

Malignant Melanoma of Vagina: A Report and Review of Literature

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About the Author



Dr. Sangeeta Pankaj is currently working as an Associate Professor at the Regional Cancer Centre, IGIMS, Patna, and her areas of interest include Gynaecological Oncology, teaching, and research work. She has been regularly doing screening for cervical and breast cancers. She has dedicated her services solely to prevent and alleviate sufferings of women with Gynaecologic Malignancies

Introduction

Melanoma is one of the most serious forms of skin cancer. Melanomas may also arise from the mucosal epithelium lining the respiratory, alimentary, and genitourinary tracts. It accounts for less than 1 % of all malignant melanoma and less than 3 % of all primary malignant tumors of the

vagina [1]. These melanomas are called mucosal melanomas, they are rare, and account for approximately 1 % of all melanomas [2]. Patients with mucosal melanomas usually belong to the older age group, and these types of melanomas occur more commonly in females compared to males, mainly due to the development of disease in the female genital tract. Melanomas arising from the female urogenital tract occur primarily in the vulva and vagina (95 and 3 %, respectively) [2]. Malignant melanoma of vagina is a rare and very aggressive tumor with an incidence of only 0.46 case per one million women per year [1] and with only around 250–300 cases reported in the literature to date. There are no significant differences, between various racial or ethnic groups [3, 4]. Vaginal primary malignant melanoma (VPMM) most commonly occurs in postmenopausal women [5, 6]. The mean age at diagnosis is 57 years [5, 6]. This is an aggressive tumor, and has a poor prognosis with 5-year-survival rate of 5–25 % only. The optimal treatment modality for vaginal melanoma is still a subject of debate.

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Case Report

An 80-year-old female presented with the chief complaints of feeling of mass in vagina, urinary hesitancy, weight loss, and severe pain in the lower abdomen radiating to her back. All these complaints had started only a couple of months earlier and were rapidly progressive, except that she had a history of uterovaginal prolapse for last 23 years. She had been in good health before these complaints started and had had no previous medical or surgical illnesses.

On examination bilateral multiple inguinal lymph nodes were palpable with the largest node on the right side measuring 7×4 cm and on the left 1×2 cm. On local examination, there was a 3×4 cm \times 3 cm hyper-pigmented growth involving the lower part of the anterior vaginal wall encasing the urethral meatus and extending to the clitoris and anterior two-thirds of labia minora with tenderness (Fig. 1). Third-degree uterovaginal prolapse was seen, and cervix and the rest of vagina were healthy. Rest of the pelvic examinations were normal, and no other palpable adnexal pathology was found. Excision biopsy was taken from the mass for histopathological examination, and FNAC was taken from the inguinal lymph nodes and sent for cytology. FNAC from inguinal lymph nodes show large number of melanocytes with large and prominent nucleoli, numerous melanophages, and intracellular and extracellular melanin pigments (Fig. 2). Microscopic sections of the biopsy from vagina show melanocytes invading the submucosal connective tissue of the vagina. There is the presence of both intracellular and extracellular melanin pigments (Fig. 3).

CT Scan revealed multiple, enlarged inguinal lymph nodes, secondaries in the liver, and a normal pelvic study. The treatment was started on concurrent chemoradiation. She received 50 Gy in 25 fractions over 5 weeks, and 2 doses of gemcitabine 200 mg weekly. Patient's condition was rapidly deteriorating despite chemotherapy and radiotherapy, and she refused to take further treatment and was lost to follow-up.

Discussion

VPMM occurs most commonly in postmenopausal women. The mean age at diagnosis is 57 years [5, 6]. The exact pathogenesis of VPMM is unknown, and it probably arises from melanocytes located aberrantly in vaginal epithelium. These melanocytes can be found in the basal layer of vaginal epithelium in 3 % of healthy women. It is thought that active junctional changes are of the initial stage in the development of malignant melanoma. Although VPMM may arise anywhere, its incidence is more common in the



