Your Diagnosis Is? Test Your Knowledge of Various Vulvovaginal Conditions

Libby Edwards, MD
Hope K. Haefner, MD
Colleen Stockdale, MD

Disclosures

• Libby Edwards, MD has nothing to disclose
• Hope Haefner, MD has nothing to disclose
• Colleen Stockdale, MD has nothing to disclose
Course Objectives

At the end of this lecture, the participant should be able to:

1. Identify the clinical features of various vulvovaginal conditions
2. Become familiar with a variety of treatments for skin diseases

Gross and Histologic Images
Test Format

The image shown represents which vulvar condition?
Test Format

The image shown represents which vulvar condition?

A. Vulvar intraepithelial neoplasia
B. Melanoma
C. Molluscum contagiosum
D. None of the above
Case Presentation

83 y.o. woman with vulvar pain
- Symptoms worsened by urination and tight clothing as well as sitting for long periods of time
- No dysuria or trouble urinating
- Firmness to palpation
Your Diagnosis Is?

A. Urethral diverticulum
B. Urethral prolapse
C. Urethral polyp
D. Urethral cancer

Urethral Diverticulum
Urethral Myoma
Urethral Cancer

Urethral cancer is more common in women than men

A. True
B. False
Urethral cancer is more common in African Americans than in Caucasians

A. True
B. False

The most common histologic type of urethral cancer is:

A. Transitional carcinoma
B. Squamous cell carcinoma
C. Adenocarcinoma
D. Melanoma
The usual initial treatment for urethral cancer is:

A. Radiation therapy
B. Surgery
C. Chemotherapy
D. Laser ablation

“Itching and terrible pain” CKS
• 85 year old with biopsy confirmed lichen sclerosus re-referred for vulvar flare refractory to treatment
• History – dementia, lives in assisted living
• Medications - clobetasol topically prn, Petrolatum 55%, Resorcinol 2% (Resinol) topically prn, fluconazole 200 mg q 3 days x 3 doses
• Social – multiple contact irritants, shared care

Exam

• Marked erythema and edema of labia, phimosis of clitoral hood and involution of labia minora (c/w LS), vestibular excoriations, + rectocele noted
• Microscopy: pH 5.0, negative whiff, 10% mature squamous cells, sparse flora/background; no yeast/clue cells/trichomonas
• Yeast culture - negative
• What is your diagnosis?

A. Contact dermatitis
B. Lichen sclerosus
C. Lichen planus
D. Vulvar cancer
• How will you manage this patient?

A. Biopsy confirmation necessary
B. Stop all contact irritants
C. Refer to Gyn Oncology
D. Topical steroids

Return 1

• Increased level of care at assisted living
• +/- compliance
• Especially with 1 nurse, shared laundry, private – prefers self care
• Using topical steroid (+/-)
• Using other topicals for comfort - Resinol, prior prescriptions, OTCs
• How will you NOW manage this patient?

A. Biopsy confirmation necessary
B. Oral steroids
C. Oral fluconazole
D. Admit to inpatient service
• Results:

A. Biopsy = features of lichen planus
B. Oral steroids = ? Better compliance
C. Oral fluconazole = still no yeast
D. Admit to inpatient service = no

Return 2+

• Better initially!
• But, pain/itching returned with steroid taper
• Now – “worse than ever”
• Noncompliant with topical steroid/assistance
• How will you NOW manage this patient?

A. Biopsy confirmation necessary
B. Oral steroids
C. Oral metronidazole
D. Oral fluconazole
• RESULTS
  – Biopsy confirmation necessary – contact dermititis
  – Oral steroids – prolonged
  – Oral metronidazole – as anti-inflammatory
  – Oral fluconazole – now has c. albicans
  – +/- topical steroid

  – Lysol wipes in bathroom (rechecked for irritants with return of symptoms)

• Better
• Normal activities
• No problem with meds
• Not using topicals....
• Complex management – dementia + assisted living

• Biopsy – rebiopsy (what am I missing!)
• Curbside consults – support!
• Next steps!

Your Diagnosis Is?

