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Recurrent Vulvar Melanoma: Diagnostic and Therapeutic Challenges – A Case Report

S.BOUMAAZA 1, Fz FDILI ALAOUI 2 , Z. TAZI , Y.BELHAJ , S.JAYI , H. CHAARA and MyAbdeillah MELHOUF

Centre Hospitalier Hassan II, Service de Gynécologie et Obstétrique II, Fès, Maroc

Abstract : *Malignant melanoma is an aggressive and potentially life-threatening tumor that can arise from both cutaneous and mucosal sites. Among these, vulvar melanoma (VM) represents a rare gynecological entity, accounting for approximately 5% of vulvar malignancies and 1% of all melanomas in women. It predominantly affects elderly, mostly Caucasian women. Its association with ultraviolet exposure remains controversial due to the mucosal nature of this location. Recurrence of vulvar melanoma, although the primary disease itself is rare, represents an even more exceptional and challenging clinical situation, owing to the intrinsic aggressiveness of the tumor and the lack of well-established, standardized management guidelines. The diagnosis is primarily based on histopathological examination following biopsy, supported by clinical, dermoscopic, and microscopic features. At the molecular level, vulvar melanoma differs from cutaneous melanoma by a higher frequency of KIT gene mutations, which can be detected through conventional sequencing, next-generation sequencing, or immunohistochemistry. Mutations in BRAF and NRAS genes may also be observed, although less consistently. These molecular alterations represent potential therapeutic targets, paving the way for targeted therapies in selected cases. To date, the management of primary vulvar melanoma relies mainly on surgery, consisting of wide local excision with or without lymph node assessment. However, the role of neoadjuvant and adjuvant therapies remains limited and insufficiently documented due to the lack of randomized controlled trials. The management of recurrent disease, which is relatively frequent in this location, remains controversial, particularly regarding the role of re-excision, radiotherapy, and systemic treatments such as immunotherapy and targeted therapy. We report the case of a 60-year-old woman with a history of left vulvar melanoma staged as pT4bN1a, initially treated with hemivulvectomy and bilateral inguinal lymph node dissection, followed by a single course of adjuvant chemotherapy. The patient was referred to our department for the appearance of a right vulvar mass, revealing an unusual pattern of recurrence and highlighting the rarity of this presentation.*

Keywords

Vulvar melanoma; Recurrent vulvar melanoma; Mucosal melanoma; Case report; Immunotherapy; Targeted therapy

Introduction:

Malignant melanoma is a highly aggressive tumor whose global incidence continues to rise [1,2]. Mucosal melanoma is itself rare, accounting for approximately 1.4% of all melanomas and only 0.03% of all newly diagnosed cancers.

Vulvar melanoma primarily affects postmenopausal women, and its long-term prognosis remains poor due to a high recurrence rate, making early detection and appropriate management particularly crucial.

Dermoscopy is a non-invasive tool that helps distinguish melanomas from other pigmented or non-pigmented skin lesions. Dermoscopic criteria have been described for the diagnosis of vulvar melanosis. However, clinical and dermoscopic differentiation between melanoma and melanosis remains challenging, and biopsy with histopathological examination remains the gold standard for diagnosis [5].

Surgery remains the mainstay of treatment for localized disease, including wide local excision with or without sentinel lymph node evaluation [1,2]. Nevertheless, due to the rarity of this condition, data regarding neoadjuvant and adjuvant therapies remain limited and poorly standardized [1,3].

A key feature of this disease is its high recurrence potential. Indeed, vulvar melanoma exhibits a particularly high recurrence rate, estimated between 40% and 60%, including local, regional, and metastatic recurrences [1,3,4]. This tendency toward recurrence is related to several factors, including the difficulty in achieving adequate surgical margins, the complex anatomy of the vulva, and the intrinsic biological characteristics of mucosal melanomas. The optimal management of recurrences remains a matter of debate, particularly regarding the role of repeat surgery, radiotherapy, immunotherapy, and targeted therapies [2–4].

Thus, vulvar melanoma represents a rare, aggressive entity with a high risk of recurrence, requiring a rigorous diagnostic approach, close follow-up, and multidisciplinary management. The aim of this article is to present a case of recurrent vulvar melanoma and to discuss recent advances in its clinical, molecular, and therapeutic features

Case Presentation:

We report the case of a 60-year-old multiparous woman with a history of treated hyperthyroidism and menopause occurring 8 years earlier. She had been followed for a melanoma of the left vulvar lip diagnosed in 2023. Initial staging classified the tumor as pT4bN1a.

