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Case Report

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Melanoma of vulva in situ, presentation of a case

Abstract

Melanoma in situ (MIS) usually arises from a junctional nevus. Clinically, the lesion appears dark brown pigmented; sometimes almost black is rare in the vulva and as a relatively slow but definite progression towards invasive melanoma. The clinical case of an 80-year-old patient with multiple comorbidities with a pigmented lesion on the vulva is described, where the histopathological study reported an MIS, and the litter is reviewed for better management.

Keywords: melanoma, vulva, pigmented lesions of the vulva, surgery, staging

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Background

Melanoma of the vulva is the second most common type of vulvar cancer.[1–10] Malignant melanoma develops from melanocytes, these are cells that contain melanosomes, and are found in the basal layer of the epidermis, which synthesize and transform the melanic pigment, with the skin being the most common site (90-91%). it also develops in the mucosa (1.3%) as in the nasal cavity, anus, vagina, and vulva.[1,2] Vulvar melanoma accounts for 3% to 10% of all vulvar neoplasms.[1] It is classified pathologically, in order of incidence, as mucosal lentiginous (27-57%), nodular (22-28%), unclassified (12-16%), amelanotic (6-27%) and superficial dissemination (4-56 %).[2,4–7] Ulcer formations, previous local radiation, human papillomavirus infection, diabetes mellitus, or immunosuppression are all predisposing factors.[6–9] The prognosis for vulvar melanoma is very poor with a 5-year survival ranging from 8–55% compared to 50–80% 5-year survival in other cutaneous melanomas.[1,4] The main prognostic factors include AJCC stage, Breslow thickness, tumor location, tumor thickness, ulceration, clinical melanosis, and lymph node status.[2,4,10]

Melanoma in situ (MIS) can develop directly, although it also usually originates from a junctional nevus. Clinically, the lesion appears highly pigmented in a dark brown color; sometimes almost black. It may be nodular, raised, ulcerated, and bleeding or as a flat, bordering lentigo lesion that may occupy a large area.[11–15]

Melanoma in situ (MIS) is rare in the vulva and as a relatively slow but definite progression towards invasive melanoma.[16,17] The ABCDE scheme for the recognition of melanoma should be considered in pigmented lesions (Asymmetry, Border irregularities, color variation, diameter>6mm, Enlargement or evolution of the change in color, shape or symptoms).[18,19] All lesions or suspicious pigments in this region should be biopsied with punch and it is the preferred method because establishing the depth of such lesions is critical. Destruction by cryosurgery, cautery, or laser is contraindicated, and all these lesions are histopathologically evaluated. Small lesions are often completely excised; when sampling hyperpigmented areas, biopsy is recommended for the thickest region.[20,21] If the diagnosis is considered within the differential diagnosis of pigmented vulvar lesions, it is easy to recognize and treat, with an excellent prognosis.[22] An excisional biopsy of the entire clinically evident lesion, with a narrow 1 to 2mm adjacent margin of normal-appearing skin, is the biopsy technique of choice when melanoma is suspected and shaving biopsies should be avoided. Incisional biopsy is acceptable for larger lesions, studies have shown that there is no worse prognosis if the initial biopsy does not remove the entire lesion, and then it is excised.[23,24] In MIS, proliferating malignant melanocytes are confined to the epidermis. Although the in situ phase exists for three of the four invasive forms of melanoma, superficial spreading melanoma, lentigo maligna melanoma, and acral lentiginous melanoma, it is clinically irrelevant as it must be removed anyway. For patients with MIS, there are no data to define the optimal extent of surgical resection; the data support the routine use of 0.5cm margins.[25,26]

Clinical case

80-year-old rural patient; With a deceased mother due to melanoma diagnosed with vulvar melanoma, she does not report smoking or alcoholism, only exposure to wood smoke in childhood; development Diabetes mellitus 4years ago and has arterial hypertension under control by internal medicine, heart disease with hypertrophy assessed by ecardiography and chronic lung disease, arrhythmias in medical management with moderate cardiovascular risk (with risk that allows surgical management), lumbar spine surgery and antecedent of hysterectomy 20 years ago probably due to benign pathology. The gynecological and obstetric history is menarche at 12years, menopause at 50 years, 6 pregnancies and 6 deliveries, mammography has never been performed.

His condition began when he saw a pigmented lesion on the right vulva, reason for the consultation, and an excisional biopsy of the vulva was performed due to suspicion of melanoma.

The presurgical evaluation by echocardiography presents hypertrophic data, adequate cardiac function, arrhythmia and management with anticoagulation is proposed without ruling out the possibility of intensive postsurgical support; CPOVID19 test that is negative is requested.

