

# NOTES PAPULOSQUAMOUS DISORDERS

# **GENERALLY, WHAT ARE THEY?**

# **PATHOLOGY & CAUSES**

- Heterogeneous skin disorders; scaly papules, plaques
  - Papule: circumscribed, solid elevation of skin < 1cm/0.39in</li>
  - Plaque: broad papule/confluence of papules ≥ 1cm/0.39in
  - Scale: dry/greasy laminated masses of keratin

### CAUSES

Inflammation

# SIGNS & SYMPTOMS

See individual disorders

### DIAGNOSIS

### OTHER DIAGNOSTICS

Rash patterns

# TREATMENT

May spontaneously resolve

# **MEDICATIONS**

- Topical (e.g. corticosteroids), nonsteroidal anti-inflammatory drugs (NSAIDs), antihistamines
- Immunosuppressants, retinoids (e.g. acitretin)

#### OTHER INTERVENTIONS

Phototherapy, colloid baths

# LICHEN PLANUS

# osms.it/lichen-planus

# **PATHOLOGY & CAUSES**

- Self-limiting chronic dermatosis
- Multifactorial pathogenesis
  - Environmental factors → genetic aberrations of immune system
  - CD8+ T cells respond to altered antigens in basal epidermidis/ dermoepidermal junction
  - Causal agent identified: lichenoid reaction (e.g. drugs)

 $\blacksquare$  Chronic inflammation in mucosal lesions  $\rightarrow$  squamous cell carcinoma

# SIGNS & SYMPTOMS

- Shiny, flat-topped, pink-purple, polygonal papules coalesce, form plaques with red scales
- Wickham striae (pathognomonic)
  - Interspersed grey-white lace-like pattern of lines

- Symmetrical peripheral distribution, esp. on flexural surfaces (e.g. wrists, elbows, ankles, shins)
- Severe pruritus
- Koebner phenomenon
  - Skin lesions induced by local trauma
- Nail involvement
  - 10% of individuals; subungual thickening/hyperpigmentation, thinning/ ridging/ grooving of nail plate, pterygium formation, onycholysis
- Mucosal involvement
- Oral mucosa
  - Asymptomatic/burning sensation, severe pain
  - Mostly bilateral, inner cheeks
  - Various types may coexist; oral variant of Wickham's striae, erosive/ulcerative, papular, plaque-like, atrophic, bullous
  - Possible secondary Candida infections
- Esophageal mucosa
  - Dysphagia/odynophagia
- Genital mucosa (glans penis, vulva/vagina)
  - Lower urinary tract symptoms, dyspareunia, itching in individuals who are biologically female



### MNEMONIC: 6 Ps

# Clinical presentation of lichen planus

**P**lanar

**P**urple

Polygonal

**P**ruritic

**P**apules

**P**laques

# **DIAGNOSIS**

### LAB RESULTS

- Typical microscopic features
  - Acanthosis: epidermidis thickening
  - Interface dermatitis: continuous infiltrate of lymphocytes along dermoepidermal junction → sawtoothing (dermoepidermal interface with zig-zag contour)

 Basal keratinocytes degenerate, appear like stratum spinosum cells (squamatization)/undergo necrosis, become incorporated into inflamed papillary dermis (Civatte bodies)

### OTHER DIAGNOSTICS

- Rash pattern distinctive at skin examination
- Skin biopsy
  - Rule out secondary malignancies

# TREATMENT

- Cutaneous lesions spontaneously resolve in nine months, longer for mucosal lesions
  - Leaves area of hyperpigmentation

#### **MEDICATIONS**

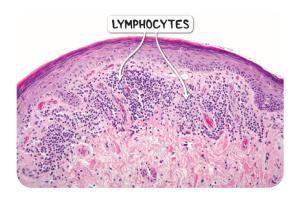
- Reduce symptoms, shorten duration
- Antihistamines (pruritus), corticosteroids
- Retinoids, immunosuppressants

#### OTHER INTERVENTIONS

- Occlusive dressings
- Phototherapy
  - Ultraviolet A radiation



**Figure 6.1** Lesions on the shins of an individual with lichen planus.



**Figure 6.2** The histological appearance of lichen planus. There is a lymphocytic infiltrate at the junction between the dermis and epidermis which is known as interface dermatitis.

# PITYRIASIS ROSEA

# osms.it/pityriasis-rosea

# PATHOLOGY & CAUSES

- Self-limiting acute dermatosis
- Unknown etiology; may be viral in origin, related to human herpesvirus 7 (HHV7)

# SIGNS & SYMPTOMS

- Upper respiratory tract infection may precede rash
- "Herald patch"
  - Solitary oval red plaque, usually located on trunk
  - First skin lesion
  - Spreads with central clearing, fading in 2–10 days
- 1–2 weeks after herald patch, multiple round/oval pink (white individuals of European descent)/dark brown (black individuals of sub-Saharan African descent) plaques with central scale appear

- Trunk, neck, upper arms, thighs; "Christmas tree" progression
  - Across chest, then rib-line
- Pruritus
- Systemic
  - Low-grade fever, headache, nausea, fatigue



**Figure 6.3** A herald patch is often seen at the onset of pityriasis rosea. It is a slightly raised, erythematous patch with superficial scaling.

