Update in Vulvar Disease

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OBJECTIVES

1. Identify clinical features of a spectrum of vulvar diseases

2. Establish therapeutic strategies for a variety of vulvar diseases
Disclosures/Conflicts of Interest

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Advisory Board of Merck, Co. Inc.

Off label use of multiple medications discussed

Written Information Available:

University of Michigan Center for Vulvar Diseases (Google)

http://obgyn.med.umich.edu/patient-care/womens-health-library/vulvar-diseases

Then, click on Information on Vulvar Diseases
University of Michigan Center for Vulvar Diseases

There are many reasons for seeing a vulvar specialist at the University of Michigan Center for Vulvar Diseases, treating only part of the solution. Our multidisciplinary approach at ensure that women receive state-of-the-art care, from cutting-edge treatment options to education and counseling to meet every individual’s needs.

The Center for Vulvar Diseases was created in 1993 to better serve and treat women with diseases of the female genitalia. Our center is one of only a handful of clinics that specializes in treating these conditions. We focus on the multidisciplinary approach to help patients improve their health.

The team approach allows us to provide the highest level of care and expertise to women who have already demonstrated a resistant and chronic illness or an unusual vulvar condition.

Many women experience different forms of vulvar pain, including vulvodynia. Vulvodynia is pain on the lips of the vulva or at the introitus with a normal appearing vulva. It is a burning, itching, or swelling. 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)
- Herpes simplex—disease of the amputated vulva, with pus-filled pockets of fluid
- Bartholin cyst—fluid-filled cyst at the base of the hymen

Published papers:
- The Vulvodynia Guideline (PDF)

Related Web Sites:
- National Vulvodynia Association
- International Society for the Study of Vulvovaginal Disease
- American Society for Cytology and Cervical Pathology
- Larry Edwards, M.D.
- American College of Obstetrics and Gynecology
- Centers for Disease Control and Prevention Treatment Guidelines
- MHS Center for Sexual Health

Lectures:
- Conquering Resistant Vulvovaginitis 2011 (PDF)
- Vulvodynia 2011
- Learn to Use the Licensor Lecture, Hawaii, April 2012 (PDF)
- Learn to Use the Licensor, Hawaii, April 2012 (written text) (DOC)
- Vulvovaginal Disease Summary 2011 (PDF)
- Vulvar Ulcers Diagnostic Algorithm 2011 (PPT)
- Vulvar Ulcers Diagnostic Algorithm 2011 (DOC)
- Your Diagnosis is... June 2012
Labium majus
Labium minus
Labia minora
Labia majora
Mons Pubis
Clitoral Hood
Clitoris
Urethra
Vestibule
Vestibular Glands
Perineum
Anus
Hymen
Hart's Line
Vaginal Opening
Left lateral or Sim's position
**Anesthesia**
- 1% lidocaine
- 27-30 gauge needle to inject 1-3 cc's of anesthetic agent
- Consider adding sodium bicarbonate to decrease pain
- Inject subepidermally

**Biopsy**
- Keyes punch
  - 3-5 mm diameter dermatologic instruments (usually 4 mm)
  - Fine suture (3.0 or 4.0 Vicryl Rapide) vs. Monsel's/Silver nitrate

**Vulvar biopsy**

**Biopsy**

**Cervical biopsy instruments that can also be used for vulvar biopsy**
Anal Intraepithelial Neoplasia (AIN 2,3)
A 79 y.o. G2P2 complains of a vulvar sore and itching that started 1 year ago. A biopsy is performed.
Paget Disease

Multifocal
Eczematoid, weeping
Brick red
Scales
Eczematoid plaque
Sharply demarcated
## Differentiating Paget From Other Conditions

Positive mucin as well as immunoperoxidase CEA staining can be used to differentiate Paget disease from melanoma

- Paget (mucin and CEA positive)
- Melanoma (mucin and CEA negative)

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### Paget’s Disease

- +mucin
- +immunoperoxidase CEA

Photo courtesy of R. Lieberman, MD
Paget Disease

Occurs most commonly on the nipple and areola, where its presence signifies an underlying adenocarcinoma of the breast
- Apocrine gland origin
- Red velvety area with white islands of hyperkeratosis and at times may be pinkish and eczematoid
# Paget Disease

