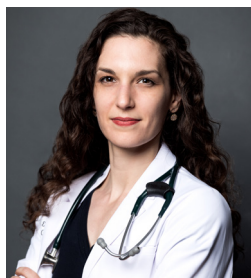


About the Author



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Dr. Delpero completed her medical training at the University of Toronto including medical school, residency in Obstetrics and Gynecology, and fellowship in vulvovaginal health. During her training she also completed a certificate in health journalism through the Dalla Lana School of Public Health and the Munk School of Global Affairs, and obtained master's in systems leadership and innovation through the Institute for Health Policy, Management and Evaluation. She is currently an assistant professor at the University of Ottawa, where she maintains a general obstetrics practice and a focussed gynecologic practice in vulvar skin disorders, colposcopy and menopause management.

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The Three Lichens: A Generalist Approach to Vulvar Skin Conditions

Emily Delpero, MD

Introduction

Vulvar skin complaints represent a significant proportion of visits to family physician, dermatology and gynecology offices. Healthcare training programs place little emphasis on exposure to these conditions, which limits providers' ability to assess and manage them. Increasing the visibility, recognition, and treatment of these conditions serves to improve patient care in Canada.

Lichen Sclerosus

Background

Vulvar lichen sclerosus (vLS) is an autoimmune condition that affects the perineal and perianal skin. It was previously classified as a rare disease which likely represents underestimation and misdiagnosis. More recent estimates indicate its prevalence as 1.7% in a gynecologist's office¹ and 2.4% in a retirement home.² This condition is strongly associated with other autoimmune diseases, with one-third of patients having at

least one other condition such as Hashimoto's thyroiditis, alopecia areata, and vitiligo.³

Currently it takes 5-15 years from onset of symptoms to diagnosis, and while most patients are diagnosed in the pre-pubescent phase of life or in menopause, there is increasing evidence that early signs and symptoms can evolve during reproductive years.⁴

Diagnosis

A diagnosis of vLS can be made based on history and physical examination. While biopsy can offer a definitive diagnosis, not all patients require this intervention. Common symptoms can include pruritus, irritation and pain, dyspareunia with recurrent tearing, and fissuring of the skin. In younger patients, simply having recurrent tearing with intercourse can be the only initial symptom.⁵ Some patients might also be asymptomatic. Physical examination signs include evolving ivory white shiny or crinkled patches on the labia and perianal skin, with petechiae and cigarette paper texture changes. These skin changes classically occur in a figure of 8 distribution but

lichen sclerosus (LS) can also arise focally and asymmetrically.

Disease Course

LS is a chronic condition with a relapsing and remitting course throughout a person's lifetime. Progression of disease includes distortion of the vulvar architecture and permanent anatomy changes such as clitoral phimosis, narrowing of the introitus, and lateral and medial agglutination of the labia minora. This can be deeply impactful to daily urinary function and sexual function of patients with vLS. Uncontrolled vLS is also associated with the development of vulvar squamous cell carcinoma in 4-7% of cases.⁶

When to Biopsy

Patients with classic lesions who respond appropriately to therapy do not require a biopsy, but healthcare providers should consider biopsy for definitive diagnosis in the following circumstances:

1. Patient preference
2. Non-classic presentation (i.e., asymptomatic or focal lesions for diagnostic clarification)
3. Any raised, hardened, ulcerated areas (i.e., concern for malignancy)
4. Condition refractory to therapy

Biopsy results can also be misleading and non-specific if an insufficient sample is taken, or if the area is pre-treated with potent steroids. Early vLS can result as non-specific vulvitis or eczema. This author recommends a 4 mm punch biopsy sample directed towards the most prominent area of abnormality.

Treatment

There are three main purposes of treating vLS:

1. Symptom control and quality of life
2. Prevention of ongoing anatomy changes
3. Lower likelihood of progression to vulvar cancer⁷

There is no cure for vLS but the gold standard of therapy is ultra-potent topical steroid ointments.⁸ Ointments are preferred over creams for the vulvar skin because creams contain more irritants like alcohol base and can easily be wiped off. The two options available in Canada include clobetasol propionate 0.05% and betamethasone dipropionate 0.05%.

There are slightly different published regimens, but recommended dosing from the British guidelines includes 0.25 g (half a fingertip

unit) to affected area nightly for 1 month, then every other night for 1 month, then twice a week.⁸

The role of maintenance therapy is currently under study but is emphasized in order to maintain disease control and address subclinical inflammation.