The patient underwent left hemi vulvectomy with bilateral inguinal lymph node dissection, followed by concomitant chemoradiotherapy. Clinical follow-up initially showed a complete response.

The patient was referred to our department for the appearance of new vulvar and cutaneous lesions.

Gynecological examination revealed, on inspection, a left vulvectomy scar with three blackish lesions measuring approximately 1 cm each, with a resectable appearance. On the right vulvar region, a 6 cm bluish mass was observed, located away from the urethral meatus, vaginal fourchette, and rectum. A 2 cm subdermal nodule was also palpable in the lower right vulvar area **(Figure 1).**

Vaginal examination was difficult due to pain but revealed no abnormalities of the cervix or vagina. Rectal examination could not be performed. Examination of lymph node areas revealed a fixed right inguinal lymphadenopathy measuring 2 cm.

Dermatological examination identified several lesions suggestive of tumor dissemination:

- An indurated bluish pigmented tumor involving the right labia majora; dermoscopy showed a blue veil pattern, consistent with progressive melanoma **(Figure 2).**
- Infiltrated blackish papules on the left labia majora.
- Multiple centimeter-sized papules located on the inner aspect of the left thigh and the pubic region, corresponding to satellite cutaneous metastases.
- Diffuse hypopigmented macules suggestive of a paraneoplastic vitiligo-like reaction, sometimes associated with melanoma.
- Multiple seborrheic keratoses and cherry angiomas, with no suspicious features.
- No palmar, plantar, or nail lesions were observed.

Pelvic ultrasound showed a hypotrophic uterus consistent with menopausal status, with homogeneous myometrium and a thin endometrium. Both ovaries were not visualized.

A surgical biopsy of the right vulvar lesion was performed. Histological and immunohistochemical analysis were consistent with nodular melanoma, with a mitotic index of 5 mitoses/mm², without ulceration, vascular emboli, or inflammatory infiltrate. Breslow thickness and Clark level could not be assessed on this biopsy specimen.

Pelvic MRI demonstrated right vulvar thickening with contrast enhancement, along with suspicious right inguinal lymphadenopathy. Incidental hepatorenal polycystic disease was also noted.

Thoraco-abdomino-pelvic CT scan performed for staging (Figure 3) revealed:

- Right vulvar tumoral thickening with nodular infiltration of the surrounding fat.
- Bilateral involvement of the posterior vulvar regions.
- Right inguinal lymphadenopathy.
- Multiple bilateral pulmonary nodules and micronodules, consistent with secondary metastases.
- A presternal subcutaneous tissue nodule suggestive of cutaneous metastasis.

The case was discussed at a multidisciplinary tumor board.

Given the documented locoregional and metastatic progression, the decision was to urgently refer the patient to medical oncology for initiation of systemic therapy

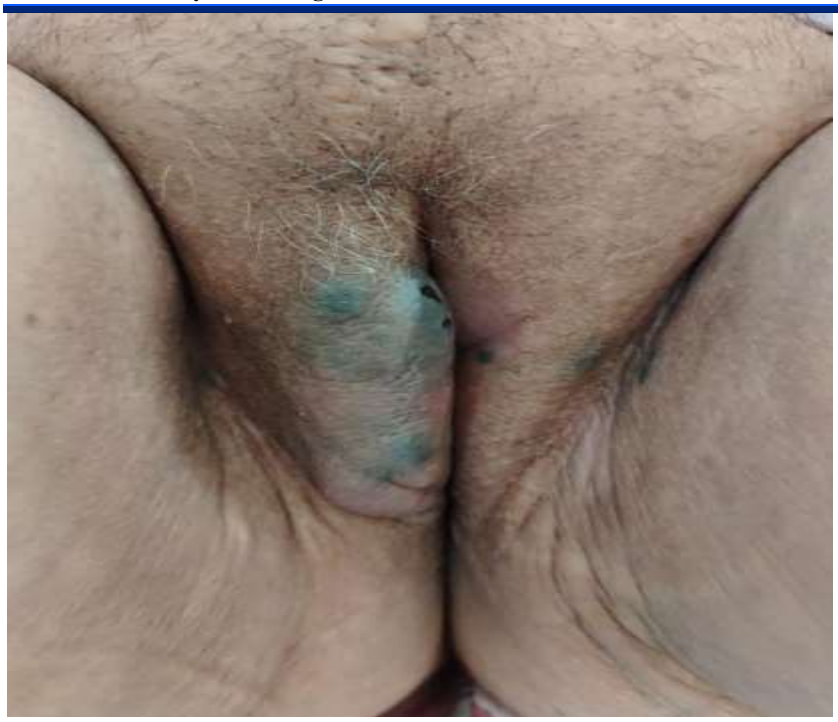

Figure 1 : Bluish pigmented vulvar lesion measuring 6 cm, asymmetric with irregular borders, located in the right vulvar cleft.

