

CASE REPORT

A RARE CASE OF PIGMENTED CUTANEOUS MALIGNANT MELANOMA OF THE VULVA

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ABSTRACT

Introduction. Malignant skin melanoma accounts for 1% of all human malignancies. Vulvar melanoma constitutes less than 1% of all melanomas and it is the second most common malignant tumour of the vulva, accounting for 10% of all vulvar malignancies. Usually, it is detected in a later stage compared to other skin melanomas because of its anatomical location and represents a real diagnostic and treatment challenge for dermatologists and gynaecologists.

Case presentation. We present a rare clinical case of a 27-year-old woman with pigmented malignant melanoma affecting the left labium majora.

Conclusions Malignant vulvar melanoma in women under the age of 45 years is rare. It is more aggressive than other types of malignant skin melanomas because of its relatively late diagnosis. Knowledge of the disease will improve the medical care provided and patients' prognosis.

RÉSUMÉ

Un cas rare de mélanome cutané malin pigmentaire de la vulve

Introduction. Le mélanome cutané malin représente 1% de toutes les tumeurs malignes humaines. Le mélanome vulvaire représente moins de 1% de tous les mélanomes. C'est la deuxième tumeur maligne de la vulve la plus fréquente, représentant 10% de toutes les tumeurs malignes vulvaires. Habituellement, il est détecté dans un stade avancé par rapport aux autres mélanomes cutanés en raison de sa localisation anatomique, et représente un véritable défi de diagnostic et de traitement pour les dermatologues et les gynécologues.

Présentation du cas. Nous présentons un cas clinique rare d'une femme de 27 ans avec un mélanome malin pigmentaire affectant le labium majora gauche.

Conclusion. Le mélanome vulvaire malin chez les femmes de moins de 45 ans est très rare. Sa malignité est plus prononcée que dans les autres types de mélanomes

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cutanés malins en raison de son diagnostic relativement tardif. La connaissance de la maladie améliorera les soins médicaux dispensés et le pronostic des patients.

Mots-clés: mélanome, mélanome vulvaire, mélanome pigmentaire.a

INTRODUCTION

Malignant skin melanoma accounts for 1% of all human malignancies¹. Vulvar melanoma constitutes less than 1% of all melanomas and it is the second most common malignant tumour of the vulva, accounting for 10% of all vulvar malignancies². Usually, it is detected in a later stage compared to other skin melanomas, because of its anatomical location and represents a real diagnostic and treatment challenge for dermatologists and gynaecologists.

We present a rare clinical case of a 27-year-old woman with pigmented malignant melanoma affecting the left labium majora.

CASE PRESENTATION

We present a 27-year-old woman without comorbidities, who presented to the dermatologist with complaints of a black, ulcerative, often bleeding, bulging tumour, 1/1 cm sized, in the middle third of the left labia majora. The tumour had developed on a long-standing naevus. A decision for surgical biopsy was made after dermatoscopy. Biopsy was performed several days later by a gynaecologist, who removed the entire tumour, along with 2 mm healthy skin and sent it for histological examination. In addition to the microscopic examination of the haematoxylin-eosin stained preparation, histological test included immunohistochemical staining with antibodies for S100, HMB45, with the following result: nodular pigmented malignant melanoma with ulceration, Breslow thickness 1.24 mm, Clark III, with no vascular invasion.

An Oncology Committee, including an oncologist, a radiotherapist and a gynaecologist, reviewed the case and made a decision for sentinel lymph node biopsy (SLNB) with radical re-excision to be performed and to re-examine the case, reviewing the histological results. The patient was immediately hospitalized in the Department of Gynaecological Oncology, „Dr. Georgi Stranski“ University Hospital Pleven, Bulgaria. The gynaecological examination revealed only a scar on the middle third of the left labia majora, a result of the melanoma biopsy conducted 14 days earlier. The performed blood tests, chest X-Ray and abdominal ultrasound showed no pathological changes.

The patient was referred to the Nuclear Medicine Unit, where ^{99m}Tc-sulphur colloid was injected intradermally around the biopsy scar. Gamma-camera scintigraphy imaging of the regional lymph pool, performed 15 minutes later, determined a small sentinel lymph node, located superficially in the left inguinal region. Node location was marked on the skin with a marker and the patient was directed to the surgery unit. 1 mL of Patent blue V dye was injected intradermally in 10 places, in the biopsy scar area of the left labium majora. Fifteen minutes later, sentinel lymph node biopsy of the left inguinal region was performed, with longitudinal cutting in the skin area marked during scintigraphy. A sentinel lymph node stained in blue was found in the group of superficial inguinal lymph nodes; its location and size matched exactly the lymphoscintigraphic mapping data (Fig. 1). The stained lymph node was removed. A radical re-excision was conducted at the melanoma area of the left labium majora; it was performed within 1 cm radius of the scar from the previous biopsy. The skin and subcutis were surgically removed along with the superficial fascia. Surgical wounds were closed with single sutures after extensive haemostasis and disinfection. The sentinel lymph node and re-excision flap were sent for histological examination, which showed no lymph node metastases and no residual tumour in the skin flap. The Oncology Committee reviewed subsequently the case: the patient was staged IB stage (T_{1b} N₀ M₀) with

