



Case Report

Vulvar melanoma: A diagnostic challenge for young women - a case report

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ARTICLE INFO

Keywords:

Vulvar melanoma
 Immunohistochemicals analysis
 Prevention
 Prognosis
 Self-examination

ABSTRACT

Vulvar melanoma is a rare malignant tumor of the female genital sphere, representing postmenopausal women's prerogative, the diagnosis is based on immunohistochemicals analysis, and treatment requires a multidisciplinary approach. On account of its high metastatic potential as well as the late diagnosis given that it has non-specific clinical signs, the prognosis remains poor.

In this study, we report the case of a woman of childbearing who presented a vaginal mass associated to chronic pelvic pain. Paraclinical investigations revealed a right vulvar tumoral process with pathological-looking inguinal adenomegalies on the right side with a necrotic center measuring 16.7 mm on the short axis, micronodules and secondary pulmonary nodules. The patient has been put under palliative chemotherapy, then passed out 8 months later.

By this work, we attempt to review the diagnostic circumstances to better understand this delay, also to encourage self-examination and self-screening of abnormal lesions, as well as leveling the awareness of health professionals on this rare disease.

1. Introduction

Vulvar melanoma is a rare illness developed from melanocytes and has a high incidence of metastatic spread; representing only 2–9% of vulvar malignancies. It usually arises after menopause in the sixth decade [1]. It has a non-specific clinical appearance, with general symptoms including bleeding, pruritus, and pelvic mass varying in severity.

A histopathological investigation is paramount to confirm the diagnosis. The treatment requires a multidisciplinary consultation; nonetheless, the prognosis is very poor.

We report a case of vulvar melanoma concerning a 49-years-old female patient. Based on a literature review, the diagnostic circumstances, pathogenesis, treatment and its prevention will be reviewed. This case has been reported following the SCARE criteria [16].

2. Case presentation

We report a 49-year-old female (gravida 3, parity 3) with no significant past medical or surgical history, which has showed to our department with chronic pelvic pain, associated to a vaginal tumor.

None of pruritus, bleeding neither fever, tiredness nor weight loss symptoms were present. There was no drug history, toxic habits or allergies. Also, the history of malignancy or a family history of a comparable clinical condition was missing.

Clinically, the patient was afebrile and hemodynamically and respiratory stable, with a blood pressure at 120/70 Hg mm, a heart rate at 80 beats per minute and a respiratory rate at 24 cycle per minute. Her body mass index (BMI) was 20.7 kg/m². A huge, firm, non-pulsatile and irregular mass of the right labia taking on the hard, uneven sidewall of the vagina, measuring approximately 4 cm, was discovered during gynecological examination. The abdominal examination revealed nothing unusual. A painless 1.5 cm right inguinal adenomegaly was discovered during a lymph node examination.

The patient reported a 6 months delay between the appearance of signs and the consultation. By this work, we intend to focus on the necessity of screening all women in the childbearing age, for a better prognosis.

A biopsy of the mass was conducted using Tru-cut® based on the clinical findings. A thinned squamous lining was massively infiltrated by an undifferentiated carcinomatous proliferation, which was made up of solid masses and thickened cords of cells with manifestly atypical,

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hyperchromatic anisokaryotic nuclei richly mitotic and strongly nucleolated, occasionally showing nuclear monstrosities, cytoplasm relatively abundant basophils, and fibro-inflammatory stroma. Anti-protein S100 antibodies (Fig. 1) and anti-Melan A antibodies (Fig. 2) were positive in immunohistochemical staining, whereas anti-CK5/6 antibodies were negative (Fig. 3). The diagnosis of vulvar melanoma was confirmed by these pathological and immunohistochemical findings.

In order to stage the disease, we completed by a cerebral, abdominopelvic and thoracic computed tomography that showed a right vulvar tumoral process, with pathological-looking inguinal adenomegalies on the right side with a necrotic center of 16.7 mm on the short axis, micronodules, and pulmonary nodules of secondary appearance (Fig. 4).

The case was examined by a multidisciplinary committee, which recommended that the patient get 30Gy (3Gy in 10 portions) of palliative analgesia radiation on the pelvis, followed by Dacarbazine-based palliative chemotherapy. The patient died eight months later.

3. Discussion

The vulvar melanoma is a rare and aggressive entity of the female genital tract [1–3]. It represents 2%–9% of vulvar cancers; classed as the second histological type after squamous cell carcinoma with a high risk of metastatic location [4].

