Your Diagnosis Is?

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ISSVD September, 2014
Chicago, Illinois

Conflicts of Interest for Drs. Haefner and Margesson

Hope Haefner, MD is on the Advisory Board of Merck Co., Inc.

Lynette Margesson, MD has no conflicts of interest
OBJECTIVES

1. Identify clinical features of a spectrum of vulvovaginal diseases
2. Establish therapeutic strategies for a variety of vulvovaginal diseases

Little evidence-based treatment
Too few studies done in vulvar diseases

Most treatments discussed are “off-label”
Written Information Available:

University of Michigan Center for Vulvar Diseases

Then, click on Information on Vulvar Diseases

http://obgyn.med.umich.edu/patient-care/womens-health-library/vulvar-diseases
- References for Vulvar Diseases: Recognizing and Treating VIN (PDF)
- Your Diagnosis Is (PPT)
- Your Diagnosis Is (PDF)

Argentina, November 14 and 15, 2013

- Vulvovaginal Surgery (PPT PDF)
- Desquamative Inflammatory Vaginitis (PPT PDF)
- Desquamative Inflammatory Vaginitis (Word PDF)
- Update in Vulvar Disease (PPT PDF)
- Update in Vulvar Disease (Word PDF)

Connecticut, December 12, 2013

- Learn to Like the Lichens (PPT PDF)
- Learn to Like the Lichens Handout (PDF)

Troy, MI, February, 2014

- Dermatologic Vulvovaginal Conditions (PPT PDF)
- Dermatologic Vulvovaginal Conditions Written Handout (PDF)
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
A 29 y.o. G3P3 presents with vulvar irritation.
What is your diagnosis?

A  Candidiasis
B  Bacterial vaginosis
C  Group B streptococcus
D  Desquamative inflammatory vaginitis
Atrophic vaginitis

What other conditions does DIV have a similar microscopic appearance to?
DIV

Etiology is unknown; past theories
- Lichen planus
- Nonspecific term for any erosive mucosal disease (LP, pemphigus vulgaris, cicatricial pemphigoid)
- Group B streptococcal infection

Current theory
Specific sterile inflammatory vaginitis, probably autoimmune

Common picture of several uncharacterized diseases

DIV

Therapy
- Intravaginal clindamycin cream vs. intravaginal hydrocortisone suppositories
- If that fails, clindamycin cream and hydrocortisone suppositories
- If no response, compound a high dose intravaginal corticosteroid and 2% clindamycin
50 year old woman developed a red facial rash that spread to entire body. Diagnosed as eczema (treated with UV therapy). All resolved but the vulva.

She developed breast cancer, and was treated with surgery, chemo (Taxotere & Cytoxan), radiation. Vulvar rash always flared with chemo.

The vulvar rash persists with swelling, pruritus, burning stabbing pain, dysuria..
Your Diagnosis Is?

A. Tinea Cruris (fungus)
B. Drug Reaction
C. Spreading pyoderma / cellulitis
D. Contact Dermatitis
Contact Dermatitis

70 mg IM Triamcinolone
200 mg fluconazole PO daily X 14
Cefadroxil 500 mg PO BID X 7
4 mg Ciproheptadine PO TID prn
petroleum jelly
Hibiclens

She was better for a few days and relapsed

Herpes culture obtained
Herpes Type I & II IgG titers sent
Yeast culture obtained
Acyclovir 800 mg PO BID X 5 days
Herpes and yeast cultures and titers negative, bacterial culture
Beta Strep and MRSA, Bactrim added
What NOT To Do?

