Case Report

Hymenal tag; a clue to the diagnosis of vaginal polyp: A case report of primary vaginal melanoma mimicking an undifferentiated pleomorphic sarcoma

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ABSTRACT

Primary malignant melanoma of the vagina accounts for less than 0.3% of all melanomas and approximately 4% of vaginal malignancies. We present a case of a 61-year-old female who presented with complaints of difficulty in urination and a polyp arising from the vagina. Local examination showed a polyp arising from the lateral vaginal wall and a hymenal tag. Histopathological examination of the polyp showed a neoplasm simulating a pleomorphic sarcoma. The hymenal tag showed classical features of malignant melanoma. Immunohistochemistry with S100 and HMB45 confirmed malignant melanoma in the vaginal polyp. We report this case, on account of its rarity and unusual phenotypic profile, which may lead to a delayed or alternate diagnosis. Primary malignant melanoma of the vagina needs early accurate diagnosis and management due to its aggressive nature.

1. Introduction

Primary malignant melanoma of the vagina is a rare and aggressive malignancy with a high risk of local recurrence and distant metastasis.

It accounts for less than 0.3% of all melanomas and approximately 4% of vaginal malignancies. Most of the patients are post-menopausal with a mean age of around 60 years. The most common symptom is vaginal bleeding followed by vaginal mass and discharge. The tumour mass is usually located in the anterior and lateral walls of the lower third of the vaginal wall. Macroscopically most are polypoidal or nodular, with an average size of 2-3 cms. Most of the cases are pigmented while a few are amelanotic. The unusual phenotypic profiles of melanomas, including those that mimic the non- melanocytic lineage, may pose a diagnostic challenge. Here we present a case report and literature review of a case of primary vaginal malignant melanoma, the morphology of which mimicked an undifferentiated pleomorphic sarcoma. The clue to diagnosis came from a hymenal mucosal tag specimen of the same patient which showed classical features of malignant melanoma.

2. Case Report

A 61-year-old female presented to the gynaecology outpatient department of our hospital, with complaints of difficulty in urination, increased frequency of micturition, and a polyp arising from the vagina.

2.1. Clinical history

She gave a history of total abdominal hysterectomy 20 years back for fibroid uterus. There was no history of malignancy in the family.

2.2. Local examination

On examination, a pedunculated swelling of 5x3 cm arising from the lateral vaginal wall, at 2 o’clock position close
to the hymenal remnant and extending to the suburethral region was identified. A mucosal tag, measuring 6x4 mm was noted at 8 o’clock position; protruding from the hymenal remnant. There were no palpable inguinal lymph nodes.

2.3. Investigations
Preoperative ultrasonogram confirmed the presence of a heterogeneous lesion with internal vascularity in the lateral vaginal wall, of possibly neoplastic etiology.

2.4. Microscopy
Vaginal polyp and hymenal tag were excised and sent for histopathology. Microscopically, the vaginal polyp showed an infiltrating neoplasm composed of pleomorphic cells with hyperchromatic-vesicular nuclei and bizarre multinucleate giant cells. The neoplasm showed ulceration of the overlying stratified squamous epithelium. Histopathological examination of the hymenal tag, showed atypical melanocytes with intracytoplasmic brownish-black pigment, exhibiting a lentiginous pattern of growth along the base of the lining epithelium and infiltrating the underlying connective tissue. This gave the clue to the diagnosis of malignant melanoma in the vaginal polyp. The neoplastic cells in the vaginal polyp were strongly positive for S100 and HMB45 immunostains; thereby confirming the diagnosis.

The patient underwent PET-CT for staging which showed randomly distributed lung parenchymal nodules suspicious for pulmonary metastasis. No lymph nodal metastasis was identified.

3. Discussion
Melanoma is a malignancy that most commonly arises in the sun-exposed regions of the trunk and extremities. The lower extremities are the commonest site in women. The melanomas occurring in the female genital tract are biologically aggressive and account for 3% to 7% of all malignancies. The gamut of surgical options for vaginal melanomas includes extirpation with en bloc removal of the involved pelvic organs. Several treatment options are advocated, but none of them are considered to be a standard approach. Surgical resection is considered to be the first choice as improved overall survival has been demonstrated in patients treated with surgery when compared to those treated exclusively with chemoradiation. The stage of the disease and the histopathological variant also play a role in prognosis. Our patient had a tumor size of more than 3 cm, causing concern for dismal prognosis. Most of the vaginal melanomas are aggressive in nature and tend to be diagnosed at late stages with a 5-year survival rate of 0%–25%; and this dwindles to 5% in 3 years with lymph node involvement. Tumor thickness and increased dermal mitotic rate ($\geq 2/mm^2$) were independent predictors for reduced survival in vulvar melanoma.

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Fig. 1: Gross and Microscopy images. **A:** Vaginal polyp gross photograph, arrow-hymenal pigmented tag; **B:** Hymenal tag microscopy 100x, H&E. shows nests of tumour cells in the epithelium and subepithelial tissue; **C:** Hymenal tag microscopy 400x, H&E. Tumour cells showing intracytoplasmic brownish black pigment; **D:** Vaginal polyp, H&E 400X, shows bizarre multinucleated giant cells; **E:** Immunohistochemistry HMB 45 on vaginal polyp 400x, Shows strong membranous and cytoplasmic positivity in the neoplastic cells; **F:** Immunohistochemistry S100 on vaginal polyp 400x. Shows strong nuclear and cytoplasmic positivity in the neoplastic cells

patient was lost to follow up after surgical excision of the tumor.

4. Conclusion

Malignant melanoma of the vagina is a rare and aggressive disease affecting women in the 6th and 7th decades of life. The varying phenotypic profile, both gross and microscopy, can lead to an alternate or late diagnosis, which can have an impact on the prognosis of the disease. Hence a high index of suspicion should be maintained by clinicians and pathologists to avoid diagnostic pitfalls.

5. Source of Funding

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6. Conflict of Interest

None.

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