Options and Discussion of Difficult Cases

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Disclosures

• No financial relationships or conflicts of interest to disclose
Course 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

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
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 sclerosis or lichen planus—chronic inflammatory skin disorders
- Vulvar intraepithelial neoplasia—precancerous condition, often associated with a virus, the human papilloma virus (HPV)
- Herpes simplex virus—a disease of the amputs 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.

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**
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)

Make Your Selection

A
B
C
D
32 y.o. G0 with **painless** vulvar irritation and erythema (presented her at this meeting last year, now with new findings)
• Vulvar irritation began in fall 2015 when she experienced new watery odorless discharge and her PCP noted whitening of the left perineum

• She went on to develop a "growth" of the left perineal region

• Initially treated with topical steroids; numerous courses of oral antivirals. Inpatient admission for IV acyclovir.

• She also used an oral antibiotic regimen, which brought no significant improvement to her symptoms

• On January 12, 2016, patient underwent excision of the perineal lesion at an outside facility
  • Pathology demonstrated ulceration, pseudoepitheliomatous hyperplasia, an underlying polypoid granulation tissue

• On January 26, 2016, patient noted regrowth of the perineal lesion over the previous scar
Your Diagnosis Is?

A. Squamous cell carcinoma
B. Aphthous ulcers
C. Pseudotumor herpes
D. Factitial
Time for a new treatment...

A. Valacylovir
B. Famcyclovir
C. Thalidomide
D. Surgical resection
What to do Now
Medication started July, 2016

Thalidomide

- Inhibits TNF-α, IL-6, IL-10 and IL-12 production
- Modulates the production of IFN-γ
- Enhances the production of IL-2, IL-4 and IL-5 by immune cells
- Inhibits NF-κB and COX-2 activity
- It increases lymphocyte count, costimulates T cells and modulates natural killer cell cytotoxicity
Additional Medications

• IUD prior to thalidomide
• Topical imiquimod
• Topical cidofovir

March, 2017
6 months after completing thalidomide, 3 small erosions noted on vulva

What Workup Would You Recommend?
What Treatment Do You Recommend?

Should she be on an antiviral during pregnancy?
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 as all burn on application.
Do they have the same condition?

A  YES
B  NO
Which is lichen sclerosus?

A

B

C

Everything white is Lichen Sclerosus?
White Vulvar Conditions

Lichen Sclerosus
Lichen Planus
Lichen Simplex Chronicus
Mucous Membrane Pemphigoid
Vulvar Intraepithelial Neoplasia
Vitiligo

Biopsy to confirm diagnosis
Always examine vagina, perianal area, mouth and skin

Perianal LP
lacy pattern

Lichen
Simplex
Chronicus
Diagnosis?

“Everything white is **NOT** Lichen Sclerosus”
Case

- 30 y.o. noticed bulge on right superior labium majus when wearing tight pants
- PMH: BMI 37, otherwise healthy
- Exam
  - 5 cm
  - Soft, well-circumscribed, translucent, cystic, nontender
  - Easily reducible
Question: The most likely diagnosis is ...

A. Inclusion cyst  
B. Cyst of the Canal of Nuck  
C. Congenital lymphatic malformation  
D. Labial edema
Relevant work-up

- Ultrasound
  - 5 x 2 x 2 cm fluid-filled cyst without color flow
- MRI
  - 8 x 3 x 2 cm cystic mass with nodularity and septations
  - Smaller similar mass on left

To the OR

- Inguinal approach
- To our surprise, cyst was OUTSIDE the inguinal canal

Benign mesothelial lined fibromuscular tissue
37 y.o. G3P3 with Rash and Pruritus

• Pathology revealing focal acantholytic dyskeratosis on July 19, 1996, as well as September 2004
• She has been treated with an antifungal (ciclopirox) in the past as well as metronidazole
• She does wake herself in the middle of the night with itching/scratching
Your Diagnosis Is?

A. Darier’s disease
B. Fox Fordyce disease
C. Ectopic breast tissue
D. Molluscum contagiosum
Darier Disease

- Rare genetic disorder that is manifested predominantly by scaly or crusted papules
- Occurs worldwide with a prevalence of 1 to 4 per 100,000
- Autosomal dominant pattern (*ATP2A2*, found on chromosome 12q23-24.1)
- Classified as a hereditary acantholytic dermatosis
- Exacerbating factors include heat, sweat, sun exposure, friction, or infection
  - Avoid lithium or calcium channel blockers

Treatment

For early disease, topical medications:

- Topical corticosteroids – low- to medium-potency
- Topical retinoids (irritation is common -reduce this by alternating applications with emollients)
- There are isolated reports of response to treatment with topical 5-fluorouracil, tacrolimus, pimecrolimus, tacalcitol, and diclofenac sodium 3% gel
Treatment- Severe Disease

- Oral retinoids

Case History  LJM

28-year-old woman with history of tender vulvar ulcer x 3 months with apareunia
   She has had a non-healing ulcer perianally x 3 years after surgery for “atypical pilonidal sinus”

No other symptoms
Your diagnosis is?

