Diagnosis and Management of Vulvar Disorders

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Disclosure of relationships with industry
Principal Investigator: Janssen, Celgene, Lilly
None of these are relevant to this talk

Disclaimers
Most medications discussed in this lecture are not FDA approved for these diseases. For example, there are NO topical corticosteroids formulated for the vagina. These are common, but orphan, diseases with very little adequate research.

Acknowledgements

Dr. Libby Edwards
http://www.midcharlottedermatologyresearch.com/
Objectives

• Recognize normal variations in vulvar anatomy
• Recognize the clinical features of the most common vulvar disorders
• Differentiate amongst vulvar disorders based on appearance and symptoms
• Understand treatment options for the most common vulvar disorders

Basic Premises

• Normal variants can be confusing

Normal Hyperpigmentation
Basic Premises

• Look carefully for subtle abnormalities
Basic Premises

- Presentations of skin diseases are often atypical or non-specific compared to dry skin
Basic Premises

• Any inflammatory dermatosis can produce resorption of normal vulvar architecture

Tips for Diagnosis

• Look at other mucous membranes and skin surfaces
Tips for Diagnosis

- Biopsy specific lesion
- Send to a dermatopathologist with your presumptive diagnoses

Genital Biopsies

- Inform patients that these are not a lab test, and a diagnostic result is not assured
- Chances of yield depend on:
  - Site sampled
  - Age of lesion
  - Information provided to pathologist
  - Expertise of pathologist
  - Luck
Genital Biopsies

- Offer topical anesthetic
- Use lidocaine with epinephrine
  - Wait 10 minutes for vasoconstriction
- Punch (4 mm) or shave

Shave biopsy technique
Genital Biopsies

- Re-biopsy serially for concerns about SCC
- Re-biopsy for non-diagnosis if appearance changes
- Re-biopsy even if diagnostic if inadequate response to treatment or appearance changes

Therapeutic Principles for Chronic Genital Disease

- Explanation of the disease, treatment and expectations
- Treat all possible factors, including trivial abnormalities
- Anticipate iatrogenic disease
  - Prevent yeast, irritant contact, etc.

Therapeutic Principles for Chronic Genital Disease

- When in doubt, use a tricyclic for pain, sedation, itching
- Avoid cream vehicles on eroded, painful, and prepubertal skin
- The vulva is relatively corticosteroid resistant, often requiring ultrapotent medication
Specific Vulvar Diseases

- Vulvar Dermatitis
  - Irritant Contact
  - Allergic Contact
  - LSC
  - Psoriasis
  - Infectious
- Lichen Sclerosus
- Lichen Planus

Vulvar Dermatitis

- Acute
  - Irritant Contact Dermatitis
  - Allergic Contact Dermatitis
  - Infectious
- Subacute/Chronic
  - 50% of chronic vulvovaginal pruritus is ACD or ICD
  - Eczema/Lichen Simplex Chronicus
  - Psoriasis

Acute Contact Dermatitis

- Morphology
  - Edema
  - Erythema
  - Vesicles with prompt erosions
- Irritant Contact Dermatitis
  - Burning
  - Irritation
- Allergic Contact Dermatitis
  - Itching

Irritant Contact Dermatitis

- Direct damage to keratinocytes
- No prior sensitization
- Irritation, rawness, burning
- Itching less than Allergic contact
- Vulvar ICD
  - Topical treatments for underlying disease
  - Skin care habits
  - Inflammation = facilitate passage of irritants

Common Vulvar Irritants

- Urine
- Feces
- Sweat
- Abnormal vaginal discharge
- Excessive hygiene
- Feminine hygiene products
  - Lubricants, pads, wipes
- Soaps and detergents
- Hair dryer
- Medications
  - Alcohol-based creams and gels, spermicides, propylene glycol

Irritant Contact Dermatitis

- Edema
- Erythema
- Erosions

Irritant contact dermatitis

From Trichloroacetic acid

Irritant contact dermatitis

From vaginal fluorouracil
Irritant Contact Dermatitis
From Lysol

Allergic Contact Dermatitis

• Incidence
  – Estimated between 15 and 30%\(^3,4\)
  – Relevant patch tests in 25%\(^4\)

• Anatomical Considerations
  – Occlusion, hydration, and friction
  – More permeable than exposed skin\(^5\)
  – Higher incidence of contact sensitivity
  – Study of vulval pruritus = 44% with 1 or more relevant contact allergens\(^6\)

