

CASE REPORT

A RARE CASE OF PRIMARY MALIGNANT MELANOMA OF THE VAGINA IN A YOUNG LADY: A REPORT

Sangeeta Pankaj¹, Vijayanand Choudhary², Rajesh Kumar Singh³

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ABSTRACT: Malignant melanoma of vagina is a rare tumor with incidence rate of only 0.46 cases per one million women per year and less than 250 cases have been reported in medical literature till date. It has a notoriously aggressive behavior and is associated with a high risk of local recurrence and distant metastasis. At present, there are various treatment options for this disease but no standard guideline. It accounts for less than 1% of all malignant melanoma and less than 3% of all primary malignant tumors of the vagina. Conventional imaging techniques i.e. computed tomography, may be sub-optimal in evaluating a small volume of tumor, which may lead to inaccurate staging of a loco-regional tumor. The optimal treatment for vaginal melanoma is a subject of debate. Malignant melanoma of the vagina is a disease of postmenopausal women, with 75% of women being more than 50 years of age. Here, we present a case of primary vaginal malignant melanoma in a 35 year woman located in lower 1/3rd of the vagina.

KEY WORDS: Malignant Melanoma, Vagina, Melanocytes.

INTRODUCTION: Malignant melanoma is an aggressive disease which affects both skin and mucosal surfaces. Melanoma of the vagina and urethra is an extraordinarily rare tumor accounting for less than 10% of all female genital tract melanomas, and 2.6% of all vaginal cancers [1]. Malignant melanoma of the vagina is a disease of postmenopausal women with 75% of women being over 50 years of age [2]. It may arise anywhere in the vagina with a predilection for the lower third [1]. Due to the scarcity of cases of malignant melanoma of the vagina, there is little evidence-based literature for guiding clinicians to understand the importance of disease staging via noninvasive imaging strategy. This is a contentious issue which affects the way this tumour is managed appropriately, whereby tumour size, regional infiltration and groin lymphatic involvement are the cornerstone for disease prognostications [3,4].

CASE REPORT: A 35 year old woman, Para 3, post-surgical menopause by 3 years, had undergone hysterectomy for Dysfunctional Uterine Bleeding. She presented in gynecological oncology department with complaints of white discharge and recurrent bleeding per vaginum for last 14 months. Per speculum examination revealed, two blue to brown coloured lesion measuring 2 cm² each, in the lower 1/3rd of the vagina placed 3 cms apart, on the postero-lateral wall of vagina. Another 1 cm² lesion was found below the urethra. Around two third of vaginal wall was having a bluish flush. On per vaginal examination uterus was not palpable and both the adnexa was also not palpable. Rectum was normal and there was no inguinal lymphadenopathy. Wide excision biopsy (with free margin of 1 cm) was done from all the three lesions and sample sent for histopathological examination.

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MRI pelvis revealed a 1.8 × 1.6 cm ill defined heterogenous enhancing lesion in the lower end of vagina which was also involving the external urethral meatus. Rest of the vagina, urethra, bladder and rectum showed normal morphology and no lymphadenopathy. On cystoscopy, external meatus was found to be indurated but bladder and urethra was normal. Sigmoidoscopy was normal.

Histopathological report diagnosed the tumor as malignant melanoma involving the vaginal mucosa. The tumour was composed of epithelioid cells arranged in irregular sheets invading the submucosal stroma diffusely. These cells had large pleomorphic nucleus and prominent nucleoli. The cytoplasm was scanty to moderate in amounts. The cytoplasm contained abundant golden brown granular melanin pigment. Melanin pigment was also present extra-cellularly.

The patient was treated with external beam radiation (40 Gy in 20 fractions; 5 fractions per week) with palliative intent, to the tumor bed, followed by chemotherapy with six cycles of Temozolomide, 250 mg orally from D1 to D5 three weekly for 6 cycles. The patient is alive after 1 year and is under follow up.

DISCUSSION: Malignant melanoma of the vagina is a very rare tumor, accounting for 0.3 - 0.8% of all melanomas in women and 2 - 5% of female genital tract melanomas, and less than 3% of vaginal tumors. [5] The tumor typically presents in the sixth to seventh decade of life and occurs more commonly in the lower third of the vagina and involves the anterior wall of vagina.[5,6] The appearance of tumors is almost always pigmented with only 10 - 20% being amelanotic. [7] The most common symptom is abnormal vaginal bleeding. [6].

