

BENIGN VULVAR SKIN DISORDERS

Week 40

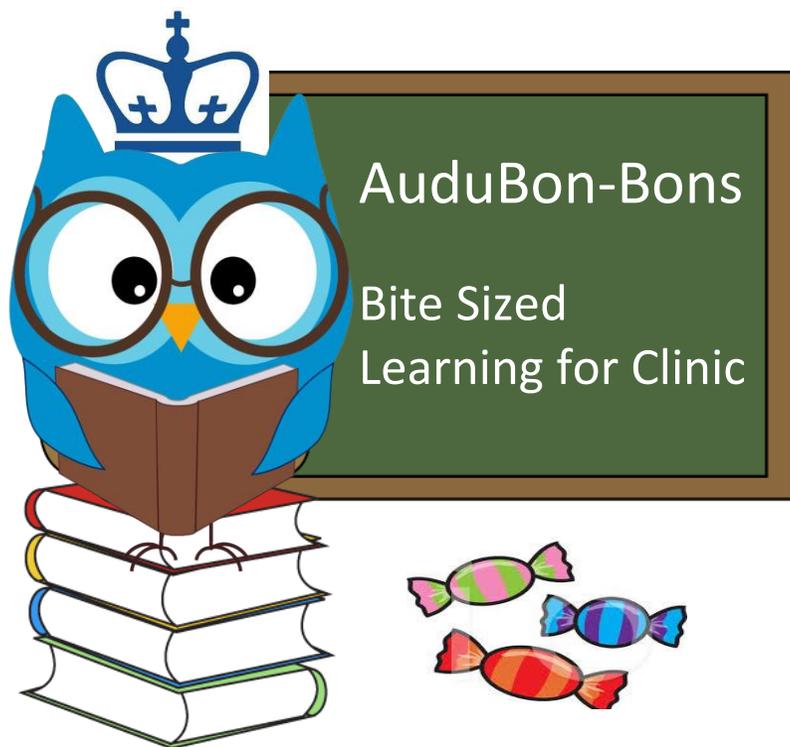
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Reading Assignment:

ACOG Practice Bulletin No. 93: diagnosis and management of vulvar skin conditions (May 2008)



LEARNING OBJECTIVES



- To formulate a differential diagnosis for vulvar lesions using a morphology-based system
- To correctly identify lichen sclerosis on physical exam
- To understand the risk of progression to malignancy for lichen sclerosis
- To feel comfortable treating lichen sclerosis



CASE VIGNETTE

- A 70 yo G2P2 woman presents with 6 weeks of difficulty sleeping due to intensely “itchy genitals.” She reports having some pain with defecation and intercourse for the past two weeks.



FOCUSED HISTORY

What elements of this patient's history are most relevant?

- **PMH:** HTN, HLD
- **PSH:** None
- **POBH:** G2P2, NSVDx2, no complications
- **PGYNH:** No abnormal Paps. Menopause at age 53
- **MEDS:** Propanolol, atorvastatin, fish oil
- **ALL:** None
- **FAM:** No family history of eczema or psoriasis
- **SOCIAL:** Retired teacher, lives with husband, no new sexual partners



PERTINENT PHYSICAL EXAM FINDINGS

What elements of this patient's physical exam are most relevant?

- **General:** Well-appearing woman
- **HEENT:** PERRLA, no nasal lesions, no lesions in the mouth
- **Abdominal Exam:** +BS, non-tender to palpation in all four quadrants, no hepatosplenomegaly
- **External Exam:**
 - **Vulva:** Appropriate hair distribution, edema of labia minora with excoriations, multiple white plaques, interlabial fold fissuring
 - **Vagina:** Atrophy, no lesions, no evidence of cystocele/rectocele, narrow introitus
 - **Urethra:** No masses, tenderness, or scarring
 - **Anus:** Peri-anal excoriations
- **Speculum/Internal Exam:**
 - Significant discomfort with insertion of speculum
 - **Cervix:** Nontender, multiparous, no discharge
- **Bimanual Exam:**
 - **Uterus:** Mobile uterus with regular contour
 - **Adnexa:** No adnexal masses palpated bilaterally, no tenderness
- **Rectovaginal Exam:** No masses or tenderness



FOCUSED PHYSICAL EXAM FINDINGS

This patient has multiple white plaques on vulvar exam.

