Your Diagnosis Is?
Test Your Knowledge of Various Vulvovaginal Conditions

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Hope Haefner, MD was previously on an advisory board for Merck Co., Inc. Colleen K. Stockdale, MD, MS and Libby Edwards, MD have nothing to declare relevant to this presentation.
Learning Objectives

After this lecture you should be able to:

• Diagnose common and unusual vulvovaginal conditions
• Identify the clinical feature of various vulvovaginal conditions
• Discuss the various treatments available for a variety of vulvovaginal diseases
Please Interrupt for Questions
Additional Information Available:

University of Michigan Center for Vulvar Diseases (Google)

Then, click on Information on Vulvar Diseases

http://obgyn.med.umich.edu/patient-care/womens-health-library/vulvar-diseases
University of Michigan Center for Vulvar Diseases

There are many reasons for seeing a vulvar disease specialist at the University of Michigan Center for Vulvar Diseases, treating them is only part of the solution. Our multidisciplinary approach at ensures women receive total care, from cutting-edge treatment options to education and counseling to meet every individual's needs.

The Center for Vulvar Diseases was created in 1993 to better serve and treat women with diseases of the external genitalia. Our center is one of only a handful of clinics that specialize in treating these conditions. We focus on the multidisciplinary approach to help patients improve their health.

The team approach allows us to provide a higher intensity of care and expertise to women who have already demonstrated a resistant and chronic illness or an unusual vulvar condition.

Many women experience different forms of vulvar pain, including vulvodynia. Vulvodynia is pain on the lips of the vulva or upon intercourse with a normal appearing vulva. It is a burning, stinging irritation. Some patients are unable to accept sexual penetration due to muscle spasms and tenderness. Other conditions associated with vulvar pain include:

- Lichen sclerosus or lichen planus—chronic inflammatory skin disorders
- Vulvar intraepithelial neoplasia—a precancerous condition, often associated with a virus, the human papillomavirus (HPV)
- Hidradenitis suppurativa—a disease of the apocrine glands and vulva, with pus filled pockets of fluid
- Bartholin cysts—fluid filled cysts at the base of the entranceway
Gross and histologic images
Sample Question

Format

• Gross or histologic picture
• Question (Your diagnosis is?)
• Type in correct answer
Your Diagnosis Is?

A. Vitiligo
B. Atrophy
C. Lichen sclerosus
D. None of the above
Your diagnosis is........
CASE

• 42 year old woman presents with chronic itching, recalcitrant to antifungal therapy
• She has failed superpotent topical corticosteroids and nighttime sedation
• Wears scented panty liners
• Otherwise healthy
Is this?

- contact dermatitis
- lichen simplex chronicus
- syringomas
- subclinical yeast infection
Is this?

- contact dermatitis
- lichen simplex chronicus
- syringomas
- subclinical yeast infection
SYRINGOMAS

- Benign tumors of eccrine sweat glands
- Most often recognized on eyelids
- More common on women than men
- When occurring on the vulva, often present with recalcitrant pruritus
Syringomas - Cause

- Genetic
- ? Hormonal
- ? Diabetes
Syringomas – Treatment

- Destruction
- Excision
- Google (crazy stuff, don’t pay attention)
  - Aloe, lemon juice, apple cider vinegar, pineapple juice
  - Caster oil, almond oil, iodine
  - Avoid alcohol
  - Avoid salty food
  - Onion juice
  - Garlic
  - Wash more
36 y.o. presents with a recent complaint of ripping pain with intercourse, sometimes with spotting of bright red blood.
Vulvar Fissures

Two main varieties

• Posterior fourchette fissures which occur with intercourse
• Skin fold fissures which patients describe as “paper cuts”
List Conditions Associated with Fissures

A.

B.

C.

D.
Treatment

• Rule out candida, lichen sclerosus, atrophy, herpes, Crohn’s

• Reduce friction
  – Ample lubricants (water, silicone, oil based)
  – Position changes

• Treat atrophic vaginitis with local estrogen and dilator

• Treat vestibulodynia and vaginismus

• Surgical excision as last resort
Treat with Dilators
Thoughts on Drying Racks
Surgical Therapy for Fissures

• Close anterior to posterior
When you are in the Trenches....

