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**Case Report**

# Primary amelanotic vaginal melanoma masquerading as pleomorphic sarcoma: a diagnostic dilemma

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## ABSTRACT

Primary amelanotic vaginal melanoma is an exceptionally rare and aggressive malignancy that poses a diagnostic challenge because of its lack of pigmentation and resemblance to other poorly differentiated malignancies. A 73-year-old postmenopausal woman presented with vaginal bleeding and a vaginal mass. Histopathological examination of the excised vaginal mass suggested a high-grade pleomorphic sarcoma with positive margins. She subsequently underwent wide local excision, and histopathological examination revealed no residual malignancy. Immunohistochemistry demonstrated positivity for vimentin, S-100, and HMB-45, confirming the diagnosis of primary amelanotic vaginal melanoma. Six weeks later, she developed right inguinal lymph node metastasis and underwent right ilioinguinal lymphadenectomy. During follow-up, the disease progressed with contralateral (left) inguinal lymph node metastasis. She subsequently received palliative external beam radiotherapy to the inguinopelvic region (40 Gy in 15 fractions), which was tolerated well. This case underscores the importance of immunohistochemistry in establishing the diagnosis of primary amelanotic vaginal melanoma, and highlights its highly aggressive behaviour,

**Keywords:** Vaginal Neoplasms, Amelanotic melanoma, Immunohistochemistry

## INTRODUCTION

Primary vaginal malignant melanoma (PVMM) is a rare neoplasm arising from the vaginal wall without involvement of the vulva or cervix.[1] It accounts for less than 3% of vaginal malignancies and 0.3–0.8% of melanomas in women.[2] PVMM is believed to originate from aberrant melanocytes located in the basal layer of the vaginal epithelium.[1]

Most PVMMs are pigmented; however, amelanotic variants are exceptionally rare and frequently mimic poorly differentiated carcinomas, sarcomas, or lymphomas because of the absence of melanin pigment, creating a significant diagnostic challenge.[3] These tumours are highly aggressive, with early local recurrence and metastatic spread, resulting in poor outcomes.[4] We report a rare case of primary amelanotic vaginal melanoma in a postmenopausal woman initially misdiagnosed as pleomorphic sarcoma.

## CASE REPORT

A 73-year-old multiparous woman (P3L3A0), postmenopausal for 28 years, presented with postmenopausal bleeding and a protruding vaginal mass of 6 weeks' duration. She had no significant comorbidities or personal or family history of malignancy. General and systemic examination was unremarkable, with no palpable lymphadenopathy. Local examination revealed a polypoidal mass protruding through the introitus, with areas of sloughing and contact bleeding. Written informed consent was obtained from the patient for publication of this case report.

Ultrasonography of the abdomen and pelvis was unremarkable. Contrast-enhanced computed tomography (CECT) abdomen and pelvis demonstrated a heterogeneously enhancing soft-tissue mass (4.3×4.1×2 cm) involving the distal vagina, without pelvic or retroperitoneal lymphadenopathy.

Examination under anaesthesia revealed a greyish-pink, multilobulated mass arising from the right lateral wall of the lower one-third of the vagina. (Figure 1).

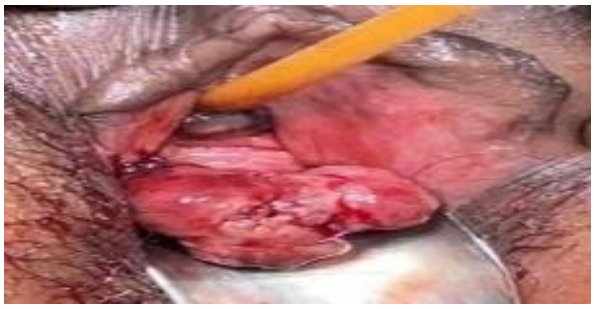


**Figure 1: Clinical photograph showing a polypoidal, multilobulated mass arising from the right lateral wall of the lower one-third of the vagina.**

The mass was friable and bled on contact. The cervix appeared normal, the uterus was small and mobile, and no adnexal mass was palpable.

