Papulosquamous disease

DR.SALEH ALRASHEED
ASST.PROF.&CONSULTANT
DERMATOLOGY

Papulosquamous diseases

- Group of skin diseases that share similar features : papule or macule lesions with scales

The category of papulosquamous disease classically includes :

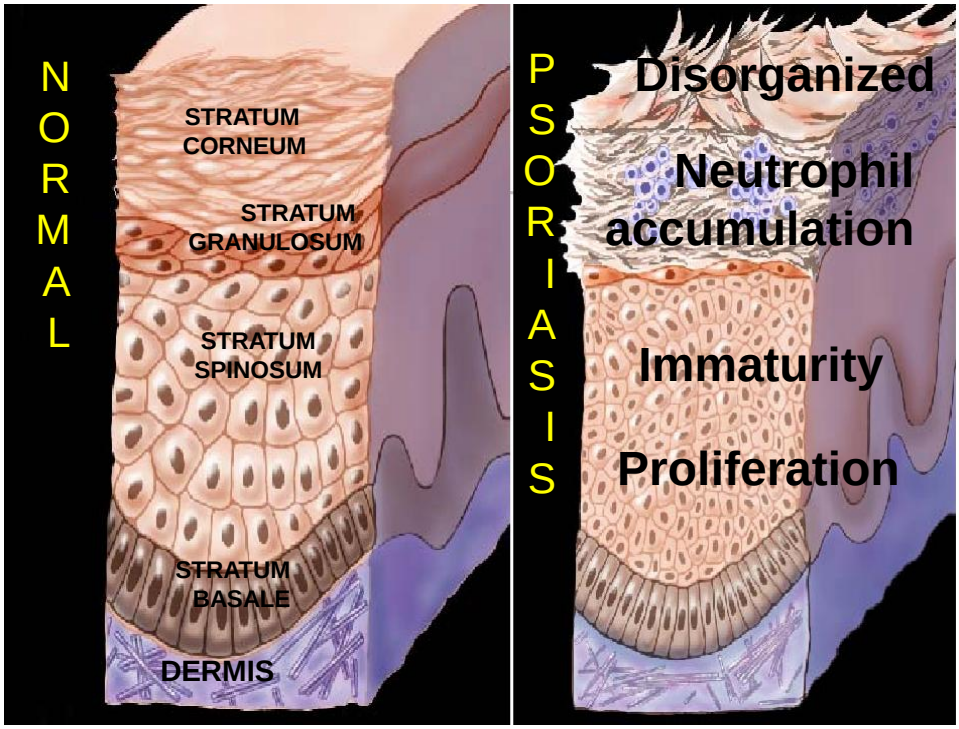
psoriasis
pityriasis rosea
planus lichen

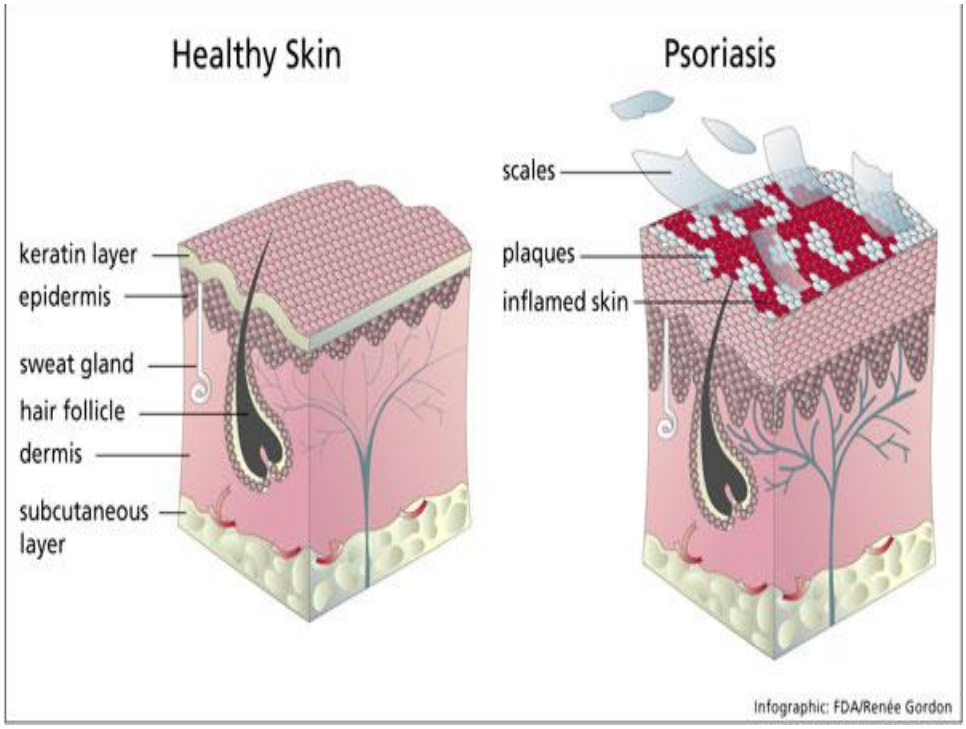
Psoriasis

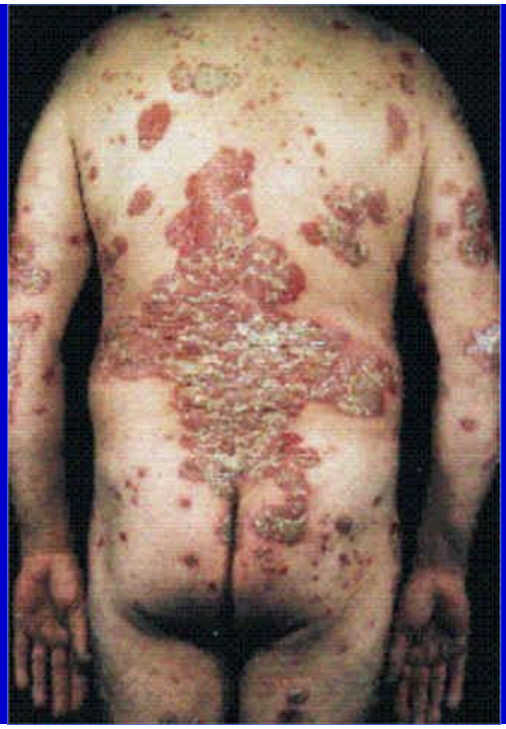
- Ps. is a common, chronic, recurrent inflammatory disease
- Often inherited (autosomal dominant - HLA B13-17-27)
- Sometimes associated with disorders of the joints and nails
- Usually symmetrical, well defined, rounded circumscribed, erythematous, dry, scaling patches covered by grayish white or silvery scales
- Site ➡ scalp, nails, extensor surfaces of the limbs elbows, knees & sacral region
- Subjective symptoms such as itching or burning may be present

Etiology

- The cause of Ps. is still unknown
- The course of Ps. is inconstant
- Tendency to recur and to persist
- Koeber reaction (phenomenon)
- Auspitz sign: is pin point bleeding when a psoriatic scale is removed. (Severe thinning of the epidermis over the tips of dermal papillae)
- The psoriatic basal-cell is shed in about 4 days where as normal cell in 28 days
- The erythema is due to the dilatation & proliferation of the capillaries in the papillary dermis


NORMAL
STRATUM CORNEUM
STRATUM GRANULOSUM
STRATUM SPINOSUM
STRATUM BASALE
DERMIS
PSORIASIS
Disorganized
Neutrophil accumulation
Immaturity
Proliferation


Healthy Skin
keratin layer
epidermis
sweat gland
hair follicle
dermis
subcutaneous layer
Psoriasis
scales
plaques
inflamed skin
Infographic: FDA/Renée Gordon



Prevalence

- Psoriasis occurs in 2% of the world's population
- Prevalence in the U.S may be as high as 4.6%
- Highest in Caucasians
- In Africans, African Americans and Asians between 0.4% and 0.7%

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Prevalence

- Equal frequency in males and females
- May occur at any age from infancy to the 10th decade of life
- First signs of psoriasis
  - Females mean age of 27 years
  - Males mean age of 29 years

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Prevalence

- Two Peaks of Occurrence
  - One at 20-30 years
  - One at 50-60 years
- Psoriasis in children
  - Low – between 0.5 and 1.1% in children 16 years old and younger
  - Mean age of onset - between 8 and 12.5 years

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Prevalence

- Two-thirds of patients have mild disease
- One-third have moderate to severe disease
- Early onset (prior to age 15)
  - Associated with more severe disease
  - More likely to have a positive family history
- Life-long disease
  - Remitting and relapsing unpredictably
  - Spontaneous remissions of up to 5 years have been reported in approximately 5% of patients

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Genetics and Pathogenesis

- Psoriasis and the Immune System
  - The major histocompatibility complex (MHC)
    - Short arm of chromosome 6
  - Histocompatibility Antigens (HLA)
    - HLA-Cw6
    - HLA-B13, -B17, -B37, -Bw16
  - T-lymphocyte-mediated mechanism

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Psoriasis as a Systemic Disease

- Koebner Phenomenon
- Elevated ESR
- Increased uric acid levels → gout
- Mild anemia
- Elevated α_2-macroglobulin
- Elevated IgA levels
- Increased quantities of Immune Complexes

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Psoriasis as a Systemic Disease

- Psoriatic arthropathy
- Aggravation of psoriasis by systemic factors
  - Medication
  - Focal infections
  - Stress
- Life-threatening forms of psoriasis