Vulvar melanoma mainly affects elderly Caucasian women with a median age of 61.6 years. The most common site of appearance is the labia majora, followed by the labia minora and then the clitoris. Overall, their pathogenesis is still poorly understood.

Vulvar melanoma is a multifactorial disease caused by the involvement of a number of factors, including genetic and environmental ones. The risk factors listed in the literature are: age (risk increases with age) [5], family history of skin melanoma [6], ethnicity (whites have three times as many melanomas as other races) [7], indirect involvement of UV radiation [8], chronic inflammatory diseases like lichen sclerosus, viruses, and irritants [9].

The clinical symptoms of vulvar melanoma are non-specific and might cause a range from asymptomatic to pruritus, vulvar bleeding, nodule, and mass. Here comes the interest for a complete clinical examination of the entire skin covering, as well as the mucous membranes, in particular, the genital tract is indisputable, consequently monitoring, yet biopsy any single or multiple pigmented lesions. It is also important to educate patients on self-examination and consulting immediately as sooner as any atypical pigmented lesions appear, in order to avoid any possible delay in diagnosis.

The clinical evaluation of lesions suspected vulvar melanoma is based on the ABCDE rule; -A-: means Asymmetry, -B-: means usually irregular Border, while nevus has smoother border, -C-: means Color,



Fig. 1. Immunohistochemical analysis showed diffuse strong PS100 positivity.

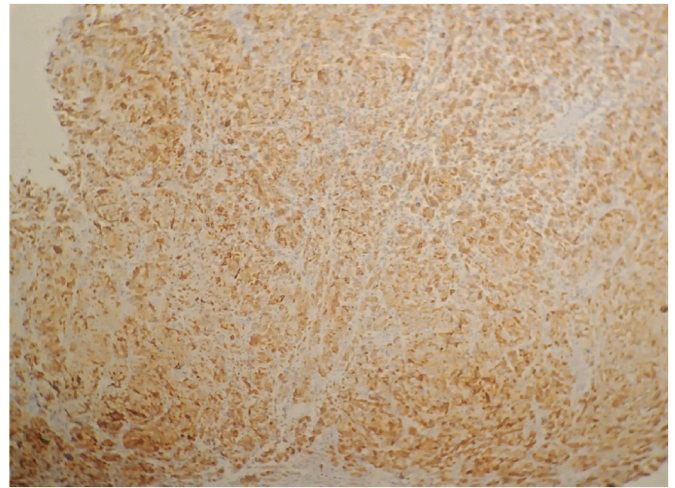


Fig. 2. Immunohistochemical analysis showed diffuse strong anti-Melan A positivity.

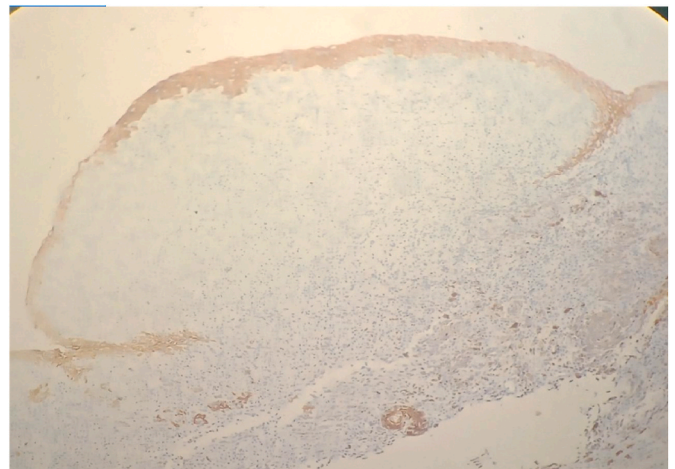


Fig. 3. Immunohistochemical analysis showed diffuse strong anti-CK5/6 negativity.