- **Infection**
- **Trauma**
- **Dermatosis**
- **Malignancy**
- **Combination**
A 53-year-old registered nurse (RN) is referred for consultation for vulvar pain, irritation and non-healing lesions. She has seen her primary care (family practice), gynecologist, dermatologist, and most recently gyn oncologist (former resident – referred to me).

Started 2 years ago as single lesion. Spread (2 areas) 1 year ago – saw derm (biopsy = acute and chronic inflammation). Topicals = nothing. This year = 3 areas = gyn onc = me.
Had 50 year check-up including colonoscopy = all good (10 year clearance).
Routine mammo/cytology = all good (current)
No meds
Newly married!
Husband ok (no penile problems!)
ROS = NEGATIVE
EXAM
No oral lesions
Part 1
Would you repeat the biopsy(s)?

A. Yes
B. No
Part 2
What is your initial diagnosis?

A. Basal cell cancer
B. Squamous cell cancer
C. Crohn’s
D. Behcet’s
E. Lichen Planus
She had biopsies taken 1 year ago by derm; acute and chronic inflammation.
Given classical “knife cut” lesion at right peri clitoral aspect recommended colonoscopy first, repeat biopsy second (she agreed .... Second opinion helped).
Though perplexed by negative ROS and colonoscopy less than 3 years prior!
Histology
(noncaseating granulomas)

D Heller images
Part 3
What is your FINAL diagnosis?

A. Basal cell cancer
B. Squamous cell cancer
C. Crohn’s
D. Behcet’s
E. Lichen Planus
Local GI repeated colonoscopy and upper endoscopy - identified Crohn's disease (terminal ileum).
Vulvar Crohn’s Disease

• Rare (2% of women will have vulvovaginal manifestations of Crohn’s disease)
• Occurs prior to GI Crohn’s in 25%
• Average age at presentation is 34,
  • but has been reported in children
• Vulvar involvement can be contiguous
  or metastatic (most common)
• Findings:
  • Knife-like ulcers
  • Chronic vulvar edema, hypertrophy
  • Aphthous ulcers
Crohn’s

“Knife Cut” Ulcers
Management of Vulvar Crohn’s Disease

Control of bowel disease vital for anogenital disease control

Systemic treatment:
Metronidazole, sulfasalazine, mesalazine, prednisone, azathioprine, cyclosporine
TNF alpha inhibitors – thalidomide, infliximab, adalimumab

Topical treatment:
Topical and intralesional corticosteroids, calcineurin inhibitors (tacrolimus)
Surgery
34 year old AA woman presents to your office with a 6 year history of recurrent boils of the vulva and medial thighs. She brings cultures which have shown group B streptococcus, klebsiella, bacteroides, MRSA, and enterococcus at various times.
CASE

• She reports that she improves briefly with each course of antibiotics, but experiences prompt relapse
• Her mother, who lives with her, has a similar condition
• She is frightened because an infectious disease doctor thinks she may have HIV causing her to have infections, but she refuses testing
Is this?

- MRSA furunculosis
- Hidradenitis suppurativa
- Evolving polymicrobial infections in patient with undiagnosed HIV
- Job’s syndrome
Is this?

- MRSA furunculosis
- Hidradenitis suppurativa
- Evolving polymicrobial infections in patient with undiagnosed HIV
- Job’s syndrome
HIDRADENITIS SUPPURATIVA  
(inverse acne)

• Occurs in one or more areas of apocrine glands (in the milk line)
• Multiple outlet follicles, resulting in keratin obstruction of follicles/comedones
• Distention of follicles producing cysts
• Eventual leakage of keratin debris and resulting foreign body inflammation
HIDRADENITIS SUPPURATIVA differentiation from infection

- Chronic nature
- Presence of comedones
- Scarring, sinus tracts
- Location
- Poor response to antibiotics
- Negative or variable culture results
HIDRADENITIS SUPPURATIVA differentiation from infection

- More common and more severe in individual of African genetic background
- Obese patients
- Smokers
- Men
HIDRADENITIS SUPPURATIVA therapy