## Workup

**History and PE**
- Symptoms include itching, burning (soreness)
- Signs include velvety appearance and bleeding

Papanicolaou smear  
Mammogram  
Cystoscopy  
Colonoscopy

## Paget Disease Treatment

**Wide local excision (how far?)**
Recurrent Paget’s Disease

Laser (recurrent)
Imiquimod (recurrent)

Lichen Sclerosis
Introduction

• Common chronic vulvar disease
• Inflammation present
• Prevalence 1 in 300 to 1 in 1,000
• Age range from childhood to elderly (bimodal distribution)
Lichen Sclerosus

Figure of Eight – Hour Glass
Treatment of Lichen Sclerosus

• Superpotent steroid ointment (clobetasol propionate 0.05%)
  – Twice daily in a thin, invisible film for 1 month then daily for two months
  – Maintain twice weekly Class 1 VERSUS
  – Decrease to Class IV steroid
Steroid Medications

Clobetasol propionate ointment 0.05%
Sig: apply to vulva bid x 1 month, then qd x 2 months  Disp: 30 gms

Triamcinolone acetonide ointment 0.1%
Sig: apply to vulva qd to bid  Disp: 80 gms

Consider decreasing gradually to triamcinolone acetonide ointment 0.025% qd to bid

Steroid Medications

• Oral steroids
  Prednisolone
  Prednisone
  Methyl prednisolone
• Rarely required
• Significant side effects
  Occasionally intralesional or intramuscular steroids
Surgical Treatment

- Limited role (high rate of recurrence)
- Surgical division of mucosal adhesions helpful in clitoral phimosis, introital narrowing
A 45 y.o. G2P1 presents with complaints of vulvar pruritus. It awakens her at night. A yeast culture was negative. She has been intermittently treated without success with Class 1 topical steroids for over a year.
For Severe Itch-Scratch Cycle

Oral steroids (short term)
Cefadroxil 500 mg po bid x 7 days
Amitriptyline for few weeks (25 mg, increase to 50 mg if needed) vs.
Atarax (25 to 50 mg po qid prn)
White cotton gloves
Erosive Lichen Planus
Lichen Planus

• Diagnosis
  – Biopsy when indicated; often nonspecific
  – Biopsy white epithelium; otherwise the edge of an erosion
  – Consider immunofluorescent study

• Treatment
  – Intravaginal dilator
  – Topical corticosteroids
    • Hydrocortisone acetate suppositories (Anusol HC) 25 to 50 mg per vagina
    or
    • Temovate ointment inserted per vagina
    Taper
Lichen Planus
Other treatments

- Anti-inflammatory antibiotics
- Misoprostol
- Hydroxychloroquine (Plaquenil)
- Retinoids
- Cyclosporine
- Cyclophosphamide
- Azathioprine
- Etanercept (Enbrel)
- Mycophenolate mofetil (CellCept)
- Methotrexate
27 y.o. with 1 1/2 year history of vulvar irritation. History of genital herpes. PMH significant for hypothyroidism.
She had tried multiple agents for her condition including topical steroids, Vagisil, antibiotics, and Diflucan. She used oral steroids but developed knee pain. Protopic has not helped.
MICROSCOPIC DIAGNOSIS:

1. Vulva, biopsy: Hyperkeratosis consistent with *lichen simplex chronicus*.
2. Right labium majus, biopsy: Scar with overlying and adjacent *lichen simplex chronicus*.
3. Vulva, left bottom, biopsy: Excoriation with *lichen simplex chronicus*. 
Skin biopsy: Site: Vulva; Lesional status: Lesional: No evidence for pemphigus/pemphigoid. Occasional cytoid bodies suggestive of lichen planus.

Your diagnosis is?

- A Lichen planus
- B Pemphigoid
- C Molluscum
- D Severe contact dermatitis
Which agent has she reacted to?

A. Diflucan
B. Topical steroids
C. Vagisil
D. Antibiotics
Vulvar pain related to a specific disorder

- **Infectious** (e.g. candidiasis, herpes, etc.)
- **Inflammatory** (e.g. lichen planus, immunobullous disorders, etc.)
- **Neoplastic** (e.g. Paget’s disease, squamous cell carcinoma, etc.)
- **Neurologic** (e.g. herpes neuralgia, spinal nerve compression, etc.)