The vaginal mass was excised. Gross examination revealed a greyish-pink, multilobulated mass measuring 3. ×3.5×2 cm, with areas of necrosis and sloughing (Figure 2).



**Figure 2: Gross appearance of the excised vaginal mass.**

The histopathological examination of the excised vaginal mass was suggestive of a high-grade pleomorphic sarcoma with positive surgical margins (Figure 3).

In view of the positive margins, the patient subsequently underwent a wide local excision. Histopathological examination of the resected specimen showed no residual malignancy.

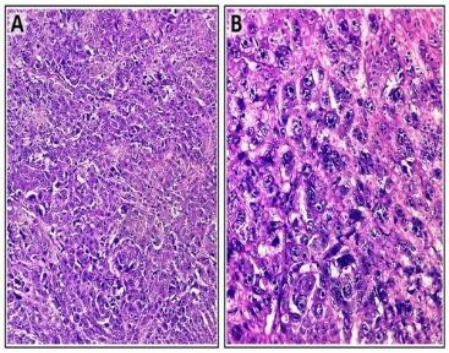



**Figure 3: Histopathological photomicrographs of the vaginal tumour showing a high-grade neoplasm arranged predominantly in sheets; (A) H&E,×100) and marked cellular and nuclear pleomorphism with prominent nucleoli; (B) H&E,×400).**

Immunohistochemistry (IHC) demonstrated positivity for vimentin, S-100, and Human Melanoma Black-45 (HMB-45), with a Ki-67 proliferative index of 50%, while being negative for other lineage-specific markers, including Pan-cytokeratin (PanCK), Cluster of Differentiation 34(CD34), Cluster of Differentiation 99(CD99), p40, Smooth Muscle Actin (SMA), and desmin, confirming the diagnosis of amelanotic melanoma. (Figure 4)

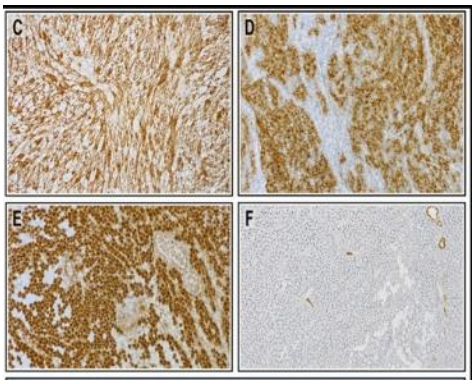



**Figure 4: Immunohistochemical staining showing tumour cell positivity for vimentin; (C) ×400), S-100; (D) ×400), and HMB-45; (E) ×400), with negative staining for SMA; (F,) ×400), confirming the diagnosis of amelanotic melanoma.**

Six weeks later, the patient developed right inguinal lymphadenopathy. Positron emission tomography-computed tomography (PET-CT) demonstrated metabolically active pelvic and inguinal lymph nodes. Ultrasound-guided fine-needle aspiration cytology (FNAC) of the right inguinal lymph node confirmed lymph node metastasis. Right ilioinguinal lymphadenectomy revealed metastatic involvement of the superficial, deep, and Cloquet's lymph nodes. Subsequently, ilioinguinal pelvic radiotherapy was planned. However, the simulation scan revealed left inguinal nodal enlargement, and FNAC confirmed metastatic disease.

Following a multidisciplinary tumour board discussion, systemic therapy was offered, but declined by the patient and her attendants because of logistic constraints. After detailed counselling regarding the treatment options, expected outcomes, potential toxicities, and prognosis, palliative radiotherapy to the inguinopelvic region was initiated. The patient received external beam radiotherapy (EBRT) to the inguinopelvic region at a dose of 40 Gy in 15 fractions using image-guided radiation therapy (IGRT) on a 6 MV linear accelerator (LINAC). The treatment was well tolerated, with only Grade 1 gastrointestinal toxicity. The patient is scheduled for the first follow-up visit 4 weeks after completion of EBRT.

