VULVAR DISEASE UPDATE

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NEOGS, April 27, 2011

Conflicts of interest

Honorarium from Taro

Little evidence based treatment
Too few studies done in vulvar disease.

Most treatments discussed are "off-label"

Why Do Vulvar Disease ?

VULVAR CARE IS COMMONLY UNAVAILABLE

For women this is devastating

- **suffer** with undiagnosed symptoms
- **waste** millions of dollars on anti-yeasts
- **hide** and scratch
- **endure** vulvar pain and dyspareunia
- **are desperate** for help

VULVA !

What is that?

Down there?

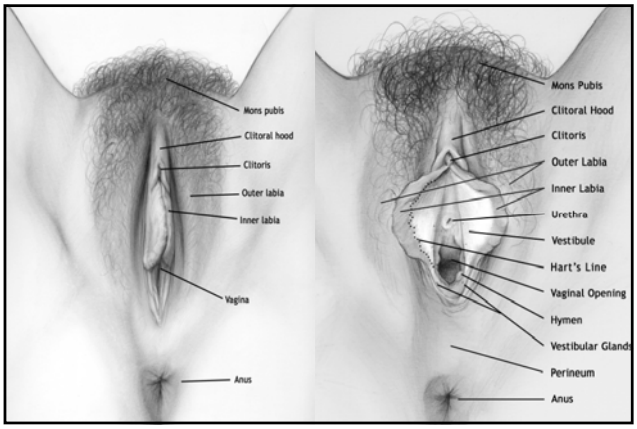
Vulvar Education

Lets eliminate the

"Down there" generation

Use diagrams and handouts

See www.issvd.org - patient education



BASIC PREMISES

- Patient anxiety and depression
- Normal anatomy can be confusing
- Look carefully for subtle abnormalities

Punch Biopsy

Pre-anesthesia -

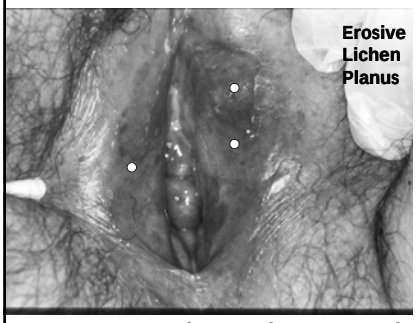
use a topical anesthetic

- **2.5% prilocaine / lidocaine cr**
- **5% lidocaine oint**
- **15 min under plastic wrap**

Use a punch, shave or scissor biopsy

Stop bleeding:

Monsel's or ferric chloride
5 or 6-0 Vicryl suture



Biopsy –
Site ○
Number 1-3
size 4 mm
H&E + IF

Any question – do more than 1 biopsy

Herpes Simplex Virus (HSV)

Commonest cause of vulvar erosions / ulcers

Primary HSV is uncommon
HSV I increasing
Reversal of HSV II/I ratio - for new cases 70-80% HSV I
Usually spread from an asymptomatic partner

Women are unaware of their infection and most often present with recurrent HSV with no primary HSV history

Herpes Simplex Virus (HSV)

Think of HSV in a patient with a non-healing genital very painful ulcer
Onset as erosions that deepen and extend with punched out ulcers or ulcerated nodules.