What characteristics of the vulvar lesion should be evaluated?

- Lesion morphology
- Number, location, and distribution of multiple lesions: scattered, grouped, linear
- Edge: clearly demarcated vs poorly defined
- Color
- Consistency and feel: tenderness, thickness, hard/soft/firm/fluctuant
- Presence of secondary changes: excoriation, lichenification, edema, scale, crust, fissure, erosion, bleeding, hyperpigmentation, hypopigmentation, atrophy, scar
- Acute inflammation: edema, pain, erythema



FOCUSED PHYSICAL EXAM FINDINGS

Differential diagnosis can be established using a lesion morphology-based system (most common benign diagnoses included)

| | |
|------------------------------------|--|
| Red patches and plaques | candidiasis, atopic dermatitis/eczema, lichen simplex chronicus, irritant contact dermatitis, psoriasis |
| Erosions | excoriation, fissures, lichen simplex, herpes simplex, candidiasis, erosive lichen sclerosus, erosive lichen planus, impetigo, irritant contact dermatitis |
| Red papules and nodules | folliculitis, cherry angioma, angiokeratoma, hemangioma, furunculosis, hidradenitis suppurativa, hemangioma, vestibular papillomatosis |
| Skin-colored lesions | vulvar papillomatosis, skin tag, HPV wart, molluscum contagiosum, lichen simplex, scar, epidermal cyst |
| White papules and nodules | Fordyce spots, milium, epidermal cyst, molluscum contagiosum, HPV infection |
| Yellow papules and pustules | candidiasis, folliculitis, furunculosis, hidradenitis suppurativa, molluscum contagiosum |
| Ulcerative lesions | HSV, HIV, aphthous ulcers, contact dermatitis, Crohn disease |
| Edematous lesions | postsurgical/postradiation edema, Crohn disease, hidradenitis suppurative, pregnancy, edema due to infection |
| White patches and plaques | vitiligo, postinflammatory hypopigmentation, lichen sclerosus, lichen planus, lichen simplex, candidiasis, HPV wart |
| Brown blue or black lesions | physiologic hyperpigmentation, postinflammatory pigmentation, HPV wart, melanocytic nevus, seborrheic keratosis, angiokeratoma, skin tags |
| Vesicles and bullae | HSV, herpes zoster, impetigo, contact dermatitis |

FOCUSED PHYSICAL EXAM FINDINGS

Your patient has the following findings on pelvic exam:



DIFFERENTIAL DIAGNOSIS

What are diagnoses on the differential?

