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Primary Malignant Melanoma of the Vagina: A Rare Case Report

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Abstract: *Primary malignant melanoma of thevagina is a very rare but aggressive tumor with poor prognosis and accounts for less than 1% of all malignant melanoma and less than 3% of all primary malignant tumors of the vagina. A 71 year old post menopausal female patient presented with complaints of dark colored discharge per vagina and difficulty in passing stool for last 3 months. Pelvic examination revealed a lobulated, raised, partly ulcerated and irregular vaginal growth involving upto the lower third of vagina and urethral orifice. Histopathology confirmed malignant melanoma with IHC marker HMB-45 and S-100 positive. Patient was treated with radiotherapy. However, the overall prognosis is very poor despite the treatment modality as most cases are diagnosed at a late stage.*

Keywords: *Primary, Malignant melanoma, Vagina*

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I. Introduction

Vaginal malignant melanoma is a rare form of non-cutaneous melanoma[1]. It accounts for less than 1% of all malignant melanoma and less than 3% of all primary malignant tumors of the vagina[2]. It is thought that VPMM arises from melanocytes present in the vaginal epithelium as a result of metaplasia or misplacement of mesodermal and epithelial tissue[3]. The disease presents poor prognosis even in the case of localized lesions, whereas lymph node and distant metastasis have been linked to reduced chance of survival[4]. It presents as a blue‐ black soft polypoidal ulcerated mass[5]. It affects menopausal women with an average age of 55 years[6]. The optimal treatment modality for vaginal melanoma is still a subject of debate[7].Although there is no effective treatment strategy yet in place, the current treatment plan includes wide local excision (WLE), radical surgery, chemotherapy, immunotherapy, combination therapeutics and palliative care. Radiotherapy is used as adjuvant therapy, as data suggest that radiotherapy does not bring overall benefit as sole therapy. Evidence from case reports suggests that the overall 5-year survival rate of patients with primary malignant vaginal melanoma is 0-25%, irrespective of the type of treatment followed[4.]

II. Case Report

A 71 year old post menopausal female presented to our Department with complaints of dark colored discharge per vagina and difficulty in passing stool for last 3 months. Her past and familial history were unremarkable. On pelvic examination, there was a lobulated, raised, part ulcerated and irregular vaginal growth involving upto the lower third of vagina and urethral orifice. There were no palpable lymph nodes and the rest of pelvic and physical examination was normal.CT scan of whole abdomen showed ill-defined fat plane at cervix and upper vagina suggestive of carcinoma cervix. Further histopathological examination of the biopsy specimen from vaginal growth proved the growth as malignant melanoma. Immunohistochemistry revealed that the tumoral cells were positive forS-100 and HMB-45. Her routine investigations including hemogram, renal and hepatic functions, x-ray chest, ultrasonography of the abdomen and pelvis were well within normal limits. A final diagnosis of primary malignant melanoma ofvagina was made based on histopathological and immunohistochemistry reports. Patient was treated with External Beam Radiation Therapy using Theratron 780C to the pelvis by AP:PA field to a total dose of 50Gy in 20 fractions 5 days a week for 4 weeks. Patient is having partial response with symptom relief and is under follow up for the last one year.

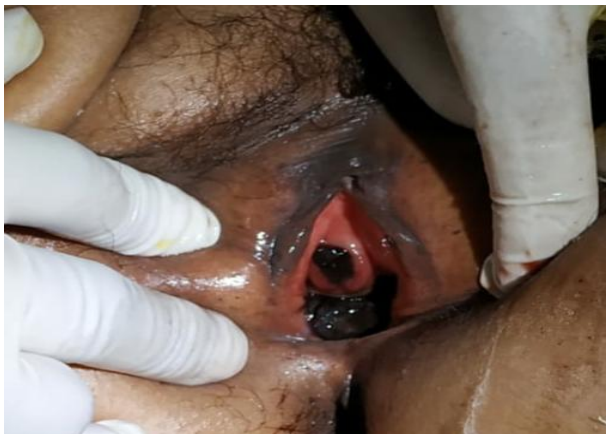

Fig 1: Blackish pigmentation of vaginal mucosa

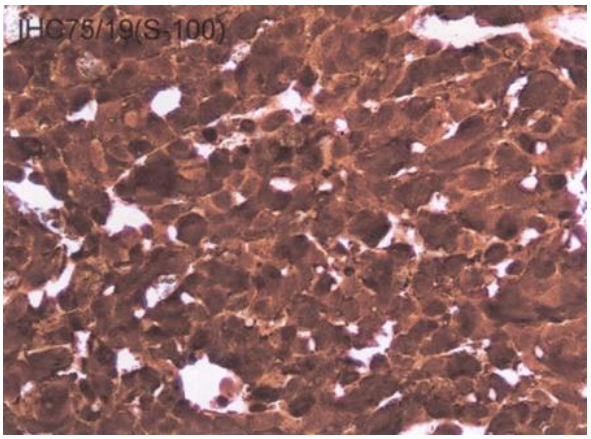

Fig 3: Malignant melanoma positive for S-100.

