

## Primary vaginal melanoma- A rare presentation

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### Abstract

A 60 years old postmenopausal female with breast cancer presented with complaint of discharge and bleeding per vaginum since. She was a known case of left breast cancer (pT2N0), for which she underwent modified radical mastectomy and completed adjuvant chemotherapy and radiotherapy. During this visit, per vaginum and per speculum examination revealed pedunculated vaginal polyp which bled easily on touch. She underwent polypectomy and specimen was subjected to histopathological examination and immunohistochemical examination. HPR was suggestive of Rhabdomyosarcoma. IHC was suggestive of malignant melanoma and was HMB 45+, MELAN A +, S100 +, Ki 67 was 50-60% whereas it was negative for myogenin, CD34, CK and Desmin. Dacarbazine based adjuvant chemotherapy regimen was initiated and patients was advised BRAF mutation study, for which she was not willing. Patient was given 6 cycle of Adjuvant Dacarbazine 200 mg/m<sup>2</sup>. Patient showed no sign of disease recurrence.

**Keywords:** melanoma, vaginal, rare, aggressive

### Introduction

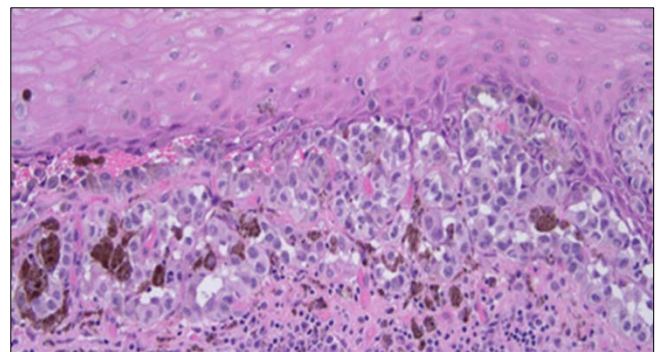
Primary vaginal melanoma is extremely rare malignant disorder affecting female genital tract, contributing to less than 0.2% of all melanomas. These tumors are aggressive in nature and arise from resident melanocytes in the mucous membranes [1]. Vulva and vagina are commonly involved in primary genital melanoma whereas cervix, uterus, and ovary involvement is extremely rare [2]. Vaginal melanoma is much more aggressive than its cutaneous counterparts and 5 year survival rate has been reported to be less than 30% [3-5]. As the disease is extremely rare and data is scarce, it is challenging to stage and treat the disease. We reported a rare case of primary vaginal melanoma in 60 years old postmenopausal female as little is known about this carcinoma.

### Case details

A 60 years old postmenopausal female with breast cancer presented with complaint of discharge and bleeding per vaginum since 2-3 months. She was a known case of left breast cancer (pT2N0), for which she underwent modified radical mastectomy and completed adjuvant chemotherapy and radiotherapy with 6# CAF and 25# respectively, 6 years back. Since then she was on adjuvant hormonal therapy for 5 years.

On examination, she was afebrile, her vitals were stable. Per vaginum and per speculum examination revealed pedunculated vaginal polyp which bled easily on touch. She underwent polypectomy. The inguinal nodes were not removed since there were no signs of lymphadenopathy. The specimen was subjected to histopathological examination and immunohistochemical examination. HPR was suggestive of Rhabdomyosarcoma with atypical melanocytes showing lentiginous pattern of growth along the basal layer of vaginal epithelium, invading underlying

connective tissue. Figure 1 reveal melanocytes within the basal layer of the squamous epithelium along with cytological atypia, increased nuclear-cytoplasmic ratios, pleomorphism and prominent nucleoli. The tumour cells had melanin which appeared as clumps of brown cytoplasmic pigment. Apart from this, invasion into adjacent cell was regarded as diagnostic feature of malignant melanoma.



**Fig 1:** Histopathological finding in our case

IHC was suggestive of malignant melanoma and was HMB 45+, MELAN A +, S100 +, Ki 67 was 50-60% whereas it was negative for myogenin, CD34, CK and desmin. Further, she was subjected to CECT of abdomen and pelvis post surgery which revealed no focal abnormality or lesion.