Libby Edwards, MD
LibbyEdwardsMD.com
DISCLOSURES
None

DISCLAIMER
Most medications discussed in this lecture are not FDA approved for these diseases. For example, there are NO topical corticosteroids formulated for the vagina. These are common diseases, with very little adequate research

WEBSITE for handouts, etc
WWW.libbyedwardsmd.com

CASE
• 62 year old woman with a 3 year history of burning, rawness and severe dyspareunia due to lichen sclerosus
• Biopsy confirmation 3 years ago
• She has been treated with daily clobetasol ointment for several months, then tacrolimus daily, and, most recently, fractional CO2 laser (MLT)
CASE

• She wants to know about your expertise with PRP/platelet rich plasma infusions.
• Other medical problems
  – hypothyroidism
  – hypertension
  – hypercholesterolemia
  – irritable bowel syndrome
  – fibromyalgia
You tell her?

- PRP is a reasonable therapy and discuss this in detail
- PRP is a poorly reported therapy and suggest more aggressive traditional concomitant alternative superpotent topical corticosteroid and tacrolimus
- Her discomfort may not be due to LS
- That she would probably respond to intralesional corticosteroids
Other possibilities include all except?

- Squamous cell carcinoma
- Atrophic vagina
- Vulvodynia
- Subtle lichen sclerosus
All now are reasonable steps except?

- Wet mount
- Reassurance with one year follow-up and discontinuation of her clobetasol
- Pelvic floor evaluation
- Evaluation of area of pain

All are reasonable steps except?

- Wet mount
- Reassurance with one year follow-up and discontinuation of her clobetasol
- Pelvic floor evaluation
- Evaluation of area of pain
Parabasal cells, no lactobacilli = atrophic vagina and pH - 7

PELVIC FLOOR

- Tender to palpation
- Poor contractions
- Poor endurance
Area of pain = vestibule

Your diagnosis is?

- Undertreated LS
- Atrophic vagina
- Vulvodynia
- Vulvodynia with factors of LS and atrophic vagina
Your diagnosis is?

- Undertreated LS
- Atrophic vagina
- Vulvodynia
- Vulvodynia with factors of LS and atrophic vagina

LESSON

- Be willing to let go of a diagnosis
- Many chronic symptoms are multifactorial in origin
22-year-old G0 with severe, generalized, erosive skin and mucosal eruption

She is diagnosed with Steven’s Johnson syndrome

Your service is consulted for management recommendations
Your management includes all of the following except:

A. Daily vaginal dilation
B. Intravaginal steroids
C. Intravaginal antibiotics
D. Menstrual suppression to prevent adenosis

17-year-old G0 with severe, generalized, erosive skin and mucosal eruption

- She had just started treatment for recent onset of seizures (phenytoin)
- Rapid onset of a diffuse maculopapular purple rash
- In 24 hours she had severe malaise, fever and blistering on skin (40% of BSA) and genital area with dysuria
Your Diagnosis Is?

A. Staph scalded skin syndrome
B. Steven’s Johnson syndrome
C. Graft vs. host disease
D. Toxic epidermal necrolysis
Toxic Epidermal Necrolysis (TEN)

• A serious, life-threatening allergic skin rash

Clinical:

Severe pain
Diffuse sloughing of skin and mucous membranes
Severe conjunctivitis, stomatitis, esophagitis, bronchitis, vulvovaginitis

Definitions

• SJS is the less severe condition- <10 percent of the body surface
  – Mucous membranes are affected in over 90 percent of patients, usually at two or more distinct sites (ocular, oral, and genital)

• TEN involves detachment of >30 percent of the body surface area
  – Mucous membranes are also involved in over 90 percent of patients.

• SJS/TEN overlap describes patients with skin detachment of 10 to 30 percent of body surface area
Etiologies

Drugs (limited to first 8 weeks)
- Allopurinol
- Aromatic anticonvulsants (lamotrigine)
- Antibacterial sulfonamides
- Nevirapine
- Oxicam nonsteroidal anti-inflammatory drugs (NSAIDs)

In children, antimicrobials- sulfonamide anticonvulsants- phenobarbital, carbamazepine and lamotrigine
Etiologies

Infection

*Mycoplasma pneumoniae* infection is the next most common trigger

Other

Rarely reported and debatable causes of SJS/TEN include vaccinations, systemic diseases, contrast medium, external chemical exposure, herbal medicines, and foods.

