral cavity – the most frequently affected site – but also the skin, male and female external genitalia, cervix, bladder, esophagus, and anal canal have also been reported later on.[6]

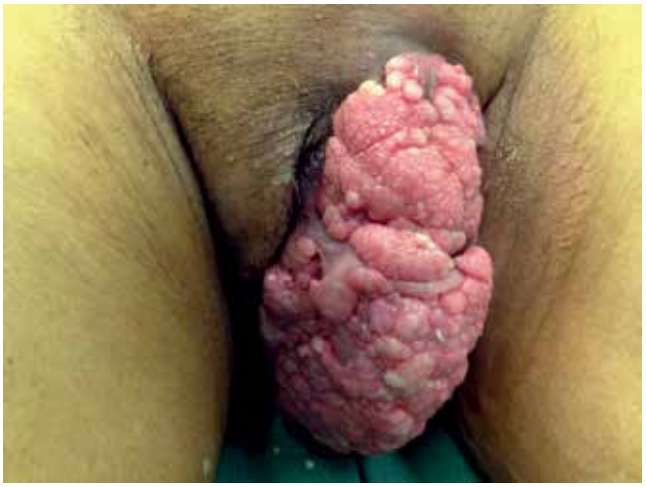

Figure 1: Vulvar lesion with approximately 15cm in diameter, sessile, fixed at its 7cm base to the left labia majora, and extending to the clitoris

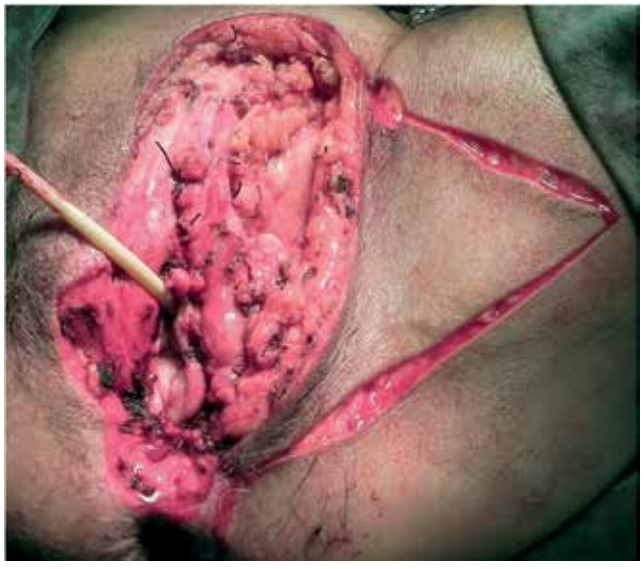

Figure 2: Radical vulvectomy with 2cm margins

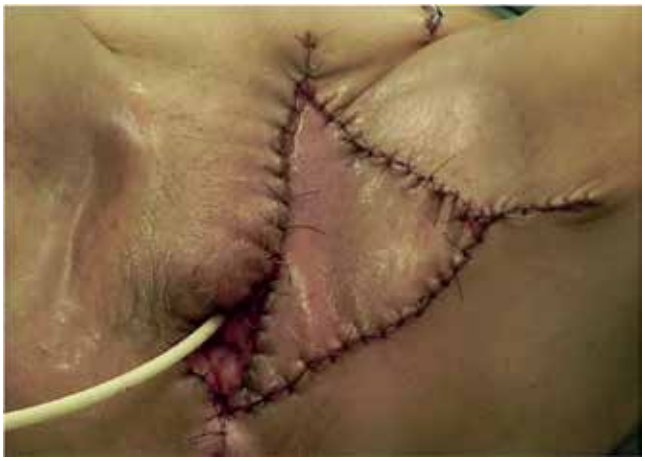

Figure 3: Immediate post-operative care: advancement flap (V-Y technique)

