What’s Up, Down There?
A Review of Vulvar Dystrophies

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I have no conflicts of interest to declare.
Objectives

- Review and discuss the categories of vulvar dystrophies.
- Understand and recognize the clinical and microscopic features of Vulvar dystrophies necessary for adequate diagnosis.
- Recommend a therapeutic regimen for vulvar dystrophy for practical use.
Fact & Disclaimers

- Data suggests average of 6 months delay in accurate diagnosis of vulvar disease due to hesitation to biopsy

- Limited evidence based research on vulvar disorders
  - Especially regarding treatment & outcomes

- Nothing is FDA-approved for the treatment of specific disorders
  - Excluding infectious & atrophic
  - “Off-label uses”
Lichen: a fungus which grows symbiotically with algae, forming a crustlike, branching patches...
Vulvar Dermatoses
(old nomenclature- “dystrophy”)

Four Main Groups:
- Lichen Planus
- Lichen Simplex Chronicus (neurodermatitis)
- Lichen Sclerosis
- Mixed Dystrophy
# Key Points: the Lichens

<table>
<thead>
<tr>
<th>Sclerosis</th>
<th>Planus</th>
<th>Simplex (eczema)</th>
</tr>
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<tbody>
<tr>
<td>Itch or burn</td>
<td>Itch or burn</td>
<td>Itch, Itch, ITCH</td>
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<tr>
<td>Scars</td>
<td>Scars</td>
<td>No scarring</td>
</tr>
<tr>
<td>Not in Vagina</td>
<td>Vagina and Mouth</td>
<td>Not in Vagina</td>
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Common Characteristics

- Vulvar dermatoses share some common characteristics:
  - They are all benign, idiopathic, pruritic, & affect multiple body sites.
  - Although, they can occur in younger females, they are most commonly found in post-menopausal women.
  - Diagnosis is usually confirmed by vulvar biopsy, & although no cure is available, supportive steroid therapy is often successful in alleviating the symptoms.
Diagnosis

- Symptoms
- Clinical examination
- Biopsy-Biopsy-Biopsy!
  - Colposcopy with toluidine blue
Lichen Planus

- Lichen planus is an uncommon eruption of small papules that can involve the wrists, ankles, groin, genitalia, & oral mucosa.
- On the vulva the disease often manifests itself as annular erosive lesions with clear margins.
- The condition often lasts between 6 months to 2 years.
Lichen Planus

- Approximately 1% of population has oral
- In women w/oral disease, 20-25% have genital
- Named for cutaneous findings-reticulated white, flat-topped papules & plaques (Wickham’s striae)
- Erosive changes w/ less clearly defined reticulated, white & violaceous plaques on oral and vulvar skin

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Etiology of Lichen Planus

Unknown

? Autoimmune triggered by exogenous antigens, possibly cell-mediated
- viral
- bacterial (superantigen)
- chemical
- drug
- trauma
Clinical manifestations of Lichen Planus

SYMPTOMS
Most often there is irritation with burning & soreness
Can be very itchy, & scratching flares it
With scratching, the vulva gets very thick & scarred
Stretching of scarring causes dyspareunia
Symptoms depend on extent of disease - e.g. when vagina is involved with erosions, there is discharge, burning, etc.
Histopathology of Lichen Planus

- Hyperkeratosis w/o parakeratosis
- Beading of granular layer
- Epidermal atrophy
- Infiltrate (PMNs)
Lichen planus
Lichen Planus

- Skin and tongue
Treatment

- High to ultra-potent topical steroids
- Calcineurin inhibitors (pimecrolimus)
- Sitz baths (w/aluminum acetate solution)
- Antihistamines
- Oral Retinoids
- Cyclosporine
- Methotrexate
- Azathioprine (Imuran)
- Hydroxychloroquine (Plaquenil)

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Daily Treatment

- Comfort care/Daily skin protectant
- Steroid therapy (cream/ointment)
- Avoidance of irritants
- Phototherapy
- Immune modulator
  - 0.1% tacrolimus (Protopic) ointment – external
  - 0.1% pimecrolimus (Elidil) cream – vaginal
Desquamative Inflammatory Vaginitis

- Controversial (is it really erosive lichen planus?)
- Profuse purulent vaginal discharge & severe vulvovaginal inflammation
- Unknown etiology
- Treatment:
  - Local steroids +/- local antibiotics +/- local estrogen
  - Hydrocortisone acetate suppository (25mg)
  - Clindamycycin cream 2% or metronidazole gel 0.75%

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Lichen Simplex Chronicus

- Persistent, unrelenting, unexplained itching and rash characterize lichen simplex chronicus also referred to as neurodermatitis, eczema or hyperplastic dystrophy.

- The itching is usually intense, and the rash often involves the perineum of both sexes, the side of the neck in women, and the ankles in men.