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Psoriasis

- T-cell mediated inflammatory dz
  - Epidermal hyperproliferation 2^{O} to activation of immune system
  - Altered maturation of skin
  - Inflammation
  - Vascular changes

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Psoriasis : clinical features

4 Prominent features

- Sharply demarcated with clear cut borders
- Covering with noncoherent silvery scales
- Homogenous erythema under the scales
- Auspitz sign positive

Characteristic Lesion of Psoriasis

- Sharply demarcated erythematous plaque with micaceous silvery white scale
- Histopathology
  - Thickening of the epidermis
  - Tortuous and dilated blood vessels
  - Inflammatory infiltrate primarily of lymphocytes

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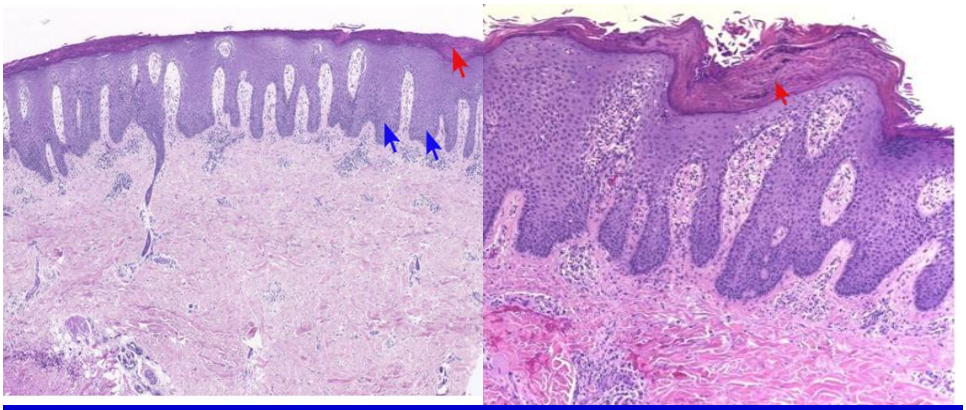
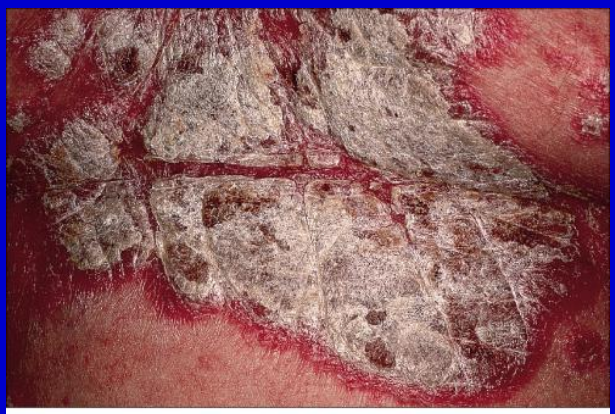


The histopathology ---- showed parakeratosis, hyperkeratosis and regular elongation of reteridges (regular psoriasiform hyperplasia).

Psoriatic Plaque



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Severity of Disease

- Three Cardinal Signs of Psoriatic Lesions
  - Plaque elevation
  - Erythema
  - Scale
- Body Surface Area

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Clinical Variants of Psoriasis

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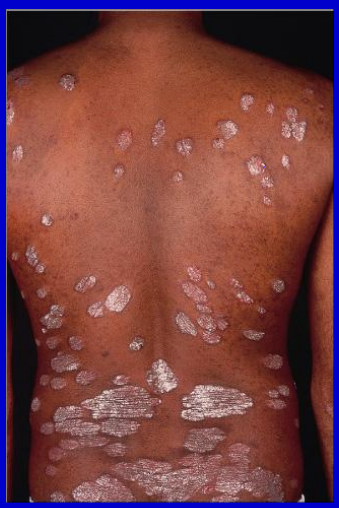
Chronic Plaque Psoriasis



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Chronic Plaque Psoriasis

- Most Common Variant
- Plaques may be as large as 20 cm
- Symmetrical disease
- Sites of Predilection
  - Elbows
  - Knees
  - Presacrum
  - Scalp
  - Hands and Feet

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Chronic Plaque Psoriasis

- May be widespread – up to 90% BSA
- Genitalia involved in up to 30% of patients
- Most patients have nail changes
  - Nail pitting
  - "Oil Spots"
  - Involvement of the entire nail bed
    - Onychodystrophy
    - Loss of nail plate

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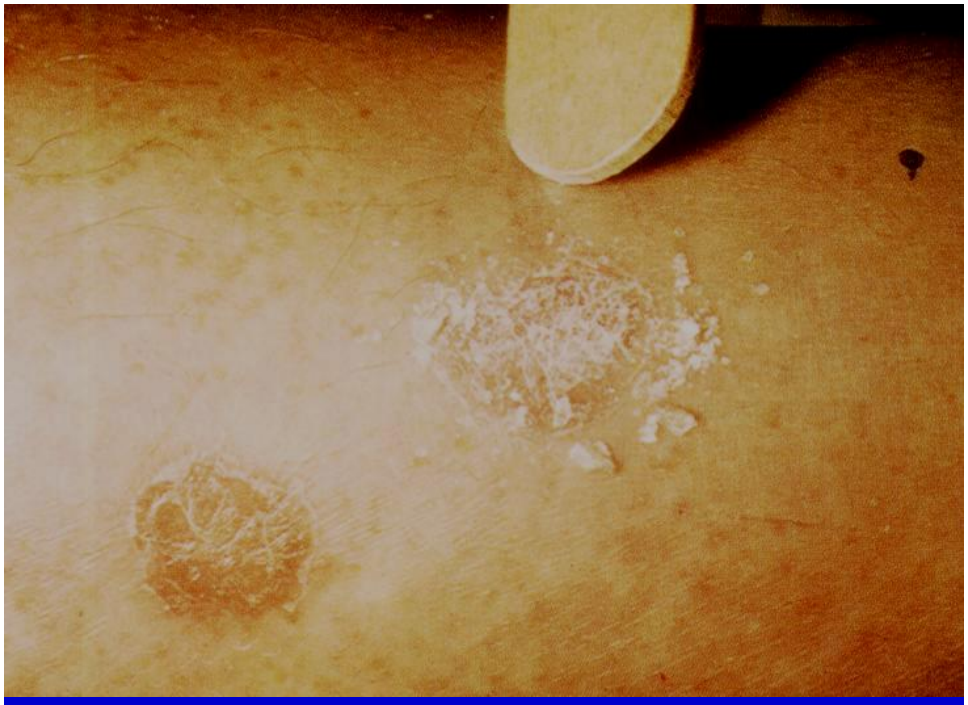
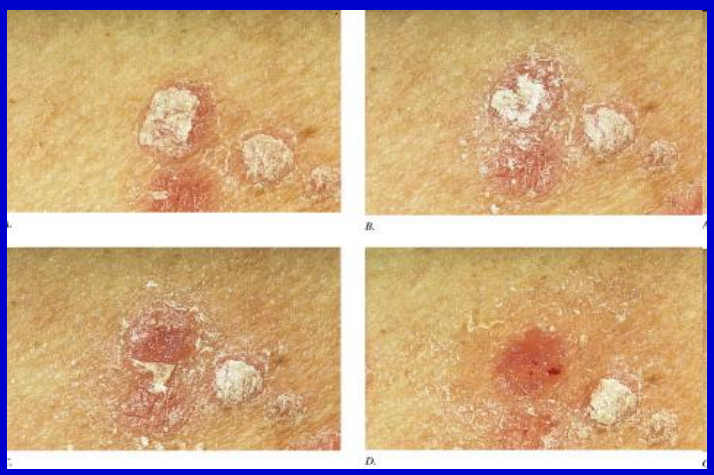
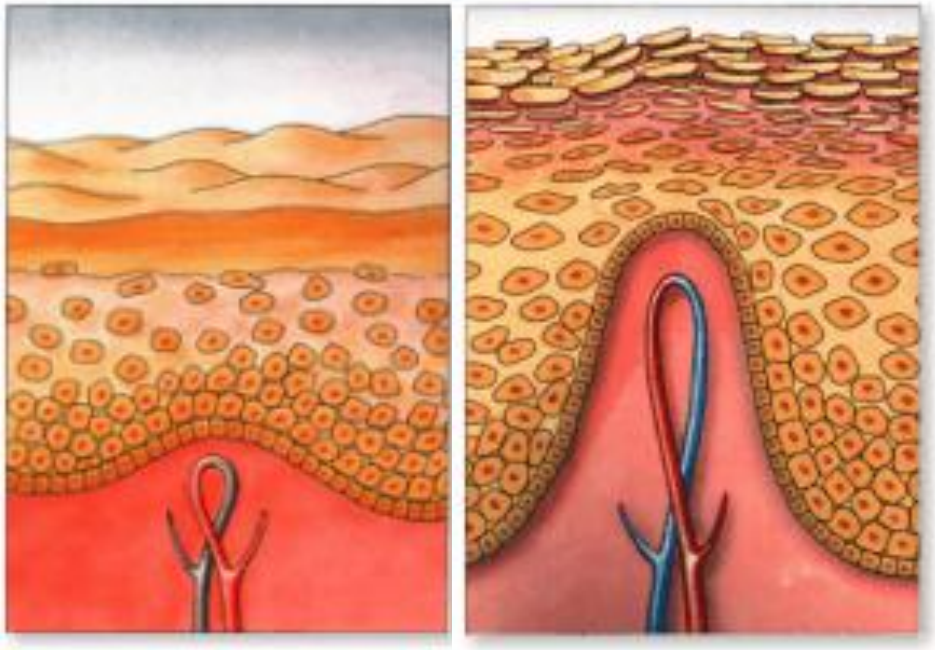
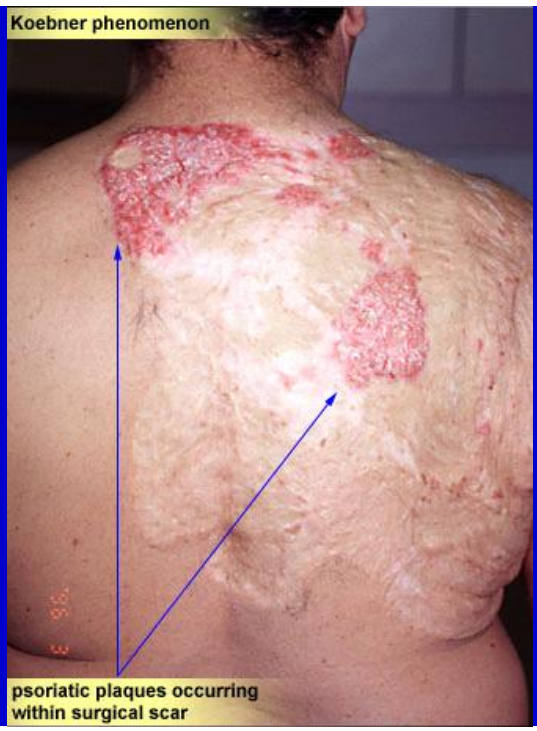
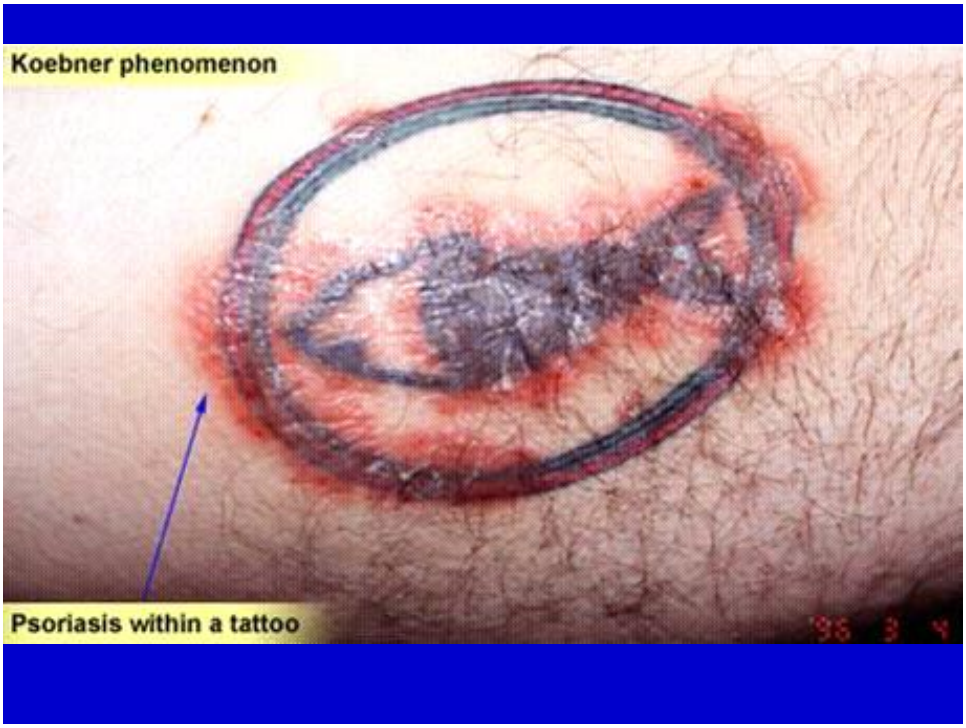


