



## WHAT SHOULD I KNOW? Vulvar and Vaginal Melanoma



### DESCRIPTION OF THE TUMOR/CANCER TYPE

Vulvar and vaginal melanomas are rare accounting for 5% of vulvar malignancies and 2.4-2.8% of all vaginal malignancies. They develop by malignant transformation of melanocyte cells that are formed in early development as an embryo. Vulvar and vaginal melanomas are considered mucosal melanomas and are more likely to have recurrence and an aggressive clinical course than cutaneous melanomas. Prognostic factors include age, tumor size, thickness of the tumor and lymph node involvement.

### SIGNS & SYMPTOMS

Vulvar and vaginal melanomas can present with a vulvar lesion, vulvar bleeding, irritation or pain. The majority of vulvar and vaginal melanomas are pigmented but may be colorless or have other appearances. Melanomas can have irregular margins or can increase in size.

### SCREENING

There are not screening guidelines for vulvar or vaginal melanoma, but any lesion that is increasing in size, pigmented, has irregular margin especially in a postmenopausal patient should be considered for biopsy or excision.

### TREATMENT AND FOLLOW UP

**Role of Surgery:** Surgery has been the standard for treatment of vulvar and vaginal melanoma. For cutaneous melanomas a margin of 1-2 cm is recommended but recent data suggest no improvement in survival in vulvar and vaginal melanoma based on the extent of negative margin. Evaluation of inguinal lymph nodes by sentinel lymph node evaluation is typically done for cutaneous melanomas but because of the rarity of vulvar and vaginal melanoma specific data concerning sentinel lymph node evaluation is limited.

**Role of Radiation Therapy:** Radiation therapy has been utilized in vulvar and vaginal melanoma for patients with lymph node

involvement at high risk of recurrence as well as in the setting of recurrence or unresectable disease for relief of symptoms or for patients who are not surgical candidates

**Role of Chemotherapy:** Targeted therapies or immunotherapy are preferred, but selected chemotherapies are options for patients who are not candidates for targeted therapies or immunotherapy.

**Targeted Therapy options:** In patients with BRAFV600 mutation in the tumor combination therapy with dabrafenib/trametinib versus vemurafenib/cobimetinib versus Encorafenib/binimetinib are options  
Tumors with activating mutations of KIT can be treated with Imatinib  
Larotrectinib or entrectinib for NTRK gene-fusion positive tumors

NRAS mutated tumors if progression on immunotherapy can be treated with binimetinib

**Immunotherapy options:** If systemic therapy is indicated immunotherapies are the preferred regimens including pembrolizumab or nivolumab versus nivolumab/ipilimumab. Pembrolizumab/low-dose ipilimumab can be used if progression after anti-PD-1 therapy. Other options include ipilimumab or high dose IL-2 therapy

### QUESTIONS TO ASK YOUR CARE TEAM

Ask about eligibility for clinical trials if applicable.

Clinical and family history may identify patients who are at risk for other melanomas or malignancies and who may benefit from genetic testing

If you are not already being treated by a gynecologic oncologist, consider seeking a second opinion.

Things to consider when getting a second opinion

include getting a pathology review by a gynecologic or dermatopathologist as well as presentation at a tumor board.