Your Diagnosis Is?

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Disclosures/Conflicts of Interest

- Libby Edwards has nothing to disclose
- Hope Haefner Advisory Board Prestige, Inc.
- Colleen Stockdale has nothing to disclose

- EXCEPT - Very little regarding the treatment of vulvar disorders is evidence based
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 vulvovaginal conditions
Please Interrupt for Questions

Make Your Selection

A  B
C  D
Your Diagnosis Is?

A. Two rocks
B. Two sea turtles
C. A whale
D. None of the above
22 y.o. with painful discolored clitoris
**Your Diagnosis Is?**

A. Hemangioma  
B. Pyogenic granuloma  
C. Varicosity  
D. Melanoma

**What do you recommend as the treatment for her pyogenic granuloma?**

A. Sharp excision of clitoris  
B. Cryotherapy  
C. Fine point electrocautery to remove lesion  
D. Trichloroacetic acid
Pyogenic granulomas are misnamed (neither infectious nor granulomatous)

A. True  
B. False

Pyogenic granulomas of the genitals are more common in males than in females

A. True  
B. False
Pyogenic granulomas of the genitalia are more common in males than in females

A. True
B. False

Equal overall (except oral mucosal lesions 2 x more common in females).

To Prevent Recurrence- Avoid Trauma
67 year old presents for evaluation of vulvar appearance following annual exam with her primary care provider. She has a history of rectal cancer treated 11 years ago. She has no history of abnormal pap smears / testing. She is otherwise well and has no concerns.
What are the skin changes from?
A. Atrophy
B. Trauma/ ecchymoses
C. Radiation
D. Normal aging
How will you treat her?
A. No treatment needed
B. Exams every 4 months
C. Topical steroid
D. Retin A topical

radiation dermatitis

- Side effect of external beam ionizing radiation
- Irradiation of the skin leads to complex pattern of direct tissue injury with damage to the epidermis and endothelial cells
  - Characterized by dermal fibrosis and poikilodermatous skin changes, including hyper-and hypopigmentation, fragility, atrophy, and telangiectasias
Radiation Dermatitis

- Onset of chronic radiation dermatitis may occur from 15 days to 10 years or more after the beginning of radiation therapy.
- Extension of the acute process and involves continued skin inflammation.

Risk Factors

- Poor nutrition
- Pre-existing skin disease
- Obesity
- Prolonged or multiple treatments
  - Total radiation > 55 Gy
- Connective tissue diseases
- Epidermal DNA genetic disorders
- Ataxia telangiectasia
- HIV
- Diabetes mellitus
- Preceding cellular damage (radiosensitizing agent)
Management (chronic)

- Avoid sun exposure
- Avoid contact irritants
  - general skin care measures

- limited evidence:
  - vitamin D3
  - topical steroids (typically short course for acute)
CASE

• 31 yo woman noted black spot on vulva this AM
• Reports no symptoms, but when directly questioned, thinks she might be a little irritated
• Otherwise healthy, no personal or family history of importance
Is This?

A. Melanocytic nevus
B. Melanoma
C. Lichen sclerosus
D. Lichen planus
CASE

- *Purple*, irregular and poorly demarcated patch, with hemorrhagic blister confirmed by puncturing it
- Mild edema and hypopigmentation of the clitoral hood
- Biopsy confirms LS
- Clears in one month

Edema and hypopigmentation
• 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
• 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
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%**

UpToDate. Clinical Manifestations
pathologic features, and diagnosis of LCH

When diagnosed in children, the most common age is:

A. 1 to 3 years
B. 5 to 9 years
C. 10 to 13 years
D. 14 to 18 years
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
68 year old female referred for evaluation of vulvar and perianal lesions. She has noted “hemorrhoids” for quite some time.

Her history is notable for well controlled hypertension with HTCZ
What is the likely diagnosis?
A. Seborrheic keratosis
B. Melanosis
C. Angiokeratomas
D. HSIL or dVIN

What do you do?
A. Biopsy lesion(s)
B. Reassure
C. Treat with TCA/ cryo
D. Treat with steroid
Seborrheic keratosis:

- Typically develop after 35 to 40 years of age
- Benign, warty growth occur anywhere on the body (torso most common)
- Restricted to nonmucosal keratinizing skin
- Shave biopsy is diagnostic
CASE

- 4 year old girl with Crohn’s disease complains of irritation and rawness
- Mom sees rash and takes her to the pediatrician
- Diagnosed with genital warts that are unusually large due to her prednisone for diarrhea
Is this?