Differential Diagnosis SJS/TEN

- Erythema multiforme
- Erythroderma and erythematous drug eruptions
- Acute generalized exanthematous pustulosis
- Generalized bullous fixed drug eruption
- Phototoxic eruptions
- Staphylococcal scalded skin syndrome
- Paraneoplastic pemphigus
- Linear IgA
Mortality with TEN is:

A. 10%
B. 20%
C. 30%
D. 40%
Your management includes all of the following except:

A. Treatment in a burn unit
B. Sedation
C. IVIG
D. Pulse IV steroids

Summary
Stevens-Johnson Syndrome or Toxic Epidermal Necrolysis

• Examine the vulva and vagina (narrow speculum)
• If vagina involved, or disease close to vagina, start on daily vaginal dilation
  – The dilator does not need to be left in for an extended period of time with each dilation
Summary, continued

• If the vulva is involved, start on clobetasol 0.05% ointment qhs
• If the vagina is involved, start patient on 50 mg hydrocortisone (ii 25 mg suppositories qhs (insert after dilation done)

Summary, continued

• The patient should continue daily vaginal dilation for 2 months (or longer if the vagina is not healed by 2 months)
• Arrange follow up 2 weeks after patient is discharged
Case CKS

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  - Lynne Margesson
**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)

- **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
CASE

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

1. d-VIN
2. Recalcitrant, erosive LS
3. Zoon’s vulvitis (plasma cell vulvitis)
4. HSIL
5. Can’t tell

Is this?

1. d-VIN
2. Recalcitrant, erosive LS
3. Zoon’s vulvitis (plasma cell vulvitis)
4. HSIL
5. Can’t tell
Biopsy shows differentiated vulvar intraepithelial neoplasia

- D-VIN is full thickness intraepithelial neoplasia in a setting of LS or LP,
- Very high risk for invasion and metastasis, unlike HSIL on vulva

Differentiated VIN

- Full thickness dysplasia (previously intraepithelial neoplasia II-III)
- In a setting of lichen sclerosus or lichen planus
- Older women
- Histology shows well differentiated squamous dysplasia
- Invades and metastasizes quickly
Woman with LS, d-VIN (white morphology)

D-VIN in patient with LS
HSIL (high grade squamous intraepithelial lesion)
- Full thickness dysplasia (formerly intraepithelial neoplasia II-III)
- Produced by HPV
- Often younger women
- Usually multifocal
- Indolent, often does not invade

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)
HSIL
morphology

• Resemble genital warts
• Most often
  - flat, pigmented, or
  - very large
Patient with HIV (0 viral load) and HSIL (eventually developed SCC on medial labium minus)
HSIL/(VIN associated with HPV)
How do you know when to biopsy a wart?

- Biopsy flat pigmented genital warts (but genital warts in patients of color are normally pigmented)
- Very large warts
- Any atypical morphology
- Immunosuppressed patients
76 y.o. hx of vulvar irritation and lichen sclerosus

- Added onto clinic for new onset of severe pain in her vulvar area
- Intense burning pain over her vulva and buttock
- Primary MD saw her 2 days previously-recommended that she use warm or cold compresses and topical lidocaine
Your Diagnosis Is?

A. Aphthous ulcers
B. Shingles
C. HSV 1 or 2
D. Erosive lichen planus

How many different types of herpes viruses exist that affect humans with disease?