The stable clinical data, with a negative 90% O_2 saturation, the lesion in the vulva in the middle third of the right lip (figures) did not affect the urethra, vagina or rectum; macroscopic surgical margins are delimited at 1cm radial (Figure 1), left inguinal region without clinically and ultrasonographically suspicious lymphadenopathy; Sentinel lymph node biopsy is deferred as it does not have a probe range or radiolabeling. A wide excision is performed (Figure 2) with 50cc bleeding ipsilateral inguinal dissection, drainage is placed.

Figure 1 Pigmented lesion on the vulva in the lower third of the labia majora.

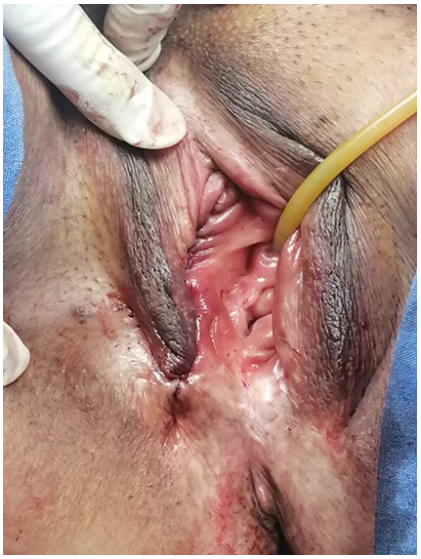

Figure 2 Wide local resection.

Pre, trans and post anesthetic management with intensive monitoring, epidural block and catheter, intravenous sedation and cardiovascular control. He is discharged to recovery and later to the floor, discharge due to improvement with drainage of the inguinal region.

The definitive report with a macroscopic description of a 0.9cm vulva lesion and lesion-free margins. The pathological diagnosis was melanoma in situ of the vulva of 0.9cm level I Clark, Breslow NA with free surgical margins (the closest to 0.4cm on the medial border) without satellite implants or lymphovascular invasion; 8 nodes without evidence of metastasis and with adipose infiltration; surgical specimen, Figure 3.

On the epidermal surface a macular lesion is observed, measuring 0.9 x 0.4cm, it is irregular in shape, well defined, dark brown in color and similar in consistency to the surrounding skin. On section, the dermis and subcutaneous cellular tissue are morphologically normal; the lesion is located 1.4 cm from the upper surgical edge (1 silk), 1.7cm from the lower surgical edge (2 silks), 0.4cm from the medial surgical edge (1 long silk and 1 short silk) and 0.6cm from the edge lateral surgical. The histopathological description of the surgical specimen, Figure 4 and Figure 5.

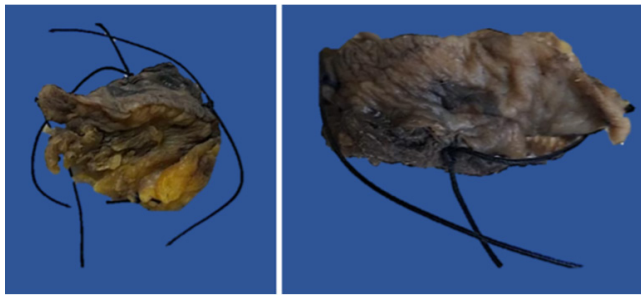

Figure 3 Macroscopic description: melanocytic lesion in the vulva region in the skin spindle, measuring 4.6x1.6cm and 3.0cm deep.

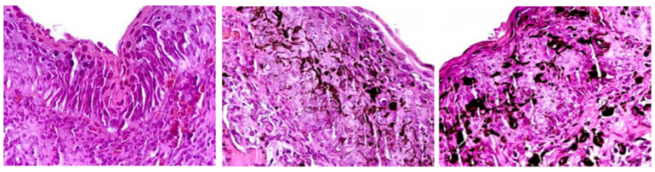

Figure 4 Microscopic description of melanoma in situ.

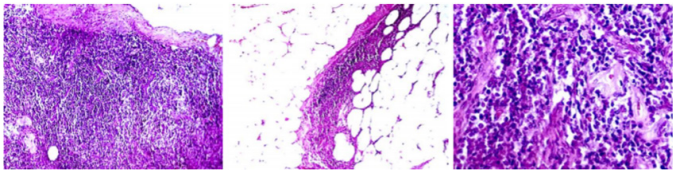

Figure 5 Description of the regional nodes.

Photomicrography (s) of neoplasia in the vulvar region: photomicrography (s) lymphatic nands:

Histopathological Report

Diagnosis: Vulvar Region Skin + Lymphatic Ganglia.

Melanoma In situ of the vulva; Integrity Of The Specimen: Intact.

Neoplasia size: Measures 0.9X0.4cm.

