VULVAR MELANOMA- A CASE REPORT AND SHORT LITERATURE REVIEW

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Abstract
Malignant melanomas of the vulva and vagina are rare tumors located in areas of the body not exposed to ultraviolet radiation. It is the second most common vulvar malignancy but represents less than 1% of all melanomas. Because the overall prognosis is poor, it represents a significant issue in women’s health. Because of the inherent difficulty in performing large studies of vulvar melanoma, many questions about the biology, etiology, risk factors, and optimal management remain unanswered. Research for vulvar cancer is ongoing. We are presenting here a case report and short literature review of the case

Keywords: Malignant melanomas; ultraviolet radiation; Cancer

1. Introduction
Cancer of the vulva is not a common disease. It accounts for approximately 4% of cancers in the female reproductive organs and 0.6% of all cancers in women¹. It occurs in about 1.5 per 100,000 women-years in developed countries but is 2-3 times more frequent in underdeveloped countries. There are about 4,000 new cases each year in the United States. Although it can occur in women in the third and fourth decade it is usually diagnosed in older women between 65 to 75 years. Over 95% of vulvar cancers arise from the squamous epithelium. The remainder are mostly melanomas². Vulvar melanoma can appear as a darkly pigmented growth. A change in a mole that has been present for years can also indicate melanoma. Although certain signs and symptoms may strongly suggest vulvar cancer, many of them can be caused by conditions that aren’t cancer. The only way to be certain that cancer is present is to do a biopsy. Vulvar melanoma is no different from melanomas that occur elsewhere on the body. They are unpredictable and can be very aggressive. As the overall prognosis is poor, it represents a significant issue in women’s health. They are treated surgically if possible. The regional lymph nodes are usually removed at surgery³. A small primary lesion on the vulva (ie, < 2 cm) with superficial invasion (ie, < 1 mm from the epithelial stromal junction of the adjacent, most superficial dermal papillae) has essentially no risk of lymph node metastasis. Consequently, these lesions can be treated with wide local excision, ensuring that adequate surgical margins are present (not only on the skin but also deep margins). In larger lesions (ie, stage IB or greater or with stromal invasion >1 mm), the incidence of ipsilateral inguinal lymph node involvement increases as the depth of invasion, as well as the gross size, increases. Consequently, inguinal lymphadenectomy is part of the primary surgical procedure. This can be performed through a small separate inguinal incision, removing the lymph nodes above the cribriform fascia and in the opening of the fascia at the fossa ovalis. If the results are negative on frozen section of these lymph nodes, then a modified partial vulvectomy is the only treatment necessary. If the results on frozen section of the ipsilateral lymph nodes are positive, then most physicians suggest removing the lymph nodes on the contralateral inguinal area as well⁴.
The lesion itself can be treated conservatively, with a partial vulvectomy. Performing complete vulvectomy is an outdated treatment unless the cancer is present bilaterally. If clitoral involvement is present, lymphatic drainage can be direct to the pelvic lymph nodes. Studies have demonstrated that, even with clitoral involvement, the deep lymph nodes are not involved unless the inguinal nodes have evidence of metastasis also. Pelvic lymphadenectomy is largely discontinued, even in cases of lymph node involvement. A large, prospective, randomized study conducted by the Gynecologic Oncology Group (GOG) noted that patient survival rates are better if the pelvic and inguinal area is treated with radiation postsurgically, as compared to patients treated with pelvic lymphadenectomies, even when the pelvic nodes are not involved\(^5,6\). Incidence of lymph node metastasis seems to be increased if vascular lymphatic space is involved. In women with locally advanced vulvar cancer, no significant differences in survival or adverse events are found when primary chemoradiation or neoadjuvant chemoradiation were compared with primary surgery\(^7\).

