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WNL we never looked: vulvar carcinoma incidence after screening cutoff

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ABSTRACT

The incidence of vulvar carcinoma increases with age, though elderly women receive less aggressive cancer therapies and fewer strategies aimed at cancer prevention. Furthermore, elderly women dual enrolled in Medicaid-Medicare experience poor survival rates for vulvar carcinoma. Herein, we provide recommendations for the prevention of and guidelines for the multidisciplinary care of vulvar carcinoma. Prevention of vulvar carcinoma can be categorized into primary, secondary, and tertiary prevention. Primary prevention consists of vaccination, secondary prevention consists of screening, and tertiary prevention is aimed at the management of premalignant and early-stage lesions.

Keywords: Medicaid, Medicare, National Comprehensive Cancer Network, prevention, vulvar carcinoma, vulvar lichen sclerosus, vulvar melanoma, vulvar squamous cell carcinoma

Vulvar carcinoma accounts for 5% of gynecologic malignancies and usually affects women over 65 years of age.[1–5] In 2019, there were 6,070 new cases of vulvar carcinoma and 1,280 deaths.[5–7] The incidence of vulvar carcinoma has been increasing by an average of 0.6% per year for the past 10 years, while relative survival is decreasing.[5] Although the incidence of vulvar carcinoma increases with age, and there is a projected increase in the elderly population (70% of United States cancer patients will be over age 65 by 2030), elderly women receive less aggressive cancer therapies, as well as fewer strategies aimed at cancer prevention.[8,9]

Squamous cell carcinoma (SCC) accounts for more than 90% of vulvar carcinomas.[1,2,4–6,10] Vulvar SCC arises from 2 pathways: (a) high-grade squamous intraepithelial lesion (HSIL) associated with cigarette smoking and human papillomavirus infection, and (b) vulvar intraepithelial neoplasia (VIN) associated with chronic inflammatory states such as lichen sclerosus (LS) or lichen planus (LP).[1–3,5,6,11] The risk of neoplastic transformation is non-negligible with an estimated incidence of vulvar SCC in genital LS between 3.5 and 7%, with a peak of increased risk in the first 1 to 3 years after diagnosis.[12–14] One study estimated the mean time between diagnosis of LS and VIN to be 0.6 years, while another noted a cumulative probability of progression to carcinoma of 1.2% at 24 months and 36.8% at 300 months.[12,13] This demonstrates the importance of lifelong surveillance.[13] In a review study of 14,268 women with vulvar LP, the rates of carcinoma, VIN, and HSIL were 0.3%, 2.5%, and 1.4%, respectively.[14] While HSIL is common in younger patients, VIN arises in women in their 60s to 80s.[6,11] It is estimated that 59% of vulvar SCC patients have localized disease, 30% regional spread to lymph nodes, 6% distant metastases, and 5% are unstaged.[5] A study of 6965 patients risk stratified by age found a 5-year survival rate of 87.5% for younger women (<50 years old) and 80.7% for older women ($P < .001$).[11] After controlling for race, stage, grade, and surgical treatment, older patients had a hazard ratio of 3.9 (95% CI [3.2–4.7]) for death.[8,11]

Melanoma is the second most common vulvar carcinoma, compromising 5 to 10% of cases, and portends a worse prognosis as compared to SCC.[5,6,10,15] Vulvar melanomas usually affect Caucasian (>85%) women in their 50s to70s.[5,6,15–18] The 5-year survival is estimated to be between 10 and 63%, with a median survival of 16 months in Black patients compared to 39 months in non-Black patients.[5,6,10,16,19] Most cases (51–77%) are localized at presentation, with nodal involvement occurring in 9 to 23%.[17]

Treatment for vulvar carcinoma ranges from wide local excision (WLE) with or without sentinel lymph node biopsy or dissection to radical vulvectomy, with or without radiation, chemotherapy, or immunotherapy.[2–4,6,7,20] For early-stage vulvar SCC (T1a, <1 mm invasion), WLE with 1.0 cm margins is recommended, though there is no recommendation for sentinel node

What is known about this subject in regard to women and their families?

