ADVANCED MALIGNANT MELANOMA OF THE VULVA IN A 70 YEARS OLD WOMAN AT TIKUR ANBESSA SPECIALIZED HOSPITAL, ADDIS ABABA, ETHIOPIA: CASE REPORT

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ABSTRACT

BACKGROUND: Vulvar melanoma is a rare malignant tumor of the female genital tract that tends to occur in older women, with a high tendency to metastasize due to delayed diagnosis.[1] In this case report, we present a 70-year-old para 8 woman who presented with a progressively increasing vulvar swelling over two years and had multiple visits to healthcare institutions. Examination revealed a 7 cm diameter multi-nodular mass with areas of dark and dark blue-violet color, filling the introitus and involving the clitoris, labia minora, the lower third of the anterior vagina, and urethra. An incisional biopsy from the mass showed malignant melanoma. Imaging with a CT scan revealed bilateral lung metastasis. Palliative chemotherapy was planned, but the patient was lost to follow-up.

KEYWORDS: Case report, vulvar cancer, metastasis

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INTRODUCTION

Malignant melanoma of the female genital tract is rare, accounting for 1-3% of all melanomas in women, and is associated with a poor prognosis.[2] Vulvar malignant melanoma (VMM) accounts for 5-10% of all primary vulvar malignancies.[1,2] It predominantly occurs in postmenopausal white women, most commonly affecting the clitoris and labia. In addition to age, chronic inflammatory diseases and human papillomavirus (HPV) infection are proposed risk factors for vulvar melanoma. Activating alterations in the KIT gene are frequently associated with vulvar melanoma.[3,4,5] Symptoms in women with malignant melanoma of the vulva include color changes, lumps or swelling, itching, bleeding or discharge, and pain, with lesions presenting in various colors. Diagnosis is based on histopathologic examination of excisional or punch biopsy from the lesion. Comprehensive skin and eye examinations are necessary to rule out other primary sites. Whole-body positron emission tomography (PET)/CT scans should be conducted to assess the extent of disease and inform therapeutic decisions.[6,7] Surgery is the cornerstone of treatment for malignant melanoma, while systemic chemotherapy is used for metastatic and recurrent disease.[1,6] This case report describes a woman with a delayed diagnosis of malignant melanoma.

Case and Observation

We present a 70-year-old para 8 woman referred for a complaint of a two-year history of vulvar swelling. The mass had been increasing in size rapidly and was associated with bleeding over the past two months, requiring one or two pad changes per day. She reported no vulvar itching, vaginal discharge, or changes in bowel or bladder habits and had multiple visits to healthcare facilities before this referral. She had undergone a total abdominal hysterectomy three years ago for cervical intraepithelial neoplasia grade 3 and had no known chronic illnesses. She also reported no family history of malignancy.

On physical examination, the patient was in good functional status. Pelvic examination revealed a 7 cm diameter multi-nodular mass with dark and dark blue-violet areas filling the introitus, involving the clitoris, labia minora, the lower third of the anterior vagina, and the urethra. Examination of the breast, inguinal lymph nodes, eyes, skin, and abdomen showed no remarkable findings. Cystourethroscopy was attempted, but the urethral meatus could not be assessed due to obstruction by the mass. Histopathologic evaluation of excisional biopsies from the mass revealed tissue composed of stratified squamous epithelium with surface ulceration, underlying stroma consisting of sheets of pleomorphic oval to spindle cells with prominent nuclei and intracytoplasmic melanin pigment with pagetoid spread. Mitosis was also observed (Figure 2).

An MRI of the pelvis and abdomen revealed a 5 cm by 4 cm mass with heterogeneous hyperintense signal in both T2 and T1 sequences, infiltrating the anterior wall of the vagina and vulva and encasing the urethra, without regional nodal or distant metastasis. Chest CT showed bilateral lung metastasis. Palliative chemotherapy was planned with dacarbazine at a dose of 250 mg/m² IV daily for five days, to be repeated every three weeks for six cycles, with an evaluation of chemotherapy response after three cycles. However, the patient was lost to follow-up as she was from a war-torn area with limited access to transportation.

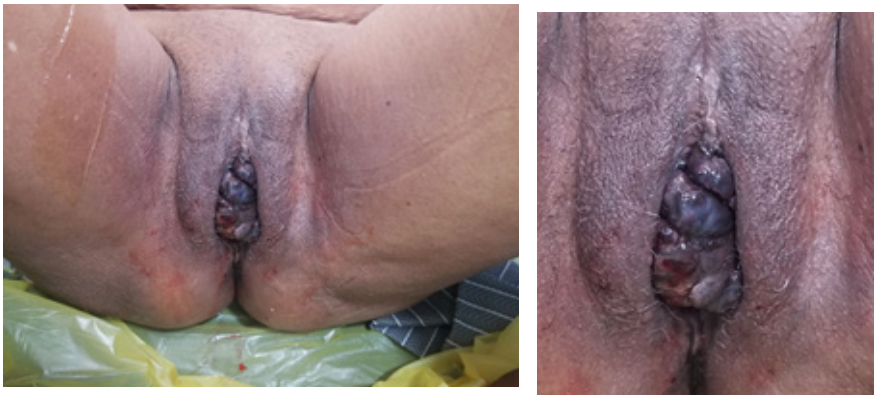

Figure 1. Dark red nodular 6 by 7cm Vulvar mass involving clitoris, lower third of anterior vagina and lower third of urethra is visible.

