DISCLOSURE OF RELEVANT RELATIONSHIPS WITH INDUSTRY

Vulvar Inflammatory Dermatoses

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I do not have any relevant relationships with industry.
Vulvar Inflammatory Dermatoses

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Terminology

- **Chondrodermatitis Nodularis Helicis:**

  A benign tender nodule in the cartilaginous portion of the ear “helix” (thus helicis) on biopsy shows inflammation involving dermis and cartilage.

- **Chondrodermatitis nodularis chronica antehelicis**
Basic Patterns in Dermatopathology
Variation in Size & Shape: beak, body, wing & tail
Superficial dermatitis

Nodular dermatitis

Vesicular dermatitis

Sup & deep dermatitis

Diffuse dermatitis

Vasculitis
NODULAR AND DIFFUSE DERMATITIS

Nodular dermatitis
- Lymphocytes predominate
  - Lymphocytes mostly
    - Lymphocytoma cutis (CLICK HERE to read more about this disease)
  - Abnormal lymphocytes monopolize, at least in loci
    - Lymphoma (Neoplastic disease that simulates an inflammatory disease.)
  - Collections of epithelioid histiocytes
- Neutrophils predominate
- Neutrophils, nuclear "dust" of neutrophils, eosinophils, and plasma cells
- Eosinophils often
- Histiocytes predominate

Diffuse dermatitis
- Lymphocytes predominate
- Abnormal lymphocytes predominate, at least in foci
- Neutrophils predominate
- Neutrophils, nuclear "dust" of neutrophils, eosinophils, and plasma cells
- Eosinophils and plasma cells in addition to
  - Plasma cells prominent
  - Mast cells monopolize
- Abnormal leukocytes
- Histiocytes predominate
- Langerhans' cells predominate
Biopsy Type

- Punch biopsy
- Shave biopsy
- Deep incisional biopsy
- Complete excision
- Curettage
Shave Biopsy
Shave Biopsy

- Raised lesions that are clearly benign
- Pathology confined to the epidermis
  - SK, AK, tags, warts, superficial BCC & SCC
- Not for melanomas
- No sutures
Punch Biopsy

- Inflammatory lesions
- Combined diagnostic and therapeutic
- Multiple biopsies
- Prognostic biopsy: leprosy, MF, etc.
- Prerequisites (lidocaine & consent)
Lupus

Fixed Drug
Rudimentary Information (5Ds)

• Description
• Demographics
• Duration
• Diameter
• Differential Diagnosis
Ancillary Tests

- Light microscope
- Immunohistochemistry
- Immunofluorescent studies
- Electron microscopy
- Molecular biology

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<th>SPECIMEN HANDLING</th>
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<td>Routine microscopy</td>
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<tr>
<td>Direct immunofluorescence</td>
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<td>Immunoperoxidase</td>
<td>Formalin, fresh* or Michel’s medium</td>
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<td>Culture for bacteria, mycobacteria or fungi</td>
<td>Fresh* or minced in sterile culture/carrier medium appropriate for organism (usually performed by laboratory)</td>
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<td>Culture for viruses</td>
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The International Society for the Study of Vulvovaginal Disease (ISSVD)
The International Society for the Study of Vulvovaginal Disease (ISSVD)

- Facilitate the exchange between clinician and pathologist
- Often “diagnosis” on a biopsy result is not the name of disease, but rather a description of the microscopic findings “Pattern”
Modified version of the 2006 ISSVD classification of vulvar dermatoses: pathologic subsets and their clinical correlates

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Spongiotic Pattern
Spongiotic Dermatitis

- Microvesicular + Lym
- Microvesicular + Eos
- Microvesicular + PMNs
- Slight Spongiosis
Dermatophyte
Spongiotic Pattern

- **Eczematous dermatitis** (atopic, contact, and allergic)
- All ages
- Difficult to determine clinically the underlining cause
- Histological features similar regarding the etiology
Atopic Dermatitis

• The most endogenous dermatitis
• History of atopic diathesis (eczema, asthma, allergic seasonal rhinitis)
• Decrease with age
• Histology identical to other spongiotic dermatitis
Acanthotic/Psoriasiform Pattern
Acanthotic/Psoriasiform Dermatitis

Irregular

Regular

[Images of histological sections showing irregular and regular patterns]
Acanthotic Pattern

Lichen simplex chronicus
Psoriasis
Lichen Simplex Chronicus/ Prurigo Nodularis
Lichen Simplex Chronicus

- Common lesion
- Repeated rubbing &/or scratching
- Usually solitary
- thickened and erythematous scaly plaque.
- Labia majorus
- Biopsy not necessary
Lichen simplex chronicus
Psoriasis
Psoriasis

• Multifactorial chronic relapsing dermatosis
• Diagnosed in 5% of women presenting to dermatologist with persistent vulvar symptoms
• Most of the patients have extra-genital manifestations
• Koebner phenomenon in 20% of the patients.
• Hair-bearing areas: mons pubis & labia majora
Psoriasis
Lichenoid Pattern

Lichen Sclerosus
Lichen Planus
Lichen Sclerosus
Lichen Sclerosus

- Common chronic vulvar dermatosis
- Idiopathic
- “et atrophicus”: no longer used.
- Occasionally associate with VIN
- Bimodal peak incidence: pre-puberty & menopause.
- Intense pruritic nature.
Lichen Sclerosus
Lichen Planus
Lichen Planus

