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Case Report

Surgical Management of Vulvar Melanoma: A Case Series

Manuel Sánchez-Prieto[a] Francesc Fargas[a] Francesc Tresserra[b]
María González-Cao[c] Sonia Baulies[a] Rafael Fábregas[a]

[a]Department of Obstetrics and Gynecology, Instituto Universitario Dexeus, Barcelona, Spain; [b]Department of Pathology, Instituto Universitario Dexeus, Barcelona, Spain; [c]Translational Cancer Research Unit, Instituto Oncológico Dr Rosell, Dexeus University Hospital, Barcelona, Spain

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Vulvar melanoma · Mucosal melanoma

Abstract
Vulvar malignant melanoma is the second most common subtype of vulvar cancer, accounting for 5–10% of all vulvar cancers. The prognosis is still very poor, although some advances have been achieved in the last years. One of the most significant changes in its management has been the development of less invasive surgical techniques that diminish the risk of postoperative morbidity and long-lasting sequelae. In this article, we review the surgical management of the pathology, based on the comment of 3 cases with vulvar melanoma treated at our institution.

Introduction

Vulvar cancers, although infrequent, are an increasingly serious threat to women's health. Vulvar cancer accounted for 0.3% of all new cancers in the USA in 2019, with 6,070 newly diagnosed cases [1]. Vulvar cancer generally affects women between 65 and 70 years. Vulvar malignant melanoma (VMM) is the second most common subtype of all vulvar cancers, accounting for 5–10% of all cases [2], while squamous cell carcinoma represents up to 90% of all vulvar cancers. VMM is the strain of vulvar cancer with the worst prognosis [3], and it is frequently diagnosed at late stages [4]. Its surgical treatment has evolved from radical vulvectomy to more conservative surgical procedures, such as wide local excision (WLE) [4, 5]. We present here the surgical management of 3 cases of primary VMM treated at our institution.

Correspondence to:
Manuel Sánchez-Prieto, mansan@dexeus.com

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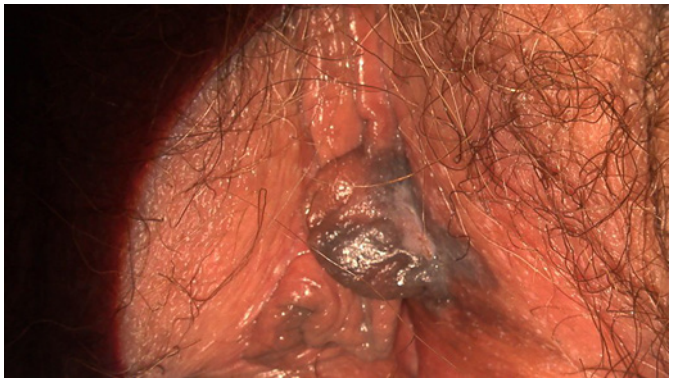

Fig. 1. Vulvar lesion. 2.5-cm irregular pigmented lesion in the upper third of the left lower vulvar lip.

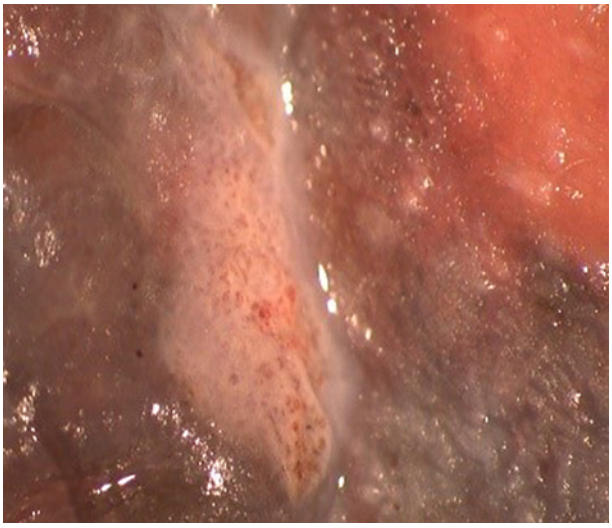

Fig. 2. Raised interior, rough, acetowhite lesion.

Clinical Case 1

A 68-year-old woman consulted because of a left vulvar lesion. As a pathological antecedent of interest, she had presented multifocal motor neuropathy in the left upper limb that was treated with rituximab.

During the gynaecological examination, a 2.5-cm irregular pigmented lesion was observed in the upper third of the left lower lip (Fig. 1) with a raised and rough acetowhite lesion inside of it (Fig. 2). There were no palpable inguinal nodes.