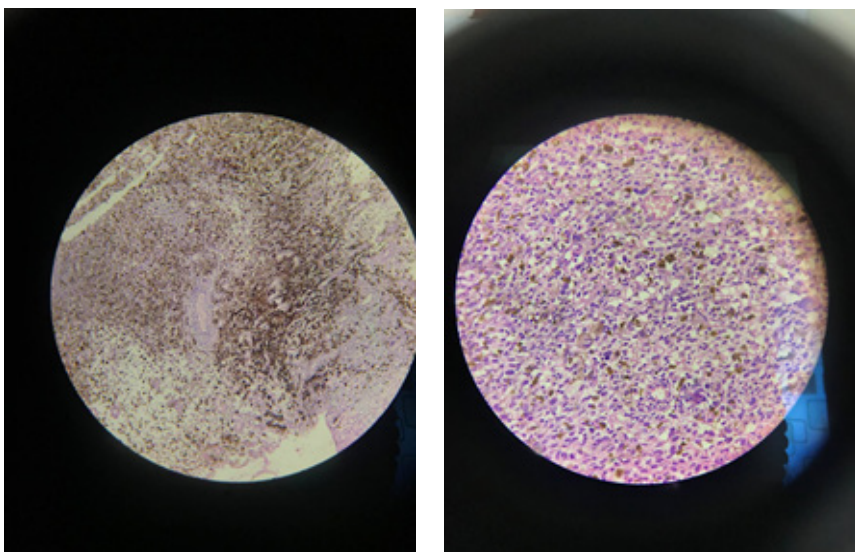

Figure 2. pleomorphic oval to spindle cells having prominent nuclei and intra cytoplasmic melanin pigment with Pagetoid spread

DISCUSSION

Female genital tract malignant melanoma, a devastating disease, is rare, accounting for 1-3% of all melanomas in women.[2] It has a poor prognosis, with a 15% 5-year survival rate.[1, 2, 8] Vulvar malignant melanoma (VMM) is responsible for 5-10% of all primary vulvar malignancies.[1 2] Most cases of vulvar malignant melanoma present at age 68 and above, although cases in younger, premenopausal HIV-positive women have been reported.[1] The risk of onset increases with age; cases rise from 0.11 per 1 million inhabitants in the 15–29-year age range to 3.5 per 1 million inhabitants in those over 60 years old.[1] In addition to age, chronic inflammatory disease and human papillomavirus (HPV) infection are proposed risk factors for the development of vulvar melanoma. Activating alterations in the KIT gene are commonly associated with vulvar melanoma.[3 4 5]

There is no specific screening test for vulvar cancer in general. Early detection of any vulvar lesion and biopsy by clinicians during gynecologic exams can prevent or reduce the incidence of malignant vulvar lesions. [10] Any pigmented vulvar lesion, unless it has been present for some time without change, should be biopsied for diagnosis.[11] Most patients with vulvar malignant melanomas are asymptomatic and diagnosed late, which may be due to the anatomic position of the lesion. Some women, however, present with symptoms such as lump or swelling, itching, bleeding, discharge, and pain, particularly as the disease progresses.[1] Approximately one-third of patients present with lymph node metastasis.[1]

Our patient presented at age 70 with distant metastasis to the lungs. She had previously undergone screening for cervical cancer and a hysterectomy for cervical intraepithelial neoplasia grade 3 a year before her current symptoms. This could have been an opportunity for early detection of the vulvar lesion in this case. Although she sought care for genital complaints, a diagnosis was not reached, possibly contributing to the advanced stage at presentation. Amelanotic melanomas account for only 2% of all vulvar melanomas.[1,6,7]

Complete skin and eye examinations are necessary to rule out other primary sites. A whole-body positron emission tomography (PET)/CT scan should be obtained to assess the disease extent and plan the therapeutic approach.[6,7] The staging of vulvar melanoma is based on the 8th edition of the American Joint Committee on Cancer (AJCC) staging system.[12] Surgery is the mainstay treatment for vulvar malignant melanoma. Systemic dacarbazine-based chemotherapy is recommended for advanced stages.[1,6] Typically, follow-up appointments are recommended every 3 to 6 months for the first 2 years and then every 6 to 12 months thereafter.

The overall prognosis is poor. The 5-year disease-specific survival rates for those with localized, regional, and distant disease are 75.5%, 38.7%, and 22.1%, respectively.[6,7,12] This case presented at an advanced stage, which entails a poor prognosis. Similar to this case, Ramesh et al. reported a case of vulvar melanoma in a 32-year-old patient managed with surgery followed by radiotherapy. In another study, Soumiya et al. reported an advanced, inoperable vulvar melanoma in a postmenopausal woman, describing an aggressive tumor similar to this case. In another case series, most patients were treated surgically, as most presented at an early stage.[13,14,15]

Conclusion

Neglecting a thorough gynecologic examination and failing to diagnose malignant vulvar melanoma can result in a devastating disease with a poor outcome. Patients attending cervical cancer screening may benefit from a comprehensive genital examination. Early diagnosis in patients with vulvar complaints can alter the treatment plan and improve patient outcomes. We recommend routine ano-genital examinations for patients presenting to outpatient clinics for various gynecologic concerns.

Sources of funding
Nothing to declare.

Consent
For the purpose of publishing this case report and the associated photographs, the patient's written informed consent was acquired. The Editor-in-Chief of this journal can examine a copy of the written consent upon request.

Declaration of competing interest
Nothing to declare.

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