Concerning Vulvar Lesions

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Disclosures

• Hope K. Haefner, MD: No financial relationships or conflict of interest to disclose
• E.J. Mayeaux, MD, DABFP, DABPM, FAAFP: No financial relationships or conflict of interest to disclose
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• Colleen Stockdale, MD, MS: No financial relationships or conflict of interest to disclose
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 Prolapse

Urethral Polyp
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

Case Presentation  LJM

- 38 year old lady has severe vulvar itching for 2 years and poor response to therapy
- She scratches and rubs during the day and is awakened scratching at night
- Topical clobetasol burns and is not tolerated
- She has been given several diagnoses, many treatments for “yeast”, eczema and she is fed up as no better
Your Diagnosis Is?

A. Lichen sclerosus
B. Lichen sclerosus and psoriasis
C. Psoriasis
D. Lichen simplex chronicus
Case 1

How would you treat the condition shown?

A. Trichloroacetic acid
B. Podophyllotoxin
C. Reassurance
D. Surgical biopsy/ excision

Courtesy of Tom Cox, MD
Micropapillomatosis Labialis

- Tiny papillary growths on the vestibule
- Usually symmetric
- May turn acetowhite
- Not HPV-related
- NORMAL variant!
- Reassurance only

Biopsy will almost always diagnose HPV lesions versus micropapillomatosis.

A. True
B. False
Micropapillations

• Biopsies often reported as HPV
  • HPV-DNA not routinely assoc.
  • If biopsy done, may be ideal to do in-situ hybridization as well as hematoxylin-eosin stains
• Three patterns: micropapillae, vestibular papillomatosis, and papillary HPV disease

Micropapillations

• Normal micropapillae are congenital but may be accentuated by inflammatory conditions

• Normal micropapillae

Courtesy of Dr. E.J. Mayeaux, Jr., M.D.
Separate Disease from Normal

• Two important lessons
  1. Do a good examination – don’t knee jerk
  2. Don’t assume everything is bad
• Guard against over-diagnosis
• Significant morbidity from overly aggressive treatment of low-grade or benign disease and of equivocal changes

“Vulvar hematomas”  CS

• 67 yr old female referred for recurrent vulvar hematomas x 8 months
  – Initially diagnosed as folliculitis, subsequently treated with I&D x 2 with recurrent cyst formation (described as large blood-filled pockets)
• Patient reports mild to moderate pain / awareness with bleeding typically weekly = no intercourse
• History of melanoma excised 4 years prior
• Multiple black raised lesions involving mons (1.8 cm), left inferior lateral labium minus 2.5 cm), right inferior labium majus (0.5 cm)

What is your DDx?
  – Hematoma
  – Melanoma
  – Angiosarcoma
  – Bullous pemphigoid

Diagnosis
• Angiosarcoma (referred to Gyn Onc/Heme Onc)
• Case reports – rare neoplasm comprise less than 2% of all sarcomas, are commonly aggressive, and disseminate widely – rarely occur in the female genital tract
• Poor prognosis – especially with vulvar sarcomas
• Our patient died within 1 year of diagnosis despite chemotherapy/radiation therapy
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...
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
What is the highest reported mortality with TEN?

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 Presentation LJM

- 67 year old lady has severe vulvar burning for almost 1 year
- She has had lichen sclerosus since menopause age 53 years
- Topical clobetasol has been used for years but was not working well so she has been using it once or twice a day as the burning is worse if she stops it
- No estrogens are being used
Your Diagnosis Is?

A. Lichen sclerosus
B. Lichen sclerosus and contact dermatitis
C. Steroid dermatitis
D. Lichen planus
Same Problem

E. J. Mayeaux, MD
Case 2
A 50 yo patient with whitening on buttock that is somewhat symmetric without itching, pain, or tissue loss.

Your diagnosis is:

A. Lichen sclerosus
B. Lichen planus
C. Lichen simplex chronicus
D. Vitiligo

Vitiligo

- Unpigmented, sharply defined macules
- Face, neck, dorsal hands, genitalia, body folds, & axillae

Courtesy of Dr. E.J. Mayeaux, Jr.
True statements about vitiligo include:

A. Affects about 1% of the general population
B. Family history present in ~5-15%
C. Sudden onset related to illness
D. 80% >40 years of age

Vitiligo

• Loss of skin pigmentation
• Immune-mediated melanocyte destruction
  • Affects all skin - cosmetic condition
  • Significant psychological distress
• 1% of the general population
  • Family history in 25 to 30%
  • Insidious onset, stress related, illness
  • 50% ≤20 years of age
## Treatment of Vitiligo

