Inflammatory Conditions of the Vulva

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Vulvar dermatoses

Genital inflammatory disorders can be diagnostically challenging

- Knowledge base
  Less studied than other pelvic structures
  Not integral part of gyn / derm / urology training

- Inherent difference between genital and non genital skin
  3 types of epithelium;
    keratinized skin
    mucocutaneous skin
    mucous membrane
  Moist environment

- Disorders unique to genital skin

Can have a HUGE impact on a woman’s quality of life
Categories of Inflammatory Disorders

- Conditions unique to genital skin
  - Zoon’s vulvitis

- Dermatoses with a predilection for genital skin
  - Erosive lichen planus
  - Lichen sclerosus

- Common skin disorders
  - Psoriasis
  - Atopic dermatitis
  - Lichen simplex chronicus
  - Contact dermatitis
Categories of Inflammatory Disorders

- Genital ulcers
  - Behcets disease
  - Complex aphthosis
  - Crohns disease
  - Auto immune bullous disease; pemphigus vulgaris
  - Lipshutz ulcer

- Vulvar inflammation secondary to vaginal infections
  - Bacterial vaginosis
  - Candida
Vulvar Anatomy

- Mons pubis
- Clitoris
- Labium majus
- Labium minus
- Vaginal introitus
Lichen Sclerosus (et Atrophicus)

Chronic destructive inflammatory skin condition with a predilection for genital skin.

One of the most common chronic vulvar conditions

Epidemiology

- Precise incidence unknown
- Favors women in a 10:1 ratio
- May begin at any age but tends to have a bimodal dist.
  - Pre pubertal girls
  - Postmenopausal women
- Estrogen may be protective
Etiology of Lichen Sclerosus

- Unknown, probably multifactorial
- Genetic predisposition
- Low estrogen
- Infection, possible link with Lyme disease
- Autoimmune

Patients with LS are at greater risk of developing:
  - Thyroid disease
  - Vitiligo
  - Alopecia areata
Symptoms of Lichen Sclerosus

- **Pruritus**: Intense
  - Scratching often worsens symptoms
  - Can disrupt sleep

- **Burning**

- **Fissures** – posterior fourchette and perianal area

- **Superficial dyspareunia**

- **Pain with defecation**

- **10-30% asymptomatic**
Distribution of Lichen Sclerosus
Clinical Features of Lichen Sclerosus

Acute / active disease
- Hypopigmentation
- Erythema
- “Tissue paper” wrinkling
- Purpura
- Fissures
Clinical Features of Lichen Sclerosus

Chronic changes

- Destruction of vulvar architecture
  - Resorption of labia minora
  - Fusion of clitoral hood
  - Burying of clitoris
  - Narrowing of introitus
Rationale for Treatment

- Provide symptomatic relief
- Halt / prevent architectural change
- Reduce risk of malignant change
  
  Increased risk of vulvar SCC (5% vs 2-3%)
Management of Lichen Sclerosus

- Biopsy – 4mm to confirm diagnosis
- If peri / post menopausal, consider topical estrogen
- Clobetasol 0.05% ointment
  - bid x 3 mths
  - qhs x 3 mths
  - 3x / week x 3 mths
  - 2x / week x 3 mths
  - 1x / week x 3 mths
  - Alternatives; Protopic ointment / Elidel cream
- Aquaphor or alternative emollient (olive oil)
- 3-6 monthly follow up
Erosive Lichen Planus

- Chronic destructive inflammatory condition that targets mucous membranes

Incidence
- Unknown, very rare
- Generally affects women over 40 years (range 29-82)
- Very rare in childhood

Etiology
- Unknown, probably multifactorial
- Possible link with Hepatitis C
- Possible aberrant cell mediated immune response to antigens in skin and mucosa
Symptoms of Erosive LP

- Extremely painful mucosal erosions – burning, raw
  Oral
  Vulvo-vaginal

- Intense pruritus – scratching worsens symptoms

- Dysuria

- Dyspareunia and tearing during intercourse
  SI - progressively more painful
  Overtime, may become impossible due to;
    Vulvo–vaginal adhesions
    Progressive vaginal stenosis

- Irritating vaginal discharge
Distribution of Erosive LP
Clinical Features of Erosive LP