- Diagnosis is clinical

- Occurs primarily in mid- to late-adult life
Lichen Simplex Chronicus

- Large patches of thickened, scaly, and occasionally hyper pigmented skin (lichenification) is commonly found.
  - Foci of atypical changes or cancer may lie within these areas.
- Symptoms often develop or worsen during stress, but once a lesion resolves, recurrence is uncommon.
- Sleep disruption due to itching/scratching
Etiology of Lichen Simplex Chronicus

Scratching and rubbing damage the skin so it loses its protective coating / barrier

Result is: susceptibility to infection, ease of irritation, more itching
Histology of Lichen Simplex Chronicus

- Hyperkeratosis
- Acanthosis
- Spongiosis
Lichen Simplex Chronicus
Treatment

- High to ultra potent steroid creams
  - Systemic vs. Topical
  - With or without antipruritic (scabicide)
    - Crotamiton (Eurax)
    - Pramoxine

- Avoidance of irritants (perfumes/dyes)

- Break “itch – scratch cycle”-
  - Tricyclic antidepressant (amitriptyline)
  - Antihistamine (hydroxyzine)

- Treat underlying conditions (yeast)

- Sitz baths (tepid soaks)

- Daily skin protection (zinc, vaseline, veggie oil)

- Vaginal dilators

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Reduce Inflammation

- Topical steroids
  - Nystatin + triamcinolone 0.1% ointment
    - Anti-fungal
  - Praxmoxine-hydrocortisone 1-2.5% ointment
    - Anti-pruritic
  - May require more potent corticosteroid for symptom control

Regimen--

- BID X 2 weeks
- HS X 2 weeks
- M-W-F @ HS X 2 weeks
- Follow up 6-8 weeks
Systemic Steroids

- Not a typical Medrol dose pack
- Oral prednisone: 40mg Q am X 5 days, then 20mg Q am X 10 days
- IM triamcinolone (Kenalog®): 80mg IM, may repeat in 2 months prn

- Key Point- eliminate the trigger
**LSC Important Points**

- Considerable overlap w/contact dermatitis, atopic dermatitis, & lichen simplex chronicus

- May exacerbate other underlying disorders (psoriasis, lichen sclerosis)

- Follow up and repeat evaluation are paramount for optimal outcome
Lichen Sclerosis

- Lichen sclerosis is a common disease of the vulva that can occur at any age, but is most common in post-menopausal women (mean age 50-60).
- The neck, trunk, and extremities can also be involved. (13%)
- The disease causes a gradual fusion & phimosis of the labia and prepuce of the clitoris.
- The thinned-out skin becomes pale and wrinkled like a parchment.

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Lichen Sclerosis

- Foci of atypical tissue or cancer should always be sought at the time of initial evaluation. These changes are symmetric and recurrence is common. “Waxes & Wanes”
- Pernicious anemia, achlorhydria, and autoimmune disorders have occasionally been associated with lichen sclerosis
- Vagina is NOT involved
Etiology Of Lichen Sclerosis

Unknown ?
Multifactorial:  - genetic
  - autoimmune
  - environmental factors

NOTE: Often associated with autoimmune conditions, e.g. thyroid disease, vitiligo, etc.
Familial cases have been reported
Prevalence up to 1 in 300
Commonest cause of chronic vulvar disease
Lichen Sclerosis

- Diagnosis is made by Biopsy
- Other concerns?
  - Autoimmune disorders
  - Progression to cancer - 2-5% risk
  - Loss of consort - apareunia
  - Bleeding
  - Surgical therapy?
Histology of Lichen Sclerosis

- Loss of rete ridges
- Dermis acellular/homogenous
- Chronic inflammatory cells
- +/- hyper or parakeratosis
Lichen Sclerosis
Lichen Sclerosis
Treatment

- High Potency Steroid ointment (clobetasol)
- Injectable triamcinolone (debilitating itching)
- Stop the “itch – scratch cycle”
- Treat underlying conditions (ie, yeast)
- Antidepressants
- Calcineurin inhibitors (pimecrolimus –Elidel®, or tacrolimus-Protopic®)
- Vaginal dilators
- Estrogen cream-use with some caution
- Testosterone or Progesterone cream: NO
Mixed Dystrophy

Occasionally patients present with mixed vulvar changes of lichen sclerosis and hyperplastic dystrophy:

Areas of thinned & thickened skin lie next to each other.

Multiple biopsies are important since mixed dystrophies have a slightly higher incidence of atypia.
What’s Up, Down There?

- Key Points
  - Vulvar dystrophies among the “Top Ten” reasons for seeking gynecologic care
  - Biopsy sooner rather than later
    - Don’t hesitate to rebiopsy if indicated
  - Education & Reassurance (time & money)
    - Establish realistic expectations
  - Underlying psycho-social concerns
    - Cancer, Sexual function, Monogamy, Normality
References

Kauppila S, et al. The effect of topical pimecrolimus on inflammatory infiltrate in vulvar lichen sclerosis AJOG Feb 2010


Stockdale C, Vulvar Disease, 2011

▪ Thank You!

▪ Questions?
University of Cincinnati Medical Campus