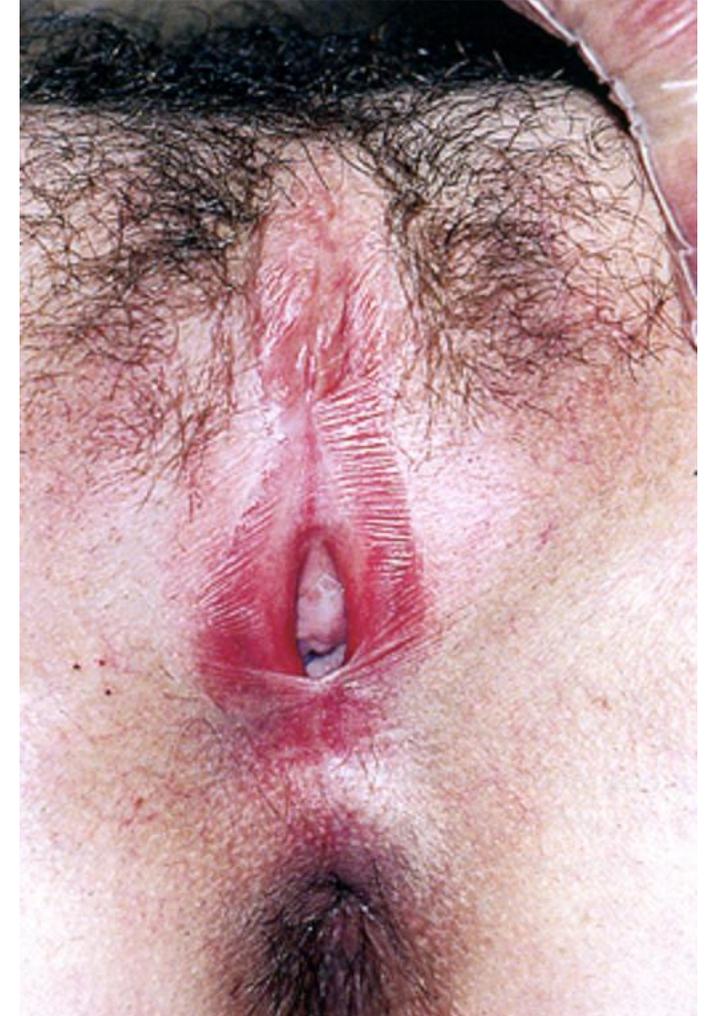
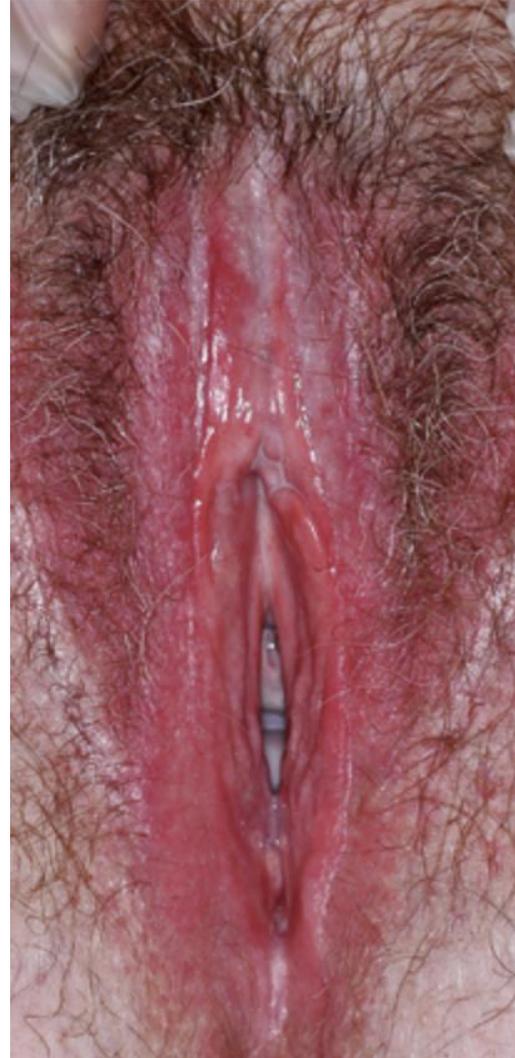
- Lichen sclerosus
- Lichen planus
- Vulvar dermatitis
- Vitiligo
- Mucous membrane pemphigoid
- Candidiasis
- Estrogen deficiency 2/2 menopause

LICHEN SCLEROSUS - OVERVIEW

- A benign, chronic, progressive dermatologic condition characterized by marked **inflammation, epithelial thinning, and distinctive dermal changes accompanied by symptoms of pruritus and pain**
- Epidemiology:
 - Two peaks of onset: **prepubertal girls** and **perimenopausal/postmenopausal women**¹
 - Prevalence: **~1 in 59 women** in general gynecology practice²
- Etiology:
 - Unknown

LICHEN SCLEROSUS – PHYSICAL EXAM FINDINGS

- Classic vulvar lichen sclerosis is expressed as **white, atrophic papules that may coalesce into plaques**
- Lesions most frequently affect the **labia minor** and/or **labia majora**, although the whitening may extend over the perineum and around the anus in a **keyhole** fashion
- **Fissuring** is frequently seen at the posterior fourchette, perianally, in the interlabial folds, or around the clitoris
- Scratching may result in **excoriations**, often associated with edema of the labia minora and prepuce
- Vulvar architecture remains intact early in the course of the disease
 - As the disease progresses, the distinction between the labia minora is lost and the clitoris becomes buried under the fused prepuce. Shrinkage of the introitus and perineum causes dyspareunia
- At the end-stages of lichen sclerosis, the **vulva is pallid and featureless** due to **midline fusion**, which leaves only a posterior pinhole orifice



LICHEN SCLEROSUS – EXTRA-VULVAR MANIFESTATIONS

Both lichen sclerosus and lichen planus cause intense itching and loss of architecture. Which disease usually involves both the vulva and the vagina?

