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A RARE CASE OF VULVAL DESMOPLASTIC MELANOMA

Gynaecology

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ABSTRACT

Desmoplastic Melanoma is a rare variant of spindle cell Melonoma. Vulval Desmoplastic Melanoma is malignant cancer of the vulva accounts for approximately 1% of all cases of Melanoma.

Materials and Methods: This is a case report of 60 years old female presented with pigmented lesion over the vulval region.

Treatment: Bilateral vulvectomy with groin dissection was done followed by Intensity Modulated Radiotherapy (IMRT)

Results: Histopathological examination showed pT4aN0M0 stage Desmoplastic Melanoma. The tumor was positive for S-100 & SOX-10, Ki-67+.

The spindle shape of neoplastic melanocytes, the prominent desmoplasia and the frequent neurotropism of neoplastic melanocytes are its most characteristic histopathological features.

KEYWORDS

VMM, Vulvectomy and groin dissetion, IMRT.

INTRODUCTION:

Malignant melanomas of the vulva are mucocutaneous Melanocytic tumors located in areas of the body not exposed to ultra violet radiation, accounts for around 10% of all malignant tumors involving the vulva.

Cancer of the vulva is a rare entity and accounts for 1%-5% of all genital tract cancers. Vulvar melanoma is the second most common vulvar malignancy and represents a significant women's health issue. Vulvar malignant melanoma accounts for 3%-5% of all vulval tumors. Desmoplastic melanoma accounts for approximately 1% of all cases of melanoma.

The incidence of vulvar melanoma is 0.1 per 1,00,000 individuals presenting typically in post menopausal women. Patients with vulvar Melanoma due to the lack of awareness and neglect usually present with the disease at a late stage and have a poor overall prognosis, with reported 5-year survival rates ranging from 8% - 61%.

Vulvar Malignant Melanoma (VMM) is a rare disease, accounting for 5% of all vulvar malignancies and is characterized by low survival and high recurrence rates. Prognostic factors are higher age, advanced Breslow thickness and lymphnode involvement. Central localisation and ulceration status are still under debate. Surgery is the treatment for primary VMM. Elective lymphnode dissection and the value of sentinal lymphnode biopsy in VMM is still being studied. Radiation therapy and Chemotherapy as adjuvant treatment. In metastatic VMM Checkpoint Inhibitors and Targeted Immunotherapy have shown clinical efficacy.

CASE PRESENTATION:

60 years old female presented with complaints of pigmented lesion over the vulval region since 6 months associated with itching vulva since 2 months. She is a known case of Hypertension, Diabetes Mellitus and Psoriasis on regular treatment.

On examination, patient is conscious and coherent, pallor present, pulse rate 90/min, Blood Pressure 140/90 mm of Hg, CVS/RS: No abnormality detected. On local examination, it was a dark brown lesion measuring 3x1 cm on the right side of vulva near the fourchette, irregular surface, clear margins, not bleeding on touch, introitus is narrowed, Psoriatic lesions noted on both lower limbs, no inguinal lymphadenopathy.

P/S: Senile vaginitis, vaginal rugosity lost, cervix normal.
P/V: Uterus normal size, anteverted, no forniceal fullness, no forniceal tenderness.

TREATMENT:

Bilateral vulvectomy with right groin dissection was done. Wide excision biopsy was done and subjected to Histopathological Examination, it was Desmoplastic Melanoma of vulva, it was a pT4aN0M0 stage Desmoplastic Melanoma. The tumor was positive for S-100 and SOX-10, Ki-67+.

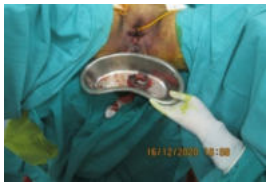

Fig.1: Vulvectomy and Wide excision biopsy

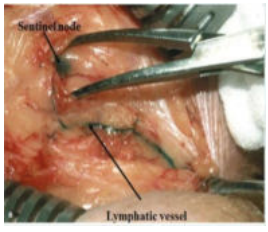

Fig-2: Groin dissection with sentinal lymphnode biopsy

Hematoxilin and Eosin stains show keratinizing stratified squamous epithelium with parakeratosis, acanthosis and basal pigmented cells. Below epithelium there is inflammatory infiltrate consisting of lymphocytes, few polymorphs and plasma cells along with fibrocollagenous connective tissue. At foci, there are few atypical cells showing moderate cytoplasm with pleomorphic, hyperchromatic irregular nuclei and few showing prominent nucleoli. Occasional atypical mitotic figures are seen. There are areas of nuclear debri admixed with inflammatory cells.

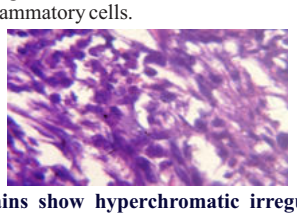

Fig-3: H&E stains show hyperchromatic irregular nuclei and prominent nucleoli

Post operatively she was given Intensity Modulated Radiotherapy (IMRT).

DISCUSSION:
In our case, patient had Hypertension, Diabetes Mellitus and Psoriasis which are Predisposing risk factors. Desmoplastic Melanoma clinically presents as a firm papule, nodule or plaque without distinguishing color characteristics and nondescript features that can easily be mistaken for a scar, fibroma or cyst. Desmoplastic Melanoma occurs most commonly on the head and neck of elderly persons, but it may occur anywhere.