Vulvodynia

- **Generalized**
  - **Provoked** (sexual, nonsexual, or both)
  - **Unprovoked**
  - **Mixed** (provoked and unprovoked)

- **Localized** (vestibulodynia, clitorodynia, hemivulvodynia, etc.)
  - **Provoked** (sexual, nonsexual, or both)
  - **Unprovoked**
  - **Mixed** (provoked and unprovoked)
8.3% of women have vulvodynia


**Diagnosis of Vulvodynia**

Define disease
- Q-tip test
- Vulvoscopy?
- Duration of pain
Generalized

Localized
Conditions to Rule Out

- Yeast (often cyclic vulvar pain)
- Desquamative inflammatory vaginitis (DIV)
- Atrophic vaginitis
- Pudendal nerve entrapment

Candidiasis
Desquamative Inflammatory Vaginitis

Atrophic Vaginitis
Pudendal Nerve Entrapment

Theories on Etiologies

- Embryologic derivation
- HPV
- Oxalates
- Hormonal changes

- Chronic inflammation
- Altered immuno-inflammatory process
- Nerve pathways
Proposed neuroimmunological mechanism of the allodynia/hyperpathia of vulvodynia

potentially inciting factors:
- infections
- irritants
- toxins
- medications
- other

increased proinflammatory cytokines:
- IL-1, IL-6, IL-8
- IFN-α
- TNF-α

nerve growth factor increased

allodynia and hyperpathia

mast cell accumulation

substance P

CGRP

increased

neuromodulation with sacral nerve stimulator

modulation of efferent signals to spinal cord

refractory pain in distribution of specific nerve root (S3 or S4)
Sacral Neuromodulations for Femal Lower Urinary Tract, Pelvic Floor, and Bowel Disorders


Physical Therapy: Initial Evaluation for Patients with Vulvodynia

- Detailed H&P
- General physical therapy assessment to evaluate ROM, LE strength and flexibility, reflexes, sensation and pain
- External vaginal assessment: sensation, cotton swab test, pelvic floor contractions and reflexes
- Internal vaginal assessment: muscle tone and strength, trigger points
- At times, biofeedback testing for objective pelvic floor strength measurement
- Pt education on posture, use of appropriate tools including dilators, vaginal cones, lubrication
What is the name of this muscle?
A. Ischiocavernosus
B. Bulbocavernosus
C. Pubococcygeus
D. Levator ani

sEMG Biofeedback

• Glazer protocol
• 60 seconds rest (pre)
• Quick contractions 5-10 with 10 sec rest
• Static Endurance holds: 10 sec hold/10 sec rest
• 60 second static hold
• 60 seconds rest (post)
• External electrodes or vaginal sensor
  – Can also use rectal sensor for extreme vaginal pain
Relax and Release

The Glazer Protocol: Evidence-Based Medicine Pelvic Floor Muscle (PFM) Surface Electromyography (SEMG) (urinary incontinence)

Transcutaneous Electrical Nerve Stimulation

Transcutaneous electrical nerve stimulation (n=80)
• Double-arm randomized placebo-controlled trial
  – Twice weekly for 20 sessions
    • Used dual channel vaginal device
• Results
  – Significantly improved VAS and SF-MPQ scores (p<0.004 and p<0.001, respectively) as well as Marinoff dyspareunia scales and FSFI
  
Murina; BJOG, 2008

Other Manual Work

• Joint mobilizations for hypomobility
• Scar Mobilization
  – Episiotomy/Vaginal Tear
  – s/p laparoscopy
  – Pfannenstiel
• External Soft Tissue Work
  – Obturator
  – Piriformis
  – Iliopsoas
  – Adductors
  – Quadratus lumborum
  – Abdominal muscles
Therapeutic Activities

• Home Manual Work
  – Will teach partners to assist with manual therapy techniques

• Dilators
  – 1x/day
  – Goal is for 10 minutes
  – Once able to tolerate #3 for 10 minutes static, begin dynamic work (2 minutes in/15 sec out)
Hart’s line

JOL DeLancey, MD collection

Vestibulectomy

Video
Surgery for Localized Vulvodynia Success Rate of 50%–90% in Previous Studies


Psychosexual Correlates of Persistent Postsurgical Pain in Patients with Vulvodynia

- 8 (22%) completely pain free after surgery at an average of 8.7 years after vestibulectomy
- 29 (78%) reported various levels of pain during intercourse
- ? Psychological distress

Evaluating Patients with Vulvar Diseases is a Team Approach

The Human Dimension