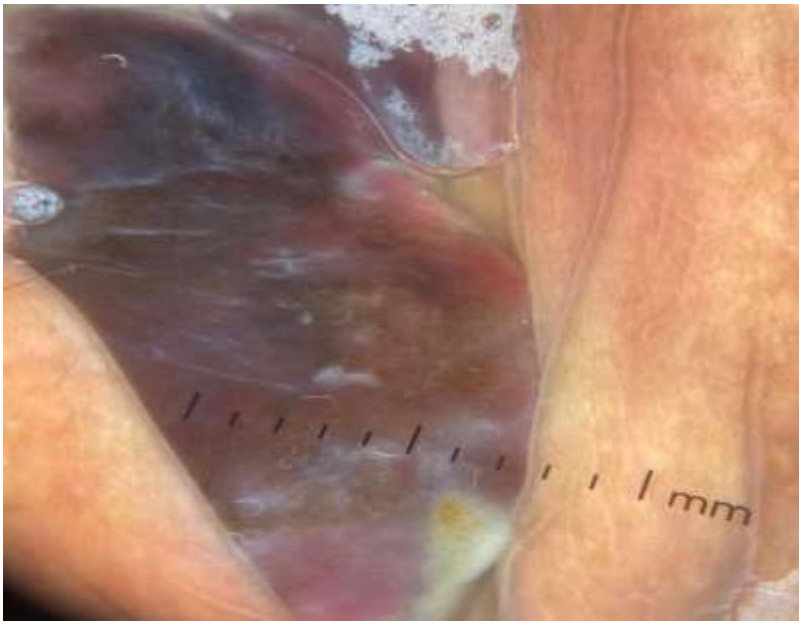

Figure 2 : Dermoscopy revealed a chaotic pattern with structureless pigmented areas, chrysalis-like structures, and linear vessels.

