Primary Malignant Vaginal Melanoma: A Case Report

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Abstract

Primary malignant vaginal melanoma is a rare tumor. It accounts for less than 5% of all primary malignant tumors of the vagina. It mainly occurs in postmenopausal women. Its diagnosis is delayed due to frequent and early recurrence. So it is important to think carefully before indicating an often heavy first surgery.

We describe a case of vaginal melanoma in a 70-year-old woman. Our aim is to present the clinical diagnosis and the therapeutic aspects of this rare entity.

Keywords: Melanoma, Vagina, Diagnosis, Treatment, Prognosis

Introduction

Primary malignant vaginal melanoma is extremely rare. Melanoma is a malignant tumor that develops from melanocytes. These come embryologically from the neural crest. They can remain aberrantly in the vaginal mucosa during their migration to the epidermis.

Vaginal melanoma is a rare disease of the genital tract. It represents less than 5% of all cancers of the vagina. It most commonly occurs in postmenopausal women. The incidence is about 0.46 cases per one million women per year [1].

Vaginal melanoma differs from cutaneous melanoma clinically and biologically. It is more aggressive with a late diagnosis and a poor prognosis [2].

This letter is designed to describe the clinical features of this rare disease and the way it is managed. The case is reported with a brief review of the literature.

Case Report

A 70-year-old woman, presented with the complaint of occasional slight vaginal bleeding for 3 months. On vaginal examination, there were three lesions of 1.5 cm at the anterior vaginal fundus, 1 cm at the right vaginal wall and 4.5 cm at the left vaginal wall flush the vulva. These tumors were brown budding, with superficial ulceration. The cervix was macroscopically normal (Figures 1 and 2).

CT scan revealed an irregular 28×20 mm mass in the left vagina without locoregional extension in inguinal region. Histologically, the biopsy confirmed the diagnosis of vaginal melanoma. The patient received palliative chemotherapy.

Discussion

Vaginal melanoma is an aggressive pathology. The average rate of recurrence-free survival, median overall survival and overall survival at 5 years are respectively close to 12 months, 20 months and 20% [3].

Malignant vaginal melanoma occurs mostly in postmenopausal women. The most common symptoms are recurrent vaginal bleeding or discharge of recent onset owing to superficial ulceration of the mass [4].

Histologically, the mucosal melanoma is different from cutaneous melanoma. The absence of epidermal component, the presence of ulceration and the lack of pigment makes the diagnosis difficult in some cases.

Immunohistochemistry, antibody anti S100 protein and HMB45 antibody were positive [5].

The thickness of the melanoma according to (Breslow) [6] and Clark's level [7] of invasion are difficult to consider because of the absence of the dermis and the presence of papillary and reticular...
architecture. Similarly, ulceration, due to the fragility of the vaginal mucosa was less indicative.

The American Joint Committee on Cancer (AJCC) revised in 2009 focuses mainly on Breslow and ulceration but it doesn’t take into account the tumor size which for many authors is a prognostic factor with a threshold value to 3 cm [8].

Surgery is the only potentially curative treatment of melanoma. Surgical margins are set according to the thickness of the lesion up to 2 cm if Breslow is above 1 mm. The data reported in the literature including rare series are insufficient to establish recommendations about such melanoma vaginal margins that are needed to achieve anterior and posterior pelvic exenteration.

Several authors advocate radical surgery up to the pelvic exenteration to improve local control and patient survival. Other authors find no benefit survival.

The low survival of vaginal melanoma justifies such a heavy surgery only in cases where PET scan is negative and unable to set healthy margins during the course of conservative treatment [5].

In the case of anterior or posterior pelvic exenteration, a reconstruction can be considered. It is preferable to emphasize on pedicled musculo-cutaneous shreds. However; complications are not to be ignored and profit loss report must be studied.

The lymph node status is an independent prognostic factor. Survival was significantly decreased with a median of 7.8 months against 30 months in patients without lymph damage.

The sentinel lymph node biopsy can avoid extensive inguinal and pelvic lymphadenectomy. Preoperative lymphoscintigraphy would make it possible to know the mapping drainage of the tumor. It always fails to respect the conventional inguinal drainage for tumors of the lower third of the vagina and the pelvic one for those located in the upper two-thirds.

Before considering a surgery, the clinical imaging is essential. The diagnosis of vaginal melanoma is often late with a higher probability of distant metastasis.

The sensitivity of PET examination for the diagnosis of distant metastases is higher than the CT and MRI. However, its impact on survival is hardly evident. Its main interest is to avoid major surgery if there is a possibility.

Thoraco abdominal pelvic and brain scanner are recommended for melanomas with poor prognosis. Surveillance is based on a quarterly check up associated with a complementary imaging (CT or PET scan) every three to six months during the first five years [9].

Melanoma is historically considered to be radio resistant but it seems that the radiotherapy associated with conservative or radical surgical treatment reduces the risk of local recurrence but it doesn’t have an effect on overall survival. Also, systemic adjuvant therapy is disappointing [3].

In case of local recurrence, surgical treatment is important; but for patients who are unable or unwilling to have surgery, clinical trials, including isolated pelvic perfusion are currently evaluated.

Metastatic Melanoma treatment relies on chemotherapy with modest response rates (less than 10%) without evidence of lengthening survival.

Currently, the development of targeted therapies anti BRAF or anti KIT and immunotherapy with CTLA4 antibody, ipilimumab has changed the therapeutic management of metastatic melanoma [10].

Conclusion

In conclusion, vaginal melanoma is a rare entity with late diagnosis and a poor prognosis. It is associated with high rate of recurrence. Therefore, we should be careful when considering an invasive surgery. In case of local or metastatic recurrences, several therapeutics are still being evaluated.

References