- Chronic cell-mediated immune reaction
- Skin & mucous membranes
- Clinical variants: hypertrophic, atrophic, actinic, linear, zosteriform, bullous, etc.
- 50% of affected women have genital involvement
- Associations: Malignancies, UC, HCV & HBV
Vulvar LP

• Two types:
  1) Mucosal (vulvovaginal):
      - Erosive
      - Reticular (Wickham striae)
  1) Cutaneous:
      - Part of generalized eruption
      - Hypertrophic

• 1-3% of cases, longstanding erosive lichen planus can result in cancer
Vulvar LP
Vesiculobullous Pattern

Bullous & Cicatricial pemphigoid
Pemphigoid gestationis
Pemphigus vulgaris & vegetans
Mechanism

Spongiosis

Ballooning

Acantholysis
Vesicular Dermatitis

- Intraepidermal
- Subepidermal
- Intra/subepidermal
Location of the vesicle

- Suprabasal
- Intaspinous
- Subcorneal
Cell Rich infiltrate

Cell Poor infiltrate
Bullous & Cicatricial pemphigoid
Bullous & Cicatricial pemphigoid

- Subepidermal autoimmune bullous disorders
- Antibodies against hemidesmosomes & type IV collagen.
- BP: tense blisters, trunk & flexural areas, Nikolsky sign (-), no scarring.
- CP: Mucosa (conjunctiva & oral cavity), scarring, Nikolsky (+).
CP more commonly involves the vulva and lesions appear erosive
Pemphigus vulgaris & vegetans
Pemphigus vulgaris & vegetans

- Rare acquired immunobullous disorders.
- Antibodies to desmoglein 3
- Painful mucosal erosions and/or flaccid bullae
- Mouth, nasal or genital mucosa
Pemphigoid Gestationis
Pemphigoid Gestationis

• Seen in 2\textsuperscript{nd} trimester of pregnancy.
• Strong association with HLA-DR3 & DR4.
• Autoantibodies bind to 180-kDa hemidesmosomes Ag (BP\textsubscript{Ag}2).
• Pruritic erythematous macules \rightarrow blisters
• Keratinizing surfaces of vulva, pubic area & around the belly button
• Linear C3 at the BM.
Acantholytic Pattern

Hailey-Hailey disease
Darier's disease
Acantholytic dermatosis of the vulvocrrural area
Hailey-Hailey Disease

- Autosomal dominant acantholytic genodermatosis
- Mutation in a calcium pump “ATP2C1”
- Flexural or intertriginous areas
- DIF: Negative
Hailey-Hailey disease

Dilapidated brick wall
Darier Disease

- Uncommon genodermatosis
- Mutation on 12q: Calcium pump protein “ATP2A2”
- 50% of cases autosomal dominant
- Seborrheic distribution
Darier disease
Acantholytic Dermatosis of the Vulvocrural Area

• Identical to Hailey-Hailey or Darier disease
• Solitary lesion
• No family history
• DIF: negative
Granulomatous Pattern

Crohn’s Disease

Melkersson-Rosenthal Syndrome
Crohn’s Disease

• Cutaneous involvement in 20-40% of cases
• Four categories:
  1. Granulomatous cutaneous disease
  2. Oral changes
  3. Nutrition related changes
  4. Miscellaneous idiopathic markers
Crohn’s Disease

• Genital involvement frequent in pediatric population:
  1. Direct extension
  2. Metastatic CD
Crohn’s Disease

• 1/3 of women CD has GYN features
  ➢ Perianal Fistula 58-76%
  ➢ Vulvar ulcers 25%
  ➢ Knifelike ulceration
  ➢ Fissures
  ➢ Skin tags
  ➢ Swelling sinus tract
  ➢ Severe scarring
Crohn’s Disease
Vasculopathic Pattern

Aphthous ulcers
Behcet’s disease
Plasma cell vulvitis
Aphthous ulcers

• Mouth > genital region
• Idiopathic: immune complex triggered by local injury/infection
• Most patients with genital ulcers has concurrent or past oral ulcers
• Aphthae major: both genital and oral ulcers
• Patients with organ involvement: Behcet disease
Behcet Disease

• Clinical triad:
  1. Oral ulcers
  2. Genital ulcers
  3. Ocular inflammations

• Idiopathic: vasculitis & autoimmune response?

• Young adults
Behcet Disease

• Vulvar manifestations:
  - Deep ulcers
  - Fenestration and gangrene
  - Complex aphthosis “Aphtha Major”
  - Secondary lesions
Pathergy Lesions
Plasma cell vulvitis

- Zoon’s vulvitis
- Vulvitis circumspecta plasmacellularis
- Plasma cell orificial mucositis
- Idiopathic lymphoplasmacellular mucositis-dermatitis
Plasma Cell Vulvitis

- Postmenopausal women (8-80 y)
- Erythematous plaques “cayenne pepper spots”
- Inner face of labia minora & periurethral mucosa
- Symmetrical and deep
Plasma Cell Vulvitis

• **Differential diagnosis:**
  - Child abuse
  - VIN
  - Extramammary Paget disease
  - Fixed drug eruption

• **Biopsy is mandatory**
In Close

• “Diagnosis” on an inflammatory vulvar biopsy may not include the name of disease, but rather the microscopic “Pattern”.

• Seven patterns have been standardized by ISSVD.

• Clinical/histologic correlation.

• Biopsy type.
Thank you