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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. [1,2] 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. [3–5] 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

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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 melanomas. The first case of malignant melanoma of the female genital tract was reported by Hewitt in 1861.[1] The most common site for gynecologic melanomas is the vulva. Around 85% of cases arise in the labia minora, clitoris, or inner side of the labia majora.[2]

Melanocytes are seen as embryological remnants of neural crest cells in three percent of healthy women. The aforesaid melanocytes beget primary malignant melanoma of the vagina.[3] Little is known about the risk factors, etiological factors, and the pathogenesis of these tumors.[4]

Vaginal melanomas usually present in the fifth to seventh decades of life. Most of the cases are seen in the lower one-third of the anterior vaginal wall. Typically,patients with vaginal melanoma experience a prodrome of vaginal bleeding (80%) vaginal discharge(25%), palpable mass(15%) in the vagina, and pain (10%).[5]

Vaginal melanomas have a polypoid to nodular macroscopic appearance in the majority of cases. Although a predominant number of these neoplasms are pigmented, 10%–23% are amelanotic.[6] The amelanotic vaginal polyp, in our case, would have caused much diagnostic confusion, if not for the pigmented hymenal tag; which makes our case noteworthy.

Vaginal melanomas have a poor prognosis. Early local recurrence, lymph node involvement, and distant metastasis are common. This might be due to the rich vascular and lymphatic network of the vaginal mucosa and a delay in diagnosis can be detrimental for the patient.[5,7,8]

Tumor size of more than 3 cm is the most important prognostic factor. Buchanan et al.[9] reported that tumors with a diameter of less than 3 cm carry a better prognosis. 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.[3] Tumor thickness and increased dermal mitotic rate ($\geq$2/mm2) were independent predictors for reduced survival in vulvar melanoma.[10]

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.[11,12] The gamut of surgical options range from conservative surgery such as wide local excision with lymph node dissection to total vaginectomy or radical extirpation with en bloc removal of the involved pelvic organs.[13] Excision with conservative margins of 1cm for tumors that are 1mm thick or less, 2cm for those 1–4mm thick with a minimum 1cm tumor-free deep margin are the current recommended standards.[13,14] Nevertheless, a significant difference in survival between patients who have radical surgical procedures and those who have more conservative surgical procedures have not been demonstrated.

Neoadjuvant treatment with chemotherapy or radiotherapy has been used in vaginal melanoma to reduce tumor burden enabling a more conservative resection.[15] Radiation therapy is indicated in surgically nonresectable disease, pathologically positive margins or metastatic nodes. Adjuvant systemic therapy is indicated in these high-risk cases.[16] Biochemotherapy is considered as a systemic option for advanced and metastatic vulvovaginal melanomas.[17] Nivolumab (a programmed death-1 (PD-1) immune checkpoint inhibitor) alone or combined with ipilimumab seemed to have greater efficacy than either agent alone, in the treatment of vaginal melanomas.[18] Our

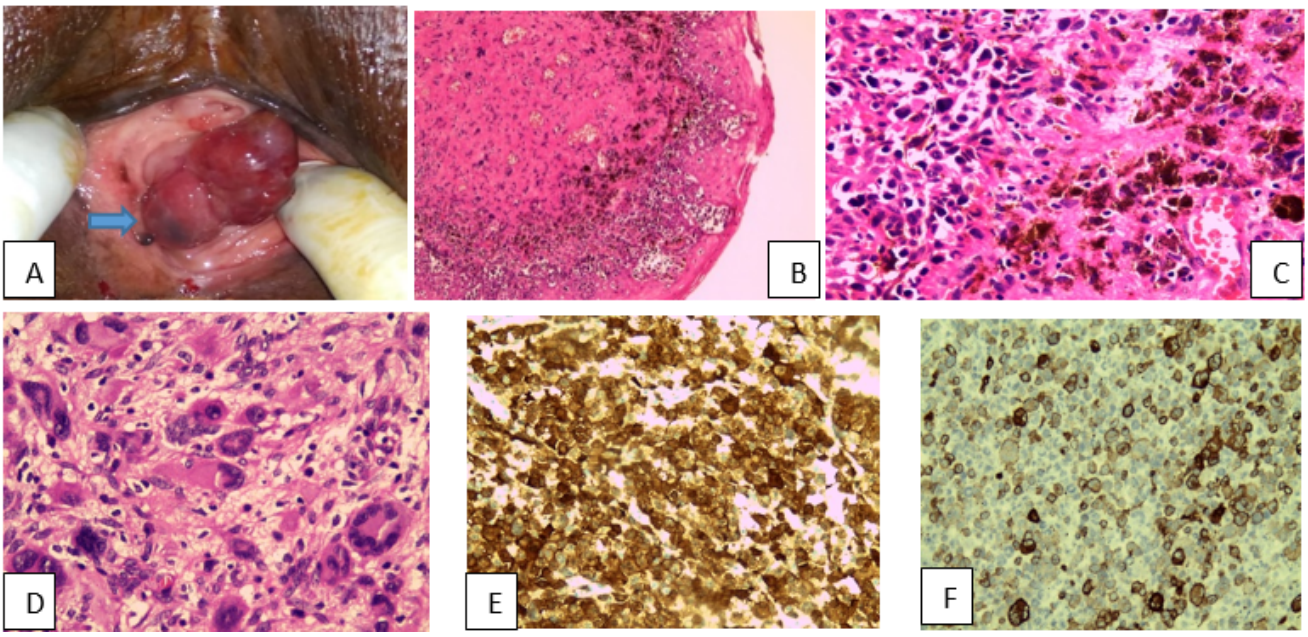

Fig. 1: Gross and Microscopy images. **A:** Vaginal polyp gross photograph, arrow-hymenal pigmented tag; **B:** Hymenal tag microscopy 100x, H&E. shows nests of tumour cells in the epithelium and subepithelial tissue; **C:** Hymenal tag microscopy 4 00x, H&E, Tumour cells showing intracytoplasmic brownish black pigment; **D:** Vaginal polyp, H&E 400X, shows bizzare 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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