“Asymptomatic” Undiagnosed Lichen Sclerosus

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Disclosures

- Dermtreat - consultant
- Up To Date - author
  No conflict with either

- Little evidence based treatment for vulvovaginal diseases
- Most information is from small open trials and clinical experience
- Most treatment discussed is “off-label”
85 year old lady presents with “bleeding” - no itch or burn

She has had minor vulvar irritation over the years that was ignored

She stopped sexual activity at menopause as vulva too dry and sore
Diagnosis -
Invasive Squamous Cell Carcinoma (SCC) and lichen sclerosus (LS)

Questions -
• Why was LS missed?
• If LS had been treated could this have been prevented?
Asymptomatic Undiagnosed LS

• How often is LS asymptomatic? **Poor Data!**

• 7% asymptomatic in childhood (Powell J, Wojnarowska F. 2001)

• 58% asymptomatic 19 new cases LS (Goldstein AT, Marinoff SC 2005)

• 15% asymptomatic of 228 LS cases
  - more symptomatic with foods like pork and spice, aging,
  - GU incontinence, 100% symptomatic with family history of LS

  (Vieira-Baptista P, Lima-Silva J 2015)
Importance of Asymptomatic LS

• Patients are not seen if asymptomatic
• Caregivers often do not recognize LS
• LS patients told to treat according to symptoms
  Resulting in poor / inadequate treatment
• Results of missing LS – progressive scarring
  sexual dysfunction
  cancer
Changing Treatment for LS

Prospective study of 507 women with biopsy-proved VLS over 6 ½ years

- 150 patients (29.6%) were partially compliant to Rx
- 357 patients (70.4%) were compliant
- Squamous cell carcinoma or vulvar intraepithelial neoplasia occurred (at follow-up) –
  - 0 of compliant patients vs 7 (4.7%) of partially compliant patients
- Symptom suppression occurred in 333 (93.3%) compliant patients
  - vs 87 (58.0%) partially compliant patients
- Adhesions and scarring seen in 12 (3.4%) compliant patients
  - vs 60 (40.0%) partially compliant patients
Prospective study of 507 women with biopsy-proved VLS over 6 ½ years

• No one plan for all
• Start with superpotent steroid until skin color and texture as normal as possible; use milder steroids for perianal areas etc
• Maintenance forever - use the most effective topical steroid at dose to prevent remission

Long - term LS treatment results -
• Improved function
• Relieved symptoms
• Reduced development or progression of scarring
• Eliminated the risk of cancer

Evidence-based (S3) guideline on (anogenital) LS

• Proposed Amendments to Guidelines:
  1. Confirmatory vulvar biopsy prior to treatment in adults.
  2. Endpoint of treatment - suppression of signs not symptoms to prevent disease progression.
  3. Any topical corticosteroid regimen that achieves skin of normal color and texture is appropriate.
  4. Adjust treatment for needs of the individual patient.
5. Early topical corticosteroid treatment will prevent scarring in nearly all patients

6. Cure is usually not possible. Patients should be counseled of the importance of the need for lifelong treatment adherence and follow-up.

7. All patients deserve regular follow-up by a doctor who understands this condition and treatment adjusted to maintain the goal of normal skin.

Evidence-based (S3) guideline on (anogenital) lichen sclerosus. JEADV. 2015; 29(10):e1-e43.
Lee A, Bradford J, Fischer G.
Goal for LS treatment - Control

Clearance in vulvar lichen sclerosus: a realistic treatment endpoint or a chimera?

• 196 patients 12 weeks Topical Rx
• 47.3% symptom free at the start
• 13.9% complete clearance symptoms - had lower symptom scores shorter disease duration

• CONCLUSION:
Despite significant improvement still have substantial residual disease

Question -
• Why is the diagnosis of LS missed?
Question -
- Why is the diagnosis of LS missed?

