Grand Rounds:
Benign Disorders of the Vulva

Taylor Bertschy, PGY-3
Vulvar Symptoms

- Often chronic
- Pruritis & pain are most common sx
- May occur w/ obvious dermatologic disease or conditions, or w/ few visible skin changes
- Infection, inflammation, neoplasia, neurologic disorders (herpes neuralgia)
- Vulvodynia: discomfort & pain in the absence of visible findings or specific, clinically identifiable neurologic disorder
## Conditions Commonly Associated With Vulvar Pruritus

**Acute**
- Infections
  - Fungal, including candidiasis and tinea cruris
  - Trichomoniasis
  - Vulvovaginal candidiasis
  - Molluscum contagiosum
  - Infestations, including scabies and pediculosis
- Contact dermatitis (allergic or irritant)

**Chronic**
- Dermatoses
  - Atopic and contact dermatitis
  - Lichen sclerosus, lichen planus, lichen simplex chronicus
  - Psoriasis
  - Genital atrophy

**Neoplasia**
- Vulvar intraepithelial neoplasia, vulvar cancer
- Paget disease

**Infection**
- Human papillomavirus infection
- Vulvar manifestations of systemic disease
- Crohn disease
Vulvar Pruritus

- Most common sx
- Acute: infectious, allergic/irritant contact dermatitis
- Chronic (gradual onset): lichen simplex chronicus, lichen sclerosus, psoriasis, HPV-related disease
Dermatitis

- Poorly demarcated, erythematous, itchy rash
- Exogenous: irritant, allergic
- Endogenous: atopic (most common), seborrheic dermatitis
Contact Dermatitis

- Irritant: detergents, soaps, perfumes, semen, propylene glycol
- Allergic: immunologically mediated
- Often avoidable
- R/o candidiasis
- Biopsy is nonspecific and of little use
- Exam: erythema, swelling, scaling, fissures, erosions, ulcers
Allergic Dermatitis

- Type IV delayed hypersensitivity
- Intermittent sx and timing of onset
- Topical medications & anesthetics are common causes
Management

- Removal of offending agent
- Correction of barrier function
  - Sitz baths, estrogen, antimicrobials, plain petrolatum
- Elimination of scratching
  - Nocturnal: Antihistamines w sedative properties (doxepin, hydroxyzine)
  - Daytime: SSRI
- Reduction of inflammation
  - Mild-high potency corticosteroid x2-3wks
  - 1% hydrocortisone prn
  - IM for refractory cases
Vulvar Hygiene

- Avoid douching, allergens, & irritants
- Use of mild soaps, avoiding soap on the vulva
- Cleansing w/ water only
- Pericare bottles, pat dry
- Apply preservative-free emollient topically
- 100% cotton menstrual pads
- Adequate lubrication for intercourse
Lichen Simplex Chronicus

- Chronic eczematous disease
- Intense, unrelenting itching & scratching
- Middle-late adult life
- 65%-75% report h/o atopic disease
  - Localized variant of atopic dermatitis
- Erythematous, scaling, lichenified plaque
- Various degrees of excoriation
- Thickened, leathery skin
- Hyper or hypopigmentation
- Bx: hyperkeratosis, wide & deep rete ridges
Vulvar Dermatoses
Lichen Sclerosus

- Chronic d/o of skin most commonly seen on the vulva
- Onset 50-60yo
- Most likely genetic or autoimmune d/o
  - Highly associated w alopecia, vitiligo, thyrotoxicosis, hypothyroidism, & pernicious anemia
  - No eval for coexisting autoimmune d/o
- Increased risk of developing vulvar cancer
  - No evidence that optimal control reduces risk
Presentation

- Pruritus→irritation, burning, dysparunia, tearing
- Porcelain-white papules & plaques w/ areas of ecchymosis/purpura
- Thinned, whitened, crinkling
  - “Cigarette paper”
- Genital mucosa largely spared
Presentation

- Introital narrowing
- “figure of eight”, “hourglass” shape
- Fusion of labia minora, phimosis of clitoral hood, fissures
- Bx necessary
  - Exclude neoplasia, invasive SCC
Histology

- Epidermal atrophy
- Loss of rete ridges
- Hyperkeratosis
- Homogenization of dermal collagen
- Band-like inflammatory dermal infiltrate
Treatment