- Lichen planus

Are there extra-vulvar manifestations of lichen sclerosus?

- Very rarely
 - There are a few case reports of combined vulvar and vaginal lichen sclerosus ^{3,4}
 - The vagina, cervix, eye, mouth, nares, and intertriginous regions should be examined when the diagnosis is uncertain

LICHEN SCLEROSUS - MANAGEMENT

Physical exam and biopsy results conclude your patient has lichen sclerosis. What treatment do you offer her?

- Superpotent topical corticosteroid (**clobetasol propionate 0.05%**) applied nightly for **6-12 weeks**
 - Improves physical manifestations and patient-reported symptom scores¹⁰
- Re-evaluate after initial 6-12 weeks of therapy
 - A good response is indicated by relief of pruritus and pain, and resolution of hyperkeratosis, fissuring, and ecchymoses¹¹
- Transition to maintenance treatment: corticosteroid usage 2-3 nights/week
- With well-controlled disease, patient should be examined once yearly

Your patient asks how likely it is that she will be able to achieve remission. What do you tell her?

- In one prospective study, no patient over age 70 years had complete clinical and histologic remission¹²
- Among those who did achieve remission, 50% relapsed within 16 months, and 84% relapsed within 4 years¹²

LICHEN SCLEROSUS – MALIGNANCY

Your patient asks if this could “be serious.” Are women with vulvar lichen sclerosis at increased risk of malignancy?

- Yes ^{5,6}

Which malignancy?

- Squamous cell carcinoma of the vulva

What is the risk?

- Less than 5% ⁷

What factors reduce the risk of development of squamous cell carcinoma? ^{8,9}

- Earlier detection
- Introduction of potent topical glucocorticoids
- More liberal use of biopsy (any area that fails to respond to therapy should be biopsied)
- Excision of abnormally thickened skin resistant to treatment



SOCIAL DETERMINANTS OF HEALTH

Lichen Sclerosus: What factors cause significant impact on quality of life?

Irritation and pain

- Itching
- Burning
- Fissures from defecation

Shame and fear

- Location on intimate places

Dyspareunia & Sexual distress

- Fissures from coitus
- Anticipation of pain decreases arousal and lubrication causing pelvic muscles to contract
- Hooding of clitoris, labial fusion, introital stenosis cause pain and difficulty achieving orgasms

Diagnostic Delay

- Longer window between the start of symptoms and establishing the diagnosis associated with lower quality of life

Early diagnosis and early treatment will improve quality of life.

High number of symptoms correlates with a worse quality of life score, thus resolving one or more symptoms can improve quality of life!



Epic .phrase

.BBonLichenScler

Description: Lichen sclerosus treatment counseling

The patient was informed of the suspected diagnosis of lichen sclerosus. She was counseled that while her symptoms could be the result of many etiological factors, her history, physical exam, and ***biopsy suggest lichen sclerosus. It was explained that the goal of therapy will be a reduction of symptoms.

We discussed treatment options and will begin with topical ***clobetasol propionate 0.05% nightly for 6-12 weeks with a plan to re-evaluate after initial 6-12 weeks of therapy.

The increased risk of squamous cell carcinoma associated with lichen sclerosus was also discussed. She was counseled on the factors that reduce the risk of malignancy, including early detection, introduction of potent topical glucocorticoids, use of biopsy to evaluate any area that fails to respond to therapy, and excision of abnormally thickened skin resistant to treatment.