- Difficult in patients with severe disease
- Careful patient education regarding expectations
- Weight loss, including bariatric surgery
- Stop smoking
- Chronic anti-inflammatory antibiotics (doxycycline, minocycline, trimethoprim-sulfamethoxazole, clindamycin)
- Dairy avoidance?
HIDRADENITIS SUPPURATIVA

therapy

- Intrallesional triamcinolone 10 mg/cc, .1-.2 cc into early cyst (can teach patient to self administer)
- (perhaps hormonal therapy – OCP, spironolactone)
- Surgery – excision of small areas, or dramatic excision of large areas with grafting
- TNF alpha blockers - adalimumab (Humira ®) recently approved
HIDRADENITIS SUPPURATIVA

Management of HS is a real challenge

Short courses of antibiotics will get these patients out of your office, but will not improve the quality of their lives
A 86 year old woman complains of vulvar soreness and itching that has recurred in the last year.

CHF, recent stroke, DVT

She thinks she had a problem like this 10 years ago but has no records. Topical steroids had made no difference.

A biopsy is performed.
Your Diagnosis Is?

A. Lichen sclerosus
B. VIN differentiated
C. Lichen planus
D. Extramammary Paget’s
Recurrent Paget’s Disease
Which treatment do you recommend for her recurrent Paget’s?

A. Triamcinolone ointment
B. Laser therapy
C. 5% imiquimod cream
D. Wide local excision
5% imiquimod cream → 9 months
What is the rate of primary Paget’s disease of the vulva being associated with an underlying adenocarcinoma?

A. 1% to 25%
B. 26% to 50%
C. 51% to 75%
D. 76% to 100%
Surgery for Primary Paget’s
16 year old presents with dysuria

Recent flu-like symptoms with mild fever, malaise

Recent oral sex with new partner

PMH – healthy, no meds

What tests do you order?

A. Biopsy
B. HSV culture
C. HSV PCR
D. Bacterial culture
Causes of Vulvar Ulcers

**Worldwide** – STIs are the most common cause

- # 1 – HSV
- # 2 – Syphilis
- # 3 - Chancroid – Africa (US in MSM)

  Granuloma Inguinale - New Guinea – very rare North America

**Infectious ulcers** – mostly STIs
- HSV, syphilis, chancroid, HIV

**Non-infectious ulcers**
- Skin disorders
Herpes Simplex - Diagnosis

- **Viral culture**
  - overall sensitivity 50% (declines as lesion heal)
- **Polymerase chain reaction**
  - Enhanced sensitivity compared to viral culture
  - TaqMan PCR allows differentiation of HSV1/2
- **Direct fluorescent antibody**
- **Serology**
  - Type-specific testing allows differentiation HSV1/2
- **Tzanck smear**
Herpes Simplex Virus (HSV)

Epidemiology

- HSV I - 25-30%  or  HSV 2 - 70-75%

- Usually spread from contact with an asymptomatic partner

- Women are more susceptible

- Recurrence rate for HSV 2 89%;  HSV 1 45%

Shedding continues through life
Antiviral drugs do not stop shedding
HSV 1 vs. HSV 2

HSV-1
- Can be latent many years
- Cold sores
- Typically 1/3 of new genital cases
- 5% genital recurrences
- Does not protect against HSV-2

HSV-2
- Can be latent many years
- Genital ulcers
- Typically 2/3 of new genital cases
- 95% genital recurrences
- Protects against HSV-1

Courtesy of Paul Nyirjesy, MD, Drexel + Zane Brown, MD, University of Washington
Primary Episode HSV
2015 CDC STD Treatment Guideline

- Acyclovir 400 mg po 3 times a day
- Acyclovir 200 mg po 5 times a day
- Valacyclovir 1 g po 2 times a day
- Famciclovir 250 mg po 3 times a day
- Treat 7-10 days, longer if healing is incomplete
Erosions with various stages of healing
CASE

- 42 year old woman presents to her gynecologist for a yearly examination
- He sees irregular brown, and black macules and patches
Is this?

- Malignant melanoma
- High grade squamous intra-epithelial lesion (HSIL, HPV-associated VIN)
- Post-inflammatory hyperpigmentation
- Vulvar melanosis/lentiginosis
Is this?