Can be treatment resistant in HIV

Diagnosis –
Tsank smear, PCR, culture,
Type-specific serology, biopsy

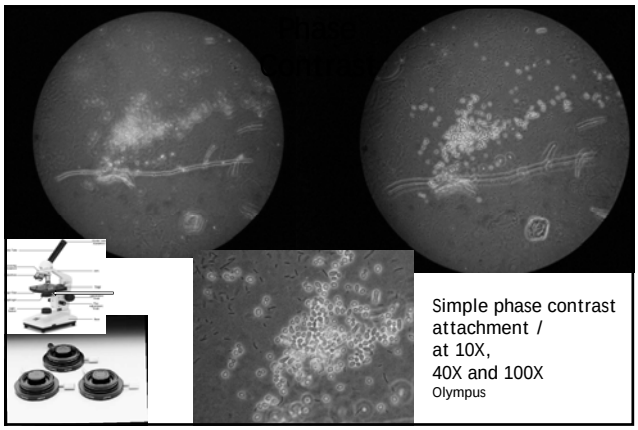
Candidiasis

Candidiasis is the commonest genital disease

Causes:	Candida albicans	75%
	Candida tropicalis, etc.	25%

NO TELEPHONE DIAGNOSIS

Candida can complicate all vulvar problems
e.g. LP, LSC, LS, Contact



Simple phase contrast attachment / at 10X, 40X and 100X
Olympus

Treatment Candidiasis

Topical imidazole cream or vag tabs– 1, 3, 7d
nystatin cream or vag tabs
Oral imidazole - fluconazole 150 mg on day 1, 3, 7
Suppression
clotrimazole 500g vag tab weekly or 200 mg twice a wk
fluconazole 150 - 200 mg orally weekly
ketoconazole 100 mg orally daily
itraconazole 100 mg orally daily
Resistant Candida
boric acid vag suppositories 600mg X 14 d

Vulvar Patients are desperate! with Itch , burn , pain

They try to :
"**Wash it away**" and "**Clean up**"
the **dirty** area
using
Soaps, cleansers, Anesthetics, Yeast Rx

Vulvar Contact Dermatitis

Primary irritant :
Prolonged or repeated exposure to caustic or physically irritating agent
This is a "chemical burn"

Very common with ALL vulvar problems
Causes:
Hygiene habits – soap, wipes, pads
Moisture - urine, feces, sweat
Topicals – lotions, antifungals

Treatment Vulvar Contact Dermatitis

Stop Contact – Irritant or Allergen

- Stop irritants
- Stop scratching
- Patch Test as indicated
- Educate patient
- Treat infection – yeast, bacteria

Control inflammation

- triamcinolone 0.1% oint twice a day for 7-10 d
- If severe, systemic corticosteroids

Vulvar Contact Dermatitis

Frequent

Complicates all vulvar conditions

Irritant contact most common

Skin barrier lost from soaps,urine,feces

BEWARE THE "DIRTY" VULVA

Psoriasis

In genital area – often atypical
moist, thin red patches in skin folds, gluteal cleft
In hairy areas – red, scaly papules, plaques
Secondary changes with infection, common
– fissuring, pustules

Often missed or hidden

Please Ask

Psoriasis Treatment

Stop irritants

Treat infection – yeast and bacteria

Stop inflammation

– topical steroids – triamcinolone 0.1% ointment am pm 5-7d then
- tacrolimus 0.1% ointment or pimecrolimus 1% cream
- if severe, systemic medications

Lichen Simplex Chronicus (LSC)

End stage of the cycle

Itch ⇨ Scratch ⇨ Itch

Worse with heat, humidity, stress and irritants

Associations: atopic dermatitis, psoriasis, contact dermatitis

Scratching feels so good

Characteristics of Lichen Simplex Chronicus

Relentless pruritus	dyspigmentation
"nothing helps"	excoriations, crusts
"years of itch"	lichenification
uni or bilateral	hair loss

The diagnosis is clinical

Treatment LSC

Optimize Epithelial Barrier function:

Control infection (cefadroxil, fluconazole)
Reducing heat, sweat, irritation
Stop irritants - Stop excessive hygiene

Immediate therapy:
Tap water soaks in tepid water
Use cool packs or compresses to deaden nerves
No hot water - No ice packs
Seal in moisture with ointments long-term

Treatment LSC

Confirm diagnosis – biopsy ?
- Educate patient
- Patch Test as indicated

Stop Itch-Scratch-Itch cycle

- Cool sitz baths/gel packs
- Sedate – doxepin or hydroxyzine PM, citalopram AM
- Topical superpotent steroids - clobetasol 0.05% oint bid x 2 wks, OD x2 wks, MWF x 2 wks
- Severe – prednisone taper or IM triamcinolone 1mg/kg up to 80 mg/dose

Look for more than one cause

Treatment Tips LSC

If water stings raw skin:
Normal saline Sitz baths:
1tsp salt in 3 cups of water
Soak for 5-7 min 2-3 times a day

For recurrent infection:
Swab skin folds and nose for C&S - R/O MRSA, Candida

Bleach Baths – 3 times a week
½ cup bleach in 10" water for 5-7 min
1 ¼ teaspoons of bleach per gallon (4 liters) of water

Lichen Sclerosus

A chronic, autoimmune, mucocutaneous disease affecting the genital skin causing whiteness, tissue thinning and scarring affecting perimenopausal women 40-50 yrs

Commonest cause of chronic vulvar disease
Prevalence 1:300 – 1:1000
670 cases in Boston – 6,600 in metropolitan area

15% cases children
15% have extragenital disease

3-5% ⟹ SCC

Murphy R. Lichen sclerosus. Dermatol Clin 2010 Oct;28(4):707-15.
Neill SM, Lewis FM, Tatnall FM, Cox NH. British Association of Dermatologists' guidelines for the management of lichen sclerosus 2010. Br J Dermatol 2010 Oct;163(4):672-82.