Some patients may benefit from added topical tacrolimus 0.1% or pimecrolimus 0.1% on days alternating with steroid application. These are considered potentially "steroid sparing" but are less effective than topical steroids at controlling inflammation.⁹ Additionally, these options are typically poorly tolerated by patients who often experience burning and irritation after application.

Rare cases that are refractory to topical therapy may require systemic medications that are beyond the scope of this article.

When to Refer

Consider referral to a vulvar skin specialist if there is diagnostic uncertainty (i.e., discrepancy between clinical presentation and histology), concerning and evolving lesions, or if symptoms are refractory to appropriate therapy.

Follow-Up

Initially, patients with vLS should have a skin examination every 6-12 months depending on response to steroids and compliance. Once stable, patients with vLS should have annual skin checks to ensure no new lesions, and be advised to seek medical attention if they evolve into new growths, change in symptoms, or new symptoms refractory to their usual therapy.

Lichen Planus

Vulvar lichen planus (vLP) is a less common autoimmune vulvar dermatosis that can mimic LS. It is diagnosed in the same way, and is characterized more by vulvar burning and irritation than pruritus although both can be present. Classically, patients present with red erosions at the introitus surrounded by white lacy striae and anatomy changes including resorption of the labia minora, clitoral phimosis, and/or narrowing of the introitus. Patients with lichen planus (LP) often have lesions elsewhere, particularly oral erosions. Unlike LS, lichen planus can involve the vaginal mucosa as well and lead to an obliterated or stenosed vaginal canal. Examining patients with vLP does involve a speculum examination to assess for vaginal involvement.^{10,11}

Clinically, distinguishing vulvar LS from LP may be challenging, and can be aided by biopsy. However, the principles of treatment are the same. These include a topical steroid taper (using ointment on the vulva, and cream for vaginal involvement) and possibly a steroid-sparing agent depending on symptoms. Similar to vLS, refractory cases may require systemic therapy.

The clinical course of vLP is less well-characterized, therefore the role of maintenance therapy is less robust in LP than LS, as is the link between LP and the evolution of vulvar cancer.¹²

When to Biopsy

If the need arises (similar to the indications highlighted for LS) for diagnosis, consider a biopsy from the edge of the erosion.

When to Refer

The reasons to refer are similar to those for vLS and include diagnostic uncertainty (i.e., discrepancy between clinical presentation and histology), concerning and evolving lesions, or if symptoms are refractory to appropriate therapy.

Lichen Simplex Chronicus

This skin condition affects all people of all ages, and there is an association and overlap with atopic dermatitis. Some patients present with symptoms elsewhere, and some have symptoms only on their vulvar skin. Anogenital lichen simplex chronicus (LSC) is quite common, with a reported prevalence of upwards of 10%.¹³

Clinically, LSC presents as an “itch-scratch cycle”. There might be a history of an identifiable trigger (i.e. prior yeast infection, recent shaving, exposure to new soap, or prolonged exposure to damp material), but symptoms of pruritus can also arise spontaneously. Other vulvar dermatoses like LS can predispose to LSC as well. Whatever the cause, the itchy skin is rubbed and scratched, which in turn leads to inflammation and propagates the pruritus. On history, patients might not endorse scratching, but they may be rubbing the skin with a towel after the shower, toilet paper after the bathroom, or at night while they are asleep.

On examination, the vulva can appear symmetrically or focally affected with erythematous, poorly demarcated scaling papules/plaques, epithelial disruption, and lichenification. If the patient has a light skin tone¹⁴, these areas can

appear whitened because the thickened skin holds onto moisture. In patients with darker skin tones, the changes can appear hyperpigmented.