- Malignant melanoma
- High grade squamous intra-epithelial lesion (HSIL, HPV-associated VIN)
- Post-inflammatory hyperpigmentation
- Vulvar melanosis/lentiginosis
VULVAR
MELANOSIS/LENTIGINOSIS

• Brown macules and patches, usually variegate color, usually multifocal, usually asymptomatic
• Generally on the modified mucous membranes of the skin
• Seen in association with lichen sclerosus or on otherwise normal skin
Indistinguishable from early melanoma, second most common vulvar malignancy

Photo thanks to Dr. Ron Jones
HSIL
HSIL and SCC
• So BIOPSY!!!
A 3 y.o. complains of a year and a half of burning pain (prepuce and clitoris). She is crying and this is significantly affecting her life.
Your Diagnosis Is?

A. Lichen sclerosus
B. Debris build up under prepuce
C. Yeast infection
D. Sexual abuse
14 year old presents with dysuria.
Felt ill past 2 days: mild fever, headache, malaise.
PMH – healthy
Not sexually active

What test do you order?
A. Biopsy
B. HSV culture
C. HSV PCR
D. Bacterial culture
Evaluation Vulvar Aphthae

Thorough history and physical – eye, oral, genital

*Rule out HSV  (further testing if indicated)

For Acute Prodrome if indicated
  CBC, diff
  Serology for HSV, HIV, EBV, syphilis, CMV, Mycoplasma pneumoniae
  Influenza – swab PCR
  For Strep -throat swab and antistreptolysin O titer

For recurrent ulcers – consider GI investigation
  for inflammatory bowel disease and celiac disease

Diagnosis of exclusion - etiology often not found

No Biopsy Needed typically
Follow-up 48 hours later

HSV, viral studies negative

Initial Tx – comfort care, acyclovir, +/- prednisone
Vulvar Aphthous Ulcers

“Canker Sores” on the Vulva

Can be acute, chronic +/- recurrent - more commonly acute

- Average age is 14 (9-19) yrs
- Sudden onset
- Usually multiple, painful, well demarcated punched-out ulcers
- Size: most <1cm; can be 1-3 cm
- Prodrome - flu-like with mild fever, headache, malaise
- Duration 1-3 weeks, can last months
- One episode, less common recurrent
- Past history of oral aphthae – canker sores
- Rarely Behcet’s in North America
Vulvar Aphthae – Management

- **Pain control** – topical, systemic
- **Prednisone** - 40 – 60 mg each morning until pain resolves
  (3-5 days, then ½ dose 3-5 days)
  - ultrapotent corticosteroid
- **Educate** - Most often a one-time event, can recur
  - If not controlled – recurrent:
    - Intraleisional triamcinolone 5-10 mg/ml
    - doxycycline 50-100 mg daily
    - colchicine 0.6 mg bid-tid if tolerated
    - dapsone 50-150 mg per day
    - pentoxyfylline
    - cyclosporine
    - thalidomide
19 year old with complex aphthous ulcer (past history of oral aphthae)
Mt Etna
CASE

• 55 year old woman presents to your office with an 8 month history of red, weeping plaques in the groin and under the breasts

• Has been treated multiple times with topical antifungals with slight improvement

• Skin, hair and nails normal except for actinic damage
CASE

• KOH positive for Candida albicans
• Routine culture shows only yeast, no pathogenic bacteria
• Treated with fluconazole 200 mg daily, soaks, and barrier creams
• Minimal improvement
Is this?

- Resistant Candidiasis
- Intertrigo dermatitis
- Inverse psoriasis
- Contact dermatitis to medications
Is this?

- Resistant Candidiasis
- Intertrigo dermatitis
- Inverse psoriasis
- Contact dermatitis to medications
INVERSE PSORIASIS

• Often unassociated with other signs of psoriasis
• Often poorly demarcated
• Often colonized or secondarily infected with yeast
• Often biopsy is nonspecific
• Trial of topical corticosteroids
Biopsy

• Psoriasiform dermatitis (don’t expect diagnostic biopsy)
• Therapy - you already know. And if treatment for LSC/LS doesn’t work, refer to dermatologist for systemic therapy
81 y.o. G2P2 presents with vulvar itching and irritation. She had previously undergone a biopsy of the lesion, revealing verruca.

