

Vulvar malignant melanoma: a rare tumor with worse prognosis

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ABSTRACT

Malignant melanoma, which has a highly malignant potential, is a tumor of the skin and mucosal membranes. Malignant melanomas of the female genital tract, including the vulva and vagina, are rare. Their overall prognosis is worse. A 75 year old woman presented with complaint of growth in vulvar region since 4 months. There was history of itching in vulvar region over growth. Surgery is still the best available treatment for the control and potential cure of malignant melanomas.

Keywords: Malignant melanoma, Vulva, Vagina

INTRODUCTION

Melanoma of the vulva is the second most common malignancy arising within the vulva and accounts for 8-10% of all vulvar malignancies. Vulvar melanoma is a disease of the elderly, and its incidence peaks in the fifth to eighth decades of life.¹ It develops more commonly among Caucasians than among African-American, Asian, or other more heavily pigmented races.² Mean age of occurrence is 69 years. Also, 5 year survival is rare as it is associated with worse prognosis.

CASE REPORT

A 75 year old woman, married for 58 years, P6L6, presented to our hospital with complaint of growth in vulvar region since 4 months. Watery discharge was associated with growth since then. Also, there was history of itching in vulvar region over growth. Patient was apparently all right 4 months back. Growth was small in size initially and has progressively increased in size over 4 months. Itching has also increased. Patient also gave history of loss of weight, loss of appetite and fever. There was no history of bleeding from the growth. No history of headache, bowel and bladder complaints. Patient was a known case of hypertension and was taking medication.

Patient was menopausal since 17 years as vaginal hysterectomy was done due to prolapse.



Figure 1: Photograph of the lesion.

General physical examination showed a lean and thin built, cachexic lady weighing 48kg with no pallor, edema, cyanosis, clubbing. Her vitals at the time of admission were: Pulse- 82, BP- 146/94mmHg; Heart and

lungs, no abnormality detected. Inguinal lymph nodes were non tender and not enlarged. On local examination, a polypoidal growth of approx. 1.5 x 2cm seen near posterior fourchette; firm in feel (Figure 1). Per speculum examination showed senile vaginitis. Rectal examination was normal.

Her routine investigations were within normal limits. All viral markers (HIV, HBsAg, VDRL and HCV) were non reactive. Imaging studies revealed no evidence of metastasis. Thus, an excision biopsy of the lesion was done. Specimen was sent for histopathological examination. Histopathology report showed malignant melanoma (Figure 2A, 2B). Thus, final diagnosis of vulvar malignant melanoma with stage IB (FIGO staging) was made. The patient was followed up later after 3 months and showed no signs or symptoms of any recurrent disease.



Figure 2A: H&E; X100; Showing malignant cells with pigment in the cytoplasm.



Figure 2B: H&E; X400; Higher magnification showing highly pleomorphic tumor cells with pigment.

DISCUSSION

Vulvar melanomas are rare. Malignant vulvar melanoma will arise most commonly from the labia minora, labia majora, or clitoris.³ In addition, pigmented vulvar neoplasia may include VIN, squamous carcinoma, and Paget disease. Thus, tissue sampling is necessary, and immunohistochemical studies and electron microscopy may help to clarify the diagnosis.

The Table 1 shows histological incidence of types of vulvar malignancies.

Table 1: Histological incidence of types of vulvar malignancies.

Tumor Type	Percent
Epidermoid	86.2
Melanoma	4.8
Sarcoma	2.2
Basal cell	1.4
Bartholin gland	
Squamous	0.4
Adenocarcinoma	0.6
Adenocarcinoma	0.6
Undifferentiated	3.9

Vulvar melanomas have been staged by a variety of microstaging systems, including the Chung, the Clark, and the Breslow systems (Table 2). The Clark system of staging cutaneous melanomas is based on depth of invasion. Agreeing that depth of invasion is important, Breslow published an alternative list of prognostic indicators but added tumor size and used tumor thickness as the most significant measures. Both the Clark and the Breslow systems have been found to correlate with prognosis in patients with cutaneous melanoma.

Table 2: Microstaging of vulvar melanomas.⁴

	Clark levels	Chung et al	Breslow
I	Intraepithelial	Intraepithelial	<0.76 mm
II	Into papillary dermis	<= 1mm from granular layer	0.76-1.50 mm
III	Filling dermal papillae	1.1-2mm from granular layer	1.51-2.25 mm
IV	Into reticular dermis	>2mm from granular layer	2.26-3.0 mm
V	Into subcutaneous fat	Into subcutaneous fat	>3 mm

The five year survival of patients with vulvar melanoma was 35% with an average age 67 year at presentation.⁵ For vulvar melanoma, the most common patterns reported differed from cutaneous melanoma in most series, with mucosal lentiginous being the most common (27-57%) followed by nodular (22-28%), unclassified (12-16%), and superficial spreading (4-56%).⁶

The main prognostic factors are demographic characteristics, tumor localization, presence of groin node metastases, and various tumor characteristic. Age, stage and lymph node involvement were found to be significant factors affecting survival in patients with vulvar melanoma.⁷

The recommended treatment for vulvar melanoma has been radical vulvectomy with bilateral inguino-femoral lymphadenectomy, regardless of lesion size, thickness, or depth of invasion.^{8,9} The recommendations for treatment of vulva melanomas with thin lesions (<1 mm) are wide local excision with a safety distance of 1 cm and, with deeper lesions, an en bloc resection with safety distance of 2-3 cm with regional (inguino-femoral) lymphadenectomy.¹⁰

Therefore, vulvar melanoma carries a poor prognosis and show a tendency to recur locally and development distant metastasis through hematogenous dissemination. Deaths from vulvar melanoma more commonly result from the effects of widespread metastatic disease, most commonly involving the lungs, liver or brain.

There is a lack of consensus in the published literature regarding the treatment options. Surgery is still the best available treatment for the control and potential cure of malignant melanomas. However, the therapy should be tailored to meet the specific needs of individual patients. Dacarbazine is the most active chemotherapeutic agent, which can produce response rates of 15-25%.

CONCLUSION

The prognosis for malignant melanoma of the female genital tract is poor, regardless of the treatment delivered, though it can be improved if the disease is diagnosed early. As there is currently no proven standard therapy; so, therapy should be tailored to meet the specific needs of individual patients.

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