A. 2
B. 4
C. 8
D. 80
Which type causes shingles?

A. HSV 1  
B. HSV 2  
C. HSV 3  
D. HSV 8

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**Herpesviruses**

<table>
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<tr>
<th>Type Name</th>
<th>Subfamily</th>
<th>Target cell</th>
<th>Latency</th>
<th>Transmission</th>
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<td>Alphaherpesvirinae</td>
<td>mucoepithelia</td>
<td>neuron</td>
<td>contact</td>
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<tr>
<td>3 VZV</td>
<td>Alphaherpesvirinae</td>
<td>mucoepithelia</td>
<td>neuron</td>
<td>contact or respiratory</td>
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<tr>
<td>4 CMV</td>
<td>Betaherpesvirinae</td>
<td>epithelia, monocytes lymphocytes</td>
<td>monocytes</td>
<td>contact</td>
</tr>
<tr>
<td>5 EBV</td>
<td>Gammaherpesvir.</td>
<td>B lymphocyte</td>
<td>B lymphocyte</td>
<td>saliva</td>
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<tr>
<td>6,7 HLV</td>
<td>Betaherpesvirinae</td>
<td>T lymphocyte</td>
<td>T lymphocyte</td>
<td>Respiratory</td>
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<tr>
<td>8 KSHV</td>
<td>Gammaherpesvir.</td>
<td>Endothelial cells</td>
<td>Unknown</td>
<td>body fluids</td>
</tr>
</tbody>
</table>

http://en.wikipedia.org/wiki/Herpesviridae
How many people in the US develop shingles?

A. 1 out of every 3 people
B. 1 out of every 4 people
C. 1 out of every 5 people
D. 1 out of every 6 people

• Reduce the risk of developing shingles and the long-term pain from post-herpetic neuralgia (PHN) by getting vaccinated (age 60 years)
• Antiviral medicines—acyclovir, valacyclovir, and famciclovir to shorten the length and severity of the illness
• Analgesics (narcotics, gabapentin), wet compresses, calamine lotion, and colloidal oatmeal baths
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. Wetprep
B. Culture
C. Biopsy
D. Further exam
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
Case

- 68 year old woman presents with a 2 year history of dyspareunia, burning and rawness
- Gyn records report KOHs showing yeast
- Poor response to antifungals
- Estrogen replaced without improvement
Your diagnosis is?

- Zoon’s (plasma cell) vulvitis
- Lichen planus
- Contact dermatitis
- Differentiated VIN
Plasma cell vulvitis
(Zoon’s vulvitis, vulvitis plasmacellularis)
Zoon’s vulvitis

- Like pornography, I can’t define it, but I know it when I see it
- It is diagnosed by its morphologic and histologic appearance
- Cause unknown

Zoon’s mucositis- Dx

- Morphology of red/brown, purpuric or deep red patches on mucous membranes (less often on modified mucous membranes)
- Usually burns, irritated
- Confirmed by biopsy showing plasma cells, dermal hemosiderin, effacement of the epidermis, lozenge-shaped epithelial cells
Zoon’s – Rx
The literature reports good results (not I)

- Ultrapotent topical corticosteroids (of course)
- Intralesional steroids
- Clobetasol, oxytetracycline, and nystatin compounded (personal communication Lynne Margesson)
- Fudisic acid (antibiotic cream)
- Topical retinoids (ouch!)
- Imiquimod (ouch!)
- Calcineurin inhibitors (often ouch!)
- CO2 laser (ouch!)

Symposia Medicus, Kauai, 2017
CASE

• 3 year old rubs at perianal skin and wakes at night crying with perianal pain
• Perianal skin is red, fissures
• Diagnosed with yeast and treated with clotrimazole cream
• Child cries because it burns with application, changed to nystatin oint
• No improvement in 5 days, treated with oral fluconazole
• Still no improvement, derm consult
Is this?

- Candidiasis
- Irritant contact dermatitis
- Lichen simplex chronicus
- Perianal bacterial disease

Is this?

- Candidiasis
- Irritant contact dermatitis
- Lichen simplex chronicus
- Perianal bacterial disease
Perianal Streptococcal Disease

- Culture yields beta hemolytic group A streptococcus
- Given oral antibiotic (amoxicillin, penicillin, beta lactamase resistant agent), mupirocin topically
- Much more comfortable in 2 days
- Recurrence common, usually in first 6 weeks
Can affect the vagina (child had purulent vaginitis), with or without rectal involvement

Perianal Streptococcal (now bacterial) Disease

- Often mistaken for yeast or LSC/eczema
- Diagnosis by morphology and history, confirmed by culture and response to therapy
- Therapy is by oral and topical antibiotics
- May affect the vagina, may have surrounding impetigo or folliculitis
- Increasingly associated with *S. aureus*
Adults can have this also; 58 year old internist self medicating for fungus infection. + culture for strep, immediate response to therapy

S aureus
Group A Strep

S aureus
LESSON

• Scaling red plaques are not all yeast and dermatitis
CASE

• 34 year old 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, 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, and refuses testing
Is this?