A. Behcet’s disease
B. Crohn’s disease
C. Aphthae
D. Tuberculosis

Vulvar Crohn’s Disease
Chronic inflammatory bowel disease
Rare on vulva -2% women have vulvar lesions

• Contiguous
  – direct fistulae from bowel to skin
• Non-contiguous/metastatic
  - painful labial edema +/- ulcers
  - “knife cut” ulcer
  - Abscesses +/- Hidradenitis Suppurativa

Non-specific
- aphthae – oral and vulvar
- Abscesses +/- Hidradenitis Suppurativa
Crohn’s Disease
Presenting with
Severe vulvar edema
Knife cut
Ulcers

Knife cut
Ulcers after
Rx for 2 weeks
40 y.o. G2P2 presents with pain and irritation of the vulva and perianal region

Vulvar symptoms began in 2012
Tried various antibiotics and ointments
Humira in 2014 with "some improvement"
Colonoscopy negative 2014
In 2015, she was given a diagnosis of hidradenitis suppurativa and was again started on Humira by Dermatology (completed 6 months)

Given 3 months of Remicade

Some infections in her breast prior to bilateral breast reduction and no issues since that time on breast

• PAST SURGICAL HISTORY: C-section x2, abdominoplasty, Roux-en-Y gastric bypass, bilateral breast reduction

• FAMILY HISTORY: Positive for diabetes and hypertension in mother and cancer in her father

• SOCIAL HISTORY: Negative for tobacco or drug use, positive for approximately 4 drinks of alcohol per week

• MEDS: None

• ALLERGIES: None
What to do now?

Recent EGD and colonoscopy negative
What to do now?

Two vulvar biopsies were taken from the lesion at the vulvar inguinal crease at the inferior left aspect of the vulva. One punch biopsy was taken at the edge of the lesion and a second punch biopsy was taken from the center of the lesion.

Diagnosis:
A-B. Vulva, biopsies: Nonspecific acute and chronic inflammation
What to do now?

Case LJM

38 yr old woman with an increasingly wet, mildly itchy, irritated vulvar eruption for 5 to 6 years. It is infected at times. She had a rare bowel lymphoma surgically removed 10 years before.
Your diagnosis is?

A. Condylomata Acuminata
B. Nevus Sebaceous
C. Molluscum Contagiosum
D. Lymphangioma

Lymphangioma
Case

• 18 y.o. G0 with Rosai-Dorfman disease, panhypopituitarism, central diabetes insipidus, adrenal insufficiency, chronic kidney disease

• Admitted to the hospital due to hypernatremia, mental status changes

• RN noted vulvar ulcers
History

- Patient reports similar episode 6 months ago
- Burning with urination
- Stopped scrubbing, used “ointment”, resolved

- Not sexually active
- Denies abuse
Relevant work-up

- Urine culture  NEG
- HSV PCR  NEG
- STI panel  NEG
- CSF HSV, culture  NEG
- Yeast culture  Candida albicans, rare

Courtesy of pathology outlines.com
http://www.pathologyoutlines.com/images/lymph/18_02.jpg
Your Diagnosis Is

A. Severe yeast infection
B. Primary herpes
C. Paget’s disease
D. Langerhan’s histiocytosis

Biopsy results

- Langerhans cell histiocytosis
  - Positive CD1a
  - Positive S100
  - Negative acid fast organisms
  - Negative BRAF V600E, K
  - Negative direct IF
Treatment in the meantime...

- Acyclovir
- Triamcinolone ointment
- Mupirocin ointment
- Nystatin cream

Vulvar ulcers are improving

Langerhans cell histiocytosis

- Rare histiocytic disorder (children)
- Single organ or disseminated, can affect CNS
- Bone involvement in majority of patients
  - Skin involvement in 40%
- Different from this patient’s Rosai-Dorfman disease

UpToDate. Clinical Manifestations, pathologic features, and diagnosis of LCH
Treatment for LCH

- Steroids
  - Topical
  - Systemic
- Chemotherapy
- Radiation
- Surgery


Course

- OR for sacral ulcer biopsies → also LCH
- Clobetasol x 3 months
- Pack wet-to-dry sacral wound to heal
- Advised chemotherapy for systemic LCH – vincristine
3 months later

**PLAN**

- Clobetasol daily to vulva
- Pack sacral wound until healed
- Chemotherapy pending

33 y.o. G5P4 presents to the University of Michigan Center for Vulvar Diseases for consultation regarding vulvar cysts/boils
- Started at 14 years of age
- Mostly on the vulva but she has had episodes of cysts/boils in the bilateral axillae, under her breasts, and on her back
- Prior treatments include: Oral and topical antibiotics (not helpful), vulvar cyst removal in 2011 in an office setting at her 6 week postpartum visit "without anesthesia" and around the clitoral area, oral antibiotics for an axillary boil around 8 years ago
• Consulted with a plastic surgeon who told her that nothing could be done about the boils since they are so close to her clitoris
• She is currently on an OCP for suppression and thinks that this is helpful
• Additionally, she has been trying to lose weight and has currently lost 50 pounds over the past year (from about 300 lb to 250 lb) through diet and exercise

Symptoms/signs

• Current symptoms include: itching and occasional drainage of the boils

• She is most bothered by a cluster of cysts near the clitoris that has been there for 12-13 years, occasionally swell and are also quite painful
What to Do?
Surgical Pathology Report

- Benign mucous cysts
Options and Discussion of Difficult Cases

Great Job!

Questions and Answers