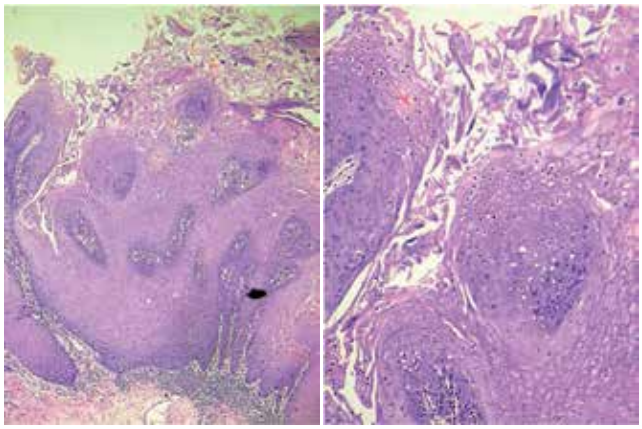

Figure 4: Histological examination - HE staining (x100 and x400) showing characteristic architecture of verrucous carcinoma

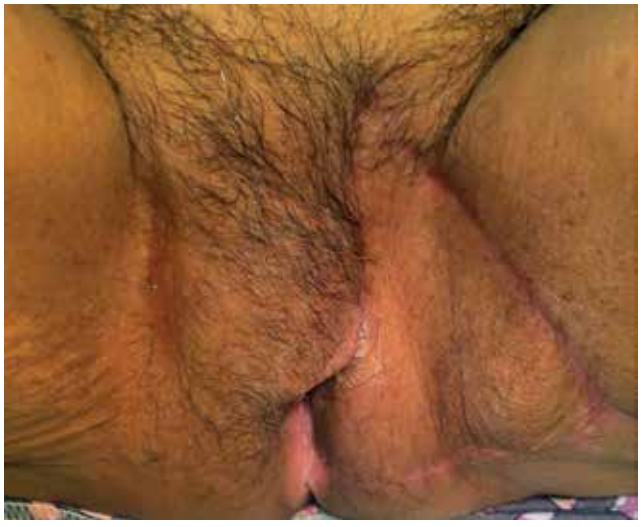

Figure 5: Clinical aspect 4 months after surgery

Verrucous carcinoma of the vulva has a peculiar behavior: it is slow-growing and rarely affects lymph nodes.[7] However, if untreated, it may reach voluminous sizes, which may be mistaken for vulvar condylomata. Therefore, this tumor classification should be kept in mind whenever a patient with condyloma does not respond to topical treatment or in cases of local ulceration.[7,8] Most cases occur in postmenopausal women, who are nevertheless younger than those affected by squamous cell carcinoma. [9]

Histological criteria for diagnosis include tumor-dermal interface with minimal stroma between the acanthotic epithelium, minimal nuclear atypia, hyperkeratotic areas on the surface of the tumor associated with low nuclear atypia, and diffuse and chronic stromal inflammation.[8] Because of this particular biological behavior, extensive biopsies of the lesion are important, including the transition between the tumor and healthy skin in order to facilitate the final histological diagnosis.

Treatment should prioritize surgical procedure, which will depend on lesion size and location (surgical excision, hemivulvectomia, or vulvectomy). A free surgical margin of at least 1cm should be used in order to avoid recurrences.[4] In cases of extensive tissue resection, grafts or flaps may be needed for local reconstruction.

Recent trends in the treatment of vulvar reconstruction demonstrate a common first choice of cutaneous and fascio-cutaneous flaps when a myocutaneous flap is not strictly required by the dimension and depth of the resection. Several flaps have been described for use in the vulvar area but the V–Y flap has received relatively little attention, particularly with regard to the satisfactory results that can be obtained. [10] V–Y flaps represent a valid option for vulvar reconstruction thanks to the simplicity and rapidity in which it is performed.

In the present case, the first diagnostic impression was that the lesion was a giant condyloma (Buschke-Löwenstein), which was discarded after the biopsy. This benign vegetating tumor would have been treated solely with local destruction. Fortunately, the patient developed a verrucous carcinoma, which helped with the differential diagnosis. Despite its extended evolution time, the invasion caused by the lesion remained restricted and there was no lymphatic dissemination. Had she presented with an invasive squamous cell carcinoma instead, there would likely have been extensive local tissue destruction, thus preventing the initial surgical treatment.

Verrucous carcinoma of the vulva is a rare slow-growing, invasive, and locally restricted tumor. Its dissemination through the lymph nodes or metastases are unusual. Clinicians should consider this type of tumor when dealing with condylomatous ulcerative lesions that do not respond to the usual treatment. ❑

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