Vulvar Disease: An Update

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Disclosure

• I have nothing relationships to disclose
• Will be discussing off label use of medications
Goals

• Identify clinical findings and associated conditions of lichenoid vulvar disease and lichen simplex chronicus
• Develop evaluation and management strategy
• Recognize pitfalls and learn how to minimize complications

Lichen Sclerosus (LS)

• Chronic dermatosis with predilection for anogenital area
  – ?Autoimmune ?Inflammatory
• Most common in post menopausal women
• Prevalence estimated at 1/300 to 1/1000 but could be as high as 1/30
• Accounts for 1/3 of patients presenting to a specialty clinic with vulvar complaints

LS: Clinical Findings

- Periclitoral edema
- Wrinkling and/or hyperkeratosis of skin
- Fusion of labia minora leading to resorption
- Fissures
- White/pallor
- Scarring of the clitoral hood
- Introital narrowing
- Sparing of mucous membranes
- Genital melanosis

Lichen Sclerosus Associated with Autoimmune Disease

- 22-28% of women with LS have associated autoimmune disease
- Most common is thyroid disease
  - 8-30% of patients
  - Check TSH
- Alopecia areata (9%)
- Vitiligo (6%)
- Pernicious anemia (2%)

**LS: Other Associations**

- Morphea \(^1\)
  - 50% of pts presenting with generalized morphea had LS
- Psoriasis \(^2\)
- Celiac \(^3\)
- IBD


**Squamous Cell Carcinoma in Lichen Sclerosus**

- Lifetime risk estimated at 5%
- Cohort study of 253 women followed over 69 months found prevalence of 3%
- Between 7-60% of vulvar SCCs occur on vulvar lichen sclerosus
- Thought to be secondary to chronic inflammation
- Patients should be advised to monitor for ulcers or lumps
- Erosion or area of hyperkeratosis not responding to therapy warrants biopsy
- Patients require long term follow up
- Treatment is thought to reduce the risk

LS: Management update

Continuous *versus* tapering application of the potent topical corticosteroid mometasone furoate in the treatment of vulvar lichen sclerosus: results from a randomized trial

A. Borghi, M. Corazza, S. Minghetti, G. Toni, A. Virgili

- Study of 67 patients
- Randomized to 5 x weekly mometasone fumarate for 12 weeks vs tapering schedule (5 per wk for 4 weeks then every other day for 4 weeks then twice weekly)
- No difference in clinical/symptom/ improvement
- No difference in adverse reaction

Long-term Management of Adult Vulvar Lichen Sclerosus
A Prospective Cohort Study of 507 Women

Andrew Lee, MBBS, Jennifer Bradford, MBBS, Gayle Fischer, MD

- Prospective longitudinal study of 507 women with biopsy proven LS
- Topical therapy tailored to degree of hyperkeratosis but most pts used potent to ultrapotent topical steroids
- Avg time to skin normalization – 4.9 month
- No SCCs in compliant pts
- 7 pts who reported they were not compliant developed SCC or VIN
- Sx did not correlate with disease progression
  - Asymptomatic progression
- Bottom line: no standardized tx for LS
- Need regular follow up until stable then maintenance therapy and then 6 month follow-up
LS: Complications

• Iatrogenic Infections
  – HSV: if pos hx, prophylactic antivirals while on clobetasol
  – Candida/tinea: itch, erythema, fissuring or scale perform KOH or culture for candida
  – Culture for strep/staph if sx not improving
• SCC
• Atrophy
  – If steroids are used correctly risk of atrophy is very low
• Steroid irritant/allergic contact dermatitis
  – Reduce potency, switch to desoximetasone 0.25% ointment, consider patch testing

Lichen Planus (LP)

• LP is an inflammatory disorder of skin, mucous membrane and nails
• Unknown prevalence
• Pathogenesis thought to be Tcell mediated immunologic response to basal cells
Types of Vulvovaginal LP

• 3 main types
  – Erosive
    • Erosions, erythema or desquamative vulvitis/vaginitis. May have surrounding wickham striae (lacy reticulations)
  – Papulosquamous
    • Pruritic, vilaceous, papules with wickham striae
  – Hypertrophic
    • Extensive, white, thick, hyperkeratotic plaques and erythematous macules and patches


Vulvovaginal LP

• Similar clinically to oral LP
• Oral LP + vulvovaginal LP = vulvovaginal-gingival syndrome (VVG)
• 43-100% of vulvovaginal cases may have oral involvement
At least 3 criteria should be present to make the diagnosis

**DDX: Erosive Vulvar Diseases**

- Lichen Planus
- Immunobullous Disease
  - Pemphigus, mucous membrane pemphigoid
- Graft Versus Host Disease
- HSV
- Aphthae
- VIN/SCC
## Vulvovaginal LP