- High potency topical steroid: Clobetasol propionate
  - Once daily, & taper
  - Complications include skin changes, adrenal suppression
  - Maintenance vs recurrence
  - Monitor q3-6 months
- Retinoids
- Short-term 2nd line therapy: Topical calcineurin inhibitors
  - Avoids dermal atrophy
  - Possible link to skin cancer, lymphoma
- Surgery
Psoriasis

- 2-3% of population
- Women of all ages
- Rapid proliferation of epidermis
  - Genetic predisposition & immunologic factors
- Loss of vulvar architecture is uncommon
- Definitive dx w/ biopsy
- Local tx w/ mid-high strength topical corticosteroids
  - Calcipotriene (Vit D derivative) may be added
  - Refractory disease: MTX, retinoids, cyclosporine
Psoriasis

- Symmetric “salmon pink” raised plaques
- Silvery scales can be absent on vulva
- Bright erythema, well defined
- Often occurs on mons pubis
Lichen Planus

- Inflammatory, mucocutaneous d/o
  - May be drug induced

- Most common & most difficult to treat is the erosive form
  - Papulosquamos, hypertrophic

- Buccal mucosa involved
  - White, reticulate, lacy striae (Wickham striae)

Up to 70% have oral disease
Erosive Lichen Planus

- Deep, painful erosions in posterior vestibule
- Agglutination & resorption of labial architecture
- Vaginal d/c has inflammatory cells, immature parabasal and basal epithelial cells
  - pH 5-6

Vaginal involvement is common in erosive lichen planus & rare in lichen sclerosis
- 55% women have personal or family h/o autoimmune d/o
- HLA-DQB1*0201 allele present in 80% of women w/ vulvovaginal lichen planus
- Biopsy shows band-like infiltrate of lymphocytes
- Treatment is not curative
  - Corticosteroids (mainstay), cyclosporine, tacrolimus, hydroxychloroquine, PO retinoids, MTX, azithroprine, & cyclophosphamidine
  - Suspect infxn w/ symptom flare
- Vaginal dilators to maintain patency
Hidradenitis Suppurativa

- Chronic process that results from occlusion of follicles and secondary inflammation of apocrine glands
- Incidence 1:300
- Sx puritus, pain, erythema, burning, local hyperhydrosis
Presentation

- Cyst formation, induration, inflammation → spontaneous rupture → sinus tract formation
  - Hyperpigmentation, scaring, pitting, & fistulous sites
- Bx neither required, nor diagnostic
- Axilla, inguinal, & anogenital areas (apocrine glands)
- Onset after puberty; relapsing & recurrent
- Poor response to typical abx
Treatment

- Prevention: avoid irritants/heat/sweat, reduce friction, weight reduction
- Abx, antiandrogens, corticosteroids, retinoids vary in success
- Surgery is primary tx
- High recurrence rate
Bartholin Cysts

- Distal obstruction of Bartholin gland
- Unilateral, tender labial mass
- Lower 1/3 of introitus b/t vestibule and labia majora
- Transected by labia minora
Treatment

- Needle aspiration
- I & D w/ or w/o Word catheter placement
  - I & D alone not recommended
- Marsupialization
- Excision of gland
  - Avoided except in women >40yo w/ recurrent disease/mass due to morbidity
- Goal is epithelialized tract & continued drainage of gland
- Antibiotics
Procedures

Step 1: A diamond-shaped portion of introital mucosa over the dome of the Bartholin gland cyst is removed. Step 2: An incision is made through the wall of the Bartholin cyst. Step 3: The wall of the Bartholin cyst is sutured to the adjacent mucosa.

Vulvar disease, Bartholin gland cyst.
Bartholin Cysts

- Routine cx for aerobic & anaerobic bacteria
- Include *Neisseria gonorrhea* & *Chlamydia trachomatis*
- Broad-spectrum abx w/ cellulitis

Consider Bartholin gland carcinoma in postmenopausal women

Incision or marsupialization w/ biopsy
Vulvodynia

- “Burning pain occurring in the absence of relevant visible findings or a specific, clinically identifiable neurologic disorder.”
- International Society for the Study of Vulvovaginal Disease
- Generalized vs localized
- Provoked vs unprovoked
- Lifetime incidence approx 16%
Factors?