Macroscopic satellite nodules: Not applicable.

Histological type: Melanoma in situ.

Breslow level: Does not apply.

Clark's anatomical level: Level 1 (Melanoma In Situ).

Ulceration: Does not apply.

Microsatellites: Does not apply.

Surgical margins:

Upper surgical edge located 1.4 cm from the neoplasia.

Lower surgical edge located 1.7cm from the neoplasia.

Medial surgical edge located 0.4cm from the neoplasia.

Surgical edge located 0.6cm from the neoplasia.

Surgical bed located 3.0cm from the neoplasia.

Mythosical index: Does not apply.

Lymphovascular invasion: Not identified.

Dissection of 8 lymphatic ganglia, without evidence of malignant neoplastic cells (0/8), with adipose infiltration.

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Pathological stage TNM: pTis, pN0, pMx.

Definitive diagnosis: EC 0 vulvar melanoma (pTis pN0 pM0 AJCC).

During the follow-up, he presented infection and dehiscence of the inguinal surgical wound, dehiscence of the vulvar wound, which was managed conservatively favorably and his clinical control continues.

His current condition began due to autodetection of a pigmented lesion in the right vulva, which is why an incisional biopsy of the vulva was performed.

Discussion

Melanoma is an aggressive skin cancer with an increasing incidence over the last 3 decades, and melanoma in situ accounts for a higher percentage of incidence.[27] Vulvar melanoma represents 6% to 10% of vulvar malignancies[28,29] - 5-year overall survival was 74.4% for vulvar melanoma in situ.

Only 4% of dermatologists included the vulva as part of their whole body skin exam, and 66% considered diagnosing vulvar melanoma part of their clinical practice, compared to 81% of practice by gynecologists. In situ and invasive melanoma show poorer overall survival than cutaneous melanoma.[30,31]

Melanoma in situ is an early non-invasive form in which the tumor is confined to the epidermis; its treatment is challenging due to the frequent subclinical microscopic spread; tumor removal is imperative to reduce the possible progression towards invasiveness and metastasis; there are multiple treatments for management, both surgical and nonsurgical, which raises dilemmas due to its size, anatomical location, and subclinical spread.[32] Melanoma in situ poses special challenges with respect to histopathology, treatment, and clinical management. The minimal mortality and normal life expectancy associated with this should guide its treatment, taking into account the possibility of its transformation to invasive melanoma, it as a significant impact on the morbidity and mortality rate.[33] Melanoma in situ is a radially growing form of melanoma in which the proliferation of malignant cells is limited to the epidermis. Histopathological characteristics are important for its diagnosis. Regression occurs when the host's immune system attacks the cells of the primary melanocytic tumor through lymphocytes infiltrated into the tumor, resulting in a fibrotic component. Several criteria have been proposed to assess the degree of histopathological regression, and some define regression based on the histopathological characteristics of the dermis, which is not appropriate for melanoma in situ.[34]

Melanoma in situ is rare in the vulva and appears to have a relatively slow but definite progression to invasive melanoma. The ABCDE scheme for recognition of melanoma should be considered in pigmented lesions (Asymmetry, Border irregularities, Color variation, Diameter >6mm, Enlargement or Evolution of color change, shape or symptoms). All suspicious pigmented lesions in this region should be biopsied, and a needle biopsy is the preferred method because establishing the depth of such lesions is critical.[35]

Comments

Melanoma in situ is a rare condition in elderly patients, as is our case, and due to the comorbidities they generally present, it should be treated individually. The vulva does not present well-defined papillary dermis in much of its skin and in all the mucosa; Most tumors only spread superficially, there are few publications of cases on the subject, melanoma of the vulva can be subclassified into 3 categories; Table 1 & Table 2.

Table 1 Clark levels

Level	definition
A.	Superficial spreading melanoma
B.	Nodular melanoma
C.	Lentiginous acral melanoma

Table 2 Staging

I.	Melanoma in situ: above the basal mb of the epidermis
II.	Extends to the papillary dermis
III.	Occupies the papillary dermis and extends to the reticular dermis without invading it
IV.	Invades the reticular dermis
V.	Extends to subcutaneous fat

Conclusion

It is a very low frequency tumor in elderly women; diagnosis is usually late. The extension can be either lymphatic (local lymph nodes) or blood (lung, brain, gastrointestinal tract). Local extension and lymph node involvement are the most important prognostic factors for patient survival. Vulvar melanomas are a unique subclass of mucosal melanomas, which are molecularly distinct from cutaneous melanomas with KIT mutations being the most common genetic alteration. A biopsy is crucial for the definitive diagnosis and surgical management in these patients can present complications.

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Conflicts of interest

The authors declare that they have no conflict of interest.

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