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<thead>
<tr>
<th>Aggravating Factors</th>
<th>Symptomatic Treatment</th>
<th>Workup</th>
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<tbody>
<tr>
<td>Soaps, washes, and cleansers Medications (topical) Antifungals, steroids, hormones</td>
<td>Identify aggravating factors History Biopsy Patch tests (delayed hypersensitivity reaction) Stop itch-scratch cycle Hydroxyzine/doxepin Wash with water only Sitz baths, cool compresses, ice packs Pat dry (do not rub) Use only cotton underwear Avoid stockings, tight-fitting pants, and jeans</td>
<td>Determine fasting blood glucose, serum iron, folate, vitamins B₆ and B₁₂, and magnesium levels, and rapid plasma reagin test Perform a direct cytologic examination (potassium hydroxide, cell smear) Obtain tissue for both histologic evaluation and immunofluorescence Culture for fungi such as Candida albicans, C. torulopsis (C. glabrata) and others and bacteria such as Chlamydia and group A streptococcus. Consider viral studies (herpes simplex virus or human papilloma virus)</td>
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<tr>
<td>Feminine hygiene products Tampons/pads, vaginal douches, hygiene sprays, suppositories, lubricants</td>
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<td>Contraception Condoms Vaginal sponge Spermicides Toilet paper Long-term moisture Normal vaginal secretions Semen Urinary incontinence Fecal incontinence Sweat</td>
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Ginat M and Goddard A.  

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### Management Strategies: LP

- Eliminate irritants/allergen
- Topicals mainstay of therapy
  - Suprapotent topical steroids (clobetasol propionate 0.05%)
  - Tacrolimus 0.1%
  - Hydrocortisone 25mg suppository (anusol)
  - Compounded hydrocortisone intravaginal cream (10%)
- Systemic medications: prednisone, methotrexate, hydroxychloroquine, acitretin, mycofenolate mofetil
- Topical/intravaginal estrogen
- Dilator therapy
- Check and recheck for candida/strep

Wet Mount

• Important to diagnose vaginal involvement and monitor for resolution
• 1wbc per squamous epithelial cell = normal

Additional Sites of Involvement

Lacrimal Canalicular Duct Scarring in Patients With Lichen Planus

Naomi K. Webber, MBBS, BSc, MRCPCH; Jane F. Setterfield, MBBS, BDS, FRCP;

Lichen planus of the esophagus: What dermatologists need to know

Lindy Peta Fox, MD, Charles J. Lightdale, MD, and Marc E. Grossman, MD
San Francisco, California, and New York, New York
• Esophageal LP (ELP) is likely under-recognized
• Predilection for middle aged women
• Associated with oral and/or genital dz
• Prevalence unknown (may be as high as 25-50%)
• Pt with oral LP and dysphagia or weight loss → EGD

Genital Tract Graft Versus Host Disease

• 60-70% of patients who receive allo transplant manifest GVHD (Lee et al. Chronic graft-versus-host disease. Biol Blood Marrow Transplant 2003)
• Incidence of female genital tract cGVHD ~50%
• Patients with genital disease more likely to have extensive cGVHD
• May be first presenting sign of GVHD

GVHD: Have a High Index of Suspicion

- Ask the patient about sx
- Diagnosis made by clinical-pathologic correlation
- Lichenoid GVH may mimic lichen planus or lichen sclerosus clinically and histologically
- Include hx of transplant on path requisition and talk to the pathologist

Lichen Simplex Chronicus (LSC)

- Clinical
  - May be subtle
  - Lichenified, thickened plaques
  - Hypopigmentation or hyperpigmentation
  - Linear excoriations (scratches), secondary erosions
**LSC**

- Due to “itch-scratch-cycle”
- Repair barrier – eliminate irritants and scratching implements, add emollient
- Rule out underlying cause of itch (yeast, irritant, other dermatosis, allergic contact dermatitis)
- Associated with atopic dermatitis
- Address both skin issue and behavioral component
  - Treat with mid to high potency topical steroids and antihistamine at night
    - Hydroxyzine 10-30mg
  - Consider addition of tricyclic or SSRI if not improving
    - Doxepin 10mg

**Vulvar Disorders: Multifactorial**

- Eliminate irritants
  - Assess for incontinence, hygiene practices, wipes etc
- Repair the skin barrier (emollient, topical estrogen)
- Correct initial infection and monitor for iatrogenic effects
  - Candida, HSV, HPV, bacteria
  - 1 dose fluconazole not enough in setting of topical steroids
  - Applying topical steroids to an infection will exacerbate it
- Any lesion that has not responded to therapy should be biopsied
Principles of Management

• The modified mucous membranes are relatively resistant to steroid atrophy
  – Keratinized skin are not
• Ointments are better tolerated than creams - a little goes a long way
• Treatment failure often due to non compliance or incorrect application
• Little risk of atrophy if use 30 gram tube over 6 -12 months

Stewart K. Clinical Care of Vulvar Pruritus, with Emphasis on One Common Cause, Lichen Simplex Chronicus Dermatol Clinic. 2010; 28(4) 669-680.
Thorstensen K., Birenbaum D. Recognition and management of vulvar dermatologic conditions: Lichen sclerosus, lichen planus and lichen simplex chronicus. JMWH. 2012

Demonstrate Where and How to Apply
Thank you!

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