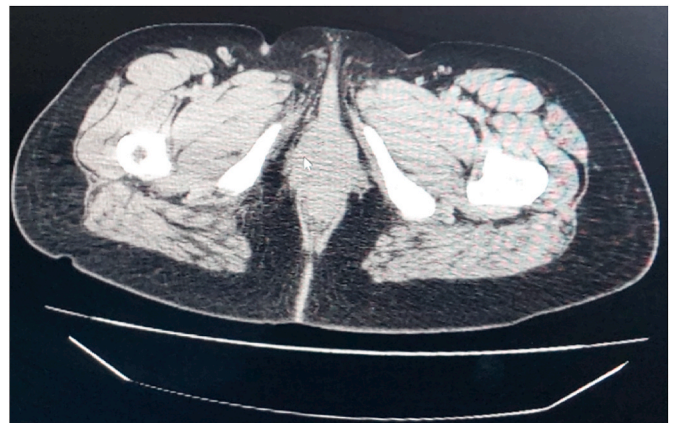


Fig. 4. Pelvic computed tomography scan shows a right vulvar tumoral mass.

multiple colors (brown, black, red, blue, white) are signs of malignancy, while benign moles are often solid brown, -D-: Diameter, -E-: Elevation or evolution and all change in shape, size, structure, color or symptom is a potential indicator of malignancy [10].

The use of dermoscopy lets the early detection of signs of malignancy in melanomas. Although the studies are insufficient, a retrospective study by the International Dermoscopy Society on genital lesions suggested that the presence of a color change (blue, gray, and white) in an area without a structure had a sensitivity of 100% and a specificity of 82.2% for detecting melanoma [11].

The immunohistochemical study is a valuable tool for validating the diagnosis based on positive immunostaining for S100 antibodies and HMB 45 and Melan A antibodies. According to sundry studies, there are several mutations that characterized the genetic profile of vulvar melanomas such as mutations of c-KIT genes. Also, the NRAS mutations and BRAF mutations have been reported in patients with metastatic melanoma. The discovery of these mutations allows using a tyrosine kinase inhibitor (Imatinib mesylate), MEK inhibitors in the event of NRAS gene mutations, or BRAF inhibitors to treat melanomas. The immune checkpoint inhibitors remain an emerging approach in the treatment of vulvar melanoma in patients who express the PD-L1 protein [12].

A local and distant extension assessment is necessary to assess the extent of this condition and plan the therapeutic approach. The staging Cerebral, abdomino-pelvic and thoracic computed tomography scan, lumbo-pelvic MRI. PET scan can be realized in some cases especially looking for lymph node involvement.

The vulvar melanoma's staging is based on the 8th edition of the American Joint Committee of Cancer (AJCC) staging system, associated to the Breslow index, ulceration of tumor, regional or metastatic lymph node involvement, secondary cutaneous or distant lymph nodes, LDH levels, metastatic involvement in the lungs, central nervous system or other sites [13].

The prognosis of vulvar melanoma is very poor with an overall 5-year survival rate of 46.6% versus 92% in cutaneous melanoma [14], depending on the thickness of the tumor (Breslow index), the depth of invasion (Clark index), lymph node status, and ulceration. However, according to several studies, the most important predictive factor is lymph node involvement, which is associated to a 5-year survival of 24% compared to 68.3% in case of negativity [15].

The complete resection with clear microscopic margins without systematic lymph node dissection is the treatment of choice for non metastatic vulvar melanoma. The sentinel technique's node remains a valid option as adjuvant treatment being considered. For the metastatic form, the treatment proposed in the first line is the immunotherapy, treatment with targeted molecular therapies can be used in case of c-KIT, NRAS, BRAF mutation. The failure of previous therapies is an indication of chemotherapy based on dacarbazine, cisplatin, or fotemustine.

Close clinical monitoring is necessary, as well as patient education on self-examination and self-screening for atypical lesions and recurrences.

4. Conclusion

In summary, vulvar melanoma is a rare gynecological melanoma with rich molecular characteristics. The diagnosis is often late. Consequently, screening and systematic examination of the genital sphere by health professionals in search of any pigmentation abnormalities is necessary in order to establish the correct diagnosis of localized forms and organize the right therapeutic plan.

Ethical approval

Approval is not necessary for case report in our locality.

Sources of funding

Nothing to declare.

Author contributions

Dr Omari Mouhsine: Written the article.

Dr Zaimi Adil: Data collection and analysis.

Pr Hadj Kacem Hanane (Radio oncology professor): interpretation of the abdomino pelvic CT of manuscript.

Pr Afqir Said (medical oncology professor): Revision and final approval of the paper.

Registration of research studies

Our paper is a case report; no registration was done for it.

Guarantor

Omari Mouhsine.

Consent

For the purpose of publishing this case report and the associated photographs, the patient's husband's written informed consent was acquired. The Editor-in-Chief of this journal can examine a copy of the written consent upon request.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Declaration of competing interest

Nothing to declare.

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