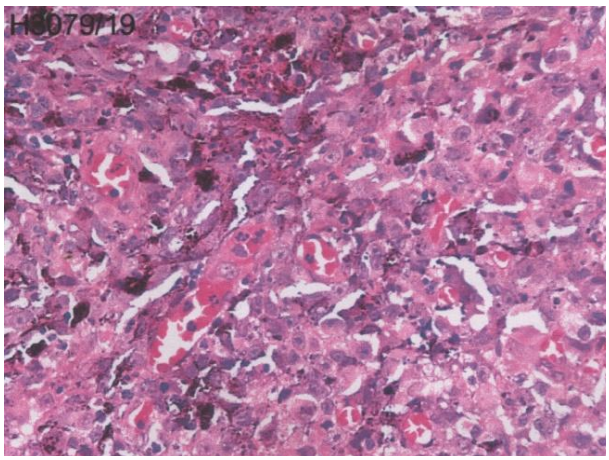

Fig 2: Malignant melanoma composed of cells arranged in sheets with intra cytoplasmic brown pigments (melanin).

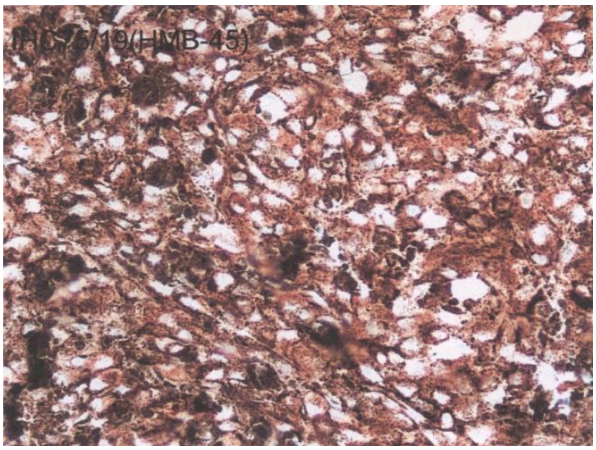

Fig 4: Malignant melanoma positive for HMB-45.

III. Discussion

The Primary malignant melanoma of vagina is a very rare tumor. It was first reported in 1887. The disease is encountered between 37 and 72 years of age, majority among postmenopausal women where about 73% of cases were older than 60 years.[3]The tumor of vaginal melanoma is most often located in the lower third (58%), and in the anterior wall (45%) of vagina.[9] The presenting symptoms are vaginal discharge, vaginal bleeding, pelvic pain, pelvic mass, and inguinal lymphadenopathy The disease is associated with a high risk of local recurrence, distant metastases, and poor clinical outcome. About 20% of mucosal melanomas are multifocal, and only 10%–23% are amelanotic melanoma.[10] Epithelioid is the most common histologic cell type of VPMM (55%). Other less common histologic cell types ofVPMM are spindled (17%) and mixed (28%).[11]

VPMM arises from melanocytes located aberrantly in vaginal epithelium. Those melanocytes can be found in the basal layer of vaginal epithelium in 3% of healthy women. It is thought that active junctional changes are the initial stage of development in malignant mucosal melanomas.[12]The theories also propose that primary malignant vaginal melanoma is a mucosal melanoma associated with *KIT* gene mutations whichencodes for a protein called c-KIT. Another theory suggests that the causative factor could be microenvironmental in nature and not associated with ultraviolet radiation.[4]

Its diagnosis mainly relies on pathological examination, typical melanoma that are commonly pigmented is easy to diagnose. However, amelanotic appearance is rare and makes diagnosis even more difficult. Consequently, immunochemical staining should often be used to supplement the diagnosis, such as protein S-100, HMB-45, Melan-A, Mart-1 and vimentin.[8] In our case, the pathological results only showed a malignant melanoma, and immunochemical analysis demonstrated HMB-45, S-100 protein positive which further confirmed the diagnosis. CT or MRI may be helpful to determine the extension, as well as the degree of cancer involvement. MRI can distinguish melanoma from other tumors because of a distinct signal pattern for melanin, i.e., high signal intensity on T1-weighted magnetic resonance images and low signal intensity on T2-weighted magnetic resonance imaging.[13]

Due to the limited number of cases in the literature, there are no standard therapy recommendations. Primary surgery is considered as the method of choice and appears to be superior to primary radiation. Various surgical procedures have been described, including wide local excision, colpectomy, radical resection with total abdominal hysterectomy, bilateral salpingo oophorectomy, and evisceration.[6] Different adjuvant chemotherapy regimens have been tried alone and in combinations to reduce recurrence rate in high-risk melanoma but none of the agents has unfortunately proven beneficial. The most promising results have been reported with Interferon alpha (IFNa), 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 IFNa, IL-2, ipilimumab, and vemurafenib in these cases, have not been completely defined.[7]

Despite aggressive therapy, the prognosis of vaginal melanoma is very poor because of the high incidence of local recurrence and regional or distant metastasis. Fifty percent of patients have positive lymph nodes, and nearly 20 percent 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. Patients with vaginal melanoma have the five year survival rate of 13 to 19%.[14] The common sites of recurrence of vaginal melanoma are vagina, vulva and groin. Once recurrence occurs, prognosis of melanoma is usually poor with limited systemic treatment modalities.[15]

IV. Conclusion

Primary malignant melanoma of vagina is a rare tumour with a poor prognosis. It should be confirmed by histopathology and immunohistochemistry markers. Though no specific treatment modality has been established but surgery is recommended as method of choice accompanied by radiation or chemotherapy. Therefore, it is necessary to initiate collaborative studies and follow up of patients in establishing a therapeutic approach that may offer the best results to the patients.

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