## DISCUSSION

Primary vaginal malignant melanoma (PVMM) may arise anywhere in the vagina; however, the anterior wall of the lower one-third is the most common site.[3,5] Lesions have also been reported in the upper one-third of the vagina.[6,7] Gökaslan et al reported tumours on the posterior vaginal wall near the vaginal introitus,[8] whereas Shah et al described pigmented tumours involving the middle and lower thirds of the vagina.[9] In this patient, the tumour originated from the right lateral wall of the lower one-third of the vagina. The most common presenting symptom is vaginal bleeding, followed by the presence of a vaginal mass, vaginal discharge, and pain.[10] This patient was presented with postmenopausal bleeding and a protruding vaginal mass.

PVMM typically presents as a polypoid, exophytic lesion with variable pigmentation ranging from red, brown, grey, black, or blue.[1,5]While pigmented lesions are more readily recognized, amelanotic (nonpigmented) lesions present a significant diagnostic challenge because of the absence of melanin pigment and their histopathological resemblance to other poorly differentiated malignancies, including carcinomas, sarcomas, and lymphomas.[3] In the present case, the tumour was initially misdiagnosed as a high-grade pleomorphic sarcoma on histopathological examination.

Definitive diagnosis relies on histopathological examination supplemented by IHC.[11]Histologically, these tumours may exhibit epithelioid, spindle cell, or mixed morphology. Strong immunoreactivity for protein S-100, HMB-45, Melan-A, and vimentin supports melanocytic differentiation, whereas negative staining for epithelial, vascular, myogenic, and other lineage-specific markers helps exclude common histological mimics.[11,12] In our case, diffuse positivity for S-100, HMB-45, and vimentin, along with negative staining for PanCK, CD34, CD99, p40, SMA, and desmin, established the diagnosis of primary amelanotic melanoma.

PVMM is characterized by aggressive biological behaviour, with a high propensity for early local recurrence, regional lymph node involvement, and distant metastasis. Common sites of local recurrence include the vagina, vulva, and groin, whereas the lungs, liver, bones, and brain are the most frequent sites of distant metastasis. Lymphatic spread depends on the location of the primary tumour: lesions arising in the upper third of the vagina typically metastasize to the external iliac lymph nodes, those in the middle third to the internal and common iliac lymph nodes, and those in the lower third to the superficial inguinal lymph nodes, consistent with our patient's early bilateral inguinal lymph node involvement

Radiological imaging, particularly CECT and PET-CT, is essential for disease staging, detecting occult metastatic disease, and monitoring patients with metastatic disease undergoing systemic treatment.

PVMM carries a poor prognosis, with reported 5-year survival rates ranging from 5% to 25%,[4] largely because of its high propensity for early recurrence and distant metastasis. Surgery remains the mainstay of treatment and ranges from wide local excision to radical procedures, such as vaginectomy with or without vulvectomy. However, achieving adequate surgical margins is often challenging because of the anatomical constraints of the vagina. Despite surgical intervention, local recurrence and metastatic spread are common. In this case, the patient developed right ilioinguinal lymph node metastasis within six weeks of undergoing wide local excision. A high Ki-67 proliferation index (>20%–30%) has been associated with aggressive tumour behaviour and poor prognosis. The Ki-67 index of 50% in this patient correlated with rapid disease progression and subsequent bilateral inguinal lymph node involvement.

Immune checkpoint inhibitors have improved outcomes in patients with advanced melanoma; however, their availability remains limited in many resource-constrained settings. In this case, the patient declined systemic therapy because of logistical constraints. Radiotherapy may provide effective local control and symptomatic palliation, although its impact on long-term survival remains limited. The patient received EBRT to the inguinopelvic region, which was well tolerated.

## CONCLUSION

Primary amelanotic vaginal melanoma is a rare and aggressive malignancy that poses a significant diagnostic challenge due to the absence of pigmentation. Immunohistochemistry plays a pivotal role in establishing the diagnosis and distinguishing it from histological mimics. A high Ki-67 proliferation index reflects the tumour's aggressive biological behaviour. Despite surgical management, the risk of recurrence and nodal metastasis remains high, underscoring the need for vigilant follow-up. Early diagnosis and timely multidisciplinary management are essential to optimize patient outcomes.

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