Lichen Sclerosus: Why Do Most Women Struggle With Their Diagnosis?

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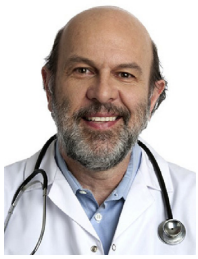

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Lichen sclerosus (LS) is a chronic inflammatory skin disease that primarily involves the vulva and surrounding tissues. It is assumed to be an autoimmune disease for which there is no cure. It affects 0.1% to 3% of the general population, most commonly postmenopausal women.[1] The spectrum of LS symptoms ranges from none to severe pain or discomfort and may evolve to include irreversible anatomical modification and other potential complications.

In 2020, the International Society for the Study of Vulvovaginal Disease declared LS to be disease of the year,[2] heralding a new era of hope and awareness for this poorly understood but treatable condition. However, many women today continue to experience physical and psychological distress related to LS. As clinicians, we must ask ourselves why this remains the case.

First, the diagnosis is all too frequently made late—often much too much late. Sometimes definitive diagnosis comes only after many unsuccessful consults with a family physician, a walk-in clinic, telehealth, or a gynaecologist, years after troublesome signs and symptoms first present. These may include severe itch, worst at night and after intercourse, if intercourse is possible; burning after urination; painful sex; or pain after sex.

Frequently seen signs are whitening of the skin that can be localized at the beginning and then spread to the interlabial sulci, perineum, and above the clitoris. Sometimes it extends to the perianal skin. Simultaneous vitiligo can make the diagnosis complex. Asymmetry or full disappearance of labia minora can be seen. A fusion line at the site of the disappeared or disappearing labia minora can sometimes be seen. Narrowing of the introitus can also be seen. Fusion of the clitoris with the clitoral hood can be partial or total, even forming an inclusion cyst. The supplementary online images show the typical presentation and complications.

Currently, the internet can help women identify LS as a possible cause for their concerns. This can aid in diagnosis, but timely diagnosis is but one challenge.

Second, treatment is not up to standard. Short-term treatment is frequently given without repetition and then stopped; for many women with LS, however, treatment will need to be lifelong. Standards of care are lacking in many jurisdictions. Where standards do exist, weekly application of potent topical steroid therapy with halobetasol or clobetasol ointment is recommended. Too frequently, however, the phenomenon of *steroidophobia*, perpetuated by pharmacy information sheets, prevails, and treatment is stopped after a few weeks because of a lack of clinical follow-up to evaluate symptoms and signs and mitigate complications.

Third, LS is poorly understood, and complications may go undiagnosed. Most family physicians and obstetrician–gynaecologists do not receive training in LS, owing to its incidence across the female lifespan, or receive only minimal training in a vulvovaginal disease clinic. The consequences of this lack of exposure can be serious. LS can be passed down in families, with the risk of heritability as high as 10%.[3] About 6% to 20% of women with LS will have extragenital disease. The lifetime risk of LS-associated

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cancer is 4%, which emphasizes the importance of biopsy for lesion(s) that do not heal, are chronically ulcerated, or are of mixed colouration and tissue quality.[4]

Fourth, complications involving sexual activity are common in LS but are rarely discussed. Issues can be hard to describe, and their causes may be complex. Rubbing of the vulva may cause vulvar pain but not necessarily pain of the vaginal wall, which manifests as itchiness during or after sex. This may inhibit vaginal penetration. In other cases, adhesion of the clitoral hood to the clitoris makes arousal difficult if not impossible. In general, counselling about sexual issues is infrequent and insufficient beyond the recommendation of a water-based lubricant. Sexual difficulties can lead to loss of productivity at work or at home; marital problems, or even the loss of a partner; loss of libido; anxiety; and depression. Referral to an experienced physical therapist will help address pelvic floor concerns, such as narrowing of the introitus and urinary issues.

Lastly, changes in the appearance of the genital area, such as discolouration, loss of labia minora, and adhesion of the clitoris to surrounding tissues can lead to lower sex drive, concerns about loss of desirability to a partner, and performance anxiety. Sexual counselling is rare and does not necessarily address all issues, such as appearance-related concerns. Biomedically trained sex therapists, psychologists, or social workers can help women make the adjustments needed after a diagnosis of LS.

The challenges associated with a diagnosis of LS are numerous, but hope is possible. Efforts to standardize the scale of severity and progression in LS, using combinations of symptoms and physical examination manifestations, is promising. Quality of life and sexual measures should be included in these measures. Furthermore, histology can be used if diagnosis is uncertain or and intraepithelial lesion or cancer is suspected. Such tools would enhance diagnosis, treatment, and research goals.

LS is considered a relatively rare disease in primary care practice, and treatments are effective, but that does not mean it is trivial. Because LS a lifelong disease, multidisciplinary care may be required to help women struggling with LS achieve a normal quality of life and sexual health regardless of age.

SUPPLEMENTARY IMAGES

Supplementary images related to this article can be found at 10.1016/j.jogc.2021.12.001.

REFERENCES

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APPENDIX

Figure A1. Typical lichen sclerosis with whitening of the vulvar skin, asymetry and atrophy of labia minora, petechiae, ecchymosis and adhesion of the clitoris by the clitoral hood.

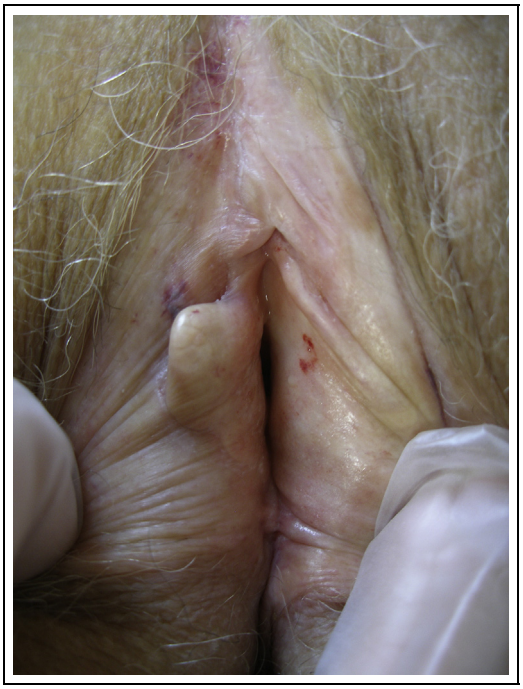
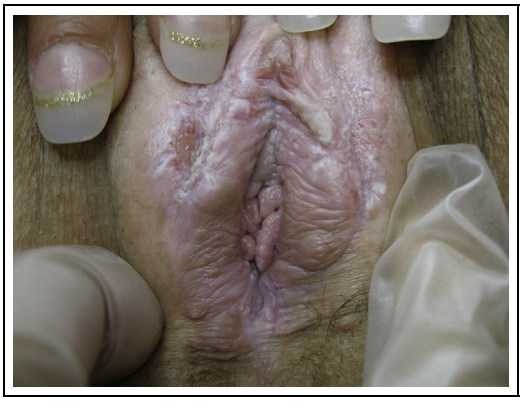


Figure A2. Wrinkled type of lichen sclerosis with heterogeneity of vulvar tissue confirmed being high grade intraepithelial neoplasia, ulcerated lesion of right interlabial sulcus confirmed being a cancer and scratch marks in the left interlabial sulcus.



Images are from the author's library of clinical slides.
Consent: Consent to publish these images was obtained from the patient.