The Vulvar Lichens
Lichen Sclerosus

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DISCLOSURE OF RELATIONSHIPS WITH INDUSTRY

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Forum F106: Vulvar Disease: What to you know?
The Vulvar Lichens: Lichen Sclerosus
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DISCLOSURES

Proctor and Gamble: member of Scientific Advisory Board
UpToDate: Royalties

Vulvar Itching

• Patient complaining of itch and some pain
• Present for a few years
• Tried monistat but did not help
• Clitoral phimosis; labia minora absent; white patches

Vulvar Lichen Sclerosus
Nomenclature

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


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 epidemiology

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

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);


Vulvar Lichen Sclerosus: clinical

• Distribution – 59% vulvar and perianal (figure of eight); vagina spared
• Extragenital 11-13% (exclusively extragenital: 6%)
• Rate of malignancy (SCCa) about 2-5 % as high as 6.7% (Bleeker)  

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

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
- Another study: 4/149 had BP180 antibody (not significant)
- Expert opinion – antibodies secondary; likely T cell mediated disease

Vulvar Lichen Sclerosus: diagnosis
- Biopsy (except in prepubertal child) vs 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

Vulvar LS: Treatment
- Follow up q 6-12 months screening exam
- Never totally stop treatment
Vulvar Lichen Sclerosus: 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