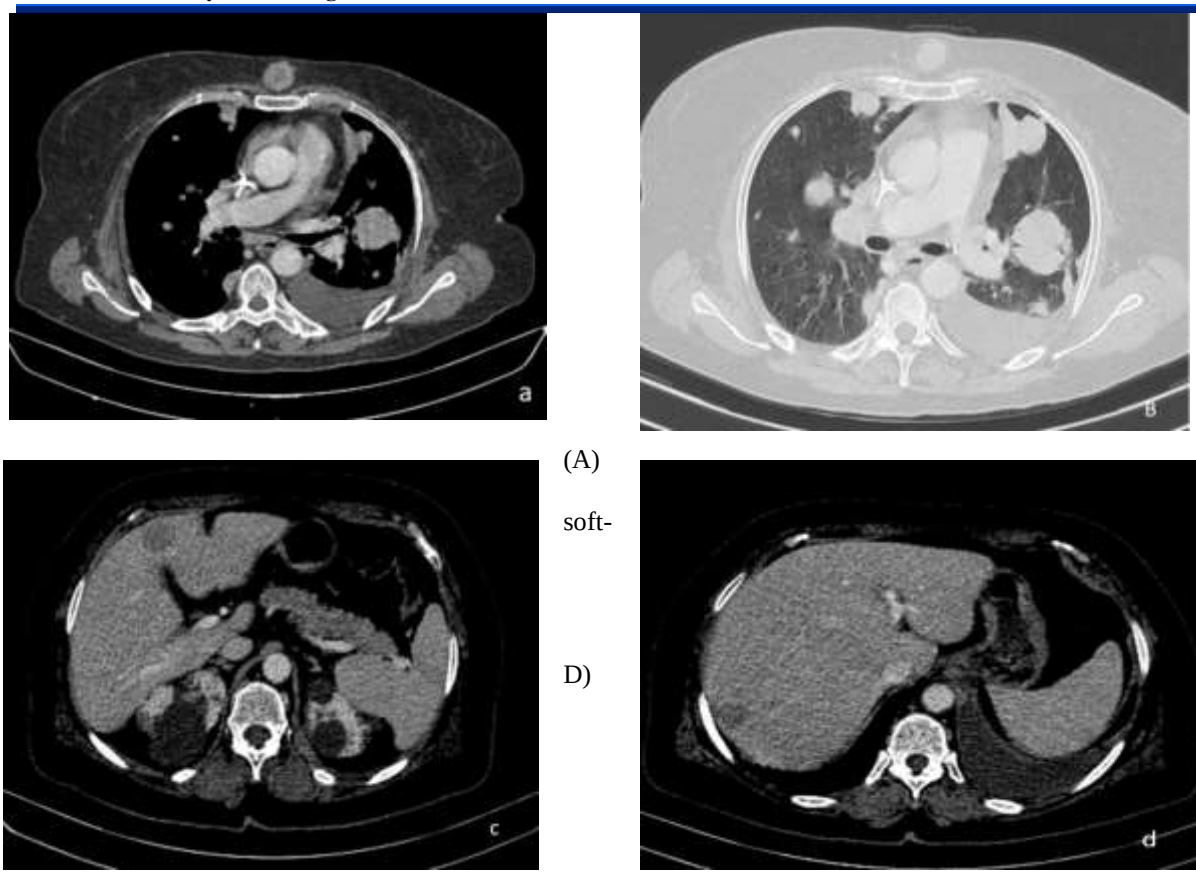

Figure 3 : CT images showing secondary metastatic lesions: (A) presternal subcutaneous soft-tissue nodule; (B) multiple bilateral pulmonary nodules and micronodules; (C, D) hepatic metastatic lesions.

Discussion:

Pigmented genital lesions are relatively common, occurring in approximately 10–12% of the general population [6]. They encompass a heterogeneous spectrum of benign and malignant entities, ranging from melanocytic nevi and vulvar melanosis to, more rarely, premalignant and malignant lesions such as melanoma.

In contrast, vulvar cancer is a rare disease, accounting for less than 1% of all female malignancies and approximately 3–5% of female genital tract cancers. Despite its rarity, it remains clinically significant due to its aggressive behavior, frequent diagnostic delay, and substantial impact on quality of life [7].

Squamous cell carcinoma represents the vast majority of vulvar malignancies (approximately 80%). Basal cell carcinoma and melanoma are considerably less frequent, accounting for approximately 8% and 6% of cases, respectively [8]. Rare tumors—including sarcomas, vulvar Paget disease, and atypical basal cell carcinoma—represent less than 2% of cases [9].

Anatomically, malignant vulvar lesions most frequently involve the labia majora (52%), followed by the labia minora (18%), clitoris (10–15%), and Bartholin's glands (1–3%) [10]. Vulvar melanoma incidence increases with age and predominantly affects postmenopausal women, consistent with the present case.

Vulvar melanoma is a rare malignancy characterized by marked histopathological heterogeneity. The most commonly reported subtypes are mucosal lentiginous (27–57%), nodular (22–28%), unclassified (12–16%), and superficial spreading melanoma (4–56%) [11].

Several predisposing factors have been reported, particularly chronic inflammatory conditions such as lichen sclerosus, which may play a facilitating role in carcinogenesis [12].

The differential diagnosis of vulvar pigmented lesions is broad and includes benign entities (lentigo simplex, vulvar melanosis, acanthosis nigricans, seborrheic keratosis, and melanocytic nevi), premalignant lesions such as pigmented vulvar intraepithelial neoplasia, and malignant tumors including squamous cell carcinoma, basal cell carcinoma, and Paget disease.

Definitive diagnosis relies on histopathological examination. According to the National Comprehensive Cancer Network (NCCN) guidelines, complete excisional biopsy is preferred whenever feasible to allow accurate assessment of Breslow thickness, the most important prognostic factor. Pathological reports should systematically include all established prognostic parameters, and staging must follow the AJCC 8th edition TNM classification [25].

European Society of Gynaecological Oncology (ESGO) guidelines similarly emphasize histological confirmation, precise lesion mapping, and appropriate staging workup tailored to disease extent [26].

Dermoscopically, features suggestive of malignancy include structureless areas combined with blue, gray, or white coloration, which are highly suggestive of melanoma. The presence of polymorphous vascular patterns further strengthens suspicion of malignancy [13].

Early-stage mucosal melanoma is often clinically silent. Without regular gynecological surveillance, progression to invasive vulvovaginal melanoma may occur, leading to symptoms such as pruritus, bleeding, discharge, dyspareunia, or a palpable mass [14]. In the present case, the appearance of a bluish lesion prompted medical consultation.

Histopathological reporting should include tumor size, Breslow depth, ulceration status, histological subtype, perineural and lymphovascular invasion, involvement of adjacent structures, and margin status. Immunohistochemical confirmation is mandatory [15]. Microstaging relies on Clark, Breslow, and Chung classifications, whereas macrostaging follows the AJCC 8th edition system [16,17].