A. Trial of clobetasol ointment 0.05%
B. Review history and stop all topicals and, as possible, stop medications
C. Patch Testing
D. Biopsy

Histology:
Acute Spongiotic Dermatitis

Allergic dermatitis – drug, contact
Allergy Testing Results

- Balsam of Peru
- Fragrance resin
- Ethylchloroisothiazolinone
- Methylisothiazolinone (latex emulsions)
- Oleamidopropyl dimethylamine
- Nickel sulfate
- Formaldehyde
Fixed Drug Eruption from Red Dyes in Medications

- Prilosec: Red # 40, 28, 7
- GasX: Red #30
- Pepto Bismol: Red # 22 and 28
- Fluconazole (Diflucan): Red # 40
- Cefadroxil (Duricef): Red # 40 and 28
- Hibiclens: Red # 40
- Robitussin: Red # 40
- Huggies Baby Wipes: Methylisothiazolinone
Vulvar Allergic Contact Dermatitis to a preservative Ethylenediamine in a cortisone cream

28 y.o. G4 P0 EAB 4 with vulvar, axillary disease. She has used Yasmin and this has not helped. GYN Hx

- Positive trichomonas
- - HIV
- Stage 3 vulvar Hidradenitis c active left axillary hidradenitis s/p resections
Hurley’s Criteria for HS Staging

**Stage I:** abscess formation, single or multiple, without sinus tracts and cicatrization/scarring.

**Stage II:** recurrent abscesses with sinus tracts and scarring, single or multiple, widely separated lesions.

**Stage III:** diffuse or almost diffuse involvement, or multiple interconnected tracts and abscesses across the entire area.

- 1-24-14 IBD panel
- Pattern consistent with IBD
  Crohn’s Disease
• 2-24-14 Colonoscopy
• Diagnosis: A-C. Terminal ileum, right and left colon, biopsies: No significant abnormality.

Endoscopy—The results were reviewed at the workstation and the image quality was good. The first gastric image was seen at the beginning of the study at 0 minutes 0 seconds. The first duodenal image was noted at 15 minutes 4 seconds. The cecum was entered at 3 hours 29 minutes 3 seconds.

The gastric emptying time could not be accurately calculated. The small bowel transit time was determined to be 3 hours 13 minutes.

The esophagus and stomach were not well visualized.

The small bowel was unremarkable for inflammatory lesions. There were some prominent white tipped villi in the duodenum and proximal jejunum such as seen at 31 minutes 41 seconds. There were no ulcers, erosions, strictures, masses, or areas of inflammation. There were no angioectasias or fresh or old heme in the lumen.
• Other PMH:
  – Negative
  – Her past surgical history is significant for the multiple excisions. The boils were removed in June 2004. Her buttock had surgery in March 2005 and April 2011. She has had other surgeries for the hidradenitis, but cannot remember the dates; however, she has never undergone an extensive resection of the vulva and buttock with skin grafting. It seems as though the tracts have just been I&Ded or single tracts excised without removal of disease in toto.

• FamHx: Family history is significant for mother with hypertension and irritable bowel syndrome. Interestingly, her mother and sister have boils; diabetes in mGM and mGF

• SocHx: She does not smoke cigarettes. She does drink up to 7 drinks a week. She does have when she is in significant pain 4 drinks at 1 time; however, she does not think drinking is a problem and can go weeks without having alcohol. We briefly discussed binge drinking. She does not use illicit drugs currently. There is a history of drug abuse with herself as well as siblings and parents.
Left Axilla

Under breasts
• MEDICATIONS: has a current medication list which includes the following prescription(s): biotin, cefuroxime, duloxetine, erythromycin with ethanol, multivits, ca, minerals/iron/fa, tretinoin, and oxycodone.

• Hepatitis testing nonreactive A, B, C

• Management options?

What are your recommendations?
• Derm recommended initiating Remicade infusions at 5 mg/kg (at 114 kg, rounded to nearest 100 mg, this is a dose of 600 mg per infusion). Infusions to occur at 0, 2, 6, and 8 week intervals followed by every 8 weeks thereafter.

• Derm recommended initiating concomitant methotrexate (goal dose of 7.5 mg weekly) in an effort to prevent antibody formation against the chimeric Remicade. Folic acid to be taken on non-methotrexate days.