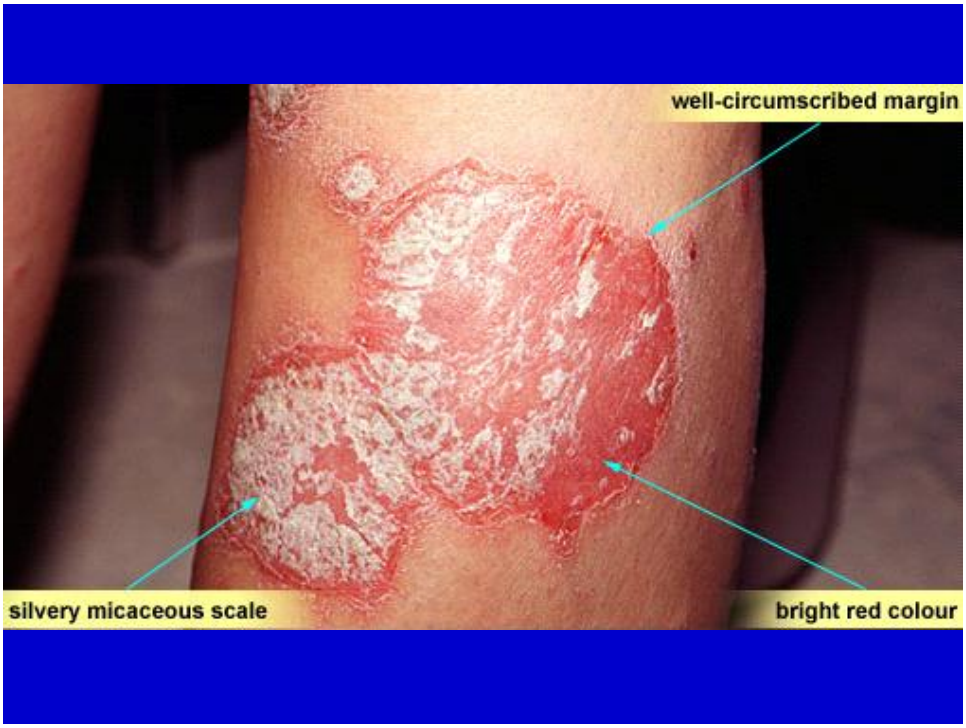
Auspitz sign

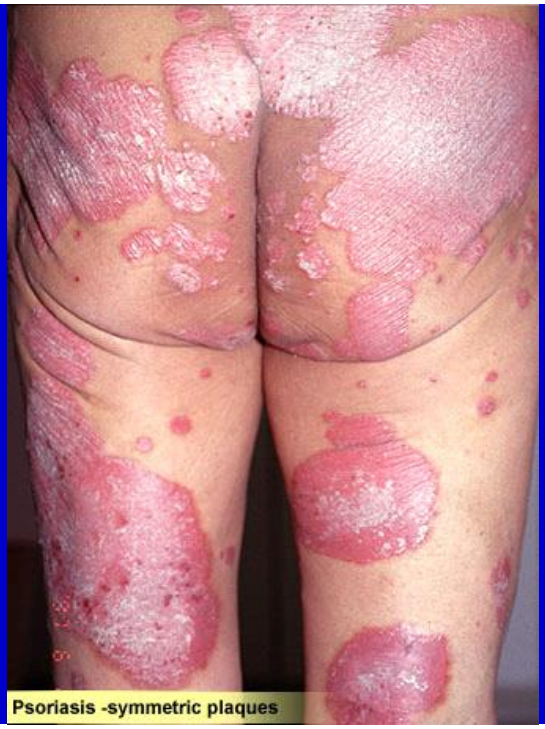




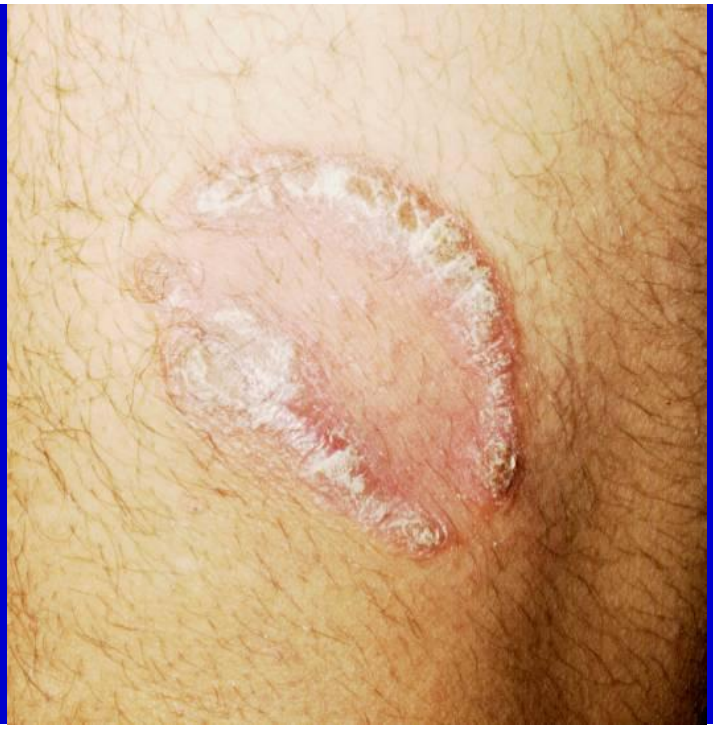

Koebner phenomenon
psoriatic plaques occurring within surgical scar

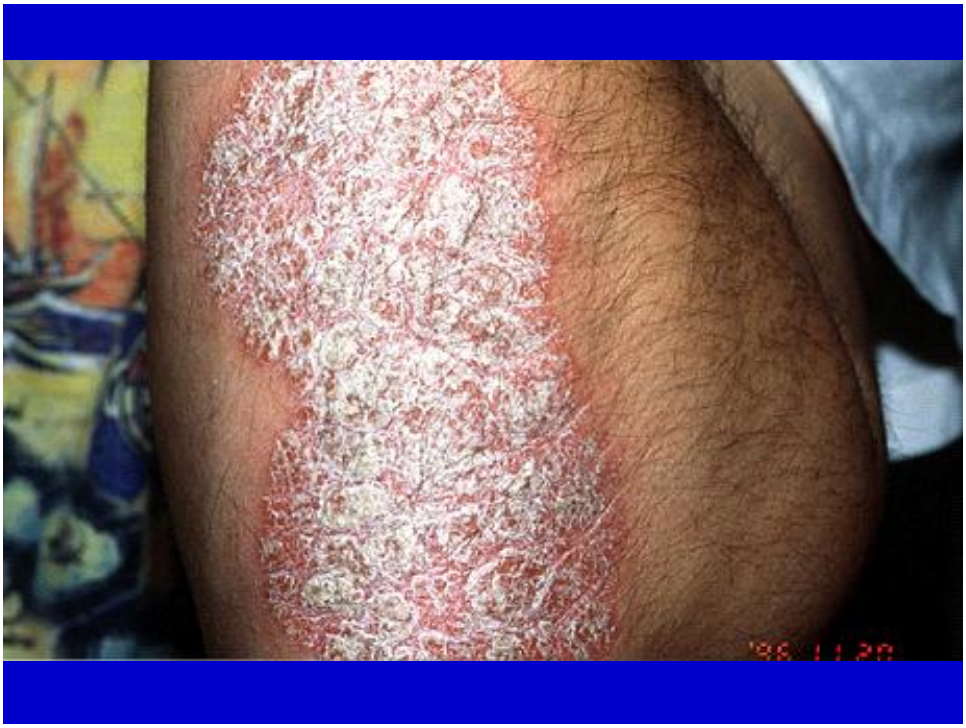

Koebner phenomenon
Psoriasis within a tattoo