In metastatic disease, molecular testing for BRAF, NRAS, and c-KIT mutations is mandatory and should be performed urgently. In non-metastatic cases, such testing is recommended in high-risk tumors. Whole-exome sequencing may be considered in refractory or therapeutically challenging cases.

Systemic staging must confirm the absence of metastatic disease prior to surgical intervention, particularly when mutilating procedures are considered.

From a therapeutic standpoint, NCCN and ESGO guidelines consistently emphasize surgery as the cornerstone of treatment. Wide local excision is recommended at diagnosis, while radical surgery should be avoided whenever possible, as it does not improve survival and is associated with increased morbidity [25,26].

Evidence indicates that radical surgery does not confer a survival benefit compared with wide local excision with adequate margins in early-stage vulvar melanoma [18], while significantly increasing postoperative complications such as infection, wound dehiscence, sexual dysfunction, and anatomical distortion [19].

Wide local excision remains the standard approach; however, optimal margins are not definitively established. NCCN recommendations suggest margins of 0.5–1 cm for melanoma in situ, 1 cm for invasive melanoma ≤1 mm Breslow thickness, 1–2 cm for 1.01–2 mm, and 2 cm for >2 mm [20]. A minimum depth of resection extending to the fascial plane is recommended. Elective lymphadenectomy is not indicated in the absence of clinically or radiologically involved nodes [15,21]. ESGO guidelines highlight the lack of vulva-specific consensus and advocate individualized surgical margins based on anatomical constraints [26].

Sentinel lymph node biopsy may be considered, particularly when adjuvant therapy is planned. It is indicated in tumors with Breslow thickness >1 mm or <1 mm with ulceration, provided no clinically suspicious lymph nodes are present. A combined technique using dye and radiotracer—or indocyanine green—may be employed via peritumoral intradermal injection.

Evidence regarding neoadjuvant and adjuvant therapies in mucosal melanoma remains limited. A small series of patients treated with biochemotherapy reported inferior survival outcomes compared with untreated patients [22].

No standard treatment exists for recurrent vulvar melanoma. Management should be individualized based on molecular profiling. Immune checkpoint inhibitors (anti–PD-1/PD-L1 and anti–CTLA-4) and targeted therapies (BRAF or KIT inhibitors) represent promising options, although evidence in vulvar melanoma remains limited. Palliative strategies include radiotherapy, electrochemotherapy for cutaneous metastases, and talimogene laherparepvec for unresectable lesions.

Follow-up depends on tumor risk stratification. For low-risk melanoma (Breslow ≤1 mm), clinical examination of the vulva, skin, and lymph nodes is recommended every 6 months for 3 years, then annually. For high-risk disease (Breslow >1 mm and/or nodal involvement), follow-up every 3 months for 3–5 years is recommended, followed by annual surveillance, with periodic lymph node ultrasound. Cross-sectional imaging (pelvic MRI, CT, PET-CT, or brain MRI) may be considered during the first 3 years in high-risk cases [23].

Recurrence is frequent and may be local, regional, or distant, particularly involving the lungs, liver, or brain. It is influenced by tumor biology and anatomical complexity, which may limit complete excision. Most recurrences occur within 2–3 years after initial treatment. Management depends on resectability and includes surgery when feasible, radiotherapy for unresectable disease, and systemic therapy with immune checkpoint inhibitors or, in selected cases, targeted therapy.

Conclusion :

Accurate and prompt diagnosis is essential in vulvar melanoma, a rare malignancy generally associated with a poor and unpredictable prognosis, characterized by a high risk of local recurrence and distant metastasis. Surgical management remains the cornerstone of treatment, and the initial procedure must be adequately performed to minimize the risk of recurrence.

Several prognostic factors influence survival, including tumor size, Breslow thickness, depth of invasion, presence of ulceration, lymph node involvement, and patient age at diagnosis. Postmenopausal women appear to have a slightly higher incidence of vulvar melanoma compared with premenopausal patients.

Overall, outcomes remain poorer than those observed in cutaneous melanoma or vulvar squamous cell carcinoma, largely due to the high propensity for both local and distant recurrence. To avoid delayed diagnosis, dermatologists and gynecologists should maintain a high index of suspicion and perform biopsy of any clinically or dermoscopically suspicious lesion, particularly in older patients.

Further large-scale studies are required to better define clinical and dermoscopic criteria that allow early differentiation between mucosal melanoma and benign vulvar pigmented lesions.

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