Fig. 1 Vaginal Melanoma

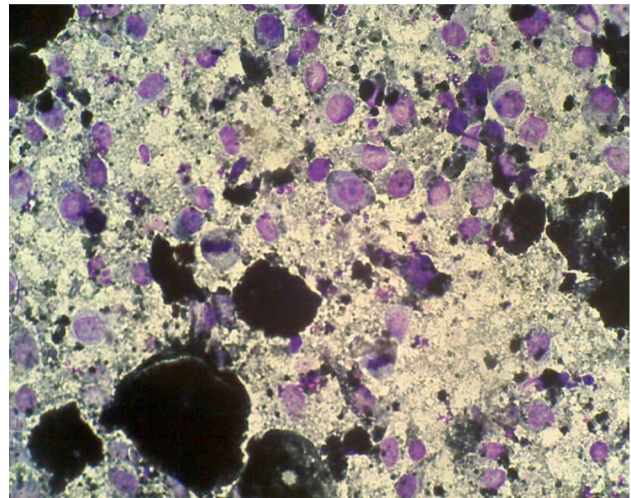


Fig. 2 FNAC from inguinal lymph node

lower one-third (34 %) and the anterior (38 %) wall of vagina. These tumors can be single or multiple, pigmented or nonpigmented. The common presenting complaints are vaginal bleeding, vaginal discharge, palpable vaginal mass, and pain. Our patient is an 80-year-old postmenopausal female with the complaints of vaginal mass and severe pain. The differential diagnoses include metastasis from other sites, poorly differentiated squamous cell carcinoma, sarcoma, lymphoma, and blue nevus.

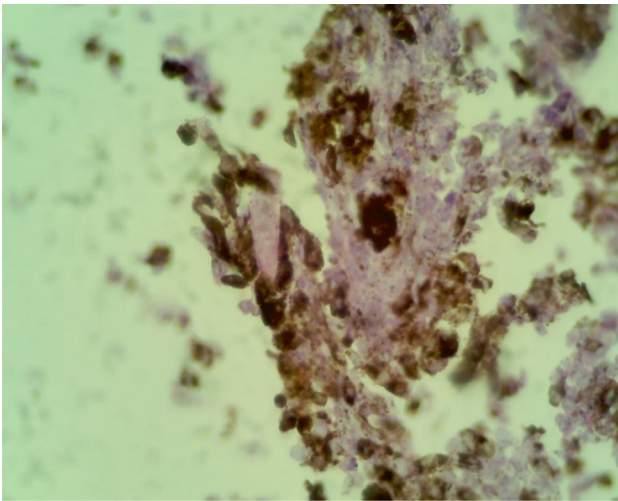


Fig. 3 HPE of biopsy from vagina

The most appropriate treatment for vaginal melanoma is still a debated subject. Some authors have found no difference in the overall 5-year-survival rates among conservative surgery, radical surgery, radiation, and chemotherapy. Surgery seemed to play the most important role in treatment providing local control of the tumor in a better way than that resulting from radiotherapy.

Achieving negative margins at surgery in these cases of VPMM can be difficult, and they require pelvic exenteration due to multifocality and anatomic constraints. No evidence, regarding the best surgical technique, is available so far. Therefore, surgery might be combined with radiotherapy or chemotherapy or chemoradiation in select cases. A number of adjuvant chemotherapy regimens have been tested alone and in combinations to reduce recurrence rate in high-risk melanoma; none of the agents has proven beneficial in randomized clinical trials. The most promising results have been reported with Interferon alpha ($IFN\alpha$), and this has become the standard of care for patients with resected node-positive cutaneous melanoma. Due to the small number of cases of VPMM, the benefits of cytotoxic chemotherapy, such as $IFN\alpha$, IL-2, ipilimumab, and vemurafenib in these cases, have not been completely defined. Because of the extreme rarity of vaginal melanoma, a clinical trial is unfortunately unlikely. There are little data

available on treatment modality of vaginal melanoma. Our patient presented with secondaries and was put on concurrent chemoradiation.

Despite aggressive therapy, the prognosis of vaginal melanoma is very poor because of the high incidence of local recurrence and regional or distant metastasis. Patients with vaginal melanoma have the 5-year-survival rate of 13–19 % only. Fifty percent of patients have positive lymph nodes, and nearly 20 % of patients have distant metastases at disease presentation. This may be explained by the extensive lymphatic and vascular supply to the lamina propria of the vaginal mucous membranes. Our patient also presented with multiple inguinal lymph nodes and liver secondaries.

Compliance with Ethical Standards

Conflict of interest All the authors declare that they have no conflict of interest and they have not received any grant.

Human and animal consent This article does not contain any studies with human or animal subjects, it is a case report.

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