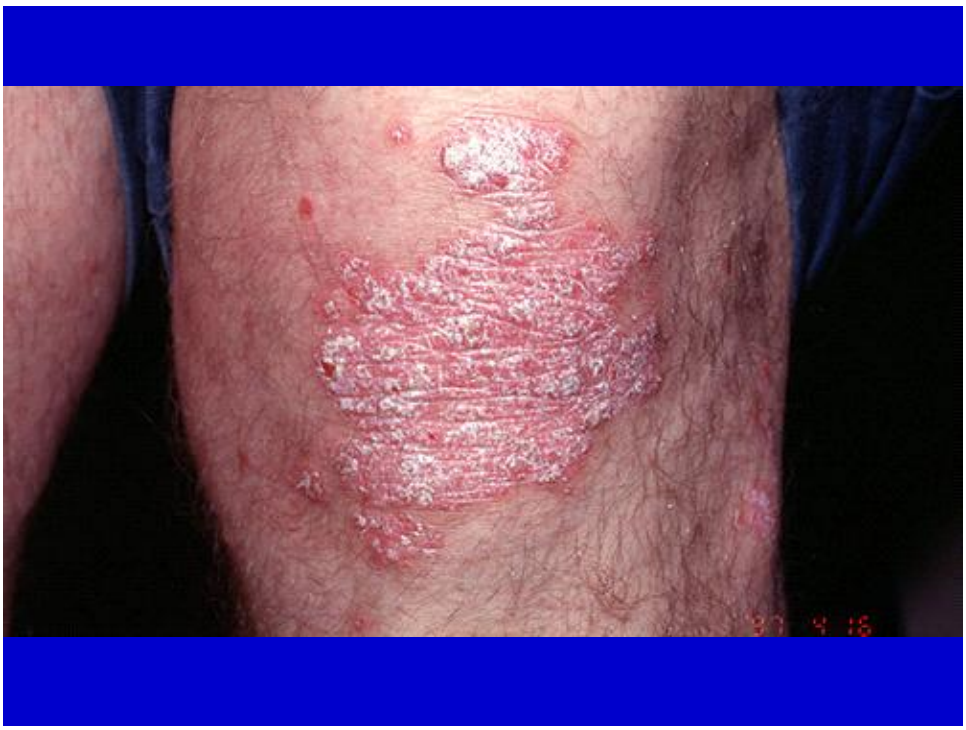

well-circumscribed margin
silvery micaceous scale
bright red colour

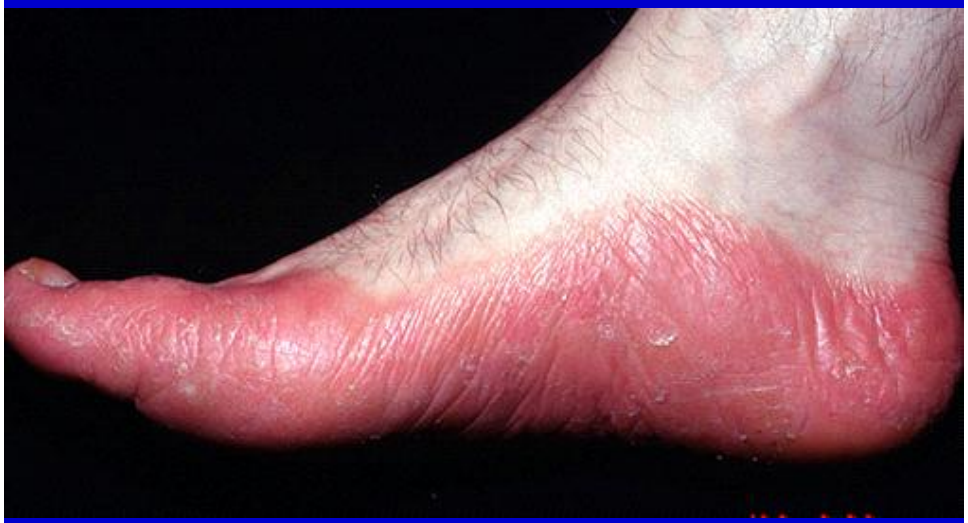


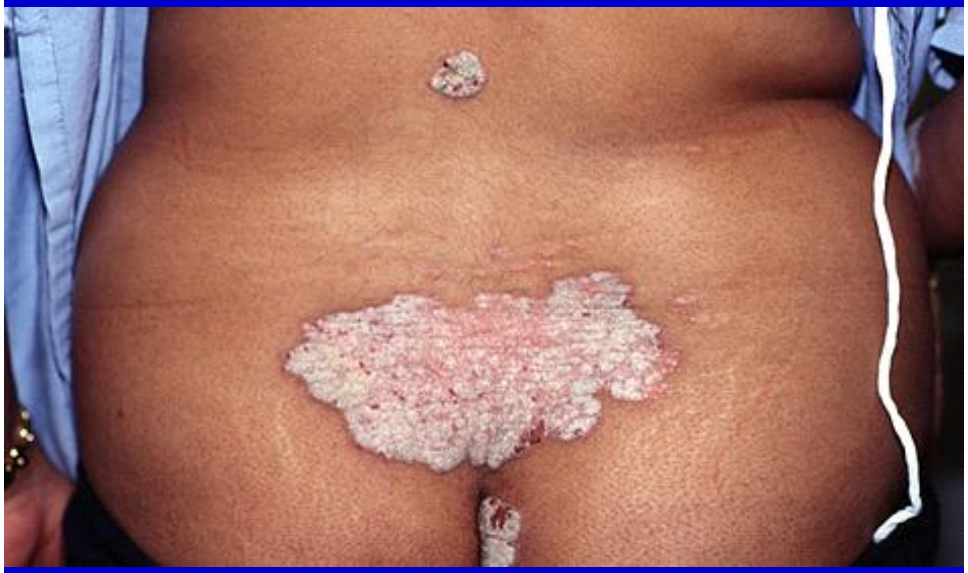
Psoriasis -symmetric plaques

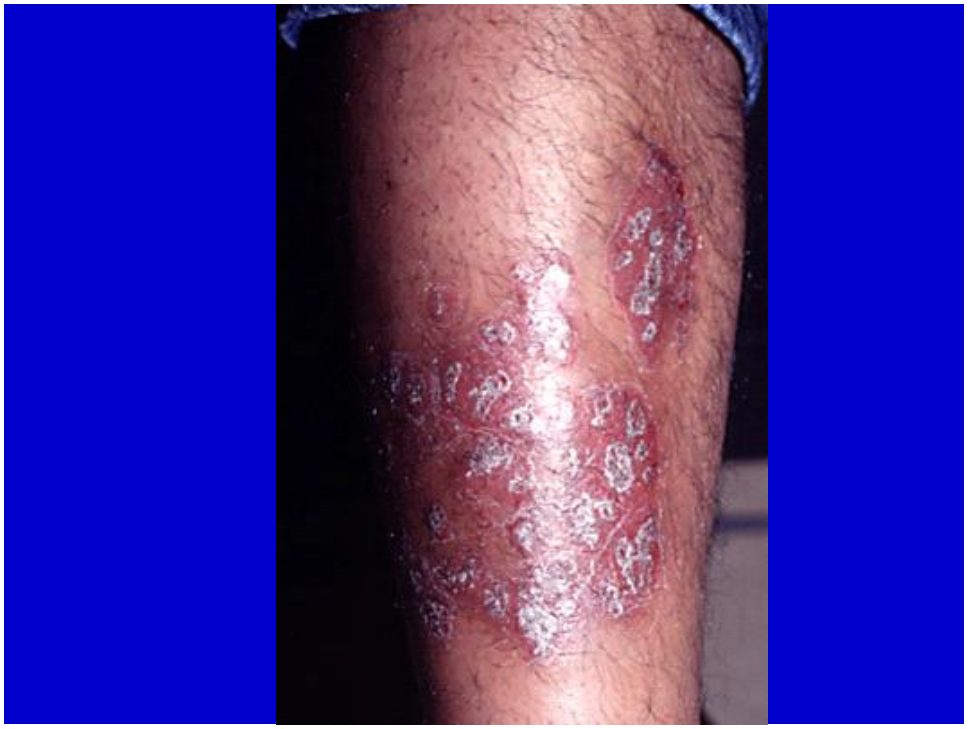






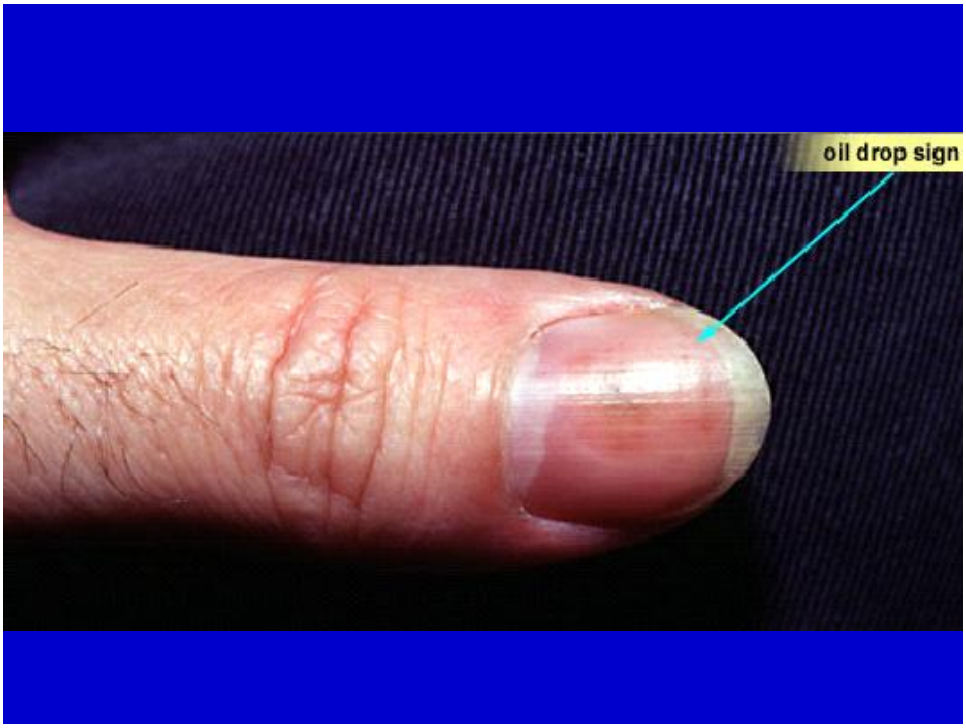