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Common Vulvar Allergens

- Topical anesthetics
  - Lidocaine, Benzocaine, Tetracaine
- Topical antibiotics
  - Neomycin, Bacitracin, Polymyxin
- Antifungals
  - Imidazole, Nystatin
- Fragrance
- Preservatives
  - MI/MCI
- Corticosteroids
- Lanolin
- Rubber-latex
- Spermicides


Allergic Contact Dermatitis

- Erythema and vesicles
- Reaction can spread outside area of contact

Allergic contact dermatitis

- Topical Diphenhydramine
Contact Dermatitis Treatment

- Patient education
  - Reassurance
  - Handouts/Written Instructions
- Stop Use of Potential Irritants
- Pharmacologic treatments
  - Treat inflammation
  - Treat itching
  - Prevent or treat infection

Contact Dermatitis Treatment

- **Potent Topical Steroids**
  - Corticosteroid ointment for 3-4 weeks
    - Betamethasone 0.05% ointment BID if severe
    - Triamcinolone 0.1% ointment BID if moderate
  - Prednisone 0.5 to 1 mg/kg tapered over 2-3 weeks if severe

- **Treat itching**
  - Anti-histamines
    - Hydroxyzine, doxepin, fexofenadine, cetirizine, loratadine

- **Prevent or treat infection**
  - Oral fluconazole 150 mg weekly while on topical steroids for Candida suppression
  - Oral antibiotics as needed for secondary impetiginization


Topical Steroid Application

- Demonstrate application
- Use photos and mirrors
- Less than a pea-sized amount

Topical Steroid Application

- Small amount twice daily
  - Taper when fully responsive
  - Don’t stop until follow-up
  - Disease dependent
- Modified mucous membranes steroid resistant
  - Labia minora, medial labia majora, vestibule, clitoris, and clitoral hood
- Hair bearing skin more sensitive
  - Lateral labia majora, inguinal area, medial thighs, perianal
Steroid Dermatitis

Milia and steroid dermatitis
From years of triamcinolone

Striae
Topical Steroid Adverse Effects

- Mostly resolve with discontinuation
- Provide reassurance
- Exception is long-term steroid under diaper occlusion

Infectious Vulvar Dermatitis

- Vulvar itching
  - Most common mistake in chronic itching is over-diagnosis of yeast
  - Elicit history of contactants for allergic contact
  - History of pleasure with scratching = LSC
- Examine for infection or skin disease
  - Wet mount
  - Culture if wet mounts persistently positive or patient unresponsive to treatment

Infectious Vulvar Dermatitis

- Candida Albicans
  - Non-Albicans Candida produces irritation > itching
- Folliculitis
  - Bacterial or fungal
- Trichomonas
- HSV
- Bacterial Vaginosis
Candida

Antibiotic

+ Clobetasol

+ New Estrogen

Subacute/Chronic Vulvar Dermatitis

- Present with eczematous changes
  - Variable Erythema
  - Sometimes lichenification
  - Excoriations
  - Fissures
  - Weeping
- Causes
  - Irritant or Allergic Contact Dermatitis
    - Covered under Acute Dermatitis
  - Eczema/Lichen Simplex Chronicus
  - Psoriasis

Lichen Simplex Chronicus

- Itch-scratch cycle
  - Precipitated by an irritant/infection
- Manifested primarily by lichenification
- Excruciating itching and pleasure with scratching
- Usually, but not always, a history of atopy
LSC Morphology

- Exam Findings
  - Erythema
  - Swelling
  - Accentuation of skin markings
- Findings can be subtle, even with significant disease
- Morphology very variable according to skin type and location

Lichen simplex chronicus

Treatment for LSC

- Patient education
- High potency corticosteroid
  - BID for a month
- "Soak and seal"
  - Tub soaks and petroleum jelly
- Eliminate irritants
  - Give a handout on the avoidance of irritants
  - Over washing, panty liners, wipes, benzocaine, latex, spermicides, condoms/diaphragms, OTC medications, certain lubricants
Treatment for LSC

- Control nighttime scratching
  - Amitriptyline
  - Start with 10 mg 2 hours before bedtime
- Treat concomitant infection if present
- Xylocaine/lidocaine 2% jelly
  - As needed for itching on modified mucous membranes

Before therapy

Baseline  Month 1  Month 2

After therapy
Resistant LSC

- Psychological factors
- Neuropathic itch
  - Treat like vulvodynia
- Rule out secondary process
  - Vaginal infection
  - Contact dermatitis
- Non-compliance

