Genital Lichen Planus

Lynette J. Margesson MD FRCPC
AAD Washington
- Mucous Membrane Symposium
Saturday, March 2, 2019

Conflicts of interest
Author Up To Date

Little evidence based treatment
Too few studies done in vulvar disease.
Most treatments discussed are "off-label"

Objectives
1. Diagnose genital lichen planus
2. Choose the best treatments
3. Locate more in depth information

Lichen Planus
• Autoimmune or cutaneous hypersensitivity reaction
• Onset - 50 - 60 years
• Affects - Skin, scalp, nails
  Mucous membranes - oral, genital, anus
  esophageal, urinary tract

1 - 3 % SCC in genital area

Always Examine the Genital Area & Mouth

Genital Lichen Planus
- Women: vulva and/or the vagina
  - Mostly have erosive disease; red, raw surface
  - Lacy white pattern is less common than in the mouth
  - Labia minora often lost; vaginal stenosis
- Men: glans, prepuce and shaft of the penis
  - Seldom have erosive disease
  - Therefore little structural damage
  - Red papules, plaques and lacy white pattern

Vulvar Lichen Planus Patterns
Variable and can be mixed - Can have normal vulva and active vaginal LP

Erosive 85%  Lacy 10%  Hypertrophic 4%
LP Clinical subtypes:

- Papular
- Reticular
- Plaque-like
- Atrophic
- Hypertrophic
- Erosive
- Bullous variants

Vulvar Lichen Planus

- On vulva, vagina, mouth often erosive
- On vulva typically non descript erosions with itching, burning, irritation and sexual dysfunction
- 10 times less common than LS

Symptoms:

- Sore, burn, ~70-80%, severe ~35%
- Itch ~50-60%
- Dyspareunia ~60-70%
- Apareunia ~30-40%
- Dysuria
- Asymptomatic ~11-57%

A cause of asymptomatic scarring

Erosive Vulvovaginal LP

- Deep red erosions, glazed erythema with thin gray edge
- Fern-like or lacy white pattern
- Variable scarring / loss of architecture
- Pain plus burning

Vaginal Lichen Planus

- Often asymptomatic
  - Inflammatory Vaginitis
    - With mucopurulent vaginal discharge - yellow, green, bloody
    - Narrowed and / or shortened vagina
    - Ring stenosis or total occlusion

Often work with gynecology and examine vagina

Vaginal involvement in genital erosive lichen planus.
Diagnosis of Lichen Planus

Morphology
- Onset: gradual, chronic
- Location: correlate with biopsy that can be reported as "lichenoid"
- Pathology often non-specific

For Diagnosis:
- Biopsy: perilesional
  - Site: number 1-2
  - Size: 3-4 mm
  - H&E
  - DIF: 56% shaggy fibrin, plus IgG, IgM, IgA, C3 at DEJ

Genital Lichen Planus

Differential diagnosis:
- Lichenoid drug reactions
- Lichen Sclerosis
- Psoriasis
- Lichen Simplex Chronicus
- Vesiculobullous disorders (ie PV, BP, BMMP, LABD)
- Graft vs Host Disease
- Squamous Cell Carcinoma
- Connective tissue disease (ie SLE, DLE)
- Erythema multiforme
- Epidermolysis bullosa acquisita

Management Principles

- Explanation of the disease process, treatments, expectations
- Handouts
- Photographs
- Treat all factors
Diagnostic Work-up for Genital Lichen Planus

- Punch biopsy: H & E, DIF studies
- Hepatitis C serologies and liver enzymes
- Discontinuation of any suspected drugs
- Referral to gynecologist for vaginal assessment
- Endoscopic examination, if indicated
- Clinical surveillance for malignant transformation

Lichen Planus Treatment

Confirm diagnosis - biopsy
- Stop irritants
- Educate patient
- Stop scratching
- Control infection
- Stop meds associated with lichenoid of fixed eruptions mimicking LP if appropriate

Control inflammation
- Topical, intralesional, vaginal or systemic corticosteroids
- topical calcineurin inhibitors

Lichen Planus Treatment

Topical therapy
- Topical/IL, vaginal steroids
- Topical immunomodulators
- Topical retinoids
- Systemic medications

Topical Steroids for Genital LP

Clobetasol, halobetasol ointment
- Use daily on non-keratinized skin until controlled.
- For keratinized skin and perianal: 2-3 weeks and taper
- If burning use Triamcinolone 0.5% ointment
- Slow or very thick: add tazarotene 0.1% cream 2-5 times a week
- Lack of estrogen: add estradiol 0.01% cream daily
- Add/substitute tacrolimus ointment if possible can be used in vagina 1 gm applicator full hs
  (No safety data!)
Symptom control: up to 90% Remission possible

Effectiveness of LP Treatment

- Clobetasol Ointment - 3 months
  - 94% improved and 71% symptom free on Rx
- In another study with a triamcinolone mix:
  - Erosions healed in 50%
  - Remission in 9%
- Overall 30% may not respond??
- Maintain on least potent corticosteroid

