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Case Report

Primary Vaginal Melanoma-A Rare Case Report Venkatraman J¹, Govindaraj T², Rathna S³

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Abstract: Primary vaginal malignant melanoma is a very rare, but very aggressive tumor. We describe a case of primary vaginal amelanotic melanoma. A 68 years postmenopausal female patient presented with painless mass coming out from the vagina with history of bleeding on and off for the past 3 months. On vaginal examination there was a polypoidal growth of size 4 cm attached to the anterior wall of vagina. Histopathology of the mass showed features of malignant amelanotic melanoma. Wide local excision was done and adjuvant therapy was given. However patient developed widespread metastasis and died 6 months after the initial diagnosis. The overall prognosis is very poor despite the treatment modality because most cases are diagnosed at a late stage.

Keywords: Melanoma, Amelanotic, HMB45

INTRODUCTION

Vaginal malignant melanoma is a rare form of non-cutaneous melanoma, and accounts for only 1.6% of female genital tract melanoma cases. Furthermore, it accounts for 2.4%-2.8% of all vaginal cancers and 0.3%-0.8% of all malignant melanomas [1] and is the second most common female genital tract melanoma. Most vaginal melanoma patients are postmenopausal, and their primary complaints tend to include vaginal bleeding and vaginal discharge and they are often only diagnosed at an advanced stage [2]. The diagnosis of malignant melanoma is easily made if melanin pigment is present. A small but important group of cutaneous melanomas can be classified as unusual variants. Many unusual variants have of these a distinct histopathological appearance; they include desmoplastic melanomas, neurotropic melanomas, and pedunculated melanomas, metastatic melanomas, and amelanotic melanomas, melanomas arising within a benign naevus, regressing melanomas, and balloon cell melanomas [3]. One large retrospective study reported that 50(1.8%) of 2881 patients with melanoma had an amelanotic primary or metastatic melanoma [3, 4]. Amelanotic melanomas can be easily misdiagnosed as carcinomas or sarcomas because of the minimal number of melanin granules. We present a case of amelanotic melanoma of the vagina, which was initially suspected to be a nonepithelial malignant tumour, but was subsequently

diagnosed by HMB-45 and S-100 protein immunohistochemistry.

CASE REPORT

A 68 year old (gravida 4, para 3) woman presented with the complaints of yellowish discharge and vaginal bleeding. On vaginal examination, a light grey to brownish polypoidal mass with superficial ulceration was found involving the upper third of the anterior vaginal wall. The polyp was not involving the cervix. The polyp was removed and sent for histopathological examination.

Histological examination of the biopsy specimen revealed a polypoidal mass covered by vaginal surface epithelium and the sub epithelium showed a tumor mass composed of diffuse infiltration of large pleomorphic and spindle shaped tumour cells arranged in solid sheets and in nested pattern (Figure 1). The tumour cells had large oval or pleomorphic hyper chromatic nuclei with a distinct nucleoli and eosinophilic cytoplasm (Figure 2). Few cells showed clear cytoplasm (Figure 1). Immunohistochemistry revealed that the tumour cells were positive for HMB-45 antibody and S-100 protein which confirmed the diagnosis of an amelanotic melanoma (Figure 3).

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DISCUSSION

Malignant melanomas are generally found in areas of skin exposed to the sun, but can also be present in non exposed sites, such as genital tract and esophagus [5]. Lower genital tract melanoma arises from melanocytic cells forming the complete spectrum of melanocytic lesions, from benign lentiginous to blue nevi to melanoma [6]. The incidence of cutaneous melanoma is increasing in the west at a greater rate than any other human cancer in the United States [7, 8]. Primary vaginal melanoma is less frequent than other melanomas of the female genital tract in both Europe and America [14, 15]. In contrast, Ikegaya et al reported that approximately 52% of female genital tract melanomas in Japan are primary vaginal melanomas due to the differences in incidence caused by the race [16].

The usual presentation of a vaginal melanoma is a polypoid exophytic mass with red, brown, grey, black, or blue in color [9], or a colorless in the case of amelanotic melanomas, which constitute up to 55% of cases at this anatomic site [10], with vaginal bleeding [11, 9, 12, 13]. Age range varies from 20 to 78 years [9], being more common between 60 and 70 years [11]. Norris and Taylor criteria are used [17] to distinguish whether it is a primary malignant melanoma of the lower genital tract [9, 10, 12, 17]: (a) presence of melanin in the cervical epithelium; (b) absence of melanoma in another site of the body; (c) presence of binding activity in the cervical epithelium near the lesion; (d) if metastatic disease is found, it should be according to the cervical carcinoma pattern.

Malignant melanoma is easily diagnosed by conventional histochemical staining; however, amelanotic melanoma, which is a unique variant of malignant melanoma, can be misdiagnosed as a carcinoma or sarcoma because of the lack of pigmentation. It has been recently reported that immunohistochemical staining with HMB-45 is useful for the cytological and histological diagnosis of amelanotic melanoma [18, 19]. Currently, there is no general recommendation for the treatment of primary vaginal melanoma [20-22]. Melanoma is frequently considered a radio-resistant tumor, and most reports favor surgery over primary radiotherapy [21]. Surgery has been associated with better clinical outcomes compared to chemotherapy approaches.



Fig 1: Tumors cells with clear cytoplasm arranged in nested pattern



Fig 2: Individual tumors cells with pleomorphic nuclei and eosinophilic cytoplasm arranged in solid sheets.



Fig 1 : Tumors cells showing positivity for s100

CONCLUSION

Primary vaginal malignant melanoma of amelanotic type is a rare disease with a poor prognosis, especially if it is not detected in early stage or if it is not treated properly. It is mandatory to do immunohistochemistry markers study to confirm amelanotic melanoma as it can mimic other epithelial tumors. To date, no specific treatment modality has been established concerning the treatment of primary melanoma of the lower genital tract, but it is recommended that this be surgical, procuring the establishment of 2-cm margins, accompanied by radioor chemotherapy. Therefore, it is necessary to initiate collaborative studies and followup of patients in establishing a therapeutic approach that may offer the best results to these unfortunate patients.

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