TAKE HOME POINTS

- The classic appearance of lichen sclerosus is thin, white, wrinkled skin localized to the labia minora/majora, with whitening extending to perineum/anus in keyhole fashion, epithelial hyperplasia, and fissuring
- Vulvar pruritus is a common symptom of lichen sclerosus
- The diagnosis of lichen sclerosus is made from physical exam and biopsy
- Treat newly diagnosed lichen sclerosus with clobetasol propionate 0.05% ointment nightly for 6-12 weeks
- There is an increased risk of squamous cell carcinoma of the vulva in patients with lichen sclerosus



CODING/BILLING

- ICD-10-CM Diagnosis Code:
 - L90.0: Lichen sclerosus et atrophicus



REFERENCES

1. Bleeker, Maaïke CG, et al. "Lichen sclerosus: incidence and risk of vulvar squamous cell carcinoma." *Cancer Epidemiology and Prevention Biomarkers* 25.8 (2016): 1224-1230
2. Goldstein, Andrew T., et al. "Prevalence of vulvar lichen sclerosus in a general gynecology practice." *The Journal of reproductive medicine* 50.7 (2005): 477-480.
3. Longinotti, Mindy, Yvette M. Schieffer, and Raymond H. Kaufman. "Lichen sclerosus involving the vagina." *Obstetrics & Gynecology* 106.5 (2005): 1217-1219.
4. Zendell, Kate, and Libby Edwards. "Lichen sclerosus with vaginal involvement: report of 2 cases and review of the literature." *JAMA dermatology* 149.10 (2013): 1199-1202.
5. Bleeker, Maaïke CG, et al. "Lichen sclerosus: incidence and risk of vulvar squamous cell carcinoma." *Cancer Epidemiology and Prevention Biomarkers* 25.8 (2016): 1224-1230.
6. Leibowitch, Michele, et al. "The epithelial changes associated with squamous cell carcinoma of the vulva: a review of the clinical, histological and viral findings in 78 women." *BJOG: An International Journal of Obstetrics & Gynaecology* 97.12 (1990): 1135-1139.
7. Neill, S. M., et al. "British Association of Dermatologists' guidelines for the management of lichen sclerosus 2010." *British Journal of Dermatology* 163.4 (2010): 672-682.
8. Jones, Ronald W., et al. "Guidelines for the follow-up of women with vulvar lichen sclerosus in specialist clinics." *American journal of obstetrics and gynecology* 198.5 (2008): 496-e1.
9. Lee, Andrew, Jennifer Bradford, and Gayle Fischer. "Long-term management of adult vulvar lichen sclerosus: a prospective cohort study of 507 women." *JAMA dermatology* 151.10 (2015): 1061-1067.
10. Bracco, G. L., et al. "Clinical and histologic effects of topical treatments of vulval lichen sclerosus. A critical evaluation." *The Journal of reproductive medicine* 38.1 (1993): 37-40.
11. Neill, S. M., et al. "British Association of Dermatologists' guidelines for the management of lichen sclerosus 2010." *British Journal of Dermatology* 163.4 (2010): 672-682.
12. Renaud-Vilmer, Catherine, et al. "Vulvar lichen sclerosus: effect of long-term topical application of a potent steroid on the course of the disease." *Archives of dermatology* 140.6 (2004): 709-712.
13. Haefner HK, Aldrich NZ, Dalton VK, et al. The impact of vulvar lichen sclerosus on sexual dysfunction. *J Womens Health (Larchmt)*. 2014;23(9):765-770. doi:10.1089/jwh.2014.4805
14. Van de Nieuwenhof HP, Meeuwis KA, Nieboer TE, Vergeer MC, Massuger LF, De Hullu JA. The effect of vulvar lichen sclerosus on quality of life and sexual functioning. *J Psychosom Obstet Gynaecol*. 2010;31(4):279-284. doi:10.3109/0167482X.2010.507890
15. Lansdorp CA, van den Hondel KE, Korfage IJ, van Gestel MJ, van der Meijden WI. Quality of life in Dutch women with lichen sclerosus. *Br J Dermatol*. 2013;168(4):787-793. doi:10.1111/bjd.12137