A. Condylomata lata
B. Jacquet’s diaper dermatitis
C. Large genital warts
D. Mollusca contagiosa in an immunosuppressed child

PSEUDOWARTS
Also called granuloma gluteale infantum, Jacquet’s diaper dermatitis

• Unusual manifestation of irritant contact dermatitis seen primarily in small children, but also occasionally in adult women (classic contactant is benzocaine - Vagisil)
• Flat-topped, skin colored to pink symmetrical papules, sometimes with overlying erosion
• Background of variable erythema
Is this?

A. Condylomata lata
B. Jacquet’s diaper dermatitis
C. Large genital warts
D. Mollusca contagiosa in an immunosuppressed child
Allergic contact dermatitis to benzocaine and irritant contact dermatitis to resorcinol in anti-itch cream; This peculiar CD morphology of eroded papules is seen only on the vulva or genital area of children.
PSEUDOWARTS

• Some feel corticosteroids are causative, others (including me) simply feel these are often used but not useful
• Liquid feces is an irritant especially likely to produce this
• Treatment consists of withdrawal of the irritant, barrier pastes
• (corticosteroids not useful)
33 y.o. G5P4 presents for consultation regarding vulvar cysts/boils near clitoris

- 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", 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

cks

• 32 year old woman who was well until 8 months ago
• Pain and tearing with sexual intercourse
• Heals in 3-4 days but always recurs
• Otherwise well, and no symptoms if not sexually active
What is your diagnosis?

A. Lichen planus
B. Mechanical fissure
C. HSV
D. Aphthous ulcer
Fishers, Erosions and Ulcers: Understanding the Difference

Fissure
- Linear-like cleavage of skin
- Extends into the dermis (can be superficial or deep)
- Results from excessive tension or decreased elasticity

Erosion
- Partial to complete loss of epidermis
- Doesn’t penetrate through basement membrane
- Red base
- Heals without scaring

Ulcer
- Full thickness epidermal loss + penetration through basement membrane to dermis
- Usually yellow-white base
- Heals with scarring
Ulcer Algorithm

A Decision-Support Tool & Novel Mobile Application

When you are in the trenches with ulcers....

- Infection
- Trauma
- Dermatosis
- Malignancy
- Combination
Mechanical Posterior Fourchette Fissure

- Unknown cause, though occasionally associated with skin disease or estrogen deficiency

- Confirm fissure; occasionally the patient reports sensation of an ulcer when it isn’t there and it can be vestibulodynia
Mechanical Posterior Fourchette Fissure

Consider trying:

- Topical lidocaine
- Woman on top
- Copious lubrication
Mechanical Posterior Fourchette Fissure

- Sometimes useful - have patient produce fissure, keep it open with dilators so the surface of the fissure re-epithelializes in the “open” position
- Most definitive therapy is surgical – perineoplasty. Excise front to back rather than side to side, and advance vaginal mucosa to cover defect

Recurrent posterior fourchette fissure
Surgery (Perineoplasty)
Fissure created prior to excision
Case

• 37 year old woman with long history of recurrent boils in the anogenital area
• Describes pain and itching
• Recently has noted growths
• Referred to gyn onc who diagnoses exuberant warts and debulks with vulvectomy
• Oops, and refers to me
Is this?

A. Genital warts
B. Lymphangioma circumscription
C. Verrucous carcinoma
D. Vesicular allergic contact dermatitis
Two Kinds of Lymphangioma Circumscriptum

• Primary tumor of lymphatics
• Severe edema that constricts lymphatic return, so that lymph vessels dilate and extend through the surface of the skin as very thin sacs of lymphatic fluid

Secondary Lymphangioma Is much more common

• She has edema from hidradenitis suppurativa
Congenital lymphedema (Milroy’s disease)
Take Home Point
• Biopsy before you chop – there is weird stuff out there
40 y.o. with painless vulvar mass
• Soft to palpation
Your Diagnosis Is?

A. Asymmetric vulva
B. Lipoma
C. Leiomyoma
D. Atypical lipoma
Lipoma of vulva

• Uncommon
• Slow growing
• May ulcerate
• Labia majora
• Well circumscribed
• Lobular
• Fibrous septae

Features of malignancy include:

A. Ring chromosomes
B. Lipoblasts
C. All of the above
D. None of the above
Atypical lipoma (aka well-differentiated liposarcoma)

- Most common type of liposarcoma
- Extremely rare on vulva
- Recurs locally
- Lipoblasts (arrows)
- Variation in cell size