- Vulvar mucosa
- Glassy erythematous erosions
- Violaceous border
- Friable mucosa / easy bleeding
- Destruction of normal vulvar architecture
  - Resorption of labia minora
  - Fusion of clitoral hood
  - Burying of clitoris
Clinical features of erosive LP

Vaginal mucosa

- Glassy erythematous erosions
- Friable vaginal epithelium / easy bleeding
- Desquamation
- Seropurulent exudate
- Adhesions of vaginal vault
- Progressive vaginal stenosis
- Obliteration of the vagina
Clinical features of erosive LP

Oral mucosa

- Gingival erythema or erosions
- Wickham’s striae
- Violaceous border
Rationale For Treatment

- Provide symptomatic relief
- Halt / prevent architectural change
- Reduce risk of malignant change

Relationship between erosive LP and SCC unclear but there does appear to be a slightly increased risk
Management of Erosive LP

- Patient education
  - Emphasis on importance of adhering to treatment regimen
  - Chronicity of disease
- General oral / vulvar / vaginal care
- Emotional support and reassurance
- Regular monitoring / follow up care
Management of Erosive LP

Vulva Care Measures

- Stop all irritants:
  - Wet wipes
  - Switch to gentle, fragrance free soap
  - Douches
- Cotton underwear
- Loose clothing
- Gentle emollient - olive oil, coconut oil, Vaseline
Management of Erosive LP

- Punch biopsy – 4mm
- If peri / post menopausal, topical estrogen
- Topical ultra-potent topical steroids -> vulva
  bid x 3 mths
  qhs x 3 mths
  3x / week x 3 mths
  2x / week x 3 mths
  1x / week x 3 mths
- Alternatives – Calcineuron inhibitor
- Topical steroid -> vagina
  Hydrocortisone suppositories
  Moderate to ultra-potent topical steroid qhs x 3 mths – taper as above
Management of Erosive LP

Oral mucosa

- Gentle oral hygiene
- Steroid gel / in orobase
- Calcineuron inhibitor
- Dexamethasone swish and spit
  +/− Doxycycline
  Nystatin
  Benadryl
- Refer to oral surgeon
Management of Recalcitrant Erosive LP

- Common problem in erosive LP
- Systemic options include
  - Hydroxychloroquine
  - Methotrexate
  - Soriatane
  - Cyclosporine
  - Mycophenalate
  - Prednisone
  - Biologics
Management of Vulvo-vaginal Adhesions

Prevention

- Erosive LP tends to flare with trauma
  - Koebner phenomenon

- Patient should avoid;
  - Traumatic activities
  - Surgical procedures
  - Vigorous SI
Management of Vulvo-vaginal Adhesions

- Adhesions can be gently massaged apart during application of topical steroid / emollient
- Vaginal dilators
- Surgical division of adhesions indicated if:
  - Narrowed introitus
  - Urinary compromise

For best outcome:
- Disease should be inactive / well controlled
- Excellent post op care

Limited role secondary to high risk of recurrence
Vulvitis Circumscripta Plasmacellularis

AKA Zoon’ vulvitus

Symptoms
- Asymptomatic
- Often associated with pruritus or burning

Clinical features
- Shiny glazed erythematous macules
  - +/- orange hue
- multiple tiny pin point macules
Vulvitis Circumscripta Plasmacellulatiss

Prognosis

- Self resolving
- No architectural change

Treatment

Only required if symptomatic

- Topical steroids
- Topical Tacrolimus / Pimecrolimus
- Topical estrogen
Psoriasis

- Chronic inflammatory condition commonly associated with thick well demarcated plaques on extensor surfaces.
- Minority of patients have genital/flexural/inverse psoriasis

Incidence
- Affects approximately 3% population

Etiology
- Multifactorial
- Genetic predisposition
Clinical Features of Psoriasis

- Pruritus, persistent discomfort – predominantly in the ‘hair bearing areas’ (lateral labia majora)

- Familiar thick silver scale of psoriasis is often absent due to flexural location

- Instead, it frequently appears as a beefy red plaque involving;
  - Mons pubis
  - Lateral labia majora
  - Perineum
  - Gluteal cleft – ‘gluteal pinkening’