- The incidence of vulvar carcinoma, especially in elderly women.
- Elderly women dual enrolled in Medicaid-Medicare experience poor survival rates for vulvar carcinoma.

What is new from this article as messages for women and their families?

- Dermatologists should be involved in the primary, secondary, and tertiary prevention of vulvar carcinoma.
- A vulvar tissue-sparing approach may be an appropriate treatment option for women with vulvar carcinoma.
- Treatment of vulvar carcinoma is multidisciplinary, and pelvic floor physical therapy is an underutilized part of the post-surgical treatment plan.

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biopsy or dissection at these early stages.[21] Historically, radical vulvectomy was performed to treat more advanced stages of vulvar SCC, though in patients with International Federation of Gynecology and Obstetrics stage IB and II disease, radial vulvectomy results in no significant benefit over conservative, modified, and individualized vulvectomies, and leads to severe genital disfigurement, complications, and a 2% surgical mortality rate.[10,18]

Similar to vulvar SCC, the surgical approach to vulvar melanoma has become more conservative.[6,15,16] Radical vulvectomy with lymph node dissection does not increase survival or clinical outcomes compared to WLE, though it does increase the incidence of complications including wound breakdown and dehiscence, stricture, dyspareunia, cystocele, rectocele, incontinence, lymphedema, sexual dysfunction, and impacts psychological wellbeing.[1,6,15,16,19,22] The annual costs of sentinel lymph node biopsy compared with lymph node dissection is $65.2 million and $76.8 million, respectively, though 3-year recurrence-free survival is similar (97.3% vs 96.9%) and lymph node dissection does not improve overall survival.[6,23] Additionally, less than one-third of patients have positive nodes, making surgery an avoidable cause of morbidity.[23]

A recent study by Kraus and coworkers in the *International Journal of Women's Dermatology* proposes a vulvar tissue-sparing technique as a treatment option for vulvar SCC to minimize psychologic, social, and sexual complications.[21] This option is proposed for HSIL and T1a vulvar SCC.[21] They base their proposal on the National Comprehensive Cancer Network guidelines, which include a section on penile tissue-sparing options such as radiation, chemotherapy, laser, and Mohs micrographic surgery.[21] The use of staged excision or Mohs micrographic surgery may also be a viable treatment option for vulvar melanoma.[6]

A cohort study of the North Carolina state cancer registry demonstrated that women >65 years old with vulvar/vaginal carcinoma dual enrolled in Medicaid-Medicare had an increase in all-cause mortality of 46% (HR, 1.93; 95% CI [1.36–2.72]) compared to the Medicare only insured group.[9] Of 11,153 vulvar SCC patients identified from the National Cancer Database from 1998 to 2004, age 60 years or older and Medicaid enrollment were associated with a greater risk of death within 5 years ($P < .01$).[7] Another query rendering 1,917 vulvar melanoma subjects from the National Cancer Database demonstrated improved survival in Medicare and private insurance enrollees (HR, 0.62; 95% CI [0.39–0.99]; $P = .05$).[10] Dual enrollment is a marker of low socioeconomic status (SES), as annual incomes at or near the federal poverty line must be met to qualify for Medicaid.[9] In the North Carolina study, a larger percentage of Black women were dual enrolled (35%) than in the Medicare-only group (10%).[9] However, in military and clinical trials where SES barriers are minimized and equal treatment is offered, vulvar/vaginal carcinoma mortality differences by race are not seen.[9] This finding highlights a disparity that makes it difficult for low-SES elderly women to survive vulvar carcinoma, which should be amenable to intervention.[9]

In referral centers, approximately two genital melanomas are diagnosed per year; however, delays may prevent patients from being referred and diagnosed in a timely manner.[20] Physicians may contribute to at least a 1-year delay in the diagnosis of vulvar carcinoma by providing treatment for the wrong diagnosis and delaying biopsy or referral.[8,22] Herein, we provide recommendations for the prevention of and guidelines for the multidisciplinary care of vulvar carcinoma.