She says since the biopsy, the lesion has increased in size significantly.
Past Medical History

- Atypical chest pain (limited to approximately 1/2 block of ambulation and 1 flight of stairs secondary to shortness of breath)
- Vertigo
- Leg cramps
- Osteoarthritis
- Lichen sclerosus
- Bilateral cataracts
- Depression
- Osteoporosis
- Basal cell carcinoma (face)
- Spinal stenosis
- Peptic ulcer disease
- Hypertension
Biopsy 1 month prior to seeing you is verruca. What do you recommend as her treatment?

A. Rub a potato on the wart and throw it over the fence
B. Cryotherapy
C. Trichloroacetic acid
D. Wide local excision
What is the Diagnosis?

A. HSIL of the vulva
B. Invasive squamous cell carcinoma
C. Verrucous carcinoma
D. Adenocarcinoma
Case

• 24 year old with complaints of vulvar pain and numbness x 3 months.
• Generalized / constant
  Waxes and wanes
• Unable to identify trigger
• No other concerns
• Partner not affected
• 24 year old with complaints of vulvar pain and numbness x 3 months.

• What is the cause?
A) vestibulodynia
B) alopecia
C) tight pants
What nerve is compressed by her tight pants?

A. Ilioinguinal
B. Obturator
C. Perineal
D. Pudendal
• The genitofemoral nerve innervates the skin of the mons pubis and labium majus in females via the genital branch, and the anteromedial skin of the thigh via the femoral branch.

• The ilioinguinal nerve supplies sensory fibers to transversus abdominis and internal oblique. It innervates the medial skin of the thigh and the skin covering the mons pubis and the adjoining labium majus in females.
## Insights into Treatment

<table>
<thead>
<tr>
<th>Lifestyle Recommendation</th>
<th>Yes</th>
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<tbody>
<tr>
<td>Wear loose-fitting clothing</td>
<td>75%</td>
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<tr>
<td>Use unscented detergents and pads, wear cotton underwear; stop using feminine hygiene products</td>
<td>68-74%</td>
</tr>
<tr>
<td>Avoid douching</td>
<td>66%</td>
</tr>
<tr>
<td>Follow low-oxalate diet</td>
<td>28%</td>
</tr>
<tr>
<td>Discontinue use of tampons</td>
<td>22%</td>
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</tbody>
</table>

*Updike and Wiesenfeld, AJOG 2005*
Behavioral Modification

• Review behaviors that can be helpful, or may be harmful:
  – Wearing non-constrictive underwear
  – Avoid pantyhose, tights, and tight pants
  – Use mild soap for bathing and water only for cleansing the vulva
  – Avoid commercial vaginal wipes, deodorants, bubble bath, scented pads / tampons
CASE

• 68 year old woman presents with a history of lichen planus and a history of a vulvectomy 8 years ago for SCC
• She says that her lichen planus is becoming more poorly controlled despite daily clobetasol
• Irritation and itching is increasing, and testosterone produced intolerable burning
Is this?

- differentiated-VIN
- Lichen planus
- Zoon’s vulvitis (plasma cell vulvitis)
- Can’t tell
Is this?

- differentiated-VIN
- Lichen planus
- Zoon’s vulvitis (plasma cell vulvitis)
- CAN’T TELL
Biopsy shows differentiated VIN

- Full thickness dysplasia (intraepithelial neoplasia II-III)
- Histology shows well differentiated squamous dysplasia
- Can invade and metastasize quickly
- Occurs in setting of LS or LP, not HPV
- Generally older women
Woman with LS, d-VIN (white morphology)
D-VIN in patient with LS
Lichen sclerosus and lichen planus confer increased risk for SCC

- Preceded by differentiated VIN
- Most often hyperkeratotic white
- Most often in older women
- Most often in long standing disease
- But can be red, thin
- Biopsy anything chronic you cannot explain
D-VIN associated with LS/LP

- Biopsy anything that you can’t explain in women with LS or LP
- Punch
- Histology – well differentiated VIN (differentiated sounds good, but this is the histology associated with rapid invasion and metastasis)
A 64 y.o. G4P4 was recently diagnosed with lichen sclerosus (no biopsy performed). She was started on clobetasol propionate. She calls complaining of vulvar pain.
Your Diagnosis Is?