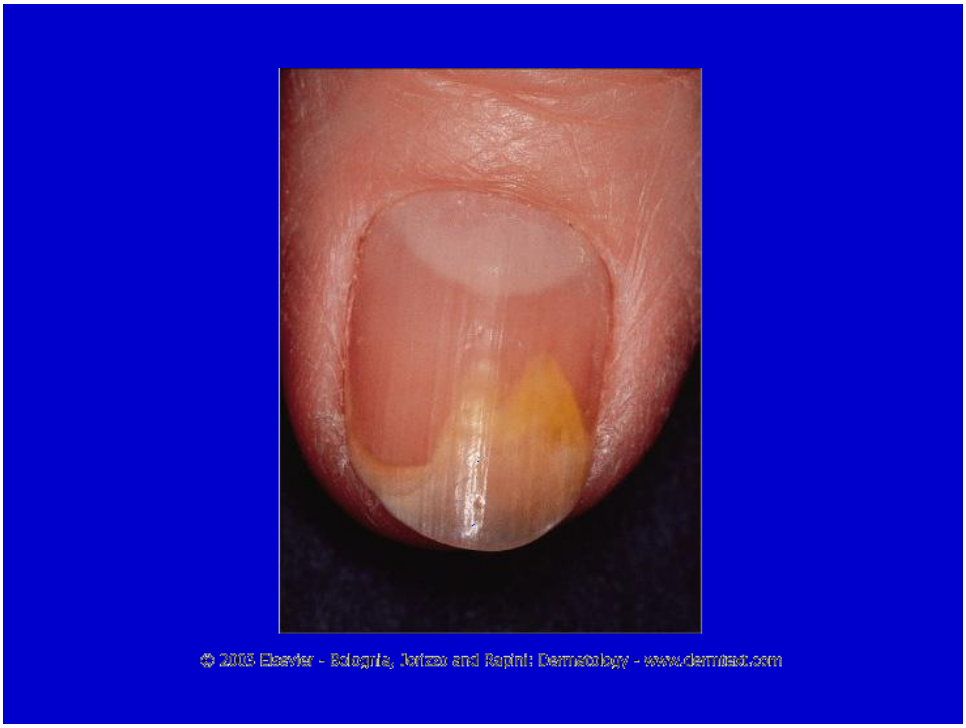





Psoriatic nail pits


Psoriatic nails
oncholysis


oil drop sign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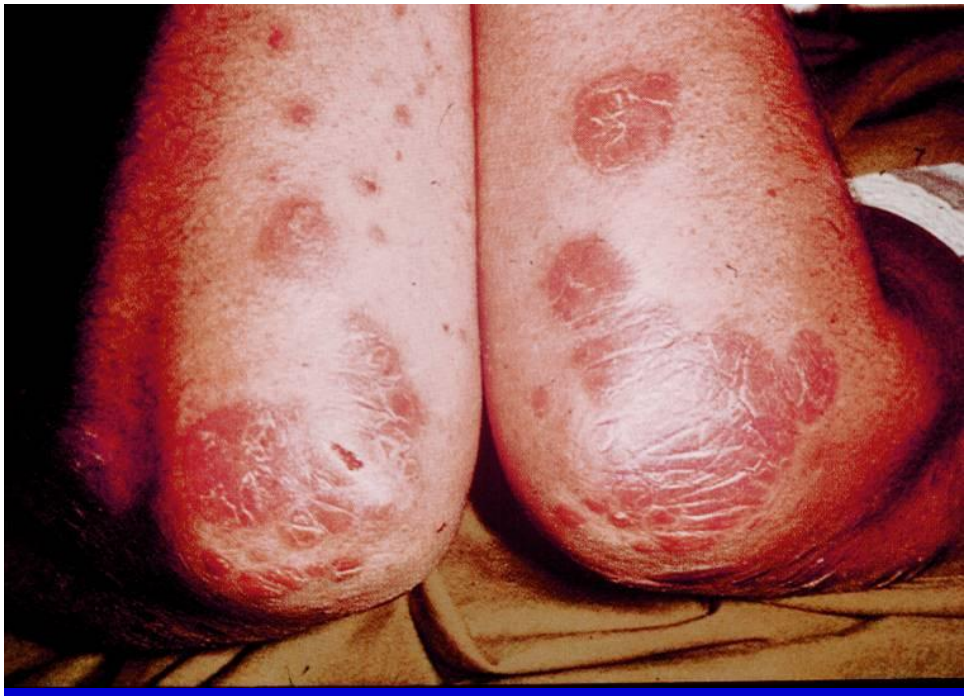
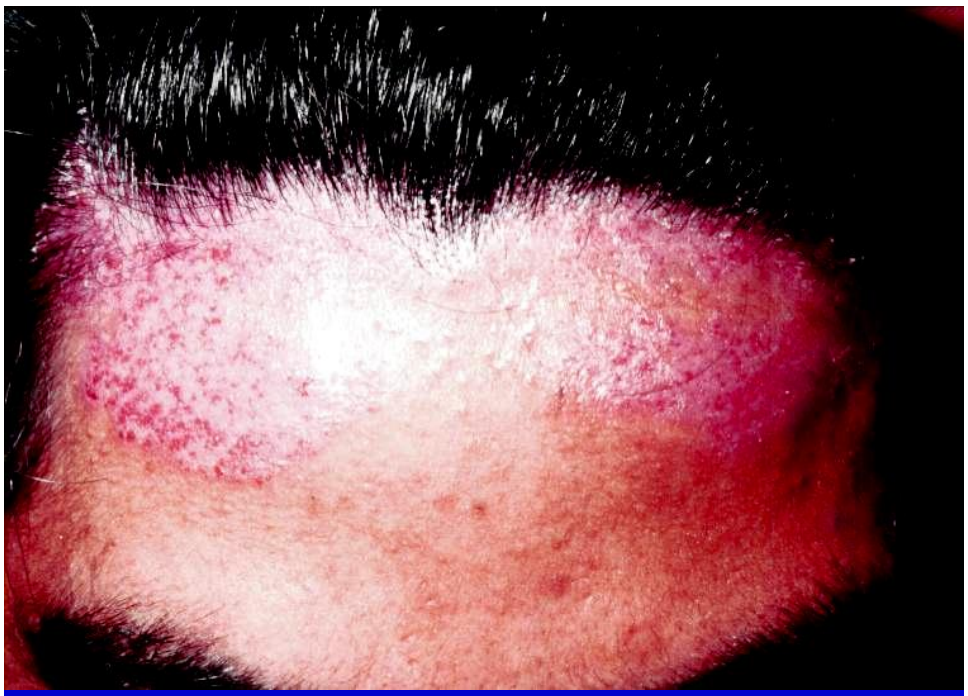
Onycholysis
Subungual hyperkeratosis