• In the interim, recommend BP wash to affected areas daily
  - Erythromycin solution daily to BID to affected areas as well
  - Continue Ceftin
  - Will likely transition to doxycycline 100 mg po BID to be used simultaneously with the above systemic and topical agents
• Comedonal acne with ice-pick scars.
- Continue tretinoin 0.1% cream at bedtime
- Discussed that she may wish to pursue treatment in the Cosmetic Dermatology and Laser Center for her scarring but did inform her that this is not covered by insurance. Her types of lesions may be amenable to TCA CROSS.
4 mos post op graft

2 years after surgery
Extensive Hidradenitis vs. Crohn’s

Recent Admission 2 Years Later
• Recommendations if medical management fails—with the potential that this may be Crohn’s?
42-year-old has recurrent acute, severe vulvar pruritus for 6 days. She has had “recurrent yeast” infections several times a year for 10 years.

She is miserably itchy

Contact Dermatitis and HSV
49 year-old woman with 6 month history of bloody vaginal discharge and vulvar irritation

- She is scheduled for EUA and D and C and any additional procedures required
- PMH significant for multiple sclerosis (wheelchair dependent), borderline diabetes mellitus, and hypothyroidism
- Suprapubic catheter for 2 years
- Difficult examination secondary to muscle spasm
After exam under anesthesia, visible pubic ramus. What Do You Suggest be Done NOW?

A. D and C only
B. Biopsy of vulvar tissue /bone debridement and culture
C. D and C and biopsy of vulvar tissue/bone debridement and culture
D. Resection of pubic bone with flap placement
Diagnosis- Osteomyelitis of Pubic Rami
What Treatment Do You Recommend?

A. Resection of pubic rami
B. Intravenous antibiotics
C. Intravenous antibiotics and serial debridement
D. Oral antibiotics
78 year old lady presented with vulvar irritation. For 1-2 years gradually worse. The upper vulva is now sore if touched. She saw her gynecologist who was concerned about some scarring and peri-clitoral irritation 1 ½ yrs ago. The biopsy was non-specific - minor inflammation. Clobetasol ointment irritated the area.
Your Diagnosis Is?

A  Contact Dermatitis
B  Paget’s Disease
C  Malignant Melanoma
D  Squamous Cell Carcinoma
How Many Biopsies?

A. One
B. Two
C. Three
D. More than one depending on induration

2, 4 mm punch biopsies
MM 1.3 mm in depth
Malignant Melanoma

Vulvar Malignant Melanoma (MM)

- 5% vulvar cancer are MM
- Found in older women > 65 years
- Site - 75% on vulvar mucosa
- Amelanotic 25%, multifocal 20%
- Atypical color - with variably red, white, or blue color: amelanotic MM pink or red
- A late diagnosis
Differential Diagnosis:

- HSIL of the vulva
- SCC
- Extramammary Paget’s Disease
- Atypical contact dermatitis
- HSV
Vulvar Melanosis

LS Melanosis
Vulvar Melanosis

Common

Pigmented macules and patches
Solitary or multifocal
Angular and asymmetrical
Usually vulvar trigone

Dx - biopsy

The Saga Continues...

61-year-old woman with T8 paraplegia, type 2 diabetes and a seizure disorder

• Fecal incontinence with colostomy bag and urinary incontinence with suprapubic catheter

• She presents with one-year history of a vulvar ulcer
• MRI revealed a large ulcer with evidence of osteomyelitis of the right inferior pubic ramus
• Also noted to have asymmetric thickening and hyper-enhancement of the right bladder wall extending to the right urethra

• Underwent exam under anesthesia, vulvar ulcer debridement and vulvar biopsies, pubic bone debridement and biopsies

• Cystourethroscopy performed by Urology did not show any involvement of the urethra or bladder
• Coagulation used on vessels on bone and under bone for hemostasis
• No active bleeding is noted at end of surgery
• Approximately 2 hours postoperatively the GYN team was contacted by nursing staff concerned that the patient was continuing to have “vaginal” bleeding
What Should Be Done Now?

- Check CBC, coagulation studies, Type and Cross
- See if one or two vessels can be isolated for suturing in recovery room
- Pack the vagina
EBL in recovery now ~750 ccs
What should be done?