### 2. Case report

A 55 year old, 5 years postmenopausal woman, Para3, with 3 living children, came with black colored swelling on vulva at right labia majora since 3 months. She was apparently alright 3 months before when she noticed a black colored swelling which suddenly increased in size in last one month. She also noticed a similar kind of swelling little smaller in size at perineum after a month of the first swelling. She did not give any history of previously existing mole. She went to the private practitioner where diagnosis of infected naevus was kept with suspicion of malignancy. Biopsy was taken and woman was referred to tertiary care center. She did not report to any hospital after that as she thought that the disease had been removed during biopsy, but in 15 days time she noticed a similar swelling much bigger in size along with another swelling at perineum, then she rushed to the present tertiary care medical institute. On examination she was cachexic, pale and under weight. There was significant inguinal lymphadenopathy, systemic examination was normal, abdominal examination too was normal. Local examination revealed two black colored masses of 4x4 cm and 3x4 cm size on labia majora and perineum respectively. Both the swellings were circumscribed, non tender, not fixed to the deeper tissue and firm in consistency. There was significant inguinal lymphadenopathy on both sides. Per speculum, bimanual and rectal examinations did not reveal any abnormality. The biopsy taken by private practioner confirmed it to be Melanoma. Operation planned was Radical vulvectomy with possible chemo-radio therapy depending on the report. Patient was thoroughly investigated where she was detected to have T wave inversions in ECG and she could not get medical fitness for surgery. So an alternative management strategy was planned for her in order to avoid risk of her mortality. Local excision of the growth keeping 1.5 cm clear margin and depth too was done and she was given external beam radiotherapy for 25 days. A close follow up was done and after two weeks of cessation of radiotherapy the clinical examination revealed good vulval scars and no lymphadenopathy. She was advised 3 monthly follow up.

### 3. Discussion

Malignant melanomas of the vulva and vagina are rare tumors located in areas of the body not exposed to ultraviolet radiation. Due to the small numbers of cases per year, most of the information is derived from retrospective analyses or literature review. A study done in Michigan University of 20 patients with vulvar malignancy revealed the mean age at diagnosis of 56.4 years. Family history was specifically recorded for 19 of 20 patients. Three (15\%) of 20 reported a family history of melanoma in a blood relative (brother, uncle, cousins). Eleven of 20 had documented status of atypical nevi elsewhere on the skin. Two (18\%) of these 11 reported at least 1 atypical nevus on nonvulvar skin (the trunk most common). The location of the primary vulvar melanoma was the labium majus (5\%), the labium minus (11\%), both labia majora and labia minora (5\%), labia not otherwise specified (16\%), clitoris or periclitoris (26\%), labia and clitoris or periclitoris (21\%), and vulva not otherwise specified (16\%). The histologic subtypes were superficial spreading (45\%), mucosal lentiginous (30\%), unclassified (15\%), and nodular (10\%). The mean Breslow depth of the 21 melanomas was 2.8 mm, with 2 (10\%) cases of in situ melanoma (range, in situ to 12.3 mm). The mean depth of the invasive lesions was 3.1 mm. Eleven (58\%) of 19
Invasive melanomas were ulcerated, according to histologic interpretation: 1 (17%) of the 6 lesions with a Breslow depth of 1 mm, 2 (50%) of the 4 with a Breslow depth of 1.1-2 mm, 3 (75%) of the 4 with a Breslow depth of 2.1-4 mm, and all (100%) of the 5 with a Breslow depth of 4 mm. Ten patients underwent sentinel lymph node biopsy with a combination of radio colloid and vital blue dye. Two (20%) underwent bilateral inguinal sentinel lymph node biopsy, while 8 (80%) underwent unilateral nodal drainage and sentinel lymph node biopsy. Melanomas initially were noted by a health care provider during routine examination in 3 (15%) of the 20 patients (in situ, 0.83 mm and 4.5 mm). The most common signs and symptoms, often multiple in the same lesion, were pruritus (23%); lump, bump, mass, swelling, or abscess (23%); bleeding (17%); lesion noted because of a visible skin change (17%); pain (7%); dysuria (7%); non healing sore or spotting (3%); and alteration of the urine stream (3%). Multiple signs and symptoms occurred in several patients, resulting in total percentages of 100%. Molecular characterization of the melanocortin type 1 receptor was performed by means of the sequencing of genomic DNA as part of an ongoing study (genes, environment, and melanoma) in one patient in our series, and a germ line mutation was found in melanocortin type 1 receptor (R151C) polymorphism. Sites for melanoma were Labium majus 27%-51% 5%, Labium minus 11%-37% 11%. Both labia majora and Minora, 21% 5% Clitoris or pericutioris 12%-31% 26%. Presenting signs and symptoms were bleeding 17%-42%, 17% Mass, lump, swelling, abscess, 28%-72% 23% Discharge 20% 0% Pruritus 2%-19% 23% Pain 7% 7% Lesion found at routine examination Not reported 15% Lesion noted visually by patient Reported without defined frequency 17% Dysuria Reported without defined frequency 7% .Nonhealing sore, spotting Reported without defined frequency 3% Alteration of urine stream Reported without defined frequency 3% Foul odor Reported without defined frequency 0% . Weinstock published data of about 203 cases of vulvar melanoma, diagnosed the incidence of 0.108 per 100,000 persons, a 5-year survival of 50%, and an average age at presentation of 66 years. Another study, by Ragnarsson-Olding et al., was a review of 219 cases of vulvar melanoma in Swedish women diagnosed between 1960 and 1984. Incidence in this study was reported as 0.27 per 100,000 in 1960-1964 and 0.14 per 100,000 in 1980-1984, a 3% decrease per year. The 5-year survival of patients with vulvar melanoma was 35%, with an average age of 67.7 years at presentation. The 1998 National American Cancer Data Base report from the national tumor registry of 84,836 cases of melanoma between 1985 and 1994 provides additional information. In this study, 1.3% of melanomas were mucosal (oral, female genital, and anorectal). Of these, only 18.0% were of the female genital tract, including vaginal melanoma, which represents approximately 0.23% of the total number of melanomas. The average age at diagnosis was 70-79 years for mucosal melanoma. Forty-nine percent of all mucosal melanomas were in patients 70 years old. This slight age discrepancy with previous studies specific for vulvar melanoma, with an average age at diagnosis of 58-67.7 years, may be the result of grouping all mucosal melanomas together. The youngest mean age reported in was 50.8 years, calculated from a study of 6 patients. Contiguous nevi are more commonly observed with superficial spreading melanoma than other histologic subtypes. Overall, only 6% of vulvar melanomas are reportedly associated with contiguous nevi. Most vulvar melanomas arise on glabrous skin.

