Vulvar Inflammatory Dermatoses

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Road Map

- Terminology
- Basic patterns
- Appropriate biopsy
- Team work
- Vulvar inflammatory disorders
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
Normal skin

Superficial dermatitis

Sup & deep dermatitis
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)
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>Proposed laboratory test</th>
<th>SPECIMEN HANDLING</th>
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<td>Routine microscopy</td>
<td>10% neutral buffered formalin</td>
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<tr>
<td>Direct immunofluorescence</td>
<td>Michel's medium or fresh*</td>
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<tr>
<td>Immunoperoxidase</td>
<td>Formalin, fresh* or Michel’s medium</td>
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<tr>
<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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<tr>
<td>Culture for viruses</td>
<td>Viral transport medium (e.g. M4RT)</td>
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<td>Electron microscopy</td>
<td>Glutaraldehyde</td>
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Team Work Environment
How many players on a soccer team?

A. 10
B. 11
C. 12
D. 13
E. 14
How many players on a soccer team?

A. 10
B. 11
C. 12
D. 13
E. 14
How many players on a soccer team?

- Attendee
- Referee
- Standby referee
- Maintaining field
- Standby players
- Trainers
- ............
How many players on a soccer team?

Hundreds
How many players you need to issue a pathology report?

A. 3
B. 4
C. 5
D. 6
E. >6
How many players you need to issue a pathology report?

A. Patient’s colleagues at work
B. The patient’s friends
C. The patient's family
D. The patient’s doctor
E. Pathology receiving office
F. Histotech
G. Residents
H. Pathologist
I. Typist
Patient Samantha
Samantha

- Pruritic white patches on external genitalia for 5 months duration
Samantha

- She had a shave biopsy by her family Dr. and the case was received by a general pathologist.
After 3 days

- Please transfer it to our skin pathologist!!!
First Reaction
First Reaction
Patient Samantha
Lichen Planus
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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<td><strong>Acanthotic pattern</strong></td>
<td>Lichen simplex chronicus (primary and secondary)</td>
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<td>Psoriasis</td>
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<td>Reiter's syndrome</td>
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<td><strong>Lichenoid pattern</strong></td>
<td>Lichen sclerosis</td>
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<td><strong>Vesiculobullous pattern</strong></td>
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<td>Cicatricial pemphigoid</td>
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<td>Pemphigoid gestationis</td>
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<td>Pemphigus vulgaris</td>
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<td>Pemphigo vegetans</td>
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<td><strong>Acantholytic pattern</strong></td>
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<td>Acantholytic dermatosis of the vulvocrural area</td>
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<td>Plasma cell vulvitis</td>
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Spongiotic Pattern
Spongiotic Dermatitis

- Microvesicular + Lym
- Microvesicular + Eos
- Microvesicular + PMNs
- Slight Spongiosis
Spongiosis

Ballooning
Dermatophyte
Spongiotic Pattern

- Irritant contact dermatitis
- Allergic contact dermatitis
Spongiotic Pattern

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

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

Irregular

Regular

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

Lichen simplex chronicus
Psoriasis
Reiter's syndrome
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

• 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
Reiter Syndrome
Reiter Syndrome

- Seronegative spondyloarthropathy
- Cutaneous erythematous plaques
- Two types:
  1) Endemic: chlamydia trachomatis
  2) Epidemic: enteric infections (Shigella, Salmonella or Yersinia)
- Vulvar involvement is rare (HIV+)
- Histologically and clinically resembles pustular 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 effected 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**
Papillary Edema
(Gossmar strands)
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
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
Plasma cell vulvitis

- Zoon’s vulvitis
- Vulvitis circumscripta 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.