Vulvar Disease: What Do You Know? An Overview

Patients who itch

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DISCLOSURE OF RELEVANT RELATIONSHIPS WITH INDUSTRY

F084-Vulvar Disease: What do You Know? An Overview
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Vulvar Lichen Sclerosus

Nomenclature

• Kraurosis Vulvae
• Vulvar Dystrophy
• White spot disease
• Guttate scleroderma
• Lichen sclerosus et atrophicus

Vulvar Lichen Sclerosus epidemiology

- Prevalence unknown – asymptomatic
- Margesson – 1/300 to 1/1000
- Age onset: girls mean 5.4 yrs; women mean 55.1 yrs; Margesson women 30-40
- In women: 78% post menopausal

Vulvar Lichen Sclerosus: epidemiology

- 28% of pts with other autoimmune dz:
  - Alopecia areata
  - Vitiligo
  - Thyroid dz (most common)
  - Pernicious anemia
  - Morphea
- Family hx: 12% (likely underreported)
- ? Asst HLA DQ7, DQ8, DQ9

Vulvar Lichen Sclerosus - clinical

• Signs – porcelain white papules/plaques; echymoses/purpura; “cigarette paper”; hyperkeratosis; ulceration; fusion of labia minora to majora; “phimosis” of clitoral hood (“burying of the clitoris”); “Closures of the commissures” (vestibule smaller – like zip closed from under clitoris)

• 70% of women with some scarring

Vulvar Lichen Sclerosus - clinical

• Symptoms – pruritus, irritation, burning, dyspareunia, tearing; constipation in girls; can be asymptomatic


Vulvar Lichen Sclerosus: clinical

• Distribution – 59% vulvar and perianal (figure of eight); vagina spared

• Extranagenital 11-13%
  (exclusively extragenital: 6%)

• Rate of malignancy (SCCa)
  about 5 %

Vulvar Lichen Sclerosus: clinical

Hyperpigmentation:

• vulvar lentiginosis:
  • can be dark and irregular
  • on bx clearly not melanoma

• nevi in LS:
  • clinically suspicious for melanoma
  • histologically suspicious for melanoma
  • tell pathologist in setting of LS

Lichen Sclerosus: pathology

• Pathognomonic changes: in papillary dermis homogenization & sclerosis with atrophy of epidermis (not always present, esp in early lesions); bx center of white lesion

Lichen Sclerosus: pathology

• Lymphocytic, lichenoid interface dermatitis in LP and LS
• Elastic stain – marked decrease in papillary dermal elastic fibers in LS c/w LP

Vulvar Lichen Sclerosus: etiology

- Unknown – thought to be autoimmune
- Sera of pts with auto antibodies:
  - 67% with antibody to extracellular matrix 1 protein
  - 30% with antibody to BP180 and BP 230
- Expert opinion – antibodies secondary; likely T cell mediated disease

Vulvar Lichen Sclerosus: diagnosis

- Biopsy (except in prepubertal child) or clinical
- Biopsy may not be diagnostic
  - Rebiopsy
  - Response to treatment
  - Look for other diseases

Vulvar Lichen Sclerosus

Treatment

• Ultra potent topical steroids:
  • 96% partial-complete relief; (65% women sx free)
  • 23% return to normal skin exam
• If not responding, consider other diagnosis

Vulvar LS: Treatment

- Tacrolimus/pimecrolimus
  - 50-94% at least partial improvement
- Pts failed topical steroids
- Burning/itching week(s); ?increase risk of SCCa
- Other: retinoids, MTX, cyclosporin, cryotherapy, PDT

Vulvar LS: Treatment

- Intrallesional steroids (esp. very hyperkeratotic)
- Follow up q 6-12 months screening exam

Vulvar Lichen Sclerosus: treatment

• Never totally stop treatment
• Lee et al:
  • suggest treatment 3 times/week
  • lower strength of topical steroid if improved
  • 0/357 compliant patients with SCCa/VIN
  • 7/150 partially compliant patients SCCa/VIN
• Molecular mechanism of LS associated SCCa – possibly down reg of IRF6 tumor suppressor gene

Lichen Simplex Chronicus

epidemiology

- “probably the single most common symptomatic condition involving the anogenital area”

Lichen Simplex Chronicus clinical

- “itch-scratch cycle”
- Lichenification on exam
- Erythema, scale
- Can have hyper- or hypopigmentation

Lichen Simplex Chronicus: etiology

• May be part of atopic dermatitis with genetic predisposition
• Heat/sweat/irritants may trigger
• Stress?

Lichen Simplex Chronicus: diagnosis

Underlying conditions:
• Contact dermatitis
• Seborrheic dermatitis/Psoriasis
• Lichen sclerosus
• Candidiasis

Lichen Simplex Chronicus: treatment

• Reduce triggers
• Repair barrier - lubrication
• Reduce inflammation- topical steroids
• Break itch-scratch cycle – sedating anti-histamine q HS
• Psych factors?

Candidiasis: epidemiology

- *Candidiasis* = *moniliasis*
- Common (75% of women at least once)
- Any age; premenopausal > post
- Candida albicans most common
- Other: C. tropicalis and C. glabrata


Candidiasis: clinical

- Acute: itching/burning; curdy discharge; erythema/edema/erosion; satellite pustule
- Recurrent and chronic: itching; +/- d/c
- Eczematous: LSC like
- Recurrent fissuring: interlabial sulci; peri-clitoral; erythema; maybe no d/c
- Cyclic vulvovaginitis: burning/itch 5-10d/mo

Vulvar Fissures

- Infections
  - Candida
  - Beta hemolytic streptococcus
  - Staphylococcus aureus

- Inflammation
  - Dermatoses: atopic, contact, seb derm
  - Lichen Sclerosus
  - Desquamative inflammatory vaginitis

Vulvar Fissures

• Infections
  • Candida
  • Beta hemolytic streptococcus
  • Staphylococcus aureus

Pediculosis pubis

- Can do “in vivo” exam with dermatoscope to see lice and nits

Pediculosis Pubis - Clinical

• Itching red papules
• Visualize lice/nits – pubic/perianal; legs, chest, forearms; eyelashes, eyebrows, beard, axillary
• Maculae cerulae – blue macules on thighs or abdomen

Pediculosis Pubis - treatment

- work up for other STIs
- wash clothing and bedding
- treat sexual partners

Pediculosis Pubis - treatment

• Treat all affected areas with:
  • malathion 0.5% lotion on dry hair for 12 hours; repeat in 1 week
  • permethrin 5% cream on wet hair; wash off in 10 minutes; repeat in 1 week
  • eyelashes:
    • permethrin 5% for 10 minutes (close eyes)
    • vaseline BID for 8-10 days