- Patient received 1 unit pRBCs intraoperatively and 500 ccs albumin
• Exam under anesthesia revealed:
  – Both bone and vaginal tissue bleeding

• Bleeding controlled with two figure of eight sutures
• Vagina was irrigated and multiple areas of bleeding coagulated with the Bovie
• Surgifoam placed over bone
• Vagina packed with Kerlix
• Transfused an additional 2 units PRBCs on POD#2
• Treated with Vancomycin and Tobramycin
• Noted to have a pseudomonas UTI and bone biopsies grew Enterobacter
• Discharged with plan for 6 weeks of IV Cefipime
• Biopsy of bone c/w granulation tissue
• Has undergone one additional debridement the first week of January...saga to be continued.

Lessons Learned
Questions to Ask in OR

• How is her blood pressure?
  – Watch the patient’s BP intraoperatively. Active bleeding may be masked by hypotension
  – This patient was 80-90s/40-50s in OR and then 120-140s/70-90s in PACU
Major Lesson Learned

Make sure they are bone dry

73 year old lady presents with bleeding from “down there” - she cannot see the area but sees blood on toilet tissue and in toilet for several weeks. There is minor irritation only. She has not been sexually active for years.
Cause of Bleeding?

A. Lichen Planus
B. SCC
C. Lichen Sclerosus
D. None of these
What Not To Do?

- Biopsy vulva
- Biopsy anal lesion
- Biopsy oral cavity
- Assess vagina

Vulvar BX
Lichen Sclerosus (LS)
Treated clobetasol 0.05%
ointment X 12 week
then 3-3 times a week

Anal hemorrhoid removed and
biopsy 12 o’clock perianal -
LS

Atrophic vagina and vulva Rx
estradiol 0.001% cream 2-3
times a week

All irritants stopped
62 y.o. woman with vulvar irritation. First noted lump on vulva in 2011. It grew and she underwent a biopsy.
Your Diagnosis Is? Part 1

A. Lichen planus
B. Lymphangiomas
C. Lichen sclerosus
D. HGSIL
Your Diagnosis Is? Part 2

A. Lichen planus
B. Lymphangiomas
C. Lichen sclerosus
D. HGSIL
They can become black in color. The theory behind this color change is secondary to:

A. Association with diabetic skin changes
B. Premalignant changes (compound nevi)
C. Scar changes from frequent rupture
D. Hemorrhage
Skin Closed with Interrupted Sutures

Doing well

No Recurrence for 1.5 years
Recently seen with 2 small “blebs”
Skin Graft
No recent follow up- was doing well but only saw her 3 months out from surgery (multiple personality disorder)
25 year old with 9 month history of painful vulvar ulcers and dysuria.

- Dx HSV (HSV cultures neg and HSV1 serology positive).
  Given valacyclovir and in severe pain - admitted to a large teaching hospital for painful ulcers and DIV.
- Rx IV acyclovir.
  No change and biopsy not helpful given prednisone, doxycycline and topical clobetasol ointment.
  She was better – almost clear, then relapsed.

- Past health – obesity, diabetes insipidus, papillary thyroid cancer, fall and head injury age 7 yrs.
What Test To Do?

A. HSV PCR
B. Biopsy
C. MRI Pelvis
D. Colonoscopy

Your Diagnosis Is?

A. HSV
B. Aphthous Ulcers
C. Langerhans Cell Histiocytosis
D. Crohn’s Disease
Langerhans Cell Histiocytosis

Also had diabetes insipidus
Lost teeth
33 year old with irritated and eroded papules in skin folds - on skin only
LANGERHANS CELL HISTIOCYTOSIS

- Rare infiltration of histiocytes forming single or multiple osteolytic bone lesions plus skin, lymph nodes, lungs, thyroid, spleen, bone marrow and central nervous system
- Usually seen in children 1-3 yrs
- Cause- myeloid dendritic Langerhans cells (controversy - reactive or neoplastic)
- Affects 1 organ in 55% (e.g. bones)

You are called to consult on a 1 day old infant delivered via NSVD. There is concern about the appearance of the vulva.
You recommend:

A  Excision with fine tip bovie
B  Follow, no need to treat
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 infection
D. Invasive squamous cell carcinoma
How many different types of herpes exist that affect humans with disease?