A vulvar biopsy was performed using a punch under local anaesthesia. The biopsy demonstrated a superficial spreading malignant melanoma. The Breslow index was 0.72 mm and Clark's level IV. The PET-CT examination did not show any metastatic lesion. Surgery consisted of vulvectomy with selective sentinel node biopsy technique. Surgery was carried out 5 days later under general anaesthesia with previous technetium 99m (Tc99m) injection. The resection limits were defined, as shown in Figure 3. Intraoperatively, permanent bladder catheterization was performed, and methylene blue was injected at the level of the lesion. A bilateral inguinal incision was made, but sentinel lymph nodes could not be identified because of the absence of Tc 99m or methylene blue, so inguinal lymphadenectomy was performed. Wide excision of the melanocytic lesion was performed, including the clitoral hood, the left lower vulvar lip, and a part of the left greater vulvar lip. The clitoris was subsequently exteriorized at the level of the upper pole. The resection of

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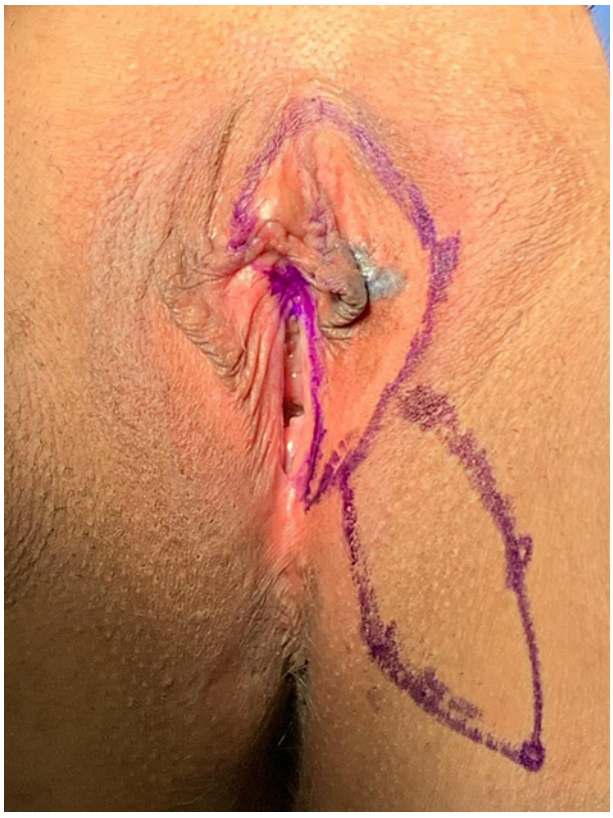

Fig. 3. Previous marking of the resection limits.

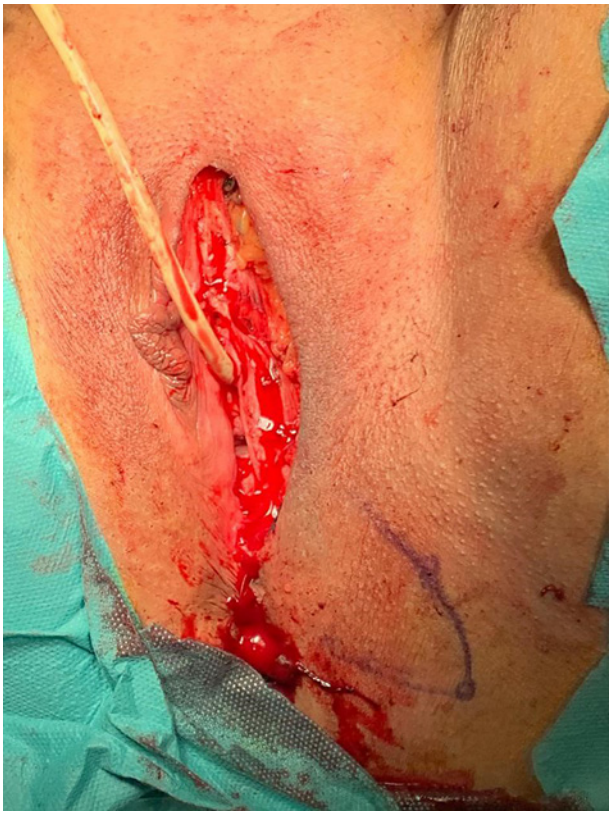

Fig. 4. WLE. Resection of the lesion and repair of the incision. WLE, wide local excision.