A. Lichen planus
B. Pemphigoid
C. Lichen sclerosus with herpes
D. Invasive squamous cell carcinoma
How many different types of herpes viruses affect humans?

A. 2
B. 4
C. 8
D. 80
### Herpesviruses

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<tr>
<th>Type</th>
<th>Name</th>
<th>Subfamily</th>
<th>Target cell</th>
<th>Latency</th>
<th>Transmission</th>
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<td>1,2</td>
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<td>Alphaherpesvirinae</td>
<td>mucoepithelia</td>
<td>neuron</td>
<td>contact</td>
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<td>3</td>
<td>VZV</td>
<td>Alphaherpesvirinae</td>
<td>mucoepithelia</td>
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<td>contact or respiratory</td>
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<td>CMV</td>
<td>Betaherpesvirinae</td>
<td>epithelia, monocytes, lymphocytes</td>
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<td></td>
<td></td>
<td></td>
<td></td>
<td>lymphocytes</td>
<td>congenital transplantation</td>
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<td>5</td>
<td>EBV</td>
<td>Gammaherpesvir.</td>
<td>B lymphocyte</td>
<td>B lymphocyte</td>
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<td>T lymphocyte</td>
<td>T lymphocyte</td>
<td>Respiratory</td>
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<tr>
<td>8</td>
<td>KSHV</td>
<td>Gammaherpesvir.</td>
<td>Endothelial cells</td>
<td>Unknown</td>
<td>body fluids</td>
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</table>

What percent of people with HSV-2 are unaware that they are infected?

A. 10-20%
B. 21 – 40%
C. 50- 70%
D. Over 80%
POP QUIZ 1

- 19 year old
- STI visit – new partner
- No current symptoms
- History of chlamydia treated 6 months prior; molluscum treated 6 months prior
What are the lesions?
A. Lipomas
B. Condylomas
C. Recurrent Molluscum
D. Scaring from TCA
E. Striae
Molluscum contagiosum

- **Poxvirus**
  - Only known host is humans
  - Causes a chronic localized infection
  - Flesh-colored, dome-shaped papules with central umbilication (2-5 mm diameter usual)
  - Spare palms and soles

- **Transmission**
  - Skin-to-skin contact, including autoinoculation
  - Also fomites, swimming pool
  - Incubation 1 week to 6 months (2-6 weeks usual)
Molluscum contagiosum

• Diagnosis
  – Characteristic appearance of lesions containing white to yellow amorphous material
  – H&E staining reveals keratinocytes containing eosinophilic cytoplasmic inclusion bodies

• Treatment
  – Self-limited, typically resolve within 6-12 months in immunocompetent patients
  – Cryotherapy
  – Curettage
  – Cantharidin
  – Podophyllotoxin
Molluscum contagiosum
A mimic....

- 67 year old female referred for recurrent yeast vulvovaginitis
- Incomplete response to topical and oral antifungals
- Pruritus affecting activities / “all consuming”
Red plaques with discrete boarders and satellite lesions
Phimosis of the clitoral hood and involution of the labia minora; thin, shiny vestibular tissue
Called for Dx opinion?

A. Wet prep
B. Culture
C. Biopsy
D. Further exam
Diffuse erythematous patches with scale on feet and lower extremities = diagnosis
What is your diagnosis?

A. Chronic yeast
B. Paget’s
C. Psoriasis
D. Contact dermatitis
Psoriasis

• Common hereditary, scaly rash
  – Vulvar = flexural psoriasis or psoriasis inverses

• Silvery white adherent scale on red plaques
  – In groin = well demarcated, moist, thin red patches
  – Often missed or hidden = look elsewhere for typical skin lesions
Psoriasis treatment

• Stop irritants
• Topical steroid ointment
• Topical tacrolimus 0.1% ointment or pimecrolimus 1% cream
  – Consider systemic medications if severe = consult dermatology
Summary

When patients do not respond to therapy

- Reconsider the diagnosis
- Check for infection - fungal, bacterial, HSV
- Consider contact dermatitis to a medication, over washing, etc.
- Evaluate for carcinoma
Thank you!!