North American Chapter of
The International Society for The Study of Vulvovaginal Disease

Your Diagnosis Is?

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Libby Edwards, MD

No conflicts of interest

Learning Objectives

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

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

https://medicine.umich.edu/dept/obgyn/patient-care-services/womens-health-library/center-vulvar-diseases/resources-providers

or search Google for

Resources for Providers University of Michigan

[Image of ISSVD logo]

North American Chapter of
The International Society for The Study of Vulvovaginal Disease

Enuresis
Family Planning
Fertility & Reproductive Health
Hysterectomy Alternatives
Minimally Invasive Gynecologic Surgery
Pediatric & Adolescent Gynecology
Pelvic Floor Disorders
Pregnancy & Childbirth
Gynecology

Center for Vulvar Diseases
Information Regarding Vulvar Diseases
Women with Disabilities Gynecology Clinic
Women's Health Program

Many women experience symptom scores or vulvar pain, including vulvodynia, vulvovaginal pain on the skin 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 papilloma virus (HPV)
- Herpesvirus vulvovaginalis—a disease of the anogenital skin
- Bartholin cysts—fluid-filled cysts at the base of the entranceway

During your first visit to the center, you will see a physician or nurse practitioner for diagnosis and development of a treatment plan. Throughout treatment, you will meet with experts from various disciplines to best meet your specific needs.

In addition to medical treatment, we also connect many patients to additional services, which may help with your condition, including sex therapy and physical therapy.

Health Library
- Vulvodynia
- Patient Education Booklet
- Resources for Providers

Contact Information:
North American Chapter of The International Society for The Study of Vulvovaginal Disease

Endometriosis
Family Planning
Fertility & Reproductive Health
Hysterectomy Alternatives
Minimally Invasive Gynecologic Surgery
Pediatric & Adolescent Gynecology
Pelvic Floor Disorders
Pregnancy & Childbirth
Gynecology

Center for Vulvar Diseases
Hair Removal Regarding Vulvar Diseases
Women With Disabilities Gynecology Clinic
Women’s Health Program

Many women experience certain forms of vulvar pain, including vulvodynia. Vulvodynia is pain on the skin 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—a chronic inflammatory skin disorder
- Vulvar intraepithelial neoplasia—a precancerous condition, often associated with a virus, the human papilloma virus (HPV)
- Herpes simplex—a disease of the anorectum and vulva, with pus filled pockets of fluid
- Bartholin cysts—fluid filled cysts at the base of the entranceway

During your first visit to the center, you will see a physician or nurse practitioner for diagnosis and development of a treatment plan. Throughout treatment, you will meet with experts from various disciplines to best meet your specific needs.

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Contact Information:

ISSVD Houston - March, 2017
- Your Diagnosis Is (PPT PDF)
- Your Diagnosis Is - Handout (PDF)

ACOG San Diego - May, 2017
- Your Diagnosis Is (PPT PDF)
- Vulvar Diseases What do you know? (PPT PDF)
- Your Diagnosis Is and Vulvar Diseases What do you know? - Handout (PDF)

Dallas, Texas - July, 2017
- Your Diagnosis Is (PPT PDF)
- Your Diagnosis Is - Handout (PDF)

Tijuana, Mexico - August, 2017
- Vaginal and Vulvar Colposcopy (PPT PDF)
- Vulvar Diagnosis and Treatment (PPT PDF)
- Vulvodynia Approach, Diagnosis, and Treatment (PPT PDF)
A 64 y.o. G5P3 a has mass on her interlabial sulcus. A biopsy is performed.
Your diagnosis is?

A. Hidradenitis suppurativa
B. Squamous cell carcinoma
C. Papillary hidradenoma
D. Adenocarcinoma of the vulva
38 y.o. G1P1 with a clitoral mass

• Increasing in size for the past 28 years
• Enlargement at clitoral location noted at her last delivery 3 years ago
• Past workup by an endocrinologist included an MRI of her adrenals/kidneys and a testosterone work up which were negative
S100 Confirmatory Staining
Your Diagnosis Is?

A. Sarcoma
B. Lipoma
C. Plexiform schwannoma
D. Normal clitoral tissue

[Video]
Your Diagnosis Is?

Libby Edwards, MD
LibbyEdwardsMD.com

DISCLOSURES
None

DISCLAIMERS
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?

A. PRP is a reasonable therapy and discuss this in detail
B. PRP is a poorly reported therapy and suggest more aggressive traditional concomitant alternative superpotent topical corticosteroid and tacrolimus
C. Her discomfort may not be due to LS
D. That she would probably respond to intralesional corticosteroids
You tell her?

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C. Her discomfort may not be due to LS
D. That she would probably respond to intralesional corticosteroids

Other possibilities include all except?

A. Squamous cell carcinoma
B. Atrophic vagina
C. Vulvodynia
D. Subtle lichen sclerosus
Other possibilities include all except?

A. Squamous cell carcinoma
B. Atrophic vagina
C. Vulvodynia
D. Subtle lichen sclerosus

All now are reasonable steps except?

A. Wet mount
B. Reassurance with one year follow-up and discontinuation of her clobetasol
C. Pelvic floor evaluation
D. Evaluation of area of pain
All now are reasonable steps except?

A. Wet mount
B. Reassurance with one year follow-up and discontinuation of her clobetasol
C. Pelvic floor evaluation
D. 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?

A. Undertreated LS
B. Atrophic vagina
C. Vulvodynia
D. Vulvodynia with factors of LS and atrophic vagina
LESSON

• Be willing to let go of a diagnosis
• Many chronic symptoms are multifactorial in origin

30 y.o. G0 with vulvar lesions for several months
Your diagnosis is?