The ABCD rule can be used to help tell a normal mole from one that could be melanoma. Asymmetry: One-half of the mole does not match the other. Border irregularity: the edges of the mole are ragged or notched, Color: The color over the mole is not the same. There may be differing shades of tan, brown, or black, and sometimes patches of red, blue, or white, Diameter: The mole is wider than 6 mm (about 1/4 inch). The most important sign of melanoma is a change in size, shape, or color of a mole. Still, not all melanomas fit the ABCD rule. Melanomas are characteristically black in color, however there are amelanotic melanomas that are not pigmented and can be confused with the usual squamous cell cancer.

The evolution and application of staging for vulvar melanoma, with far fewer numbers of patients, has lagged behind that of cutaneous melanoma. In the past, vulvar melanoma was grouped with other vulvar cancers and staged with the International Federation of Gynecology and Obstetrics system, which, though particularly appropriate for squamous cell carcinoma, does not reflect prognosis for vulvar melanoma.
Chung systems of staging have better correlation with prognosis than the International Federation of Gynecology and Obstetrics system. The new Breslow depth stratification ranges in the American Commission for Cancer staging system more accurately reflect outcomes for cutaneous and probably vulvar melanoma than the previous Breslow depth stratification ranges. Chung staging is primarily based on Clark level of tumor invasion. However no staging system is perfectly applicable nor is any one system consistently applied to the staging of vulvar melanoma.

Conclusion
Vulvar melanoma is the second most common vulvar malignancy and has an overall poor prognosis, usually with later detection than cutaneous melanoma. The data on vulvar melanoma consist almost entirely of case reports and small retrospective series. A non standardized approach to vulvar melanoma, characterized by the use of multiple staging systems and outdated prognostic parameters, exists in the literature. Vulvar melanoma behaves biologically similarly to cutaneous melanoma and has similar prognostic factors, despite having what is most likely a unique pathogenesis. It is logical to take advantage of the wealth of information gathered from large numbers of patients with cutaneous melanoma and apply this to vulvar melanoma when possible. Standardized and complete documentation of clinical and histologic findings is needed to advance knowledge and facilitate grouping of cases from different institutions for more statistically powerful conclusions. Contrary to squamous carcinoma, melanoma has a high risk of metastasis.

References
Figure 1 Showing Gross specimen of Melanoma of Vulva

Figure 2 Operative Specimen showing Vulvar Melanoma