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

Herpesviruses

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<th>Subfamily</th>
<th>Target cell</th>
<th>Latency</th>
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<td>Betaherpesvirinae</td>
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<td>T lymphocyte</td>
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<td>Gammaherpesvir.</td>
<td>Endothelial cells</td>
<td>Unknown</td>
<td>body fluids</td>
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http://en.wikipedia.org/wiki/Herpesviridae
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%
88 years old has itchy irritated vulva that has been getting a bit worse for several months. LS for years.

- She is very well on no meds.
- She never has had vulvar or genital problems.
- She has had no regular gynecology care.
Your Diagnosis Is?

A. Condyloma
B. Verrucous carcinoma
C. Condyloma and squamous cell carcinoma
D. Condyloma and verrucous carcinoma
Condyloma with Lichen Sclerosus And Squamous Cell Carcinoma
Two ladies 56 and 59 years old have similar problems. They have vulvar burning, sexual dysfunction with no penetration for over a year. They also have no tolerance to any topicals.
Do They Have the Same Condition?

A. YES
B. NO
Scarred Vulva - biopsy
Lichen Sclerosus and
Vaginal and Oral Lichen Planus

Erosive LP Vulva and Vagina

Papular Lichen Sclerosus
62 Year old lady has a chronically sore vulva.

For several years she was treated for yeast, never better.

She cannot be sexually active due to severe entry dyspareunia.

She is desperate as she was told this is VULVODYNIA!
Your Diagnosis Is?

A. Vulvodynia
B. Lichen Sclerosus
C. Premenopausal Atrophy
D. Squamous Cell Carcinoma

Lichen Sclerosus with tearing from sexual trauma and low estrogen
LS and sexual dysfunction are very common.

Introital narrowing.
Lichen Sclerosus

Complications
- SCC
- Introital narrowing resulting in dyspareunia and rarely urinary problems
- Sensory changes - clitoral sensory loss
  - vulvar hyperalgesia, vulvar pain
- Psychosexual problems

LS Sexual Complications

significant sexual dysfunction due to:

1. Sensitive, delicate, thin skin that tears easily
2. Fear of pain and poor arousal, pelvic floor dysfunction
3. Scarring with hooded clitoris, introital stenosis with poor arousal, poor or no orgasms and pain
54 yr old lady was seen for a dry, burning vulva for the last year. Her last period was at age 51yrs. The skin feels raw and all creams irritate. She was given clobetasol ointment that caused more burning. She cannot tolerate any attempts at penetration.

Treatment

• Education
• Stop all trauma
• Treat LS with clobetasol 0.05% ointment daily for 12 weeks
• Daily topical estrogen cream
• When healed dilators and pelvic floor treatment
• Consider tricyclics or gabapentin if still painful
• May need surgery
Her main problem is not?

A. Lichen Planus
B. Atrophy
C. Lichen Sclerosus
D. Lichen Simplex Chronicus
Vulvovaginal atrophy with lack of estrogen

6-8 weeks estradiol 0.001% cream 2-3 nights a week and all was better but some bleeding after sex

What is true?

A. She has Lichen Planus
B. She has LS and LP
C. She has Lichen Sclerosus
D. This is a Scarred vulvitis
61-year-old G3P3 presents with constant vulvar drainage
If you could only look at one other area of her body, where would you look?

A. Eyes
B. Colon
C. Axilla
D. Mouth
What is this?

A. Squamous cell carcinoma
B. Epithelial inclusion cyst
C. Pyogenic granuloma
D. Paget’s disease
4 months post op from skin grafts
2 years after surgery

69 year old lady has had an itchy irritated vulva for many years after menopause. Mostly she ignores it. Now she has a burning vulva with dysuria that has been getting worse for 1-2 years. Estrace cream and antiyeast creams burn and do not help. She cannot have penetration.
Your Diagnosis is?

A. Lichen Planus - LP
B. Scarred vulvitis
C. Lichen Sclerosus - LS
D. LS and LP

Sheets LS papules Bx LS
Eroded Scarred vulva Bx LP

8/5/2014
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 pre-cancer or cancer
If in Doubt, Cut it Out