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Vulvar squamous cell carcinoma in a 21-year-old virgin Case report and literature review

Saloua Krite1, Belhaj Yassine2, F.Z Fdili Aloaoui3, Sophia Jayi3, Hikmat Chaara3, Moulay Abdelah Melhouf3

1. Department of Gynecology and Obstetrics II, Hassan II Teaching Hospital, Fez.

ABSTRACT: *Vulvar cancer is considered as a disease of older women, the typical age is over 70 years. There are not enough reports of vulvar cancer cases in people under 30. These patients typically have risk factors such as human papillomavirus (HPV) infection and immunosuppression. We present here a case of invasive squamous cell cancer of the vulva in a 21 years old patient without any risk factors. Radical local excision of the vulva and bilateral inguinal sentinel lymph node was performed. The clitoris was preserved during the procedure. The patient did not receive adjuvant treatment. Follow-up after 02 months showed no signs of disease. Vulvar carcinoma in very young women can develop without any predisposing factors. Early detection and good examination with biopsy for any suspicious lesion of the vulva even outside the age group will result a better survival.*

Keywords: sentinel lymph node, 21 years old, vulvectomy, Reconstructive surgery

INTRODUCTION:

Vulvar cancer is usually a squamous cell cancer developed in the skin, occurring most often in older women. It usually manifests as a visible or palpable lesion. Diagnosis is based on biopsy. Treatment includes excision and lymph node dissection or sentinel lymph node mapping.

OBSERVATION:

We report the case of a 21 years old patient, still a virgin, without medical or surgical history, admitted for the management of a swelling of the vulva evolving for 6 months preceded by itching, having benefited from a biopsy in favor of a well-differentiated and micro-invasive squamous cell carcinoma, by a private dermatologist, then referred to the gynecology department of the Fez University Hospital for additional support. The gynecological examination carried out by PR MELHOUF revealed: a reddish swelling of 2 cm on the internal surface of the vulva with an eroded plaque just above the tumor classified T1bN0Mx. the pelvic ultrasound was without abnormalities, and the MRI revealed the presence of a 4 cm left vulvar tumor FIGO stage II, which invades the clitoris, the lower third of the vagina, comes into contact with the lower third of the meatus urethral and the right pubo rectalis muscle, without extension to the anal canal or other muscles of the perineum. Inguinal lymph node formations, with a short centimeter axis for the largest on the left, without signs of malignancy.

DISCUSSION:

Vulvar cancers are relatively uncommon, ranking as the nineteenth most common cause of cancer incidence in European women with approximately 16506 new cases in 2020, and affect predominantly elderly women [1]. The vast majority are squamous cell carcinomas. Epidemiologic risk factors associated with vulvar cancer are notably age, human papillomavirus (HPV) infection prevalence, smoking, HIV infection, vulvar intraepithelial neoplasia, and lichen sclerosis. Recently several studies have illustrated the delay in diagnosis that is often the case in patients who are referred with (the suspicion of) vulvar cancer. A study in patients from Germany showed a mean delay of vulvar cancer diagnosis ranging from 186 to 328 days [2]. This was most commonly due to a misdiagnosis of vulvovaginal inflammation. To prevent this delay, women with any vulvar complaints should undergo vulvar examination in a low-threshold manner. Diagnosis of vulvar cancer is made by a punch or incision biopsy of the vulvar lesion. For accurate treatment planning (sentinel lymph node (SLN) procedure yes/no, expected uni or bilateral lymph drainage) the localization of the primary tumor is important. Therefore, excision biopsy should be avoided. In patients with multiple vulvar lesions, all lesions should be biopsied separately to rule out multifocal disease, since patients with multifocal disease are not eligible for the SLN procedure. Because vulvar cancer is a rare disease and the outcome of, for example, the SLN procedure is related to the experience of the treating physician, treatment should be centralized in centers with adequate experience. A European study showed that patients treated in centers with low volume institutions had worse survival rates [3]. In our case the patient was diagnosed early with squamous cell carcinoma and referred to our department for an adequate surgical management, however there was a discordance between clinic and radiology which will be a problem because the technique to use it depends on the stage of the disease. The patient was staffed in multidisciplinary consultation and treated according to the clinical classification and the vulvar cancer standard: **total vulvectomy with sentinel lymph node**.

According to European Society of Gynecological Oncology Guidelines for the Management of Patients with Vulvar Cancer - Update 2023: The vulvar tumor should be removed with a radical local excision. For many years, the primary aim has been to

obtain tumor-free margins of at least 8 mm. large recent studies could not confirm the relation between tumor-free margin distance and incidence of local recurrences. The evidence for the 8 mm margin is very low. The discussion on the optimal tumor-free margin in order to reduce the risk of local recurrences is still ongoing [4] [5], [6], [7] the working group advises aiming for tumor-free margins. A pathological minimal margin of >2–3 mm seems sufficient, but the optimal margin remains to be decided. In order to achieve this, a sufficient surgical excision margin is advised; however, in case of midline tumors close to the clitoris/urethra/anus, this can compromise the margin distance. SLN dissection for early vulvar cancer appears safe, accurate, and cost effective.[8], [9] The prerequisites for SLN dissection are unchanged since the previous guidelines and would support the use of SLN dissection as an alternative to inguinofemoral lymphadenectomy for small (<4 cm), unifocal tumors without clinical suspicion of lymph node metastasis. *Reconstructive surgery*

Availability of reconstructive surgical skills as part of the multidisciplinary team is required in early as well as advanced-stage disease. The type of reconstruction is based on patient/tumor characteristics and experience of the surgical team

CONCLUSION:

We as physicians should not underestimate any vulvar lesion seen, even if the patient falls below the typical age range and does not carry any well-known risk factors such as HPV infection and immunodeficiency so as not to delay the diagnosis and possible treatment in time.

FIG 1 and 2

FIG 3

FIG 4

FIG 5

FIG 6

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Figure 3: drawing to follow for total vulvectomy
Figure 4: total vulvectomy with preservation of neighboring structures including the clitoris.
Figure 5: specimen well spread and oriented

Figure 6: after a month and half post-operation with a small lack of healing due to infection

Authors' contributions:
The authors declare no conflict of interest.
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