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Vulvar lichen sclerosus often overlooked in women of reproductive age

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By Ted Bosworth

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NEW YORK – A delayed diagnosis of vulvar lichen sclerosus is common, and the risk of this delay is permanent scarring and structural genital changes, cautioned an expert who holds teaching appointments in both dermatology and obstetrics/gynecology.

Vulvar lichen sclerosus (VLS) should be considered in females of any age complaining of vulvar itching, including sexually active women for whom other diseases may be more likely, emphasized Dr. Bethanee J. Schlosser, director of the Vulvar Mucosal Specialty Clinic at Northwestern University, Chicago. She spoke at the American Academy of Dermatology summer meeting in a session jointly sponsored by the European Academy of Dermatology and Venereology.

Dr. Bethanee J. Schlosser

"We need to recognize that when women in their 20s and 30s complain of vulvar pruritus, it is not always vulvovaginal candidiasis," Dr. Schlosser emphasized. She said some clinicians do not even consider VLS in this age group, because they have been mistakenly informed that this disease has a bimodal distribution that restricts most cases to preadolescent girls and postmenopausal women. According to Dr. Schlosser, up to 40% of cases occur in women of reproductive age.

VLS is a variation on lichen sclerosus, which is a chronic inflammatory condition associated with epithelial thinning that can occur anywhere on the body. The disease is progressive. It is not an

erosive process initially, but Dr. Schlosser presented several cases that demonstrated secondary erosions and fissures can eventually result in permanent structural damage to the anatomy.

The etiology remains incompletely understood, but Dr. Schlosser said that VLS is now considered an autoimmune condition that is commonly associated with other autoimmune diseases, particularly thyroiditis. Dr. Schlosser advised that screening VLS patients for additional autoimmune disorders is appropriate.

The key issue, however, is making the diagnosis in the first place. The waxy plaques and epidermal wrinkling that characterize this disease may not be immediately distinguishable from other dermatologic lesions, particularly as the severity varies. Pruritus is the most common symptom, but up to 30% of women are asymptomatic, according to Dr. Schlosser. Histologic evidence of hyperkeratosis on biopsy in the context of characteristic clinical signs confirms the diagnosis.

Whether in girls, women of reproductive age, or postmenopausal women, the substantial gap between the median age of onset and the median age of diagnosis is a source of concern. In peri- and postmenopausal women, Dr. Schlosser cited data suggesting that the mean delay to a diagnosis can be 5 or more years. According to Dr. Schlosser, one source of delay may be the well-known reluctance of many patients to disclose genital symptoms, but she also maintained that VLS, which has an incidence of 0.1%-0.2%, is not often considered in the initial evaluation of vulvar dermatologic complaints.

The standard first-line therapy for VLS is highly potent corticosteroids. In contrast, topical testosterone, once widely used when VLS was thought to be a product of hormonal imbalance, “has no role in this disease,” according to Dr. Schlosser. Second-line treatments for those who need an alternative to steroids include topical calcineurin inhibitors, such as tacrolimus or pimecrolimus, the synthetic vitamin D cream calcipotriene, and topical retinoids. Oral steroids can be used in difficult cases, but Dr. Schlosser said this is uncommon, estimating that she may have placed only 4 of the 150 VLS cases she has accumulated in her clinic on a systemic therapy.

Generally, topical therapies, which must be maintained indefinitely, suppress symptoms and slow or halt the progressive disease process, according to Dr. Schlosser, who cited a large study published more than 10 years ago. In this study, 66% of 255 VLS patients followed for a median of 66 months on topical steroids became symptom free (Arch Dermatol. 2004;140:702-6) <http://archderm.jamanetwork.com/article.aspx?articleid=480627> . All but 4% improved. Normal skin texture was achieved in 23%, and 68% showed partial improvement. Scarring was less common in children than adults. Squamous cell carcinoma occurred in 2.4%.

The small but clinically significant risk of squamous cell carcinoma has been documented previously, but Dr. Schlosser said there is new evidence that topical therapy may reduce the risk. In a study of 507 women with a median follow-up of 4.7 years, the risk of squamous cell carcinoma was inversely related to compliance with therapy.

"Among the women who used the assigned therapy most or all of the time, zero developed squamous cell carcinoma as opposed to seven cases in the partially compliant therapy group," she reported. Although this does not prove that treatment reduces risk of cancer, Dr. Schlosser indicated that it is strongly suggestive.

Several questions about VLS remain unanswered, according to Dr. Schlosser. More data, for example, are needed to determine whether the current order of therapies for first-line treatment as well as maintenance regimens is optimal, particularly for reducing cancer risk. However, Dr. Schlosser emphasized that the most important challenge to better outcomes is early recognition of this entity.

Dr. Schlosser reported financial disclosures with Allergan and Galderma Laboratories.

References