- Recurrent infections, OCPs, destructive tx
- Depression, anxiety and h/o sexual abuse are common w/ vulvar pain syndromes
- Higher incidence of IBS & interstitial...

Neuropathically mediated pain
Damage & loss of peripheral afferent neurons
Inflammation causes sensitization
Pelvic floor pathology
Hypertonicity→ guarding
Management

- No FDA-approved tx
- R/o dermatitis, dermatoses, infxn, neoplasia, inflammatory conditions, neurologic d/o
- Patient education in vulvar hygiene
- Referral to specialist, multidisciplinary approach
- Cotton swab test, neurosensory & muscle exam
- Fungal culture
Vulvodynia Assessment

Step 2 – Cotton-Swab Exam of the Vulva

Using a cotton swab, test for allodynia, hypo- and hyperalgesia by applying gentle pressure to the following areas:

1-2 Inner thigh
3-5 Labia majora
6-8 Interlabial sulcus
9 Clitoris | Clitoral hood
10 Perineum
11 Sites within vestibule (see next slide)

For each site, the patient:
- Rates the pain severity (VAS score)
- Describes the pain character (burning, raw, etc.)
Vulvodynia Assessment
Step 3 – Neurosensory Exam

- Instruct the patient on ‘cotton’ vs. ‘pin-prick’ sensation by touching her outer thigh (just above the knee) with the cotton portion, followed by the wooden portion, of the q-tip.
- Using the cotton-swab, gently stroke (upwards) each of the sensory dermatomes.
- Note sensation (normal, allodynia, hypo-sensitive), pain score, pain character (e.g., sharp, burning, shooting), and right-to-left-side differences.
- Break the q-tip in half and repeat exam with the sharp wooden portion of the q-tip using punctate pressure (apply light pressure for 1 sec) in each dermatome.
- Note sensation (normal, allodynia, hypo-sensitive), pain score, pain character (e.g., sharp, burning, shooting), and right-to-left-side differences.

From ongoing NIH Grant 5K23HD053631
Management

- Amitriptyline
- Gabapentin
- Behavioral therapy
- Vulvar hygiene
- Physical therapy
- Vaginal dilation
- Electrical stimulation
- Nitroglycerin
- Lidocaine
- Capsaicin
- Interferon
- Steroids
- Botulinum toxin
- Acupuncture

- Vestibulectomy has highest reported clinical cure rates
- Only indicated in cases of localized pain
Physical examination
Cutaneous or mucosal surface disease present

No

Cotton swab test

Tender, or patient describes area touched as area of burning

Yeast culture

Positive

Antifungal therapy

Inadequate relief

Inadequate relief and pain localized to vestibule

Surgery (Vestibulectomy)

Not tender, no area of vulva touched described as area of burning

Alternative diagnosis (incorrect belief that vulvodynia is present)

Inadequate relief and pain generalized

High-dose and multiple medications for neuropathic pain; consider referral to pain specialist; consider neuromodulation

Yes

Treat abnormal visible condition present (infections, dermatoses, premalignant or malignant conditions)

Positive

Antifungal therapy

Inadequate relief

Inadequate relief and pain localized to vestibule

Surgery (Vestibulectomy)

Negative

Inadequate relief

Good relief

Adequate relief

No additional treatment; stop treatment when indicated

Treatment Options
1. Vulvar care measures
2. Topical medications
3. Oral medications
4. Injections
5. Biofeedback/physical therapy
6. Dietary modifications
7. Cognitive behavioral therapy
8. Sexual counseling
Things to keep in mind...

- Vulvar manifestations of generalized conditions:
  - Acanthosis nigricans
  - Crohn's disease
  - Behcet disease

Figure 1: Multiple fleshy ulcers seen involving labia majora, minora, and clitoris. A single “knife-cut” ulcer seen in the left clitoral-labial groove.
References

- Precis: Gynecology 4th Edition
- William's Gynecology 2nd Edition
- ACOG Practice Bulletin: Diagnosis and Management of Vulvar Skin Disorders
- ACOG Committee Opinion: Vulvodynia
“That's all Folks!”