- Hidradenitis suppurativa
- Evolving polymicrobial infections in patient with undiagnosed HIV
- Job’s syndrome (hyper IgE synd)
- Gardner’s syndrome

Is this?

- Hidradenitis suppurativa
- Evolving polymicrobial infections in patient with undiagnosed HIV
- Job’s syndrome (hyper IgE synd)
- Gardner’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 therapy

- Weight loss
- Stop smoking
- Chronic anti-inflammatory antibiotics
  - takes about 3 months for benefit
  - doxycycline or minocycline 100 bid
  - clindamycin 150 bid with probiotics
  - trimethoprim sulfamethoxazole DS bid
HIDRADENITIS SUPPURATIVA

therapy

• Intralesional triamcinolone acetonide 10/cc, about .2 cc into new cyst
• (perhaps hormonal therapy – OCP, spironolactone)
• TNF alpha blockers; adalimumab (Humira) 40 mg SQ weekly
• Surgery
  • removal of individual cysts or en bloc
  • unroofing cysts and sinus tracts

LESSON

• HS is not infectious
• Short courses of antibiotics are not useful
• Surgery and adalimumab can save quality of life
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?

- Inverse psoriasis
- Intertrigo dermatitis
- Resistant Candida
- Contact dermatitis
Is this?

- **Inverse psoriasis**
- **Intertrigo dermatitis**
- **Resistant Candida**
- **Contact dermatitis**

**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
THERAPY OF PSORIASIS
(and lichen sclerosus, lichen planus and contact dermatitis)

- Patient education, handouts
- Eliminate irritants (including unnecessary topical agents, soap, panty liners – good luck with that)
- Stop nighttime scratching with sedation
- Ultrapotent corticosteroids (clobetasol) ointment bid and follow-up in a month – don’t stop it too soon
- Re-evaluate flare – infection, contact derm

THERAPY OF PSORIASIS
(and lichen sclerosus, lichen planus and contact dermatitis)

- Amitriptyline 10 mg, ½ to 3 tablets 2 hours before bed (10 hours before waking). Produces deep, deep sleep.
- Diphenhydramine (Benadryl®) or hydroxyzine (Atarax®) 25 mg, 1-3, an hour before bed. Produces light sleep.
- Antihistamines not useful for eczema/LSC
- Can stop when itching is controlled
THERAPY OF PSORIASIS
(and lichen sclerosus, lichen planus, and contact dermatitis)

• Patient education, handouts
• Eliminate irritants (including unnecessary topical agents, soap, panty liners – good luck with that)
• Stop nighttime scratching with sedation
• Ultrapotent corticosteroids (clobetasol, halobetasol) ointment bid and follow-up in a month – don’t stop it too soon
• Re-evaluate flares – infection, contact derm

THERAPY OF PSORIASIS
(and lichen sclerosus, lichen planus and contact dermatitis)

• Use ointment, not cream
• Start with superpotent (halobetasol, clobetasol) (Goodrx.com)
• Show with a mirror where to apply
• Demonstrate how much
• Reassure regarding safety, tell her to ignore information to stop it in two weeks
• But re-evaluate in one month
THERAPY OF PSORIASIS
(and lichen sclerosus, lichen planus and contact dermatitis)

• Patient education, handouts
• Eliminate irritants (including unnecessary topical agents, soap, panty liners – good luck with that)
• Stop nighttime scratching with sedation
• Ultrapotent corticosteroids (clobetasol) ointment bid and follow-up in a month – don’t stop it too soon
• When needed, systemic therapy (methotrexate, adalimumab, etanercept, etc)
• Re-evaluate flares – infection, contact derm

LESSON

• Psoriasis can mimic and co-exist with Candidiasis
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
LESSON

BIOPSY!!!

Thank you!!