UNIT 5: NEOPLASIA

Educational Topic 51:
Vulvar Neoplasms

Rationale: Early recognition and proper evaluation of vulvar neoplasms can reduce morbidity and mortality.

Intended Learning Outcomes:
A student should be able to:

- List the risk factors for vulvar neoplasms
- Describe symptoms and physical examination findings of a patient with vulvar neoplasms
- List the indications for vulvar biopsy

TEACHING CASE

CASE: A 67 year-old woman presents with the complaint of a pruritic area on the right side of her vulva. She has noticed this for about three months, and has used a variety of over-the-counter creams, including imidazole and corticosteroid preparations, without success. She underwent menopause at age 52, and tried hormone replacement therapy for three years, but discontinued this due to irregular bleeding. The bleeding stopped when she stopped the hormones. She does have a history of abnormal Pap smears, including a cervical conization at age 35. Her last Pap was approximately 7 years ago. The patient has a long-standing history of hypertension and takes a beta-blocker. She smokes 1 pack cigarettes per day, and has done so for 30 years. Her general physical examination is essentially unremarkable. Examination of her groin lymph nodes reveals no palpable adenopathy. Examination of the external genitalia reveals a 1 cm raised, firm, irregular, lesion on the right labia majora. Excoriations are also noted adjacent to this lesion. The rest of the vulva is notable only for atrophic changes. The vagina is also atrophic, and the cervix is grossly normal. The uterus is small and the ovaries are nonpalpable.

COMPETENCY-BASED DISCUSSION & KEY TEACHING POINTS:
Competencies addressed:
- Patient Care
- Medical Knowledge
1. What risk factors does this patient have for a vulvar neoplasm?

- Vulvar neoplasms encompass both pre-invasive diseases of the vulva (Vulvar Intraepithelial Neoplasia – VIN 1, 2 or 3) and invasive vulvar cancers (squamous cell carcinoma – 90%; melanoma – 5%; Bartholin gland adenocarcinoma; Paget’s disease of the vulva; sarcoma). VIN is associated with genital HPV infection, history of abnormal pap smears, immunosuppression, and smoking. Vulvar cancer can develop from progression of untreated or incompletely treated VIN and may also be associated with increasing age, chronic vulvar dermatoses and pruritis, history of VIN as well as smoking.

- The most common presenting symptom is pruritis, but other symptoms may include pain, discharge, bleeding, dysuria, or the discovery of a vulvar nodule or mass.

- Lichen sclerosis is associated with vulvar squamous cell carcinoma (up to 5% incidence), so close follow-up is recommended in these patients.

- Two types of vulvar cancer have been described including Type 1 which typically affects younger women (ages 35 – 65) and is associated with HPV, VIN, and smoking, and Type 2 which typically affects older women (ages 55 – 85) and is associated with chronic vulvar inflammation/lichen sclerosis, and less so with HPV and smoking.

2. What are the essential steps in evaluating a patient with these vulvar complaints?

- Thorough pelvic examination with complete assessment of the entire external genitalia including the mons pubis, labia majora/minora, urethra, Bartholin’s and Skene’s glands, vaginal introitus, and perineum is essential. Findings of any lesions should be detailed based on location, size, number, and visual/palpable characteristics (ulceration, excoriation, nodularity, friability, color, consistency, tenderness, mobility). Because multicentric disease is commonly encountered, particularly in younger women, a complete examination should also include the cervix, vagina, and perianal area. Up to 50% of women with VIN will have antecedent or concomitant lower genital tract disease, usually cervical or vaginal intraepithelial neoplasia (CIN or VAIN), so speculum examination should be performed with visualization of the anterior, posterior, sidewalls, and fornix of the vagina as well as the entire cervix. Pap smear should be performed per the usual guidelines, but especially in this patient population. Bimanual and rectovaginal examination are also indicated. Palpation of the inguinal/groin lymph nodes should be performed with the patient in the supine position.

- Colposcopic examination of the vulva (and vagina/cervix) may also be performed to assess the extent of any vulvar lesion that is identified grossly. This involves application of 3-5% acetic acid to the vulva for 5 minutes and thorough assessment of the entire vulva, perineum, and anal areas.

3. What is the next step in the management of this patient? When should a vulvar biopsy be performed?

- A tissue diagnosis is essential at this point, and a vulvar biopsy is indicated. Vulvar biopsies are indicated any time a vulvar lesion is identified, especially if there is any question related to the diagnosis. Since HPV is associated with a number of vulvar epithelial disorders, including genital warts, VIN, and some vulvar carcinomas, distinguishing warts from vulvar neoplasia on the basis of appearance alone is not always possible because VIN can present as red, white, dark, raised, or eroded lesions. If the patient is immunocompromised or the diagnosis is uncertain (Paget’s disease can present as multiple bright red, scaly, eczematoid plaques, whereas melanoma can range in color from brown to bluish-black), biopsy should be undertaken.

- In general, a biopsy should be performed on hyperpigmented, indurated, fixed, or ulcerative lesions, or lesions that do not respond to medical treatment or worsen during treatment (for initially suspected warts or other dermatoses). If multiple vulvar lesions are identified, a representative biopsy should be per-
formed from the most concerning lesion(s) if they appear similar, or multiple biopsies should be obtained from various sites if they appear different. The biopsy can be performed in the office under a local anesthetic.

- Treatment depends on the final diagnosis. For VIN, treatment options include excisional or destructive methods such as simple vulvectomy/wide local excision, laser vaporization, ablation, and topical therapies such as imiquimod or 5-FU (rare). Choice of therapy depends on location and extent of lesions, grade/concern for invasion, age, and desire for sexual activity. For invasive carcinoma, treatment is more extensive, depends on the stage of cancer, and may include radical local excision, radical vulvectomy, inguinal lymphadenectomy, radiation, or chemoradiation therapy.

REFERENCES


ACOG Practice Bulletin 93, Diagnosis and Management of Vulvar Skin Disorders, May 2008 (Reaffirmed 2013).