Prevention of vulvar carcinoma can be categorized into primary, secondary, and tertiary prevention.[4] Primary prevention consists of counseling on smoking cessation and vaccination, aimed at preventing the incidence of HSIL and thus vulvar SCC.[4] With the advent of the human papillomavirus infection vaccination, approved by the Federal Drug Administration in 2006 for women aged 9 to 26 years, screening for cervical cancer, and thus overall examination of the female genitalia, was given a cutoff.[3,8,24] However, women who met the upper limit of vaccination criteria initially are now only in their 40s.[3,8,24] The cutoff for cervical cancer screening with Papanicolaou smears is age 65, leaving a gap of women who neither received the vaccination nor will receive continued screening.[3,8,24] With gynecological care and the frequency of examination becoming less frequent in this elderly population, the incidence of cervical as well as vulvar carcinomas is increasing.[8,24]

Secondary prevention consists of screening with a complete history and physical examination.[2–6] We also recommend serial photographs to improve detection. Though there are no specific screening guidelines for vulvar carcinoma, we recommend an annual examination of the external genitalia, even if no longer receiving Papanicolaou smears.[8] Up to 50% of patients may be asymptomatic, others feel embarrassed to disclose their symptoms, and elderly women are less likely to conduct home self-examinations, emphasizing the need for physician surveillance.[4,8,18] Furthermore, some elderly women are unable to conduct self-examinations due to limited mobility or obesity. The regular and lifelong use of ultrapotent topical corticosteroids can reduce the risk of vulvar carcinoma in chronic inflammatory states (ie, LS, LP).[14]

We recommend a biopsy of any clinically suspicious, evolving, or growing lesion of the vulva, and serial photographs of nonworrisome lesions.[2–4,6] Clues to a malignant diagnosis include asymmetry, scarring, architectural changes, signs of inflammation (ie, erythema, edema, lichenification), fissures, erosions, ulceration, and discharge.[18] We recommend either punch or snip excision for biopsy. At least a 4 mm punch biopsy at the edge of the lesion, including vital tissue rather than just necrosis or granulation tissue, is recommended.[2] Snip excision consists of inserting a loop of suture (ie, prolene for ease of visualization) to create tension, followed by snip excision of the taut skin with iris or gradle scissors. Wide excisional biopsy is not recommended as it may interfere with further treatment.[2] Physicians should note any changes to the vulva, lesion size, number, position, mobility, presence of infiltration, and safety margins in case of further treatment.[2] In cases of multiple vulvar lesions or in cases of LS, multiple biopsies are recommended as carcinoma may be obscured.[2,18,21] Dermoscopy is a useful tool for discriminating malignant from benign lesions.[16] Additionally, lymph nodes should be palpated and assessed for size, mobility, and consistency, and the skin overlying the lymph nodes should be examined.[2]

Tertiary prevention is aimed at the management of premalignant and early-stage lesions.[4] A multidisciplinary team approach including gynecology, oncology, and psycho-social rehabilitation should be sought.[22] It is important to ensure that surgery is tissue-sparing if possible, sparing the morbidity and disfigurement of radical vulvectomy.[22] After surgery, we recommend pelvic floor physical therapy. A study in the American Journal of Obstetrics and Gynecology found that pelvic floor physical therapy was more effective than lidocaine for reducing pain during intercourse ($P < .001$) and the improvement was clinically meaningful with 79% of women in the physical therapy group report being very much or much improved compared to 39% in the lidocaine group ($P < .001$).[25]

The incidence of vulvar carcinoma is increasing; however, we as dermatologists play a substantial role in the primary, secondary, and tertiary prevention of these cases.

Conflicts of interest

None.

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AG and JG: Study conception and design. AG: Data collection. AG and JG: Analysis and interpretation of results. AG and JG: Draft manuscript preparation. All authors reviewed the results and approved the final version of the manuscript.

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