- Vagina, labia minora and vestibule NOT involved
Distribution of Genital Psoriasis
Look for “clues” or signs of psoriasis elsewhere:

- Nail dystrophy; Pitting
  - Subungual hyperkeratosis
  - Onycholysis

- Thick well demarcated silver plaques on;
  - Extensor surfaces
  - Scalp
Management of Genital Psoriasis

- Biopsy if diagnosis in doubt (4mm punch biopsy)
- Patient education – very important
- Topical steroids – mainstay of treatment
  - 1% or 2.5% Hydrocortisone ointment
  - 0.1% Triamcinolone ointment
- Calcipotriene cream / ointment
- Alternatives
  - Pimecrolimus cream
  - Tacrolimus ointment
Eczema

- Often occurs on a background of atopy – family / personal
- Frequently just limited to the inferior vulva / perineum

Symptoms
- Intense itching, often worse in evening / nighttime
  - Frequently wakes patient
  - Scratching often provides temporary relief
- Fissures
- Dysuria
- Dyspareunia
Management of Eczema

- Biopsy if diagnosis in doubt
  - Generally not required
- Patient education
- Discourage scratching and rubbing
- Mid – high potency steroids
- Tacrolimus ointment / pimecrolimus cream
- Oral antihistamines at night
- Regular emollients
Lichen Simplex Chronicus

- Chronic inflammatory disorder
- Relatively common
- Sites affected
  - Labia majora - women
  - Scrotum – men

Pathogenesis

- Itch – scratch cycle
- Itch -> urge to scratch -> release of histamine -> causes further itch -> scratch
Clinical Features of LSC

- Associated with intense pruritus, often worse at night
- Scratching provides temporary relief
- Persistent rubbing and scratching causes;
  - Progressive lichenification
  - Exaggerated skin lines
  - Alopecia in affected areas
- Typically involves, mons pubis, labia majora, perineum
Management of LSC

- Biopsy if diagnosis in doubt
  - Generally not required
- Patient education
- Discourage scratching and rubbing
- Mid – high potency steroids
- Tacrolimus ointment / pimecrolimus cream
- Oral antihistamines at night
- Regular emollients
Vulvar contact dermatitis is quite common. It may be allergic or irritant in origin. Irritant > allergic.

<table>
<thead>
<tr>
<th>Irritants</th>
<th>Allergens</th>
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<tbody>
<tr>
<td>Soap*</td>
<td>Benzocaine (Vagisil)</td>
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<tr>
<td>Urine</td>
<td>Preservatives</td>
</tr>
<tr>
<td>Feces</td>
<td>Neomycin</td>
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<tr>
<td>Sweat</td>
<td>Latex condoms</td>
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<tr>
<td>Douches</td>
<td>Lanolin</td>
</tr>
<tr>
<td>Creams (alcohol)*</td>
<td>Perfume</td>
</tr>
<tr>
<td>Spermicides</td>
<td>Pantyliners*</td>
</tr>
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Management of Contact Dermatitis

- Thorough history to elicit possible culprit
- Avoidance of allergen / trigger
- Patch testing may be required
- Gentle skin care
- Emollient; Olive oil / Aquaphor / Vaseline / coconut oil
- Mild topical steroid
- Tacrolimus ointment / Pimecrolimus cream
Vulvar biopsy

- Apply Lidocaine cream 20-30 minutes ahead of time
- Infiltrate with Lido /Epi, preferably buffered
- 4mm punch biopsy
- 5-0 silk suture
Vulvar care measures

- Stop all irritants: Wet wipes, scented soaps, douches
- White cotton underwear
- Loose clothing
- Gentle emollient – olive oil, coconut oil, Vaseline,
- Hypoallergenic detergent
- Avoid fabric softener, dryer sheets
- Switch to gentle, fragrance free soap / no soap
- Silicon based lubricant
Take home points

- Important to be familiar with normal anatomical landmarks
- Be aware of the different types of epithelium
- Flexural moist environment
- Biopsy if diagnosis is in doubt
- It’s absolutely OK to use ultra potent topical steroids for severe vulvar dermatoses
- Patient education and support is invaluable
  Very important in setting of chronic disease / long term management