Dacarbazine based adjuvant chemotherapy regimen was initiated and patients was advised BRAF mutation study, for which she was not willing. Patient was given 6 cycle of Adjuvant

Dacarbazine 200 mg/m<sup>2</sup>. Patient was followed up regularly till 18 months and showed no sign of disease recurrence.

## Discussion

Primary vaginal melanoma is extremely rare cancer, and is mainly observed in postmenopausal females belonging to age range of 60 to 80 years. The mode of presentation is usually universal i.e. postmenopausal bleeding [5, 6]. In our study, the female presented at the age of 60 years with postmenopausal bleeding and vaginal discharge. However, Jamaer *et al* reported a case of 84 year old postmenopausal female who was diagnosed to be suffering from primary vaginal melanoma during the preoperative insertion of an indwelling urinary catheter [7]. The exact etiopathogenesis of primary vaginal melanoma is unknown but genetic, environmental or their interactions have been implicated in its pathogenesis [8, 9]. In our scenario, female had past history of breast cancer. Similarly, Kühn *et al* documented a case report of 44 year old female with past history of hysterectomy for multiple uterine myomas [10]. Thus, hormonal factors may also be implicated in the pathogenesis of vaginal myomas.

Literature suggest that usually, cases with vaginal melanomas are detected in advanced stages; approximately 50% with lymph node metastasis at the time of diagnosis, and 20% have distant metastasis [11]. However, in our case, female presented in early stage, and no locoregional metastasis was observed. These tumors are considered as aggressive tumor and prognosis is even worse than their cutaneous counterparts. As these are rapidly growing tumor, their prognosis is poor and five year survival rate have been reported between 0 to 25% despite treatment [12, 13].

Vaginal melanoma could be accurately diagnosed by visual inspection along with histopathological evaluation. But in our case scenario, vaginal polyp could not be diagnosed as melanoma during the physical examination. Primary melanomas are observed in lower one-third of vagina which often bleed on touch and are polypoid in appearance, sometimes ulcerated. [14-16] Surgery is the management of choice with wide local excision, radical resection with total abdominal hysterectomy, colectomy, bilateral salpingo-oophorectomy, evisceration depending upon the extent of involvement [17]. However, in our study, initial diagnosis was vaginal polyp and thus polypectomy was done which on histopathological analysis and immunohistochemical analysis was suggestive of primary vaginal melanoma. Van Nordstr and *et al* documented superior outcome of radical surgery over “wide local excision” or radiation [18]. The histopathological findings in this patient was consistent with the features suggested in literature i.e. melanocytes within the basal layer of the squamous epithelium along with cytological atypia, increased nuclear-cytoplasmic ratios, pleomorphism and prominent nucleoli. The tumour cells had melanin which appeared as clumps of brown cytoplasmic pigment. Apart from this, invasion into adjacent cell was regarded as diagnostic feature of malignant melanoma [7]. Immunohistochemistry may also be helpful in providing reliable confirmation of a diagnosis. They may be positive for protein S-100, T311, Mart-1, Melan A, HMB-45, tyrosinase and vimentin [6, 9]. In our study, IHC was positive for HMB 45+, MELAN A +, S100 +, whereas it was negative for myogenin, CD34, CK and Desmin.

Surgery is the mainstay of treatment which is followed by

adjuvant chemotherapy. Common chemotherapeutic agents which are utilized in management of primary vaginal melanoma include dacarbazine, temozolomide and paclitaxel [9, 19]. Radiotherapy is rarely used for management of these aggressive cancer. Dacarbazine based adjuvant chemotherapy regimen was initiated in our patients which showed good results and no sign of disease recurrence could be observed till 18 months. However, literature reveal poor prognosis even after best management as majority of cases present in advanced stages [12, 13].

## Conclusion

Primary vaginal melanoma are rare aggressive tumor affecting the female genital tract. Physical examination along with histopathology and immunohistochemistry can establish the diagnosis. Although surgery remains the primary modality of treatment, adjuvant therapy should be chosen carefully which may help in improving the survival of the patient.

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