Psoriasis

- Atypical or non-specific on vulva
- Any inflammatory process can cause resorption of normal architecture
- Morphology
  - Pink
  - Poorly demarcated
  - Subtle scale
  - Can look similar to lichen simplex chronicus
Psoriasis

- Check other areas of the skin for clues
- Consider biopsy if uncertain
- Treatment
  - Topical steroids first line
  - Phototherapy
  - Biologics
  - Refer to dermatology

Lichen Sclerosis

- Probably autoimmune primarily with other influences
  - Genetic, hormonal, local environment
- Histology shows features shared with other hyper-immune/autoimmune disease
  - Lichen planus, Graft-vs-host disease, Lupus
- Increased autoantibodies
- Clusters with vitiligo, LP
Lichen Sclerosus 
Morphology

• Classic - white, crinkled plaques on the vulva, perineum, and perianal skin (*figure of 8*)
• Sometimes waxy texture
• Sometimes shiny
• Sometimes thickened hyperkeratotic texture
• Always texture change

Lichen Sclerosus 
Symptoms

• Itchy
• Pain from erosions
• Dyspareunia
• Fragility and from scarring, narrowing of introitus
• Often complicated by estrogen deficiency, secondary infection

Smooth, waxy plaque of LS producing pain and irritation
White, shiny skin with loss of architecture

Thickened, hyperkeratotic, white skin in a setting of loss of architecture
Lichen sclerosus
But could be LP or LSC

Lichen Sclerosus
• 5-10% of women have extra-gential disease
• No vaginal disease
• No oral disease
Lichen Sclerosus

- Treatment in women is an ultrapotent corticosteroid, or an ultrapotent corticosteroid, or an ultrapotent corticosteroid, or tacrolimus

Before clobetasol  4 mos clobetasol

Lichen Sclerosus

- Treat BID until texture is normal
- Then, best therapy is controversial
- Some treat intermittently guided by symptoms
- Many experts feel strongly that ongoing 2 or 3 x weekly treatment is needed to prevent scarring and decrease the risk of SCC
Lichen Sclerosus
Other Options
- Tacrolimus ointment twice daily
  - Irritating, less effective, black box warning
- Mid-potency corticosteroids such as triamcinolone relieve symptoms
  - ? Effect on scarring, texture, and color
- Tretinoin (Retin-A)
  - Generally too irritating, but has been reported (once) as useful for hyperkeratotic disease

LS with Vulvar melanosis

Lichen Sclerosus
Other Considerations
- SCC associated with LS in 3-5% of untreated patients
- Most experts believe that successful control dramatically decreases this risk
SCC arising within LS

Lichen Planus

- Autoimmune, cell-mediated disease of older women
- Genital disease more common in women, most often erosive
- Men more likely to exhibit papular disease
Lichen Planus

Morphology

- Nonspecific erosions
- White, reticulate, or fernlike striae
- Sometimes solid white epithelium that resembles LS
- Red or white papules most common in men

Non-genital Lichen Planus

Non-specific erosions
Lichen Planus

- Scarring is prominent on vulva and in vagina
- Usually, only mouth and vulvovaginal skin affected
- Men generally experience no scarring
- Diagnosis by PE & confirmed on biopsy of white skin (women) or red papules (men)

LP- Vagina and Mouth
Lichen Planus Treatment

- Stop irritants, control infection
- Corticosteroids - If prominent erosions, consider prednisone 40-60 mg PO and re-evaluate each week
- Otherwise, ultra-potent steroid (clobetasol) bid – follow for AE’s
- Protopic ointment twice daily can be added (stings), black box warning

Lichen Planus
Remember vagina & mouth!

- Topical corticosteroids and Protopic for these areas as well
- Clobetasol gel QID for mouth, ointment QHS for vagina (high risk for candidiasis) - consider systemic absorption
- Insert dilator (or intercourse) daily to prevent adhesions
Lichen Planus Treatment

- Hydroxychloroquine (Plaquenil) 200 mg*
- Topical or oral cyclosporine
- Methotrexate up to 25 mg q wk*
- Mycophenylate (CellCept) up to 3 g/d
- Etanercept (Enbrel) 50 mg SQ 2X/wk
- Azathioprine/cyclophosphamide
- Retinoids (Accutane, Soriatane)

Recalcitrant Problems

- Consider poor compliance
- Re-evaluate for infection
  - Staph, Strep, Candidiasis, HSV
- Re-evaluate for irritant/allergic contact dermatitis
- Re-evaluate for wrong/additional diagnosis
- Re-evaluate for SCC/evolving SCC
VIN in patient with LP

Thank you!!

Questions?