- Molluscum contagiosum
- Skin tags
- Neurofibromatosis
- Epithelial inclusion cysts

This condition is caused by a:

- RNA pox virus
- Herpes virus
- DNA pox virus
- Human papilloma virus
Your treatment is?

- A. No treatment needed; they self resolve
- B. Cryotherapy
- C. Curette lesions
- D. All of the above are options

Other treatments?

Cantharidin
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?

A. d-VIN
B. Recalcitrant, erosive LS
C. Zoon’s vulvitis (plasma cell vulvitis)
D. HSIL

HOLD UP ALL CARDS  Can’t tell
Is this?

A. d-VIN  
B. Recalcitrant, erosive LS  
C. Zoon’s vulvitis (plasma cell vulvitis)  
D. HSIL  

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)
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)
White and pigmented morphology
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
A 62 y.o. presents with a long-standing history of lichen sclerosus diagnosed by biopsy in her 30s presents with worsening symptoms for the past year

- Rawness, tearing, pruritus, burning and increased pain despite using clobetasol
- She has been recently placed on lidocaine prn with minimal relief
Your most likely diagnosis is?

- A Bartholin cyst
- B Lipoma
- C Sarcoma
- D Hernia

Your next step? Part A

- A Pelvic/vulvar CT
- B Pelvic/vulvar MRI
- C Pelvic/vulvar Ultrasound
- D Go straight to the OR
Your diagnosis is? Part B

- A  Bartholin cyst
- B  Lipoma
- C  Sarcoma
- D  Hernia
The labium majus is homologous to the:

- [A] Scrotum
- [B] Urethra
- [C] Testicle
- [D] Seminal duct
CASE

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

A. Malignant melanoma
B. High grade squamous intra-epithelial lesion (HSIL, HPV-associated VIN)
C. Post-inflammatory hyperpigmentation
D. 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

Hope Haefner, MD
HSIL
HSIL and SCC

LESSON

BIOPSY!!!
• 88yo G4P2 who had cobalt therapy for endometrial cancer 50 years ago
• Complains of vulvar irritation
Would you take a biopsy?

- A Yes
- B No
Your diagnosis is?

- A Squamous cell carcinoma
- B Adenocarcinoma
- C Paget disease
- D Prolapser (Bowel)
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?

A. Inverse psoriasis
B. Intertrigo dermatitis
C. Resistant Candida
D. Contact dermatitis

Is this?

A. Inverse psoriasis
B. Intertrigo dermatitis
C. Resistant Candida
D. 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

46 y.o. G0 was first noted to have unilateral labial swelling in September 2006
MRI of the Vulva

- Asymmetric enlargement of the right labium majus without a discrete lesion. Soft tissue stranding and inflammatory changes in the right labium majus fat are likely inflammatory/postinfectious in etiology. No lipoma or hernia seen. If clinically indicated, a follow up study in one year could be obtained.
Specimen Taken to Pathology

- Pathology resident informs you it does not look grossly like a lipoma
- Resident stains the outside
Your Diagnosis Is?

- A Lipoma
- B Colon
- C Fibroma
- D Leiomyosarcoma

29 y.o. G3P2 with Dyspareunia

- Pain with entry
Does She Have Localized Vulvodynia?

- A Yes
- B No
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?

A. Zoon’s (plasma cell) vulvitis
B. Lichen planus
C. Contact dermatitis
D. differentiated VIN
Your diagnosis is?

A. Zoon’s (plasma cell) vulvitis
B. Lichen planus
C. Contact dermatitis
D. 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!)
41 y.o. G2P2 c/o painful intercourse

- Present for 3 years
- Cyst was needle drained in the office
- Grown since last visit and hurts with intercourse
Your diagnosis is?

A. Bartholin duct cyst  
B. Gartner duct cyst  
C. Skene duct cyst  
D. Rectocele

Your treatment is?

A. Cyst wall biopsy with Kevorkian  
B. Marsupialization of cyst  
C. Excision of cyst  
D. No treatment needed
You are called to the OR to help perform a laser treatment for HSIL of the vulva and anus on a 17 y.o. patient.
What additional study would you recommend?

A. HIV testing
B. Coagulation studies
C. Pulmonary function tests
D. Serologic herpes testing
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?

A. Candidiasis
B. Irritant contact dermatitis
C. Lichen simplex chronicus
D. Perianal bacterial disease
Is this?

A. Candidiasis
B. Irritant contact dermatitis
C. Lichen simplex chronicus
D. Perianal bacterial disease

Perianal Streptococcal Disease

- Culture yields beta hemolytic group A streptococcus
- Given oral antibiotic (amoxicillin, penicillin, beta lactimase 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
LESSON

• Scaling red plaques are not all yeast and dermatitis
A 68 y.o. G5P5 presents with severe clitoral pain since January 2009.

Past medical history -
  Hypertension, atrial fibrillation, urinary incontinence, and rheumatic fever as a child
Pap smear June 2008 negative, satisfactory; No hx of abnormal Pap
What is ideal management?

A. Pelvic exam in clinic, bx in clinic
B. Radiate
C. Examination under anesthesia, clitoral resection
D. Sclerotherapy
Your most likely diagnosis is?

- [A] Priapism of clitoris
- [B] Severe cellulitis
- [C] Cancer
- [D] Varicosity
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?

A. Hidradenitis suppurativa
B. Evolving polymicrobial infections in patient with undiagnosed HIV
C. Job’s syndrome (hyper IgE synd)
D. 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
Poor Response to Therapy

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

Great Job!