Inflammatory Conditions of the Vulva

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Vulvar Dermatoses

- Due to location, genital inflammatory disorders can be diagnostically challenging
  - Flexural moist environment
  - Inherent difference between genital and non-genital skin

- Disorders unique to genital skin
### Categories of Vulvar Disorders

- **Conditions unique to genital skin**
  - Zoon's vulvitis

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

- **Common disorders that less commonly affect genital skin**
  - Psoriasis
  - Atopic Dermatitis
  - Lichen Simplex Chronicus
  - Contact Dermatitis

### Categories of Vulvar Disorders

- Behcets disease

- **Auto immune bullous disorders**
  - Pemphigus Vulgaris

- **Vulvar inflammation secondary to vaginal infections**
  - Bacterial Vaginosis
  - Candidiasis
Vulvar Anatomy

Vulvar Microanatomy
Lichen Sclerosus (et Atrophicus)

Chronic destructive inflammatory skin condition with a predilection for genital skin

Epidemiology

- Precise incidence unknown
- Affects both men and women but favors women in a 10:1 ratio
- May begin at any age but tends to have a bimodal dist.
  - Prepubertal girls
  - Postmenopausal women
- Estrogen / progesterone 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

- Vulvar pruritus;
  - Intense
  - Scratching often worsens symptoms
  - Often disrupts sleep
  - Commonly misdiagnosed as candidiasis especially early on
  - Frequently leads to delay in diagnosis

- Burning

- Fissures – posterior fourchette and perianal area

- Superficial dyspareunia

- Encopresis / constipation - children

Distribution of Lichen Sclerosus
### Clinical Features of Lichen Sclerosus

#### Acute / active disease
- Hypopigmentation
- "Tissue paper" wrinkling
- Purpura
- Fissures and tears

#### 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, Aquaphor, 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
  Methotrexate
  Soriatane
  Cyclosporine
  Mycophenolate
  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

**Treatment**
- Adhesions can be gently massaged apart during application of topical steroid / emollient
- Vaginal dilators
- Surgical ligation of adhesions
  - Disease must be quiescent
  - Requires post op high dose systemic steroids to reduce risk of post–op flare
**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

**Management**
- Reassurance
- Keep area as dry as possible
- Mild topical steroid if symptomatic
- Alternatives; Calcineuron inhibitors

**Prognosis**
- Generally resolves after 2 – 5 years
Psoriasis

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

**Incidence**
- Approximately 5% patients with psoriasis

**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

Clinical Features of Psoriasis

- Look for clues or signs of psoriasis elsewhere
  - Well demarcated plaques;
    - Scalp
    - Extensor surfaces
  - Nail dystrophy;
    - Pitting
    - Onycholysis
    - Subungual debris
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
- Alternative – Calcineuron inhibitor

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
- Calcineuron inhibitor
- 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
- Itching -> scratching - >histamine release -> further itching -> more scratching
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
- Calcineuron inhibitor
- Oral antihistamines at night
- Regular emollients
Vulvar Contact Dermatitis

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>
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<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>
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<tr>
<td>Creams (alcohol)*</td>
<td>Perfume</td>
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<tr>
<td>Spermicides</td>
<td>Pantyliners*</td>
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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 calcineuron inhibitor
Support and Counseling

- 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