Because
- Not recognized!
- Avoided / ignored - noncompliant
- Concomitant conditions
Recognize Normal Anatomy

- 38 year old lady presents with an itchy vulvar rash for 2 - 3 years.
- She has been treated repeatedly for “yeast” with no response.
Anatomy can be Confusing

- Recognize the missing bits
The Vulva
Better teaching Aid?
The Vulva

• **STOP LOOK SEE**

• Use diagrams and handouts

• See ISSVD.org - Patient education for

Diagram of Normal Vulva
Why Poor Patient Compliance with Rx LS?

- Education - inadequate / poor
- Fear
- Phobic re touching area
- Limited mobility
- Money
- Anxiety, depression

Biggest Issue with LS

NO SYMPTOMS
Why Poor Patient Compliance with Rx LS?

- Education - inadequate / poor
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Biggest Issue with LS

NO SYMPTOMS
No Symptoms

• 73 year old lady with biopsy proven LS and now has blood on toilet tissue
• No Rx since “it did not bother “ her
• Bx - dVIN
• She ignored/avoided LS and never went for follow up
Only “Intermittent Symptoms”

• Followed for years - 61 year old with biopsy proven LS > 20 years
• She has intermittent itch and irritation and now feels a “bump”
• Dx Invasive SCC
• Inadequate follow up and noncompliance
Fear of RX

- 42 year old who presented with vulvar itching at age 32 years and biopsy confirmed LS but never did any Rx as “a nuisance”
- Avoided follow up and only came in as motor cycle riding now irritating
- DX - invasive SCC metastatic to lymph nodes
- Fear → Noncompliance
Mobility and Depression

- SCC Vulva with surgery 4 years ago
- No follow up
- Almost 300 lbs and cannot reach vulva
- Topical Rx not possible
- Noncompliant due to mobility and depression
Concomitant Conditions

• Psoriasis and LS
• Having more than one problem confusing
• Both itchy
• LS missed
Asymptomatic

• 68 year old now sore for 4-6 weeks with biopsy proven LS 10 years but problems with sexual dysfunction for years

• Treatment variable as “not a problem”

• Bx – Invasive SCC
Anxiety and Phobia

- 54 year old lady celibate all life as sexually abused as child
- No gynecologic care
- Almost no touching of vulva
- Now intolerable itch for 2 months
- BX dVIN and LS
Asymptomatic

- 68 year old now sore for 4-6 weeks with biopsy proven LS 10 years but problems with sexual dysfunction for years
- Treatment variable as “not a problem”
- Bx – Invasive SCC
To Effectively Manage LS

• Educate providers and patients
• Treat according to signs not symptoms
• Follow for lifetime
Question -
• If LS had been treated could cancer have been prevented?
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If LS had been treated could cancer have be prevented?

• 1 study - small study... yes
Role of LS in Malignant Change

- Proposed that LS may generates a field of molecularly altered epithelium
- There is a propensity for malignant change with LS
- This is also seen in ulcerative colitis another chronically inflamed tissues disease and steroid Rx reduces that risk. (Eaden 2000)

Role of LS in Development SCC

• Still debatable whether LS is a precursor lesion of dVIN or whether dVIN gives rise to HPV-negative VSCC (Yap et al. 2017)

• Many studies have found that local vulvar recurrence in vulvar SCC were more likely to occur in the residual LS left behind after surgery (Regauer 2011; Yap et al. 2016)

• It is believed that chronic inflammation induces cumulative molecular changes to the underlying epithelium, which, over time, leads to the development of recurrence vulvar SCC


• 69 year lady lady with LS for 20 years and this is 3rd HPV, negative SCC recurrence on the vulva
• All vulvar SCC have been at different vulvar sites
• Never symptoms - just feels “lump”
Role of LS in Vulvar Intraepithelial Neoplasia and Squamous Cell Carcinoma

A survey on the use of topical steroids in patients treated for lichen sclerosus-associated vulval squamous cell carcinoma.

• Does treatment of LS prevent cancer and cancer recurrence?

• More studies needed
LS with dVIN or SCC

Usually symptomatic with variable itch, burn, irritation
Asymptomatic LS

• Not uncommon
• Need better education
• All women need yearly vulvar checkup
• LS need lifetime treatment based on treatment response
• Maybe avoid SCC development in LS cases?