<table>
<thead>
<tr>
<th>Treatment</th>
<th>Examples and comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sun protection</td>
<td>Avobenzone (Parsol 1789) or titanium dioxide, mexoryl, clothing</td>
</tr>
<tr>
<td>Cosmetic coverage</td>
<td>Concealers (e.g., Dermablend, Covermark)</td>
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<tr>
<td>Topical steroids</td>
<td>Topical steroid class II and III (betamethasone 0.05% [Diprolene], fluocinonide 0.05%; head and neck most responsive to treatment</td>
</tr>
<tr>
<td>Phototherapy</td>
<td>Topical or oral psoralens with psoralen ultraviolet A-range, narrow-band ultraviolet-B therapy</td>
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<tr>
<td>Surgical grafting</td>
<td>Mini-graft, punch-graft techniques; for localized, stable disease</td>
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### “Lump” CS

- 37 year old G2P2 with 2 week history of increasing vulvar “lump” and pain
- Smoker
- No history of abnormal Pap smears
- Biopsy locally = HSIL = VIN Usual (VIN 3)
• Exam >>>
• Firm, friable, tender 3 cm mass right inferior vulva
  – No extension on vaginal or rectal exam
  – No LNA
• Next step
  – Biopsy
  – Imiquimod
  – TCA
  – Laser

Vulvar SCCA

• Biopsy performed = Well differentiated SCCA
  – 4th most common gyn cancer USA
  – 4-6% of gynecologic cancers in the US
  – 5 year survival about 70%

• Referred to Gyn Onc for management
38-year-old 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

Plexiform Schwannoma: An Unusual Clitoral Mass - Video
Case Presentation

• 42 year old lady complains of chronic vulvar itching with wet irritating vulvar discharge so that her underwear is always damp
• She has psoriasis since her teens and it flares intermittently in the vulvar area and skin folds but is not a problem usually
• She had pelvic radiation for cervical cancer three years ago
Your Diagnosis Is?

A. Flaring psoriasis  
B. Condyloma acuminata  
C. Condylomata lata  
D. Acquired lymphangiomas
E.J. Mayeaux, MD
Case 3

65 year old who complains of some itching and pain of the vulva. History of mouth ulcers but none currently.

What is the diagnosis?

A. Lichen Planus
B. HSIL
C. Contact Dermatitis
D. Can’t tell
**Biopsy**

- **Anesthesia**
  - 1% lidocaine (sodium bicarb)
  - 27-30 gauge needle to inject 1-3 cc's of anesthetic agent
  - Inject *intradermally*
- 3-5 mm Keyes punch or cervical biopsy instrument

**Red HSIL**

Courtesy Dr. E.J. Mayeaux, Jr.

Courtesy Dr. E.J. Mayeaux, Jr.

Courtesy of Hope Haefner, MD
Treatment of HSIL: Surgical

- Cold Knife (preferred)
- LEEP with path specimen
- Laser ablation (if no concern for invasion)


Medical Treatment for HSIL

- Imiquimod 5%
  - Two to three times weekly for 12-16 weeks
  - Colpo every 4-6 weeks during treatment - Excision for failures
  - Pain and erythema may limit use

- Cidofovir 1%
  - Response similar to Imiquimod in 2016 Cochrane review
  - Hair bearing skin unresponsive. Tends to ulcerate

- Photodynamic Therapy
  - ALA sensitizer + light = cell death
  - Quality of evidence “poor” in 2016 Cochrane review

“Recurrent vulvar condyloma”

• 49 year old G3P3 referred for further evaluation and treatment given a 4 year history recurrent vulvar condyloma. Initially treated with TCA. Now with increasing lesions despite use of imiquimod. Desires “laser ablation”.

• Exam

• What is your diagnosis?
What’s your Dx?

A. Warts
B. Not warts

How would you proceed?

A. Re-treat with TCA (provider applied)
B. Re-treat with imiquimod (pt applied)
C. Proceed with laser (patient preference)
D. Perform biopsy (diagnosis)
Biopsy indicated

- Always biopsy for failure to respond to treatment for condyloma!!
- Need to rule out dysplasia / cancer
- However, in this case does not look like cancer…
- Biopsy confirmed acrochordon (skin tags)
  - Benign growth
  - Associated with skin friction, obesity, diabetes

Treatment

- Symptomatic
- Concern for associated dysplasia
- Excise
- May consider: cryotherapy, cautery, LASER, ligation