Vulvar Dermatology: Pearls
From a Specialty Clinic
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Top Ten Tips
1) When it comes to vulvar concerns, our expertise makes us well equipped to help our gynecology colleagues.
2) Look closely. Know normal anatomy. Notice if something is missing.
3) Don’t be afraid to biopsy. Easy, often helpful, never wrong.
4) Vulvar conditions affect the woman’s sexuality. Ask and utilize.
5) Vulvar papillomatosis is not HPV. Soft, monomorphous columnar flesh-toned papules at introitus.
6) Lichen sclerosus requires long-term treatment with ultrapotent topical steroids, for life.
7) When treating women with erosive gingival lichen planus, ask about genital symptoms.
8) If the vulva itches, it’s candida until proven otherwise. Treatment takes longer by the time it gets to your clinic (no one-dose treatment).
9) Vulvar Crohn’s requires systemic treatment, and is a worthy endpoint.
10) Vulvodynia is neuropathy, not a wastebasket term.

Vulvar Anatomy
Vulvar Papillomatosis

Range of Pathologic Diagnoses
- Dermatitides
- Infections
- Benign neoplasms
- Malignant neoplasms
- Trauma
- Neuropathic

Dermatitides
- Lichen Sclerosus
- Lichen Planus
- Lichen Simplex Chronicus
- Zoon's Plasmacellular Vulvitis
- Desquamative Inflammatory Vaginitis?
- Psoriasis
- Crohn's
- Nonspecific vulvitis due to vaginitis
- Contact: irritant, allergic
- Other blistering conditions not specific to the vulva
Case

- 73 year old woman with 5 year history of itch
- Treated for yeast infections, not improved

Lichen Sclerosus

- Rare, chronic, inflammatory condition affecting genital and extragenital skin
- 1:1,000 women
- Autoimmune condition occurring in genetically predisposed patients
- Chronic trauma and an occlusive moist environment may precipitate
- Higher rate of other autoimmune: AA, thyroid, vitiligo (OR 4.3)
- Bimodal peak, prepubertal and postmenopausal women, but can occur in any age
- 7-15% cases occur in kids
- 11% extragenital disease (Funaro, 2014)
Lichen Sclerosus: clinical features

- Pale, white, “waxy”, “atrophic”
- Purpura
- Pruritis, excoriations
- Fissures
- Pain, dyspareunia
- Scarring
  - Labial agglutination, resorption
  - Narrowing of introitus, tearing with intercourse
  - Adhesions to clitoris progressing to complete burying
- SCC risk 4-6% (vs 1% in controls)

Lichen Sclerosus

Lichen Sclerosus: Candida

- Candida infection commonly complicates and can make harder to manage
- Candida responsible for acute flares, nonresponse to treatment
- Many require chronic management of candida
- Index of suspicion for candida should be low
- Test as part of initial workup, retest if indicated by symptoms and to confirm clearance if initial testing positive
Lichen Sclerosus: Treatment

- Clobetasol
- Tacrolimus
- Pimecrolimus: for itch only
- Mid-potency steroids
- No role for topical testosterone, dihydrotestosterone or progesterone. *JAAD*, 67 (2), 305-312.

Scarring/agglutination is PREVENTABLE with proper management; requires long term use of ultrapotent topical steroids

- Clobetasol 0.05% OINTMENT is gold standard (not cream)
- Educate properly to use sparingly, only on affected areas.
- Pea size (chocolate chip) for entire vulva
- Start twice daily for 2 months, then follow up
- Taper to TIW if/when possible
- Goal is complete objective remission

Treatment is for a lifetime

- Never treat to symptoms alone!
- Reassure patients about steroid use. Steroid atrophy is reversible.
- Can add tacrolimus 0.1% (such as M-F, continue clobetasol S,S) if atrophy a problem
- Treat/control candida, HSV
- Good vulvar care; avoid irritants
- Follow patients closely for treatment response and to monitor for SCC
Lichen Sclerosus: Sex

- Women with lichen sclerosus have sexual dysfunction.
- Even with adequate treatment, sexual dysfunction persists.
- Dyspareunia, decreased orgasm, and decreased coital frequency compared to unaffected women. (Burrows LJ, 2011)
- Consider coexisting vulvodynia or pelvic floor dysfunction
- Refer to physical therapy and/or sex therapy

Lichen Sclerosus: Treatment Failure

- Incorrect diagnosis
  - Ulcers should be biopsied
  - Discrete thickened areas, shaggy hyperkeratosis, concern for VIN or SCC
- Missed concurrent conditions
  - Candida
  - HSV
  - Low-estrogen atrophy
  - Contact dermatitis
- Noncompliance
  - Fear of steroids
  - Treating to symptoms, not to resolution of clinical findings
  - Inadequate education
  - Physical disability
  - Inadequate follow up
Malignant Melanoma Associated with Lichen Sclerosus in the Vulva of a 10-Year-Old

Lichen Sclerosus: Systemic Treatment

- For severe, recalcitrant cases
  - Cyclosporine
  - Mycophenolate mofetil
  - Methotrexate
  - Acitretin or isotretinoin