Lichen Sclerosus Treatment

Confirm diagnosis – biopsy

- Stop irritants
- Stop scratching
- Educate patient
- Treat infection – yeast, bacteria

Control inflammation

- clobetasol or halobetasol 0.05% oint once or twice a day for 3 months then maintenance 1-3 times a week
- If very thick, consider intralesional triamcinolone

Not responding?
Reassess, rebiopsy, R/O SCC, contact

Follow forever

Lichen Sclerosus
Alternate Treatments

Calcineurin inhibitors:
pimecrolimus 1% cream bid
tacrolimus 0.1% ointment bid
Antimalarials – hydroxychloroquine 200mg bid
Methotrexate 10 -15 mg/ week po or sc + folate 1 mg/d
Cyclosporine 4-5 mg/kg/d

Compliance issues children and teens – 2.5% HC oint daily

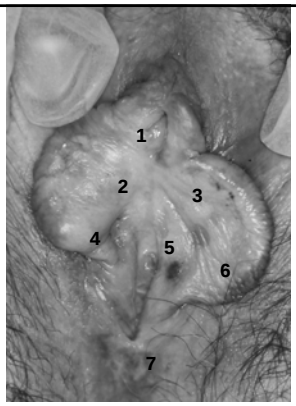
Lichen Sclerosus

For very thick poorly responding LS

Prednisone for short 2-3 week course

Intralesional triamcinolone
3.3-10 mg/ml after topical anesthetic
Monthly for up to 3 months

Sites of Injections of T/C 2.5 to 10 mg/ml



Before RX Topical 2.5% lidocaine + prilocaine for 20-30 min

Update on intralesional steroid: focus on dermatoses. Richards RN.J Cutan Med Surg. 2010 Jan-Feb;14(1):19-23.

MANAGEMENT PRINCIPLES

- **Explanation of the disease process, treatments, expectations**
- **Handouts**
- **Photographs**
- **Treat all factors**

MANAGEMENT PRINCIPLES

- Anticipate and minimize iatrogenic disease (yeast, irritant contact dermatitis)
- Avoid cream vehicles on painful, inflamed, or estrogen-deficient vulvar skin
- Avoid topical therapy in general and use oral medications, except for corticosteroids
- The vulva is relative steroid resistant – use ultrapotents

Lichen Planus

An autoimmune, mucocutaneous disorder of altered cell mediated immunity in older women - 50 – 60 years

Affects - Skin, scalp, nails
Mucous membranes - oral, genital, anus
esophageal, urinary tract

Responds to immunosuppressive therapy

2-5% ⟹ SCC

Always Examine the Vulva & Vagina & Mouth

Lichen Planus Vulvar Patterns

60% itchy 70-75% painful

Classic –
White, lacy, or fernlike topped papules

Hypertrophic -
Extensive white, thick scarring, destruction
Very itchy
Looks like LS

Erosive –
Red plaques with whitish to lacy edges
Glazed erythema, erosions, ulcers, scarring

Variable morphology

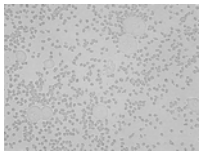
Vaginal Lichen Planus

In the vagina: may be asymptomatic

LP causes **Desquamative Inflammatory Vaginitis DIV** with:

- Pain
- Erosions
- Synechiae
- Dyspareunia
- Dysuria
- Stenosis/ Scarring
- Discharge

- Mucopurulent vaginal discharge – yellow, green, bloody



Always examine vagina

Erosive Vulvovaginal LP

Pain plus burning

Deep red erosions, glazed erythema with thin gray edge

Fern-like or lacy white pattern

Variable scarring / loss of architecture

Examine :
Vulva + Vagina + Anus + Mouth + Skin

Lichen Planus Treatment

Confirm diagnosis – biopsy

- Stop irritants
- Stop scratching
- Educate patient
- Control infection

Control inflammation

clobetasol or halobetasol 0.05% oint

intralesional, vaginal or systemic corticosteroids

topical tacrolimus (Protopic) 0.03%, 0.1% oint - burns

2 mg suppositories qhs

Corticosteroids for Lichen Planus

Systemic Corticosteroids: Use for severe LP

Triamcinolone 1 mg/kg up to 80 mg/dose IM

(Kenalog 40®) q4wks x 3

Prednisone 40-60 mg PO OD, decreasing dose

Intravaginal Hydrocortisone acetate QHS:

- suppository 25mg (available) or 100 mg (compounded)
- 10% compounded vaginal cream, 4 - 5g per dose (400-500mg/dose)

Use nightly for 14 days then Mon Wed Fri **use dilators**

Note: adrenal suppression and candidiasis

Lichen Planus Treatment

SEVERE

Systemic corticosteroids – Triamcinolone 1mg/kg IM every 4 weeks for 3-4 shots

Methotrexate 5-10mg/week PO or SC

Cyclosporine 4-5 mg/kg/d –3-4months

Mycophenolate mofetil 500mg -1.5 g bid

Etanercept 50 mg 1-2 doses / week ?

Hydroxychloroquine 200 mg bid

Acitretin (1 mg/kg)

The Lichens

LS	LP	LSC
Itch or burn	Itch or burn	++++ Itch
Scars	Scars	No Scar
Not in Vagina	In Vagina And Mouth	Not in Vagina

Causes of Treatment Failure

1) **Noncompliance**
   - **poor education**
   - Fear of topical steroids
   - **Limited mobility**

2) **Incorrect diagnosis** – was a biopsy done ?

3) **Associated problems**

LS, LP, contact, Candida, HSV, estrogen loss, SCC

Mucous Membrane Pemphigoid

Rare mucocutaneous scarring & blistering disease

Autoimmune – auto-antibodies to epidermal basement membrane

Sites:

mouth 85% **vulva 50%**

eyes 65% skin 25%

Non-healing erosions vulva LP , LS, Pemphigus
- desquamative vaginitis
- scarring

Diagnosis : histopathology and immunofluorescence

Vulvar Squamous Cell Carcinoma

Commonest vulvar malignancy

85-90% of all vulvar cancers

Classification - Intraepithelial
- Invasive

VIN and Vulvar SSC

Intraepithelial

Vulvar Intraepithelial Neoplasia (VIN 2-3, high grade VIN)

VIN, usual type **multifocal** - HPV 16, 18
VIN, warty type - in younger women
VIN, basaloid type - looks like condylomata
VIN, mixed (warty/basaloid) type

VIN, differentiated type **solitary** - in older women, associated with LS

Invasive SCC - in women > 65 yr.
- up to 40% - lichen sclerosus

Vulvar SCC

30-40% vulvar SCC occur in lichen sclerosus

Lichen sclerosus and lichen planus can develop SCC in 3-4% cases

Causes of Vulvar Ulcers in North America

Infections

Venereal

Herpes simplex

Syphilis

HIV

Non- Venereal

EBV

M.pneumonia

Dermatoses – Non-bullous

Aphthosis

Lichen planus

Lichen sclerosus

Crohn's disease

Contact

Drug

Tumors

Squamous cell carcinoma

Vulvar Aphthous Ulcers

"Canker Sores" on the Vulva

- Average age is 14 (9-19) yrs
- Sudden onset
- Usually multiple, painful, well demarcated punched-out ulcers
- Size: most <1cm; can be 1-3 cm
- Prodrome - flu-like with mild fever, headache, malaise
- Duration 1-3 weeks, can last months
- One episode, less common recurrent
- Past history of oral aphthae – canker sores
- Not Behcet's

Hupper et al.J Ped Adolesce Gynrcol 2006;195-204
Farhi et al. Arch Dermatol 2009;145;38-45

APHTHAE – Acute vs Chronic

Acute (more common)

– usually a prodrome - fever, headache, malaise, GI upset
- EBV, Mycoplasma pneumoniae, viral upper respiratory infection or gastroenteritis, influenza, Strep

Recurrent / Complex (recurrent oral and genital aphthae)

Inflammatory Bowel disease - Crohn's, Ulcerative colitis, Celiac disease
Behcet's disease
Medications – cytotoxic, NSAIDs
Myeloproliferative disease, cyclic neutropenia, lymphopenia
HIV