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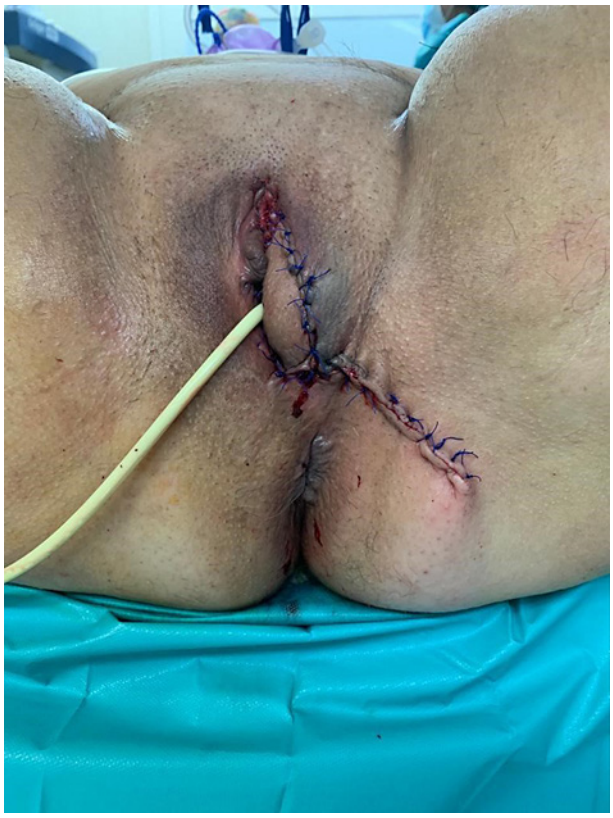

Fig. 5. Gluteal flap for coverage of the vulvar defect.

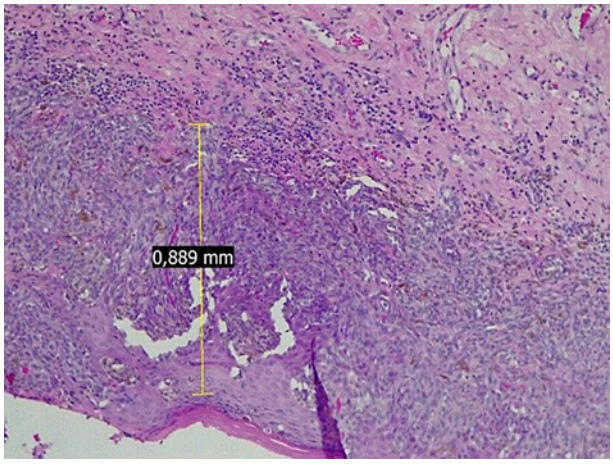

Fig. 6. Histology confirmed a vulvar melanoma of superficial extension with a Breslow of 0.089.

the mass and repair of the incision are shown in Figure 4. Subsequently, the left vulvar lip defect was covered with a left gluteal flap (Fig. 5). The histopathologic study confirmed the diagnosis of vulvar melanoma of superficial extension with a Breslow of 0.89 mm and Clark level IV with ulceration (Fig. 6). The resection margins were reported to be free of lesions. Six left inguinal lymph nodes and 5 right inguinal lymph nodes were obtained, all of which were negative for melanoma infiltration.

The postoperative evolution was satisfactory. No adjuvant therapy was indicated.

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Clinical Case 2

A 60-year-old patient with a history of vulvar atypical melanocytic hyperplasia underwent a vulvar lumpectomy. The patient had no other pathological antecedents of interest.

Five years after vulvar lumpectomy, a right periclitoral melanocytic lesion and a hyperpigmented lesion were observed inside the right labia majora. Both lesions were biopsied and diagnosed as superficial spreading melanoma, with a Breslow index of 0 and a Clark level I. A vulvar lumpectomy was performed. The resection margins were negative for melanoma infiltration. Reconstruction with a zeta-plasty flap was performed. The postoperative evolution was satisfactory. The definitive histopathologic report confirmed the diagnosis of melanoma in situ.

Clinical Case 3

A 64-year-old patient consulted because she noted a vulvar lesion with hyperpigmentation. A 1-cm hyperpigmented lesion was observed in the right lower lip. The lesion was biopsied, confirming the diagnosis of malignant melanoma of superficial extension in situ. Right hemivulvectomy and sentinel node techniques were performed with Tc99m and infiltration of methylene blue in the lesion. A 3-cm right inguinal incision was made, and dissection was performed in planes up to the sentinel node, which was dissected. A sample with 2 secondary lymph nodes was sent for analysis. The resection margins were lesion-free. Lymph nodes had no evidence of metastasis; approximation of the surgical margins of the vulvar bed with 2 double points and skin closure of both surgical wounds.

Discussion

The female genital tract represents a rare location for melanoma (<2% of melanomas, all locations combined). However, the vulvar location is the most frequent site of female genital tract melanoma [2], representing approximately 1% of melanomas [6]. VMM has a poor prognosis and is the vulvar cancer type with the worst prognosis [3], especially in those with regional and distant metastatic disease [6]. Prognostic factors include disease stage, tumour size, patient age, comorbidities, and insurance status [7].

The diagnosis of VMM often takes a long time. This is attributable to patients delaying consultation [8]. Thus, late diagnosis considerably reduces the survival [3].