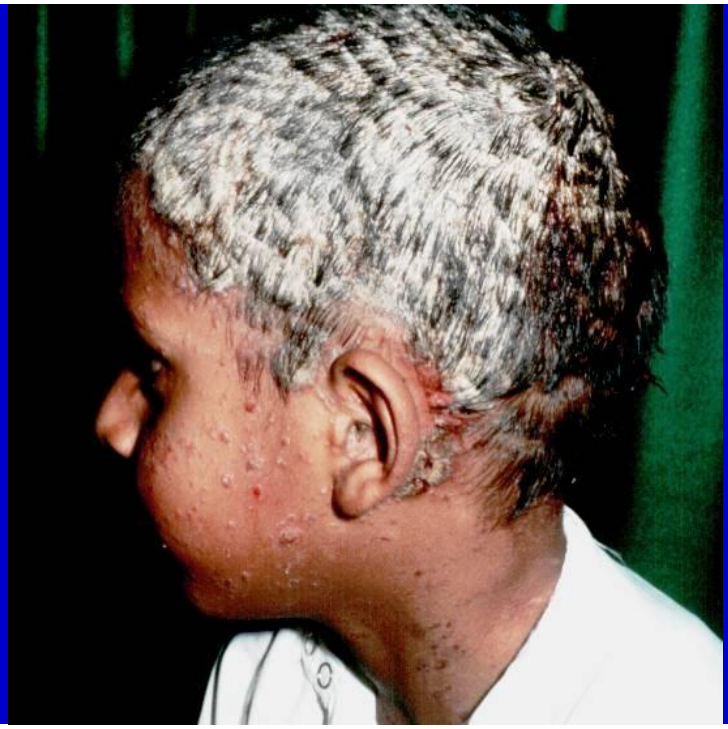

Severe nail dystrophy in psoriasis

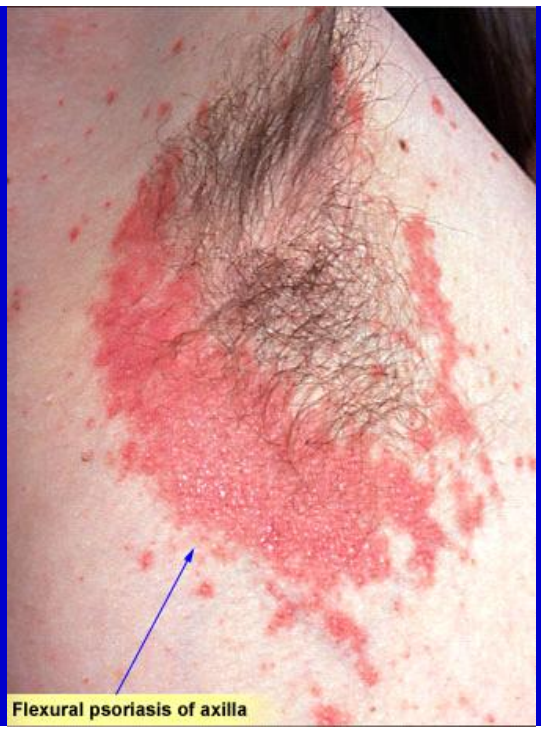
Psoriasis Vulgaris

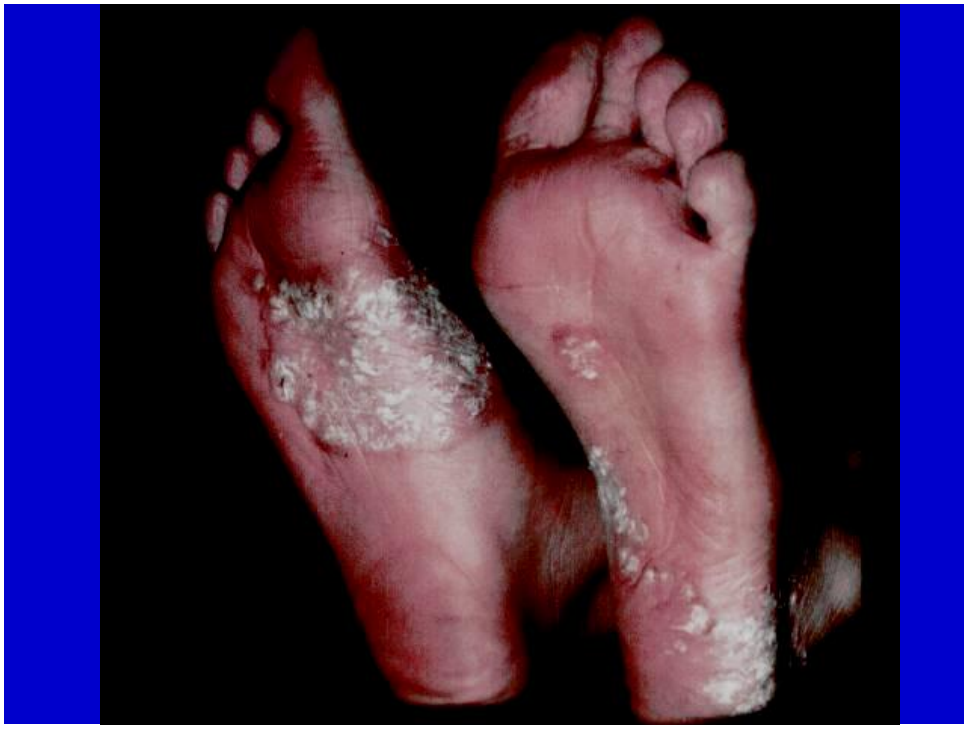
- Well defined red plaques
- White or silver scales
- Symmetrical
- Extensor surfaces
- Seborrheic Ps.
- Inverse Ps.
- Napkin Ps.
- Nails changed ➡ Oil spots or oval areas of onycholysis 2-6 mm in the nail bed not extending to the free distal border ➡ nail pitting

