

## **LICHEN SCLEROSUS**

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A mutilating disease of mysterious origin  
(photo)

Though lichen sclerosus is a disfiguring disease, the intensity of symptoms does not necessarily correlate with clinical appearance. Generally, the first change is (A) whitening of an irregular area on the labia, near the clitoris, on the perineum, and/or other vulvar areas. In some cases (A and B), inflammation can alter the anatomy of the vulva by flattening the labia minora, fusing the hood over the clitoris, effectively burying it beneath the skin, and shrinking the skin around the vaginal opening.

Lichen Sclerosus

Lifelong follow-up is a must

Although it has long been described in medical journals and textbooks, information on lichen sclerosus was often unreliable until recently, and adequate treatment guidelines were lacking. The cause still has not been fully elucidated, but a wealth of information now allows for considerable expertise in the management of this disease.

Lichen sclerosus is a chronic inflammatory and scarring disease that preferentially affects the anogenital area and is 6 to 10 times more prevalent in women than men.<sup>8</sup> Any cutaneous site may also be affected, but the vagina is never involved.

Infection? Autoimmunity? An infectious cause has been proposed but never proven. In some women, an autoimmune component is recognized: Immunoglobulin G antibodies to extracellular matrix protein I have been found in 67% of patients with lichen sclerosus, but whether these antibodies are secondary or pathogenic is unclear.<sup>9</sup>

A genetic component is suggested by the association with autoimmunity and by the link with human leukocyte antigen DQ7 in women<sup>10</sup> and girls.<sup>11</sup>

Affects 1.7%, or 1 in 60 women.<sup>12</sup>

In females, lichen sclerosus peaks in 2 populations: prepubertal girls and postmenopausal women.

No remission after age 70. Although remission of the disease has been reported, a recent study concluded that lichen sclerosus never remits after the age of 70; the average length of remission is 4.7 years, although this figure is still in question.<sup>13</sup> Only close follow-up can determine if disease is in remission.

Main symptom is itching

Pruritus is the most common symptom, but dysuria and a sore or burning sensation have also been reported. Some women have no symptoms. When erosions, fissures, or introital narrowing are present, dyspareunia may also occur.

Typical lesions are porcelain-white papules and plaques, often with areas of fissuring or ecchymosis on the vulva or extending around the anus in a figure-of-8 pattern.

Both lichen sclerosus and lichen planus may be seen on the same vulva.

Squamous cell carcinoma can arise in anogenital lichen sclerosus; risk is thought to be 5%. Instruct women in regular self-examination because carcinoma can arise between annual or semiannual visits.

## INTEGRATING EVIDENCE AND EXPERIENCE

Ultrapotent steroids: Good control, but risk of malignancy persists

Renaud-Vilmer C, Cavalier-Balloy B, Porcher R, Dubertret L. Vulvar lichen sclerosus. Arch Dermatol. 2004;140:709-712.

If we gynecologists have been assuming that lichen sclerosus is one of those nebulous, little-explored diseases out there, we need to think again. Lichen sclerosus is a chronic and mutilating condition, an obstacle to quality of life, a threat to body image, a destroyer of sexual function, and a risk for malignancy.

Cancer developed only in untreated or irregularly treated lesions

In a key study, Renaud-Vilmer and colleagues<sup>13</sup> explored remission and recurrence rates after treatment with 0.05% clobetasol propionate ointment, as well as whether the treatment reduces risk of malignant evolution.

They determined that the rate of clinical and histologic remission is related to age.

Although 72% of women under age 50 had complete remission, only 23% of women between 50 and 70 years of age had complete remission, and none of the women older than 70 did.

Relapse was noted in most women over time (50% by 18 months), and 9.6% of women were later diagnosed with invasive squamous cell carcinoma.

Although we have known since 1988 that ultrapotent steroids offer outstanding relief of symptoms and some control over the disease, the optimal length of treatment has never been clear.

The prospective study by Renaud-Vilmer et al has impressive power that derives from its 20-year duration. They demonstrated that ultrapotent steroids do not cure lichen sclerosus in women over 70.

Complete remission in younger women is only temporary, and steroid therapy offers no significant reduction in the risk of vulvar cancer—although carcinoma developed only in untreated or irregularly treated lesions.

Histologic and clinical findings were used to judge efficacy.

Because only 83 women were studied, the cohort is too small for the findings on carcinoma to be significant, but the authors emphasized that lifelong follow-up is necessary in all cases.

Lichen sclerosus never backs down after menopause

In more than 15 years of vulvovaginal specialization, I have found similar results. Older women detest the need to apply topicals to the genital area, but they are the ones who need ongoing use, because the disease never backs down after menopause.

I follow a cohort of young women in whom I detected early disease. Their clinical signs regressed and scarring was prevented by steroid treatment, but disease recurrence appears to be inevitable:

The longest remission has been just over 4 years.

I have seen cancer arise quickly even in closely supervised patients, although many cases of squamous cell carcinoma have occurred in women with undetected or poorly treated disease.

Use tacrolimus with caution

Although small trials have produced some enthusiasm for therapeutic use of tacrolimus, treatment should proceed with extreme caution, as the drug inhibits an arm of the immune system and women with lichen sclerosus are at risk for malignancy. The agent now carries a warning based on the development of malignancy in animals.

Consider treatment even without biopsy proof

Although a biopsy generally makes the diagnosis, treatment should be considered even in the face of an inconclusive or negative finding if the clinician suspects that lichen sclerosus is present. The reason treatment should proceed in these cases: Loss of the labia or fusion over the clitoris can occur if the disease progresses, as shown in the photos on page. (see above)

Powerful corticosteroids are treatment of choice

Treatment can control lichen sclerosus, relieve symptoms, and 'prevent further' anatomical changes.

Potent or ultrapotent topical corticosteroids in an ointment base are preferred. These drugs are now widely recognized for their efficacy and minimal adverse effects, although no regimen is universally advocated.<sup>14</sup> The patient applies ointment once daily for 1 to 3 months, depending on severity, and then once or twice a week.

Ointments are preferred over creams for vulvar treatment, because creams frequently contain allergens or irritants such as fragrance and propylene glycol preservative.

I continue once-weekly therapy indefinitely in postmenopausal women.

If a premenopausal woman is not comfortable using the ointment indefinitely, I will allow her to discontinue treatment but follow her every 3 to 6 months.

Treatment also requires educating the patient about the disease, instructing her in gentle local care, and showing her exactly where to apply the ointment.

In all cases, lifelong follow-up is necessary. Hyperkeratosis, ecchymoses, fissuring, and erosions resolve, but atrophy and color change remain.

Scarring usually remains unchanged, but may resolve if treated early in the course of the disease.<sup>15</sup>

Testosterone is not as effective 'as' an ultrapotent steroid,<sup>16</sup> and is no more effective than an emollient.<sup>17</sup>

Estrogen is valuable for skin integrity, but has no role in the treatment of lichen sclerosus.

Dilator work may be necessary for dyspareunia, once the disease is controlled.

Refer for help with depression and/or negative body image, if present.

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