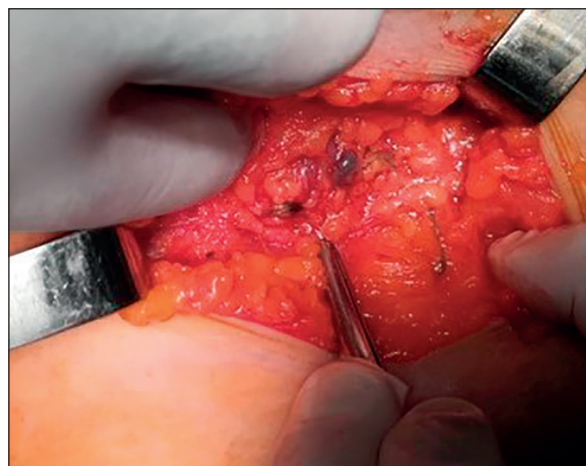


Figure 1. Stained inguinal sentinel lymph node on the left labium.

a decision for clinical observation, without additional treatment. Five years later, the patient is alive and there is no progression of the disease.

DISCUSSION

Malignant melanoma of the female genitalia is rare. Its frequency ratio to other melanomas of the skin is 1:71. Postmenopausal women are most commonly affected³. Our patient was only 27 years old.

The aetiology of vulvar melanoma has been poorly investigated. Its origin is considered to be either *de novo* or from a pre-existing pigmented naevus⁴. Our patient had a 10-year history of a naevus of the left labium majora, that had changed in a period of approx. one month and most likely triggered the malignant melanoma.

Vulvar melanoma is clinically asymptomatic in the early stages, and very often there is anamnestic evidence for a previous presence of a naevus. Ulceration of the tumour, accompanied by bleeding, appears in a later stage. The presence of enlarged superficial inguinal lymph nodes is a late sign, indicating regional metastases. The most common clinical forms of vulvar melanoma are: superficial spreading (40–58%), followed by mucosal lentiginous (27–57%), nodular (22–28%), and unclassified (12–16%) melanomas⁵. Our case presented a variant of nodular melanoma, which makes it even rarer.

The exact diagnosis of malignant vulvar melanoma is made by biopsy removal of the tumour, with 1 – 2 mm margin of healthy surrounding skin, followed by its histopathological examination⁶⁻¹⁴. These narrow margins of melanoma excision are rigorously observed and adhered to, in order to preserve lymphatic drainage from the tumour, which is crucial for the accuracy of a subsequent sentinel node biopsy.

Depending on the presence of melanin in melanoma cells, melanoma types are histologically classified as chromatic and achromatic, the latter having a more aggressive course^{1,15-21}. In our case, the melanoma was chromatic.

Sentinel lymph node biopsy of the regional lymphatic pool is performed after histological verification of the melanoma, for meeting the inclusion criteria. A double-detection method using 99m Tc-sulphocolloid and Patent blue V is applied. Re-excision of the affected area must be conducted regardless a sentinel lymph node biopsy is carried out or not. Excision margins are related to Breslow thickness of the tumour and are usually within 1 – 2 cm radius of the previous biopsy scar. The sentinel lymph node and/or re-excision flap are sent for histological examination. In case of pathoanatomically identified metastases in the sentinel lymph node or clinically detected metastasis to

the regional lymphatic basin (inguinal region), lymph node dissection with removal of all lymph nodes should be executed^{6-14,22-24}. In our case, we followed this protocol precisely, and as there were no sentinel-node metastases, no further surgery was required.

Accurate staging of the disease is essential for the proper treatment of malignant melanoma and is based on the TNM classification. Treatment strategy is determined depending on the stage: whether a particular patient needs adjuvant therapy or monitoring only²⁵⁻²⁸. Our patient was diagnosed with stage IB, which by protocol required monitoring only.

CONCLUSIONS

Malignant vulvar melanoma in women under the age of 45 years is very rare. It is more aggressive than other types of malignant skin melanomas, because of its relatively late diagnosis. Knowledge of the disease will improve the medical care provided and patients' prognosis.

Author Contributions:

M.V.S. and A.K. were responsible for the diagnostic procedures. P.V. and S.S. were responsible for the clinical diagnosis and treatment decisions. S.S. and S.K. performed the surgery. P.V. wrote the manuscript. All authors have read and agreed to the published version of the manuscript.

Compliance with Ethics Requirements:

„The authors declare no conflict of interest regarding this article“

„The authors declare that all the procedures and experiments of this study respect the ethical standards in the Helsinki Declaration of 1975, as revised in 2008(5), as well as the national law. Informed consent was obtained from the patient included in the study“

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