Because of its nondescript clinical features, Desmoplastic Melanoma is often diagnosed at more advanced levels of dermal invasion. It often involves nerve fibres, when it is called neurotropic melanoma. The malignant cells within the dermis are surrounded by fibrous tissue. It sometimes arises underneath a lentigo maligna, a type of melanoma in situ.

The ratio of Vulvar Malignant Melanomas when compared with cutaneous melanoma is 1:71. It occurs predominantly in postmenopausal women, contrary to other site melanomas which occur before 45 years.

Clinically, patients present with asymptomatic pigmented lesion, the usual clinical form is Superficial Spreading Melanoma, the much lesser common form is nodular which is more aggressive and has a poorer prognosis, the histological types in order of incidence, the superficial spreading (40-58%), mucosal lentigenous (27-57%), nodular (22-28%) and unclassified (12-16%).

STAGING: Staging is by the Breslow, Clark, and Chung systems of staging included in the AJCC (American Joint Committee on Cancer). The kaplan - Meier method is one of the method used to analyze disease specific and overall survival.

TREATMENT: The treatment of vulvar cancer depends primarily on histology and staging**.** Other variables influencing management are age, coexistence of comorbidities and performance status of the patient. Treatment is predominantly surgical, chemoradiation for advanced tumors, exenteration to achieve adequate surgical margins. Management should be individualised. Cisplatin and 5-Fluorouracil are effective in advanced vulvar cancer.

Radical vulvectomy with bilateral lymphadenectomy is becoming questionable. Alternatively, wide local excision and regional lymphadenectomy (inguinal - femoral) is recommended.

Surgery remains the primary treatment modality for all locally resetable melanomas. A surgical margin of 0.5 - 1.0cm for melanoma insitu, 1cm for invasive melanoma with a Breslow thickness $\leq$ 1mm, 1-2cm for a Breslow thickness of 1.01 - 2mm, and 2cm for a Breslow thickness of $\geq$ 2.01mm is generally recommended. The lymphnode status was the most important independent predictor of survival. All women with malignant melanoma of female genital tract > 1mm depth of invasion to undergo sentinal node biopsy. Those with positive lymphnodes may be candidates for adjuvant treatment.

ADJUVANT TREATMENT:
Adjuvant treatment with Immunotherapy, Chemotherapy and Radiotherapy may be recommended in specific cases and for local recurrences and distant metastases.

CHEMOTHERAPY:
There are limited therapeutic agents for advanced, metastatic, and/or recurrent melanomas not amenable to surgery. The only U.S Food and Drug Administration (FDA) approved agents for many years were dacarbazine and high-dose Interleukein-2 (IL-2). Recently Ipilimumab and Vemurafenib gained FDA approval.Traditional systemic cytotoxic therapies, including Platinum/Taxane regimens, can be used, but with response rates of approximately 20% at best.

TREATMENT STUDY:
1. The use of Vemurafenib in melanomas harboring a BRAF V600E mutation resulted in a dramatic improvement in progression free survival compared with dacarbazine in a phase - III trial.
2. A current phase - II trial from the Eastern Cooperative Oncology group (ECOG) is assessing the use of dasatinib, a multityrosine kinase inhibitor in melanoma harboring a C-KIT mutation (E2607)

PROGNOSIS:
The main prognostic factors include tumor size, depth of invasion, presence of ulceration and presence of lymphnode metastasis. In addition, the patient's age and tumor stage also appear to be critical. Some studies show prognosis of vulvar melanoma is worse than that of extra genital melanoma.

a) Breslow's depth,
b) An absence of any of the pathological risk factors, such as satellitosis, in-transit metastasis, Lymphovascular Space Invasion (LVSI) and dermal mitosis,
c) Removal of inguinofemoral lymphnodes,
d) Lateral margin of > 1cm, and
e) C-KIT expression as valuable prognostic predictors for disease free survival. Lichen sclerosus may be associated with vulvar melanoma.

PROGNOSIS STUDIES:
An American study demonstrated that increasing Breslow depth was associated with declining survival, whereas other histopathological features such as ulceration, increasing mitotic index and the presence of atypical melanocytic hyperplasia were not associated with a significant difference in survival. A recent Chinese study revealed that macroscopic tumor growth and treatment method were independent prognostic factors for overall survival.

CONCLUSION:
Desmoplastic Melanoma exhibits an increased risk for local recurrence after wide excision. With regard to the probability of local recurrence and related clinical management, a number of factors play a role, in particular margin clearance and perineural invasion. With positive or suboptimal surgical margins and/or perineural invasion, consideration may be given to radiation therapy. For patients with metastatic desmoplastic melanoma, immunotherapy provides hope.

Vulvovaginal melanomas must be studied in much greater detail. The role of inguino femoral lymphadenectomy needs to be determined. There is a need to find an acceptable therapeutic regimen for locally advanced vulvovaginal melanomas.The use of novel immun otherapies and targeted therapies specifically in vulvovaginal melanomas needs to be assessed.

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