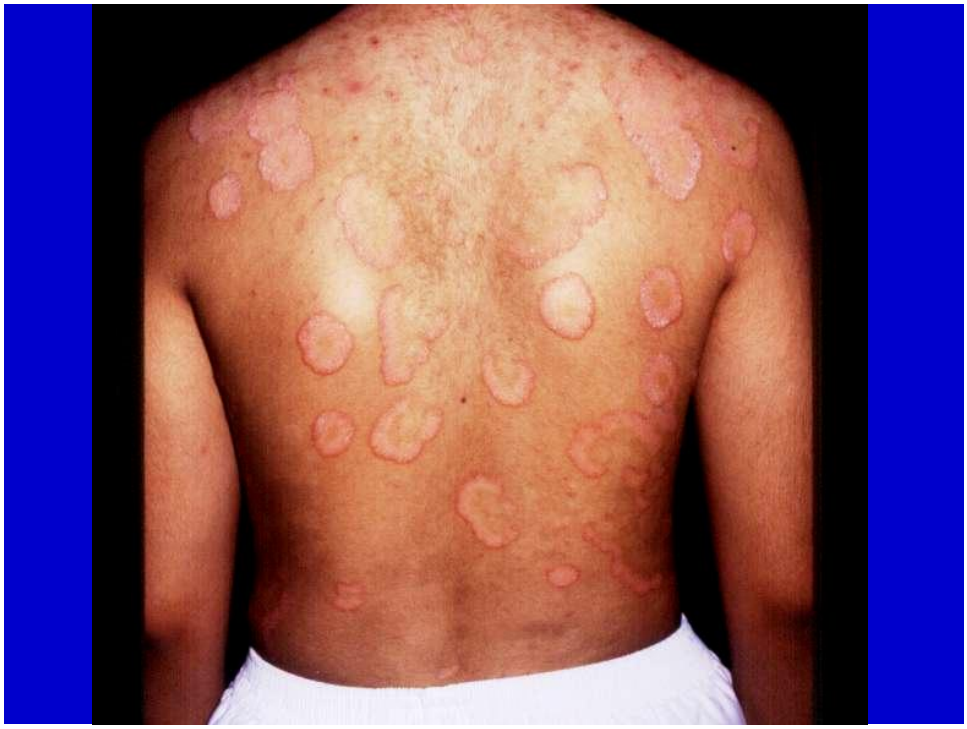


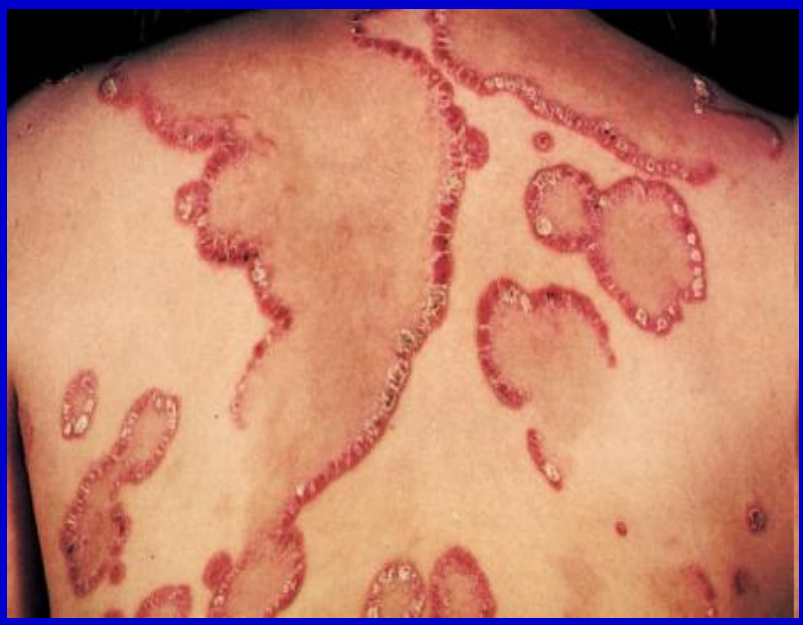





Flexural psoriasis of axilla







Life–Threatening Forms of Psoriasis

- Generalized Pustular Psoriasis
- Erythrodermic Psoriasis

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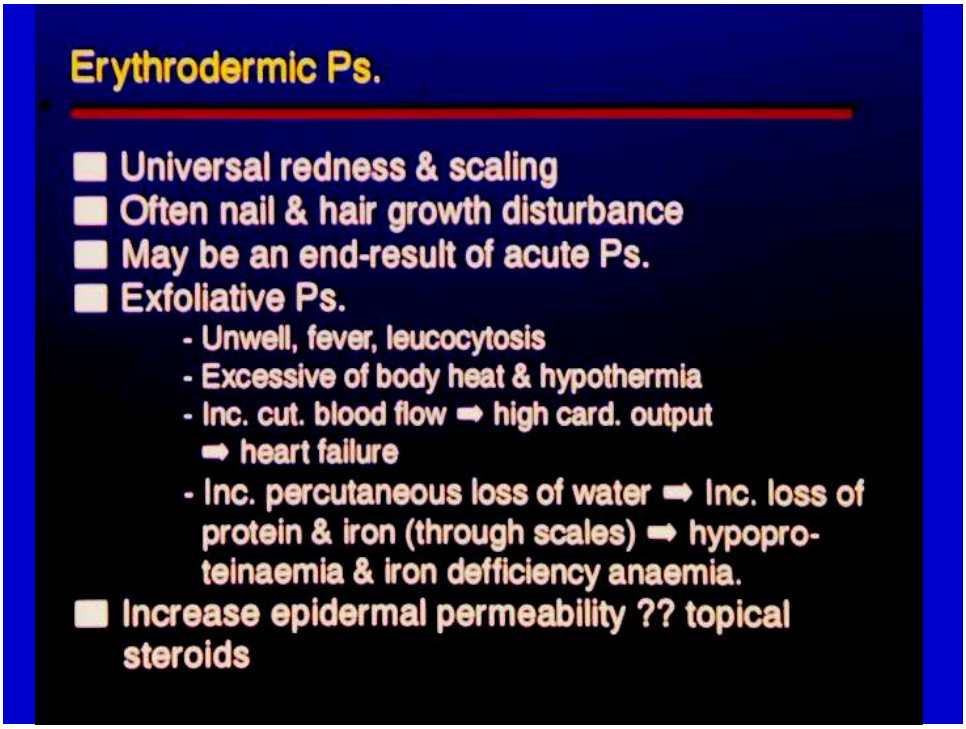

Erythrodermic Psoriasis

- Classic lesion is lost
- Entire skin surface becomes markedly erythematous with desquamative scaling.
- Often only clues to underlying psoriasis are the nail changes and usually facial sparing

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Erythrodermic Psoriasis

- Triggering Factors
  - Systemic Infection
  - Withdrawal of high potency topical or oral steroids
  - Withdrawal of Methotrexate
  - Phototoxicity
  - Irritant contact dermatitis

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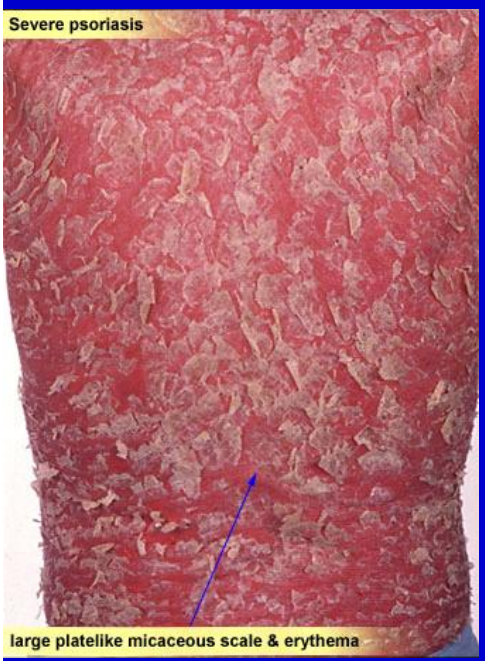

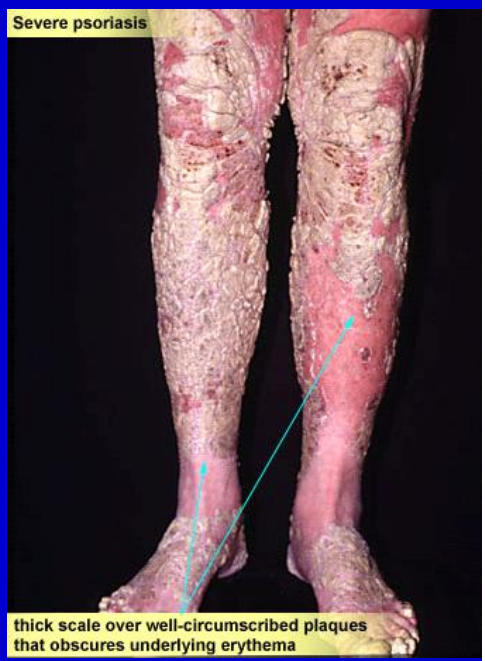

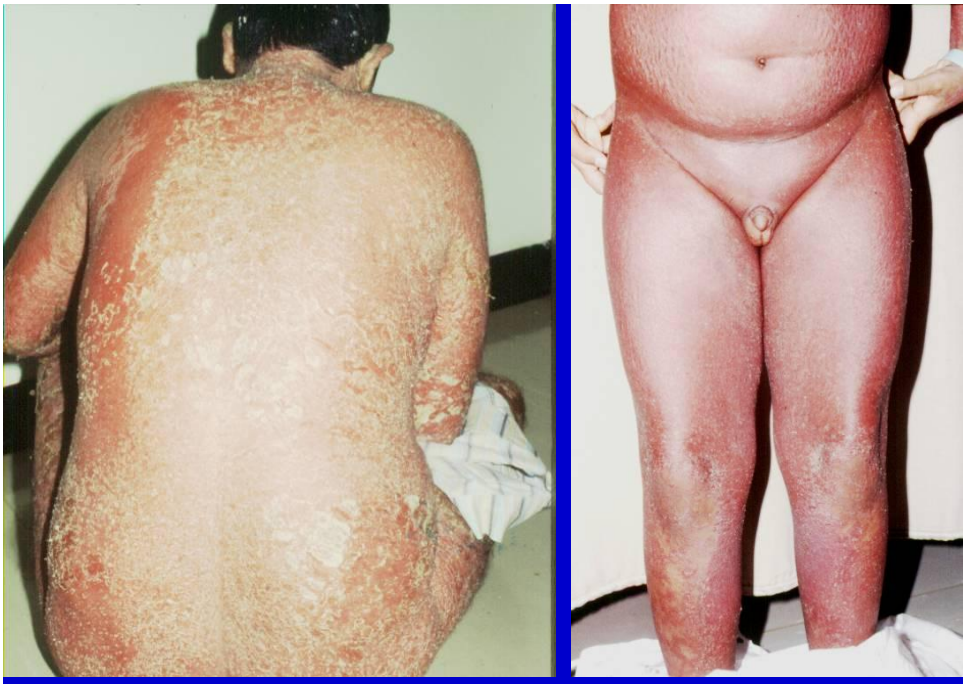

Pustular Psoriasis

- Localized (Palms & Soles)
  - Yellow pustules which turn brown
  - Occur on a red background
  - Often symmetrical
  - Palms & soles
  - There may be typical Ps. elsewhere
- Generalized (VonZumbusch)
  - High fever, leucocytosis
  - Extensive crops of small sterile pustules
  - Skin generally is bright red & sore

Generalized Pustular Psoriasis

- Unusual manifestation of psoriasis
- Can have a gradual or an acute onset
- Characterized by waves of pustules on erythematous skin often after short episodes of fever of 39˚ to 40˚C
- Weight loss
- Muscle Weakness
- Hypocalcemia
- Leukocytosis
- Elevated ESR

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Generalized Pustular Psoriasis