# **DIAGNOSIS**

# LAB RESULTS

- Skin biopsy (rare)
- Microscopic features
  - Dyskeratosis: abnormal premature keratinization
  - Extravasated erythrocytes within dermal papillae

### OTHER DIAGNOSTICS

- Rash pattern
  - Distinctive at skin examination

# TREATMENT

May spontaneously disappear in 6–8 weeks

### **MEDICATIONS**

Antihistamines for pruritis

## OTHER INTERVENTIONS

- May spontaneously disappear in 6–8 weeks
- Colloid baths for pruritis



**Figure 6.4** The clinical appearance of pityriasis rosea on the torso of an adult male.

# **PSORIASIS**

# osms.it/psoriasis

# PATHOLOGY & CAUSES

- Chronic dermatosis of skin, nails, joints
- Multifactorial pathogenesis
  - $ilde{ }$  Environmental factors o genetic abnormalities of immune system
  - CD4+ TH1, TH17, CD8+ T cells collect in epidermis, secrete cytokines (e.g. IFN-gamma, TNF-alpha, IL-17, IL-22), growth factors → abnormal microenvironment ("cytokine soup") accelerates keratinocyte proliferation → defective keratinization, epidermal thickening
- Unpredictable progression with

spontaneous remissions, sudden exacerbations (e.g. may worsen in winter—lack of sun, humidity)

- Skin abrasion, infection, drugs (e.g. lithium, beta blockers, chloroquine), psychosocial stress → exacerbations
- 10–15% of individuals develop psoriatic arthritis
  - $\ ^{\circ}$  Inflammatory cells in joint tissue  $\ \rightarrow$  synoviocyte proliferation
  - Surrounding connective tissue also involved (e.g. enthesitis)

#### **TYPES**

- Plaque psoriasis, AKA vulgar psoriasis; 90%
- Guttate (eruptive), inverse (flexural), pustular, erythrodermic

# SIGNS & SYMPTOMS

- Plaque
  - Pink, salmon-colored papules/plaques covered by loosely adherent silver-white scales
  - Any area of body, esp. extensor surfaces (e.g. knees, elbows), lumbosacral area, scalp, glans penis
  - Itching is mild/absent
- Nail involvement in 30% of individuals
  - Subungual thickening
  - Yellow-brown discolorations of nail plate (resembling oil slicks)
  - Crumbling/ridging/pitting of nail plate
  - Onycholysis: separation of nail plate from bed
- Guttate (eruptive)
  - Drop-like appearance, associated with group A streptococcus
- Inverse (flexural)
  - Skin folds
- Pustular
  - Blisters filled with non-infectious pus
- Erythrodermic
  - Total body inflammation, skin exfoliation, severe itching, swelling, pain; ability to regulate temperature, perform barrier functions impaired; possibly fatal
  - May develop from any type (e.g. plaque during corticosteroid rebound phenomenon)
- Auspitz sign
  - Pinpoint bleeding appears when scale removed
- Koebner phenomenon
  - Characteristic skin lesions induced by local trauma
- Psoriatic arthritis
  - Inflammatory arthritis: pain, red overlying area, swelling, hot to touch
  - Frequently occurs after onset of rash
  - Asymmetric peripheral oligoarthritis;

- joints of hands, feet most affected, followed by sacroiliac bone, spine
- Fusiform swelling of digits (dactylitis);
   aka "sausage digits"
- Aggressive disease with joint damage, malformations not common

# DIAGNOSIS

#### DIAGNOSTIC IMAGING

• For psoriatic arthritis

#### X-ray

 Erosive changes, "fluffy" periostitis, presence of new bone formation

#### MR

Inflammation in adjacent bone marrow and soft tissues

#### LAB RESULTS

- Skin biopsy (rare)
- Acanthosis
  - Epidermidis thickening
- Parakeratosis
  - Keratinization (retention of nuclei in stratum corneum)
- Neoangiogenesis with tortuous blood vessels below stratum corneum
- Accumulation of neutrophils in superficial epidermis (spongiform pustules), in stratum corneum (Munro microabscesses)
- Clinical diagnosis
  - Psoriasis features, clinical pattern of joint involvement
- Confirmation
  - Elevated inflammatory markers, negative rheumatoid factor (RF), anticyclic citrullinated peptide antibody (anti-CCP)

#### OTHER DIAGNOSTICS

- Rash pattern
  - Distinctive at skin examination
- Differentiation from rheumatoid arthritis
  - Minority show polyarthritic pattern with no skin lesions; note asymmetry, distal interphalangeal joint involvement, mild joint destruction

# **TREATMENT**

- No definitive cure
- Avoid triggers

### **MEDICATIONS**

- Topical corticosteroids → antiinflammatory, antiproliferative
- Vitamin D derivatives (calcipotriene, calcipotriol) → limit keratinocyte proliferation
- Anthralin → suppresses proliferation
- Combination therapy is most effective (e.g. betamethasone dipropionate + calcipotriene)
- Affected area > 10%, unsuccessful topical treatment, involves face, hands, genitals
- Immunosuppressant
  - Methotrexate, cyclosporine
- Systemic retinoids
  - $ext{-}$  Acitretin  $ext{-}$  inhibits pro-inflammatory cytokines
- Biologic therapy
  - Anti-TNF (infliximab, etanercept, adalimumab), T-cells (alefacept), IL-12/23 (ustekinumab)
- NSAIDs, immunosuppressant/biologic therapy
  - For psoriatic arthritis

#### OTHER INTERVENTIONS

- Topical
  - Coal tar → inhibits cellular mitotic activity, proliferation
  - Moisturizers, emollients
- Phototherapy
  - Ultraviolet A radiation
  - Often combined with topical tar/ systemic acitretin/psoralen/methoxsalen
  - Immunosuppressive, antiproliferative



**Figure 6.5** A large psoriatic plaque on the upper limb.



Figure 6.6 Psoriasis affecting the hand.