Grossly, the tumor appears as black or grayish black nodular, polypoid or fungating mass. Frequently, the overlying epithelium is eroded or ulcerated which explains their frequent confusion with squamous cell carcinoma. The differential diagnosis includes metastasis from other sites, poorly differentiated squamous cell carcinoma, sarcoma, lymphoma and blue nevus.

Several treatment options are administered but none of them are considered to be a standard approach. Surgical resection is considered the treatment of choice with survival benefits. [6, 7] Different surgical methods such as wide local excision, radical surgery (total vaginectomy with or without vulvectomy), and pelvic exenteration have been described. However, research has continued to demonstrate that there was no difference in survival between patients who have radical surgical procedures and those who have more conservative surgery.[8,7,9] Some authors advise for radical surgery due to the aggressive behavior of vaginal melanoma.[8,9] After surgery, patients with a high risk of recurrence (including regional lymph node involvement or thicker primary tumors, i.e. American Joint Committee on Cancer Stage IIb /IIc / III) should be considered for adjuvant therapy.

The role of cytotoxic chemotherapy in vaginal melanoma has not been completely defined because of the small number of cases. Dacarbazine has been considered the standard of treatment for metastatic or recurrent melanoma since 1972. Other useful antineoplastic agents include temozolomide, platinum analogs, nitrosoureas, and taxanes. The response rate of these single agents is 11 - 22%, with median overall survival of 5.6 -11 months. Combination chemotherapy that is most commonly used for melanoma includes Dartmouth regimen (Dacarbazine/ Carmustine/Cisplatin/Tamoxifen) and CVD regimen (Cisplatin/Vinblastine/Dacarbazine), which have proved to increase objective response rate but have no overall survival benefit.[10] Based upon

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the above observations, it may be possible to develop combination chemotherapy regimens that improve overall survival in near future.

Radiotherapy for local control has been mostly offered in the following two conditions, surgically non-resectable disease or as an adjuvant therapy in case of pathologically positive margins or positive lymphadenopathy. [6, 7] Prognosis is poor with a 5-year survival rate ranging from 10-20%. It appears to be influenced by tumor size with lesions greater than 3 cm having a bad prognosis. Age, mitotic count, stage and location of the lesion do not influence survival.

To conclude, early diagnosis of a loco-regional malignant vaginal melanoma may give patients the best chance for prolonged survival. The most accepted line of treatment of malignant melanoma of the vagina is surgery followed by post-operative radiotherapy. Primary malignant melanoma of the vagina has a poor prognosis with high risk of local recurrence and distant metastasis. In our case, local excision followed successively by radiotherapy and chemotherapy proved to be a tolerable modality.



Fig 1: Vagina showing blue to brown discoloration of the vagina and the lesion below urethra.



Fig 2: Vagina showing brown discoloration of vagina and lesion on lower third of vagina.

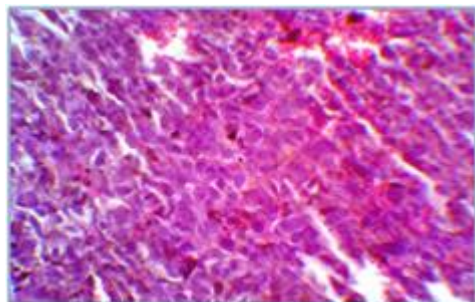


Fig 3: Numerous atypical melanocytes are seen arranged in irregular sheets. Golden brown granular melanin pigment is present both intracellularly and extracellularly.

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AUTHORS:

1. Sangeeta Pankaj
2. Vijayanand Choudhary
3. Rajesh Kumar Singh

PARTICULARS OF CONTRIBUTORS:

1. Assistant Professor, Department of Gynaecological Oncology, RCC, Indira Gandhi Institute of Medical Sciences, Patna.
2. Assistant Professor, Department of Pathology, Indira Gandhi Institute of Medical Sciences.
3. Associate Professor, Department of Gynaecology, RCC, Indira Gandhi Institute of Medical Sciences, Patna.

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Rajesh Kumar Singh,
Associate Professor, RCC,
Indira Gandhi Institute of Medical Sciences,
Patna.
Email- drrajeshsingh@yahoo.com

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