Case

52 year old woman complaining of painful intercourse for the past year. She also complains that her discharge seems "sticky".
Erosive vulvovaginal lichen planus

- Distinct subset of lichen planus
- Usually lacks the typical involvement of glabrous skin (5% have)
- 68% show clinical signs of oral LP
- 20-25% of women with oral LP have vaginal involvement
- Peri-menopausal onset typical (mean age 40-62)
- Not seen in children

- Affects modified mucosa and mucosa (medial labia minora, vestibule, introitus, vagina)
- "Glazed erythema", dusky red or reddish brown patches
- Lacy, reticulate white plaques (Wickham’s striae), can be very subtle
- Painful, tender
- Dyspareunia common
- Itchy
- Bleeds easily
- Erosions common
- Agglutination and loss of labia minora, clitoris, prepuce.
Erosive Vulvovaginal Lichen Planus

- “Usually” associated with inflammatory vaginitis
- Vaginal epithelium reddened; variable erosive patches which bleed with trauma
- Longstanding vaginal involvement results in synechiae and obliteration of vagina
- Increased secretions; may be yellowish, sticky, itchy and irritating
- PMNs, immature epithelial cells, no clue cells or yeast, no lactobacilli
- pH high, 5-6

Erosive LP: Malignant Transformation

- Uncommon. True incidence unknown.
- Vulvar carcinoma almost always occurs in setting of pre-existing skin disease.
- Suspect SCC in the chronic, indurated or granulated ulcer, or white hyperkeratotic plaque.
Erosive Lichen Planus: Diagnosis

- **Differential:**
  - Lichen sclerosus
  - Similar whitening, agglutination/scarring
  - Does not involve vagina
  - Association with LP higher than allowed by chance alone
  - Mucous membrane pemphigoid
  - Pemphigus vulgaris
  - Erythema multiforme
  - Fixed drug reaction

- **Biopsy:**
  - Edge of erosion, or un-eroded red or white papule
  - H & E, DIF and IIF

Erosive Lichen Planus: Treatment

- Clobetasol 0.05% ointment
- C-Clobetasol 0.05%, oxytetracycline 3%, nystatin 100,000u/g
- C-hydrocortisone 50-100 mg/gram inserted per vagina at HS
- Topical estrogen
- Treat concurrent yeast, HSV
- Tacrolimus 0.1% ointment BID, taper to qd as possible
- Compounded 1 mg vaginal suppository? (less irritating)
- Monitor serum levels
- Arch Dermatol. 2004;140:715-720

Erosive LP: Systemic Therapy

- For failure of topical therapy: Has topical therapy been optimized? No complicating factors?
- No one systemic treatment consistently effective
- Systemic steroids
- Plaquenil 200mg BID
- Delayed effect
- Oral cyclosporine
- Mycophenylate mofetil (500-3000 mg daily)
- Methotrexate, azathioprine, cyclophosphamide
- Retinoids?
Erosive Lichen Planus: Surgery?

- Not indicated
- Can worsen disease
- Scars recur immediately, often worse
- Dilator use preferred if issue is vaginal introitus or vagina aperture (length is harder to improve)
- If disease entirely quiet, can consider surgery, but followed by use of vaginal mold with topical steroids

Vulvar Crohn's Disease

- Uncommon extraintestinal manifestation of Crohn's; rare in a pediatric population
- Edema, induration, erythema of the labia majora and labia minora, knife-cut ulcers, perianal skin tags, anal fissures, inguinal adenopathy, suppuration
- Timing and course are independent of manifestations of intestinal Crohn's
- A valid endpoint for treatment. Responds more slowly than intestinal Crohn's
- Biopsy may be instructive (noncaseating granulomas)
- Look for symptoms of weight loss, diarrhea, aphthous stomatitis, pyoderma gangrenosum, erythema nodosum, arthritis
Vulvar Crohn’s Disease


Vulvar Crohn’s Disease in Children

Pediatric Dermatology


Vulvodynia

- Chronic neuropathic pain with no identifiable cause
- Burning, stinging, itching, raw
- Localized or generalized
- Provoked or unprovoked
- Primary or secondary
- Normal appearance
- May coexist with other vulvar disorders, cannot diagnose while those are active
Vulvodynia: possible causes

- Nerve injury or irritation
- Abnormal response of nerves to infection or trauma
- Chronic inflammation
- Hypersensitivity to candida
- Pelvic floor dysfunction

Vulvodynia: treatment

- Physical therapy and biofeedback
- Topical local anesthetics
- Topical compounded anticonvulsants, TCA, muscle relaxants
- Intravaginal valium
- Tricyclic antidepressants, anticonvulsants (gabapentin, Lyrica), SSNRI (duloxetine, venlafaxine), SSRI (citalopram)
- Pudendal nerve blocks
- Vestibulectomy (surgery)
- Sexual counseling

Vulvodynia

- Not a wastebasket term
- Diagnosis of exclusion
- Increasingly treatable
- Requires an expert approach
Thank you!