A biopsy of the area is often non-specific, and may return demonstrating “psoriasiform dermatitis” which is confusing for clinicians who may in turn diagnose the patient with psoriasis. Distinguishing psoriasis from LSC can be difficult, but patients with vulvar psoriasis will have plaques elsewhere on the body (i.e. scalp), or nail, or joint involvement.¹¹

Treatment

Treatment involves several avenues to disrupt the itch-scratch cycle.¹¹

1. Counselling patients carefully about vulvar hygiene will help reduce potential contact irritants, and exposure to heat and sweat (**see box on vulvar hygiene tips**)
2. Restore the skin barrier with petrolatum base, coconut oil or zinc ointment
3. Address the inflammation directly with moderate-to-potent steroids. These steroids may need to be used once daily for 1-3 months depending on the severity of initial presentation, then decreased to a PRN basis
 - A common regimen to consider is fluocinonide 0.05% ointment (0.5 of a fingertip unit) nightly for 1-3 months then 2-3 x weekly as needed. Alternatively, one may use desoximetasone 0.25% ointment with the same schedule.
 - If a patient has not improved after 1 month of topical therapy, systemic therapy may be required, or the diagnosis may need to be reconsidered.
4. Finally, patients should be informed that if they do not stop re-traumatizing the skin with scratching or rubbing, the condition will not resolve. If there is concern for nighttime or involuntary scratching, consider a short 1-2-month course of antihistamine at bedtime to reduce the impulse

Follow-Up

In patients with a new diagnosis of LSC, early follow-up at 3-6 months may be required to assess response, compliance with therapy, and to trouble shoot any therapy-related problems. Patient presentations may resolve completely, but if there is an underlying dermatosis (i.e., vLS) further annual follow-up is recommended as discussed above.

Vulvar hygiene tips:

- No soaps/cleansers/douches on the vulva, even “baby soap” or “sensitive skin” soaps
- Use lukewarm water on the vulva
- Avoid cleaning the vulvar skin more than once a day, and avoid scrubbing the skin
- Let the vulvar skin air dry or pat dry
- Avoid tight fitting clothing
- Remove sweaty or wet clothing as soon as possible
- Consider cotton underwear or no underwear at night
- Consider avoidance of pads, or use cotton or hypoallergenic versions

Prescribing Topical Steroids

Healthcare providers, including pharmacists and physicians, are trusted sources of information for patients. Unfortunately, they unwittingly also become sources of steroid phobia and non-compliance.¹⁵ Steroid phobia is defined as “vague negative feelings and beliefs held by patients/caregivers” which are associated with non-compliance, undertreated conditions, reliance on alternative and unproven remedies, or the need to scale to systemic medications which carry their own toxicities. This is best characterized in the field of atopic dermatitis, but it applies to patients with other skin conditions as well.

Safety

When topical steroids are used and monitored appropriately, the risk of side effects is extremely low.¹⁶ For vulvar dermatoses, the impact and harm associated with an undertreated skin condition outweigh the risk of treatment-related adverse events.

Locally, topical steroids may cause skin thinning, telangiectasia, or hypopigmentation (although untreated LS or LP can certainly damage the skin in their own right). There are potential systemic effects of immunosuppression or adrenocortical insufficiency; however, this was found in patients who were using 100 g of ultrapotent topical steroid once-weekly for 1.5 years.¹⁷

When discussing topical ointment dosing, fingertip units (0.5 g) have been described as: from the end of the finger to the first joint. The vulva requires half of a finger tip unit (0.25 g) to cover its entire surface. Using a topical steroid on

the vulva every day for 30 days would translate to a cumulative monthly dose of 7.5 g.

How to Safely and Effectively Prescribe Steroids for Vulvar Skin Conditions

1. Use ointment, not cream
2. Demonstrate to the patient the quantity to use
3. Demonstrate where to apply (i.e. via mirror, diagram, or touch)
4. Explain concerning, albeit unlikely, side effects patients might read or hear about, or be advised of by other people including other physicians and pharmacists. Be careful to place them in the context of how little product the patient will be using.
5. Encourage judicious use. If there is concern of overuse, prescribe 30 g at a time with no refills. This should be sufficient for a 4-month supply for a patient using steroids daily.
6. Follow-up with the patient as required

Other Resources for Healthcare Providers

Useful resources for additional, evidence-based information include:

- The International Society for the Study of Vulvovaginal Diseases (www.issvd.org) a global organization that promotes evidence-based research, publications and conferences.
- Vulva Diaries podcast by Dr. Amanda Selk
- Jill Krapf, MD on Instagram

Conclusion

Various types of vulvar skin conditions occur among a significant proportion of the population, and require treatment on the part of family physicians, dermatologists and gynecologists. This paper highlights the diagnosis, clinical signs and symptoms, and treatment approaches for these conditions with a view toward enhancing healthcare providers’ ability to assess and manage them.

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