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Key concepts in management of vulvar cancer

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Vulvar carcinoma is an uncommon tumor that is seen most often in older women. Subtle symptoms such as pruritus should prompt examination and targeted biopsy in all women as this disease can be successfully treated even in elderly, frail individuals. Vulvar cancer has a bimodal age distribution and is seen in both young and older women with risk factors including human papillomavirus (HPV) infection, smoking, and vulvar skin diseases (i.e., lichen sclerosus). This cancer is staged surgically, with an update in 2009 incorporating prognostic factors. The treatment of vulvar carcinoma has evolved to include more conservative surgical techniques that provide improved cure rates with emphasis on minimizing morbidity. Advanced and metastatic lesions are now treated with chemoradiation which produces substantial cure rates with decreased morbidity. Promising areas of research in vulvar cancer include refinement of sentinel lymph node biopsy, prevention of lymphedema, and preservation of sexual function following treatment.

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Introduction

Vulvar carcinoma is uncommon, representing only 5% of gynecologic malignancies and 1% of cancers in women. In 2013, there will be an estimated 4490 new cases of vulvar cancer and 950 deaths from this disease in the United States [1]. Although this cancer is most often seen in postmenopausal women, there are increasing numbers of young women with this diagnosis [2]. Human papillomavirus (HPV) infection and smoking increase risk for this cancer. Vulvar cancer can present with relatively subtle symptoms such as pruritus or dysuria. Awareness and recognition of this disease entity is particularly important because it can be successfully treated even in very frail individuals, and the success and morbidity of treatment is correlated with stage at diagnosis. Delay in the diagnosis of vulvar cancer can occur when lesions are not discussed secondary to patient embarrassment or when symptoms are treated without examination or histologic confirmation of the diagnosis.

Etiology

Over 90% of vulvar cancers are of squamous histology, although melanomas, basal cell cancers, sarcomas, and adenocarcinomas from Paget's disease or originating in the Bartholin's gland can also occur. Risk factors for the development of vulvar cancers include smoking, HPV infection, lichen sclerosus, immunosuppression, vulvar or cervical neoplasia, and northern European ancestry. Squamous cell carcinomas are the most common type of vulvar cancer and arise from one of two pathogenic mechanisms that correlate with age. Basaloid or warty type carcinomas are seen in younger women with HPV infection. Keratinizing squamous carcinomas are associated with inflammatory vulvar skin diseases such as lichen sclerosus and are generally found in older individuals. These keratinizing squamous carcinomas are not typically associated with HPV infection [3,4]. Although the incidence of vulvar cancer is fairly stable at 1% lifetime risk, the incidence of vulvar intraepithelial neoplasia (VIN) is increasing, and one study found that rates had tripled in younger women over the last two decades [2,5].

Clinical presentation

Most vulvar cancers present as a palpable lump or visible mass on the vulva with or without associated pruritus, discharge, dysuria, or bleeding. The lesion may be difficult to visualize secondary to labial agglutination from surrounding dystrophy. Rarely, vulvar cancers are asymptomatic. As coexistent lower genital tract neoplasia is found in 13% of cases, initial evaluation should include examination of the entire vulva for multifocal lesions as well as the rest of the lower genital tract (cervix and vagina) including cervical cytology [6]. Any suspicious lesion should be biopsied, and multiple biopsies of multiple sites or of the same suspicious lesions over time may be necessary to make the diagnosis. If the lesion is clinically suspicious and the pathology does not correlate with the clinical suspicion (for example, necrosis) then repeat biopsy may be warranted. The center of the lesion should be sampled for highest yield. If vulvar cancer is suspected, excision of the primary tumor prior to evaluation by an oncologist should be discouraged as it can preclude accurate estimation of tumor size, location, margin status, and make sentinel node evaluation difficult or impossible.

Evaluation prior to treatment

Evaluation of the patient prior to treatment must include a thorough assessment of the patient's medical history, functional status, and physical exam. Vulvar cancers spread by direct extension onto surrounding tissues and also by tumor emboli with lymphatic spread first to inguinal lymph nodes. The primary tumor must be carefully evaluated for its size and proximity to adjacent structures such as the urethra, vagina, and anus and fixation to bone. The vagina, cervix, and perianal skin should be evaluated for coexistent neoplasia and the bilateral groins and supraclavicular (scalene) nodes carefully examined for palpable adenopathy. Patients with significant discomfort may benefit from an exam under anesthesia, and in advanced cases, cystoscopy or proctoscopy may be of benefit to determine the extent of disease. Vulvar cancers may be multifocal and arise in a background of intraepithelial