Vaginal Primary Malignant Melanoma: A Rare and Aggressive Tumor

A Case Report

Dr. Mangilal Choudhary, Dr. Jayanti Mehta, Dr. Rekha SIRVI, Dr. Maryem Ansari, Dr. Daleep Kumar
Department of Pathology, SMS Medical College Jaipur, Rajasthan

Abstract

Vaginal primary malignant melanoma is a rare and very aggressive tumor. It most commonly occurs in postmenopausal women, with a mean age of 57 years. Our patient is a 70-year-old, postmenopausal woman presented with a complaint of abnormal vaginal bleeding. On gynecologic examination there was a pigmented, raised, ulcerated, and irregular lesion 5 x 4.5 cm in the upper third of lateral vaginal wall. She underwent a wide local excision of the lesion. The histopathology revealed vaginal primary malignant melanoma and no clear surgical margins. She denied any additional surgical interventions and underwent postoperative adjuvant radiotherapy. Follow up 5 months after initial diagnosis revealed no evidence of local recurrence or distant metastasis. The prognosis of vaginal primary malignant melanoma is very poor despite treatment modality, because most of the cases are diagnosed at advanced stage. Particularly patients with no clear surgical margins and tumor size >3 cm needed postoperative adjuvant radiotherapy.

1. Introduction

Vaginal primary malignant melanoma (VPMM) is a rare and very aggressive tumor.\(^1\) It accounts for 0.3–0.8% of all malignant melanomas, 2–5% of female genital tract melanomas, and less than 3% of all vaginal malignancies\(^2\). The estimated incidence of VPMM is 0.026/100,000 women per year. It most commonly occurs in postmenopausal women\(^3\), with a mean age of 57 years. The precise etiology of VPMM is unknown. It is thought that VPMM arises from melanocytes present in the vaginal epithelium. Our aim is to present a case of VPMM that underwent a wide local excision and postoperative adjuvant radiotherapy. This aggressive tumor has a poor prognosis with 5 year survival rate 5-25%. We present a case of primary vaginal malignant melanoma located in lower one third of vagina and review the current literature.

2. Case Presentation

The patient, an 70-year-old, gravida 3, para 2 postmenopausal woman, presented to the Department of Obstetrics and Gynecology of the SMS Medical College & attached Hospital with a complaint of abnormal vaginal bleeding. Her medical history included hypertension and...
diabetes mellitus and treatment on Telmisartan drug. She had menopause at the age of 22 year back & history of sterilization of 40 year back. Her surgical history was unremarkable. Her family history revealed no evidence of cancer among the first-degree relatives. On gynecologic examination there was a pigmented, raised, ulcerated, and irregular lesion 5 × 4.5 cm in the upper Third of lateral vaginal wall. There were no palpable inguinal lymph nodes, and the rest of pelvic examination was normal. Preoperative computer tomography (CT) of the abdomen and pelvis, abdominal ultrasound (U/S), chest X-ray, intravenous pyelography (IVP), colonoscopy, and urethra cystoscopy was normal.

She underwent a wide local excision of the lesion and biopsy received in the Department of pathology. The histopathology showed a malignant neoplasm consisting of tumor cells mainly with spindle cell morphology. There was abundant deposition of melanin and presence of multinuclear giant cells. The histological diagnosis was confirmed by positive immunohistochemistry. Tumor cells expressed melan A. However, tumor cells did not express SMA, epithelial membrane antigen (EMA). The final diagnosis was vaginal primary malignant melanoma. The patient denied any additional surgical interventions and underwent postoperative adjuvant radiotherapy.

She received only high dose rate brachytherapy (HDRB) with 192Ir due to age, performance status, and comorbidities.

Followup 5 months after initial diagnosis, with CT of the abdomen and pelvis, abdominal U/S, chest X-ray, IVP, colonoscopy, and urethrocystoscopy, revealed no evidence of local recurrence or distant metastasis.

**Figure 1a** – Low power view (10x) of histological section in H& E staining show Malignant Melanoma

**Figure 1b** – Low power view (10x) of histological section in H& E staining show Malignant Melanoma
**Figure 2a.** high power view (40x) of histological section showing Malignant Melanoma (spindle cell type)

**Figure 2b.** High power view (40x) of histological section showing Malignant Melanoma (spindle cell type)

**Figure 3** low power view (10x) of H & E Staining showing malignant melanoma after Demelanization.

**Figure 4** High power view (10x) of H & E Staining showing malignant melanoma after Demelanization.
3. Discussion

Primary malignant melanoma is a rare entity first reported by Poronas in 1887\(^4\). The tumor typically presents in the sixth and seventh decades of life and occurs more commonly in the lower 1/3\(^{rd}\) of the vagina and mostly in the anterior vaginal wall. It affects post-menopausal women and does not have any known risk factors\(^5\). The most common symptoms are vaginal bleeding, vaginal discharge and feeling a mass in the vagina.

Grossly, the tumour was grey brown polypoid to nodular soft tissue piece ms. 2.0 x0.5 cm. and microscopically section show sheets of polyhedral or spindly cells with hyperchromatic nuclei and showing pigment in the cytoplasm. The appearance of the tumor is almost always pigmented and only 10-23% are amelanotic. The natural course of malignant melanomas is marked by early local recurrence, extensions and frequent metastases to the lymph nodes, viscera and also life threatening hemorrhage, making it the most dangerous form of vaginal tumor.

Malignant melanoma localized in the vagina result from the malignant transformation of an ectopic melanocytes during menopause\(^7\). Histopathologically the neoplastic cells are either epitheloid or spindled.\(^6\) Immunohistochemically S100 protein was strongly and diffusely positive in 96%. S100 remain the most sensitive marker for these tumor.

Vaginal melanoma have a poor prognosis. The crude 5 year survival rate was 21% in the series reported by chung et al. Melanoma with a mitotic count of less than 6 mitotic figures per 10 HPF have a better prognosis than those with higher rates.\(^8\) Thus mitotic count is inversely proportional to survival but vascular invasion and depth of invasion do not correlate with survival. Tumor size is the strongest predictor of survival.

The differential diagnoses include metastasis from other sites, poorly differentiated squamous cell carcinoma, sarcoma, lymphoma and blue nevus.\(^9\) It has a high rate of recurrence and poor long term survival.

In these patients surgery remains the primary treatment of choice. The spectrum of surgery ranges from conservative (wide local excision) to radical (vaginectomy and pelvic exenteration). If wide local excision with clear margins is possible, the role of radical surgery as primary treatment for VPMM remains unjustified. However, if local excision is not possible, pelvic exenteration may be reasonable. Our patient underwent a wide local excision. Recently, the sentinel lymph node biopsy has gained popularity.\(^10\)

Radiotherapy can be applied as primary treatment for patients who are unable or unwilling to have...
surgery. It can be applied preoperative as adjuvant treatment to reduce tumor size and enable a more conservative surgery. It can be applied postoperatively as adjuvant treatment for patients with incomplete tumor resection or with pelvic metastases.

The combination of chemotherapy and immunotherapy in patients with advanced stage CMM is associated with an increased response rate. However, it has the disadvantage of increased toxicity. The role of biochemotherapy in patients with advanced stage VPMM has not been established.

**Conclusion**

The case is reported because of its rarity and attempt is made to describe its clinical features, pathological features with prognosis and its treatment.

**References**