Intravaginal Corticosteroids

Hydrocortisone base or acetate:
- Dose depends on severity
- Suppository 25mg (available) or 100 - 200 mg (compounded)
- 10% compounded vaginal cream: 1-5g per dose (100-500 mg/dose)
- Use nightly for 14 days then Mon Wed, Fri - use with dilators prn
- Note: risk of adrenal suppression and candidiasis

Intralional Triamcinolone

- 3.3 to 10 mg/ml after local anesthesia
- For refractory areas

No Compliance
Topical Immunomodulators

steroid sparing
- pimecrolimus (Elidel) 1% cream bid for mild LP
- topical tacrolimus (Protopic) 0.03%, 0.1% ointment bid
- vaginal tacrolimus hs - 2 mg suppositories or 0.1% vaginal cream 1-2 gms = 1 - 2 mg/dose
Note - use after control with topical steroid
- all can burn

Systemic Therapy for Genital LP

- Use if severe or topicals not effective in 4-6 months - do not wait too long
- Systemic corticosteroids and cyclosporine usually effective quickly
- Most others take 2-3 months to start working

Systemic Therapy for Genital LP

- Triamcinolone IM
- Prednisone
- Hydroxychloroquine 200 mg bid
- Methotrexate 2.5-15 mg/week PO or SC + folate
- Mycophenolate mofetil 500 mg - 1.5 g bid
- Cyclosporine 4 - 5 mg/kg/d - 3 - 4 months
- Azathioprine 50 - 75 mg/d to bid
- Acitretin (10 mg 5 - 7 d/wk)
- Adalimumab

Prednisone for Severe, Chronic, Recalcitrant LP

- Tapered dose of 40 to 60 mg/d over 1 - 3 weeks
- Cumulative dose of 600 - 625 mg
- Indications: Dose - am with food
  - 40 mg 4-5d, 30 mg 4-5d, 20 mg 5d, 10 mg 5d
  - 60mg 2d, 50 mg 2d, 40 mg 4d, 30 mg/4d
  - 20 mg 4d, 10 mg 4d
- Consider 1 to 2 repeat courses 4 - 6 weeks apart
- Consider combination with steroid sparing agent

Mycophenolate mofetil

- Dosing: 500-1500 mg bid
- Starting dose: 500 mg
- Incremental increases by 500 mg over 6-8 weeks to 1.5 gm bid
- Reports of sustained remission
- Duration of therapy: minimum 1 year
**Severe Lichen Planus Treatment**

- Hydroxychloroquine 200 mg bid
- Methotrexate 5-15 mg/week PO or SC + folate
- Cyclosporine 4 - 5 mg/kg/d - 3 - 4 months
- Azathioprine 50 - 75 mg/d to bid
- Acitretin (10 - 25 mg 5 - 7 d/wk)
- Adalimumab

About 30-40% vulvovaginal LP need systemic Rx

Surgery may be needed for reconstruction and cancer

---

**Risk of Cancer in Vulvar LP**

- A small increased risk of vulvar malignancy in vulvar LP is suggested but more data needed
- In one cohort (mean follow-up of 72 months), 2.4% of patients with LP had a history of or current genital malignant neoplasm
- Another series of 100 - 3% had vulvar dysplasia
- SCC in vulvar LP aggressive

---

**Complications of Vullovaginal Lichen Planus**

- Chronic pain, rarely itch - vulva, vagina
- Scarring - variable
  - Loss of labia minora,
  - Clitoris - clitoral scarring, phymosis, buried clitoris,
    - clitoral pseudocyst, hypo or supersensitivity
  - Introtal Scarring / narrowing
  - Vaginal Scarring - partial, total, ring stenosis
- Dysuria, rarely urethral stenosis
- Vaginal discharge - inflammatory vaginitis
- Vulvar pain / sexual dysfunction and psychosexual problems
- Squamous cell carcinoma

---

**Prognosis**

- Waxes and wanes!
- Remission variable.
- Women often give up on treatment as too difficult with associated depression - poor QOL
Vulvar LP - Why not Better

Her Reasons -
- Clobetasol too expensive
- Clobetasol ointment burns
- Treatment messy and not a cure
- Not using it anyway!

Causes of Treatment Failure
1) Incorrect diagnosis - was a biopsy done?
2) Missed concurrent conditions
   LS, LP, contact, Candida, HSV, estrogen loss, SCC
3) Ineffective treatment plan
4) Noncompliance
   poor education
   fear of topical steroids
   limited mobility

Look for Concurrent Conditions

Infection:
- Candida, Staph /Strep, HSV

Rashes:
- Contact dermatitis
- Lichen Simplex Chronicus
- Mix - LS and LP

Cancer:
- SCC, VIN III

Other:
- Lack of Estrogen, Trauma (scratching)

Summary for Genital LP
1) Look carefully at all skin and mucous membranes - note subtle scarring.
2) Always check vulva and vagina in LP cases in women - do not rely on history
3) Biopsy may be non-specific - find an interested pathologist
4) Do not rely on topical treatment alone - use systemic treatment if needed
5) Recognize causes of treatment failure

References
- Unexpectedly high frequency of genital involvement in women with clinical and histological features of oral lichen planus.
- Lichen planus affecting the female genitalia: A retrospective review of patients at Mayo Clinic.