

Primary Melanoma of the Vaginal Mucosa: A Case Report

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Abstract

Gynecological melanomas are rare neoplasia affecting the vulva in 4% of cases, the vagina in 2% and the cervix in less than 1%. Melanomas are usually tumors arising from melanocytes, the cells responsible for skin pigmentation, but in some cases they may also develop in mucous surfaces. In addition to being particularly rare, vaginal melanomas are extremely aggressive pathologies with a generally poor prognosis. What's more, these vaginal cancers are often detected late, as melanoma usually presents asymptotically until very advanced stages of the disease. What's more, during a gynecological examination, the speculum can also cover more than 50% of the vaginal walls, masking more pigmented areas.

Keywords: Melanoma Vaginal, Tumour, Melanocytic

Introduction

Melanoma is a malignant cancerous tumor that develops on melanocytes, embryologically derived from the neural crest. Melanocytes are embryologically derived from the neural crest, and because they fail to migrate to the epidermis, they may remain in the vaginal mucosa in an aberrant fashion [1]. Vaginal localization is rare, accounting for less than 5% of vaginal cancers, mainly in post-menopausal women [2]. Mucosal melanomas differ from cutaneous melanomas in that they have a higher mortality rate [3]. This is probably due to the fact that they are often diagnosed late, at locally advanced stages. Vaginal melanoma is therefore a highly aggressive pathology whose surgical treatment is difficult and mutilating.

Through a case study from the Ibno Rochd University

Hospital in Casablanca, we present an update on this rare pathology and its management.

Case report

The patient was 63 years old, married, multigestational, multiparous (6 children born vaginally), with an undocumented history of total hysterectomy without adnexal preservation 6 years ago. She presented to the clinic with abnormal genital bleeding for 2 months, in a context of preserved general condition. Examination revealed a median supra- and subumbilical laparotomy scar, a normal vulva and perineum; speculum examination revealed a clean vaginal slice with a hyperchromic patch on the left vaginal wall (**Figure 1**). Vaginal touch and abdominal palpation revealed no abnormalities.



Figure 1: Speculum examination of vaginal melanoma.

A suspected lesion was biopsied and found to be a malignant vaginal melanoma. A PET-SCAN scan was performed to assess distant extension, and found no suspicious extra-vaginal hypermetabolism. The decision to perform a partial colpectomy was taken at a multidisciplinary consultation meeting. The surgical specimen revealed a primitive melanocytic tumour proliferation in the vaginal mucosa measuring 1.1 x 0.3 cm. No adjuvant treatment, notably chemotherapy or radiotherapy, was indicated. A long-term monitoring plan was drawn up; no recurrence or metastasis was identified.

Discussion

Malignant mucosal melanoma is a rare tumour, accounting for 0.0% of all cancers [1]. It rarely occurs in the female genital mucosa, which accounts for less than 2% of all melanomas [1]. In this case, it can occur in the vagina, vulva and cervix. Melanocytes originate embryologically from the neural crest. During their migration to the epidermis, some melanocytes may remain aberrantly in the vaginal mucosa or in the endocervical canal. This can lead to primary vaginal, cervical or vulvar melanomas [2]. Unlike cutaneous melanomas, no risk factors have been identified.

Analysis of cases of primary vaginal melanoma reported in the literature shows that they most often occur after the age of 50 [3]. Diagnosis is generally made late, in the presence of various non-specific signs: leucorrhoea, metrorrhagia, pain, pruritus, bleeding, palpation of a mass secondary to invasion

of the mucosal epithelium with ulceration and superinfection [2].

Clinically, the tumor may be uni or multifocal, sometimes pigmented. In vaginal melanoma, the tumor is most often found in the lower third (58%), and in the anterior wall (45%) [3].

Immunohistochemical characteristics are similar to those of cutaneous melanomas. The tumour markers used to confirm the diagnosis are the anti-Melan A antibody, the anti-S100 protein antibody and the HMB45 antibody, the latter being more specific to melanoma cells. The use of both antibodies is recommended. More recently, other antibodies more selective of the melanocytic lineage are being used: NK1 /C-3 and NK1BETEB.

Once a melanoma has been diagnosed, the next step is to prove its primary nature. This involves looking for a primary melanoma in the skin, ophthalmology, ENT or digestive tract, or a history of previous excision of a pigmented skin lesion.

As with cutaneous melanomas, surgery is still the only potentially curative treatment for this tumor. Vaginal melanoma is a highly aggressive disease. Overall survival at 5 years is around 20% [4,5]. Surgery is the only potentially curative treatment for this type of melanoma, and the margins depend on the extent and thickness of the lesion, with a maximum of 2 cm [6], which is perfectly consistent with anterior and posterior pelvicotomy, or even exenteration [7].

The lack of published studies on vaginal melanoma, which are limited to a few clinical cases or small retrospective series,



makes it difficult to draw up recommendations to guide treatment. Moreover, exenteration does not increase survival according to published series, as does abdomino-pelvic amputation in anal cancer, leaving it debated by some authors [7,8].

The lymph node status of this type of melanoma is similar to that of cutaneous melanoma [9]. The sentinel lymph node has been shown to have a significant prognostic impact in cutaneous melanoma, and can be superimposed in vaginal melanoma to avoid the need for extensive inguinal and pelvic curage, which is associated with significant morbidity [10,11].

A comprehensive imaging work-up is essential, and should systematically include PET-SCAN, or even thoracoabdominopelvic and cerebral CT scans in melanomas with a poorer prognosis [12]. Surveillance continues to focus on a quarterly clinical examination, with imaging every three to six months for the first five years [5].

No adjuvant therapy has been shown to be effective in vaginal melanoma; immunotherapy, for example, is associated with significant toxicity, without increasing survival [13,14].

Conclusion

Vaginal melanoma remains a rare pathology, diagnosed late and with a poor prognosis due to its rich vascular-lymphatic network. Recurrences are frequent and early, prompting particular caution before proposing radical surgery. Several therapies are currently being evaluated. Recommendations for the management of vaginal melanoma still need to be drawn up, on the basis of several studies yet to be published.

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