Given the seriousness of this pathology, increasing patient and clinician knowledge of its symptoms will lead to an earlier diagnosis and improve patient survival. VMM commonly presents as a macula, papule, or nodule >6 mm in length, with irregular borders and coloration [9]. Some patients have itching, bleeding, ulceration, or groin adenopathy [9]. Occasionally, there are amelanotic varieties without pigmentation. In the event of vulvar discomfort and injuries, an adequate pelvic examination should be performed to identify benign or malignant diseases [10]. Any pigmented lesion on the vulva should be biopsied [11] unless it has been present and unchanged for several years. The diagnosis is based on clinical and histopathologic examination.

There is no consensus regarding the adequate staging system for vulvar melanomas [11]. The American Joint Committee on Cancer (AJCC) staging system for cutaneous melanomas is the best applicable system for the classification of VMM, as the main prognosis factor is the deep of melanoma infiltration described according to Breslow index [12, 13].

Treatment of localized VMM is surgical [4, 11]. It consists of WLE with a surgical margin of 1 cm for lesions <2 mm thick and 2 cm for lesions 2 mm thick [14]. The margin may extend to the subcutaneous fascia through subcutaneous fat [4]. As the melanoma generally affects the

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clitoris and labia minora, the resection vagino-urethral margin is a common site of recurrence. If necessary, the distal urethra can be removed to obtain a safety margin of at least 1 cm [14].

Neoadjuvant radiation therapy may be considered to reduce tumour mass in large tumours or tumours in close proximity to vital structures such as the urethra or anus [14].

Sentinel node biopsy (SNL) can help obtain information about regional involvement while avoiding extended pelvic lymph node dissection [15]. SNL is usually performed in cases of squamous vulvar tumours smaller than 4 cm with clinically normal nodes [15]. Extended pelvic lymph node dissection can be avoided in cases of negative SNL in VMM [16]. The tracer applied is Tc99m, to which a vital dye can be added, such as methylene blue [17]. Although the detection rate of SNL by Tc99m and methylene blue is reported in the literature to be approximately 96% [15], in the case presented here, neither Tc99m nor methylene blue was detected. Fluorescent imaging in open groin surgery is an alternative to identify SNL in vulvar cancer using indocyanine green together with Tc99m as a dual tracer in videoendoscopic inguinal lymphadenectomy [18]. Therefore, there is currently a growing interest in hybrid tracers such as indocyanine-99mTc-nanocolloid [19], which reaches a sensitivity of 100% [15]. In addition, the study of SNL should be completed with ultrastaging techniques using immunohistochemistry for cytokeratins in narrow sections [20].

Adjuvant treatments include radiation therapy that it is considered for better local control when there are positive margins or after positive lymph node dissection [4, 21], while adjuvant chemotherapy is not usually indicated because it has not demonstrated survival benefit in VMM [14].

The risk factors for developing a vulvar melanoma are poorly understood. The relation with a previous lichen sclerosis lesion has been previously described [22], but no other risk factors are known. Regarding one of the cases presented in this review, a possible relation with a chronic immunosuppressive treatment could be hypothesized. Rituximab is an anti-CD20 monoclonal antibody with immunomodulatory effects that is used to treat autoimmune disorders as multifocal motor neuropathy [23]. The reduction of CD20 + B lymphocytes located in the melanoma microenvironment could be involved in the development of this melanoma [24]. The influence of rituximab therapy on nevi morphology has been described [25]. However, the latest reviews on this topic conclude that there is no evidence to associate rituximab therapy with an increased risk for developing melanoma specifically or cancer in general [25, 26].

Conclusion

Genital melanomas are rare but aggressive tumours. The diagnosis is usually made by biopsy. The revised AJCC staging system is used to diagnose vulvar melanoma. WLE with adequate margins is the main treatment for early-stage primary VMM. Radiation therapy can be helpful as an adjunctive therapy.

Melanomas of the female lower genital tract must be investigated in more detail as they may be different from other subtypes of melanoma. Given that VMM is an infrequent tumour, and its treatment is complex, management of these cases should be carried out by a multidisciplinary team.

Statement of Ethics

Written informed consent for publication was obtained from the patients for publication of this case report and any accompanying images. The case series report is exempt from ethical committee approval. There is no potential risk to patient privacy.

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Conflict of Interest Statement

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Author Contributions

The manuscript has been prepared following the Instruction to Authors of the Journal and we provide assurance that (a) all the listed authors have participated actively in the conception and design, or analysis and interpretation of data; to (b) drafting the article or revising it critically for important intellectual content; and on (c) final approval of the version to be published and all the authors have revised the manuscript critically.

Availability of Data and Meterial

All data generated or analysed during this study are included in this article and its online suppl. material files; for all online suppl. material, see www.karger.com/doi/10.1159/000517820. Further enquiries can be directed to the corresponding author.

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