Vulvar Aphthae

Diagnosis

Diagnosis of exclusion
Look for associated conditions

Cultures negative, biopsies non-specific and blood work non-contributory

DDx: HSV, EBV, drug, Crohn's, Behcet's, Syphilis, HIV

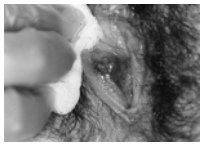
Vulvar Aphthae – Therapy

- **Pain control** – topical, systemic
- **Prednisone** 40 – 60 mg each morning until pain resolves (3-5 days, then ½ dose 3-5 days)
  - ultrapotent corticosteroid
- **Educate** -Most often a one-time event, can recur

If not controlled:

Intralesional triamcinolone (Kenalog 10) 5-10 mg/ml
colchicine 0.6 mg bid-tid if tolerated
dapsone 50-150 mg per day
dapsone + colchicine
pentoxyfylline 400 mg tid
thalidomide 100-150 mg per day

Epstein-Barr Virus (EBV)



Lymphotropic herpes virus

Age - teens to early 20s

Clinical
- acute onset
- fever, malaise
- sore throat – lasts 2 to 3 weeks
- punched out painful ulcer(s)
- recent hx of mono in family member

Serology -EBV IgM anti-VCA antibody

Vulvar Behcet's Disease

Rare

Triad - oral ulcers, genital ulcers, uveitis

Clinical -
painful, punched-out vulvar ulcers
- last 2-3 wks
- heal with scars, sinuses
- erythema nodosum
- minor to no eye problems

Good prognosis

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" ulcers
- Non-specific
  - aphthae – oral and vulvar

Drug Eruptions

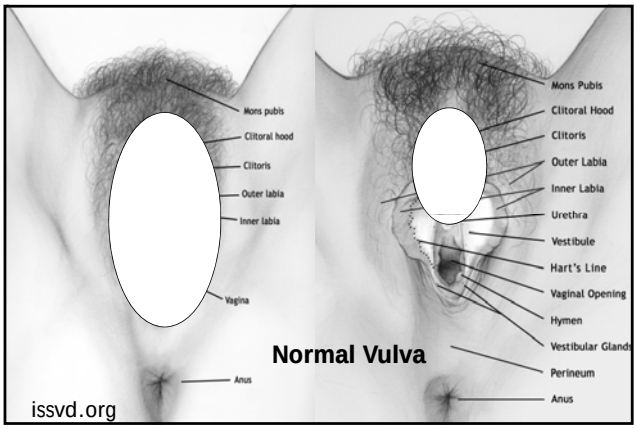
Drugs can cause 100 different skin reactions
Many can be seen on the vulva
Erosive/ulcerative types –
erythema multiforme
aphthous ulcers
Pemphigoid
fixed eruptions,
LP and LE eruptions,

Topical Treatment Instructions

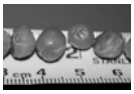
Give specific instructions for topicals

Amount to use
- show her, in the office, how much to use
- squeeze onto your finger, and demonstrate

Application site
- some women have never seen their vulva
- use a mirror, and show the exact place(s)
- give her a diagram with correct areas indicated
  www.issvd.org Patient Education



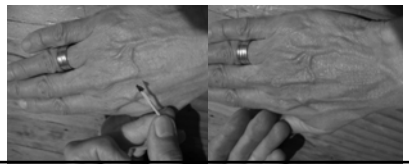
How much to apply ?



"PEA SIZED" AMOUNT ?

"TOOTH PICK" Dermatology

The amount of ointment or cream on the large end of a toothpick will cover the vulva



Commonest Missed Concurrent Vulvar Diseases

Candidiasis
Contact Dermatitis
HSV
Atrophy
Cancer

LOOK FOR MORE THAN ONE PROBLEM

Treat Vulvar Disease

Listen – review history
Biopsy and RE-BIOPSY
Look for
- infection – Candida, HSV, bacteria
- trauma from aggressive hygiene or other practices
- contact dermatitis
- squamous cell carcinoma

Educate
Support – counseling as needed
Assess compliance

LOOK FOR MORE THAN ONE PROBLEM

Vulvar Disease Treatment Handout

Handout : **Your Diagnosis Is? Test Your Knowledge of Vulvovaginal Disorders**

Available at:
http://www.med.umich.edu/obgyn/cvd/ref_phys.htm
Click on Summary of Vulvovaginal Diseases

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Vulvar Disease References

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See Dermatologic Therapy Diagnosis and treatment of vulvar and Vaginal Disorders Vol 23 Oct 2010
Dermatologic Clinics Vulvovaginal Dermatology Vol 28 Oct 2010