- Cause is obscure
- Triggering Factors
  - Infection
  - Pregnancy
  - Lithium
  - Hypocalcemia secondary to hypoalbuminemia
  - Irritant contact dermatitis
  - Withdrawal of glucocorticosteroids, primarily systemic

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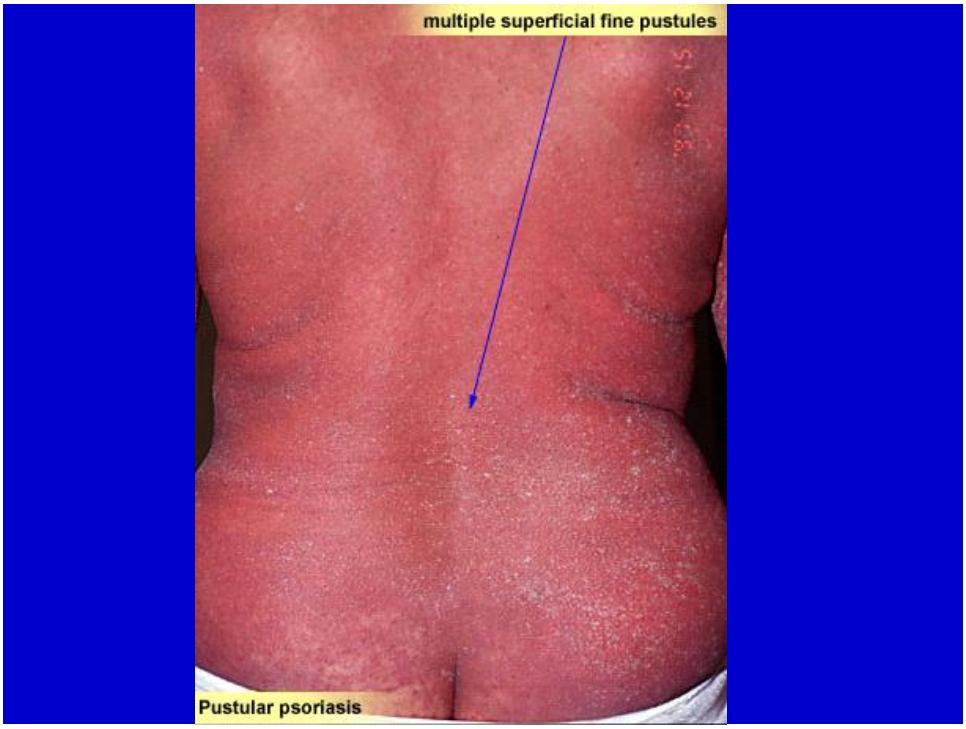
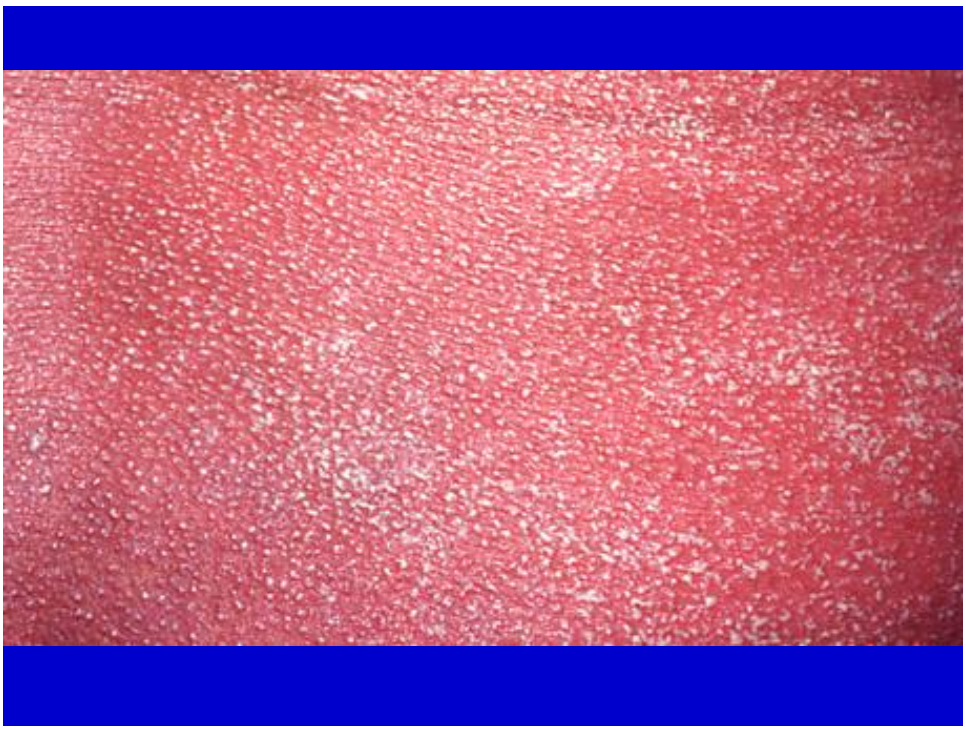
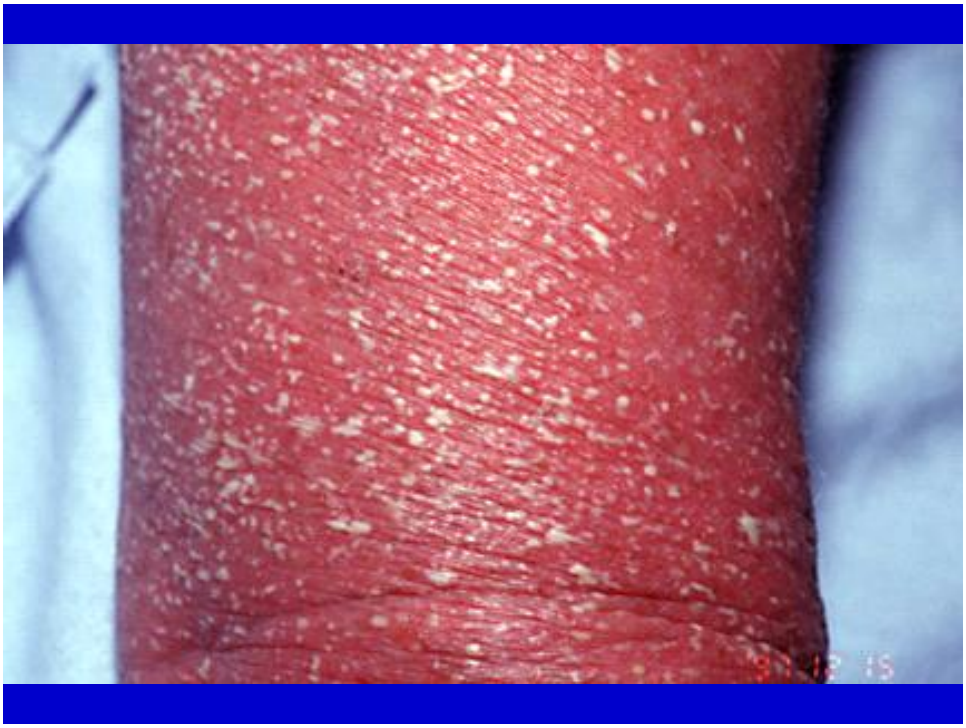
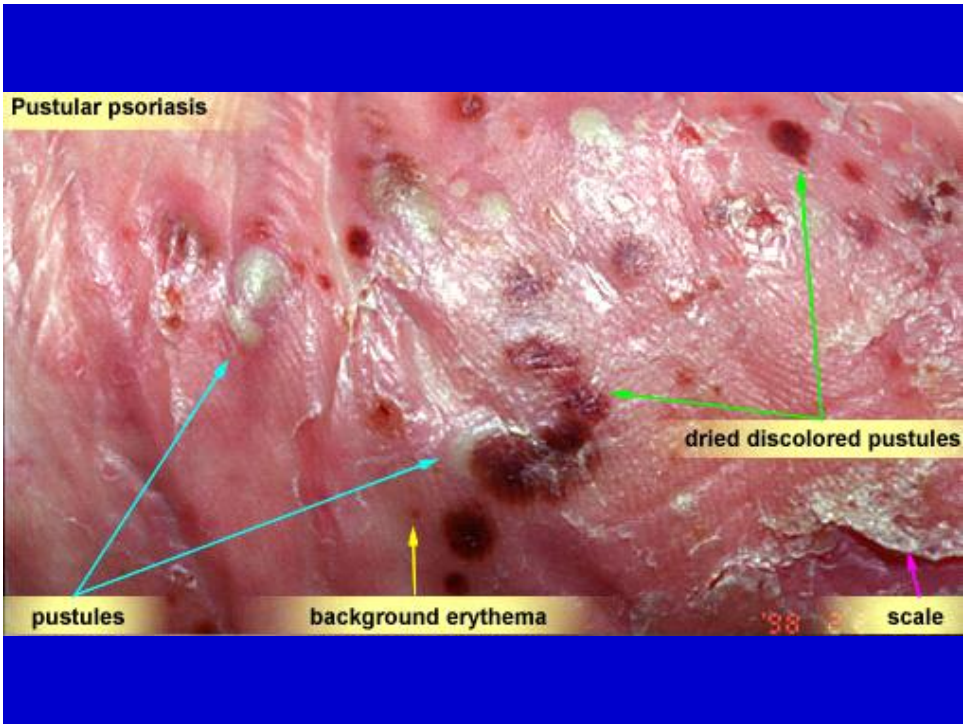
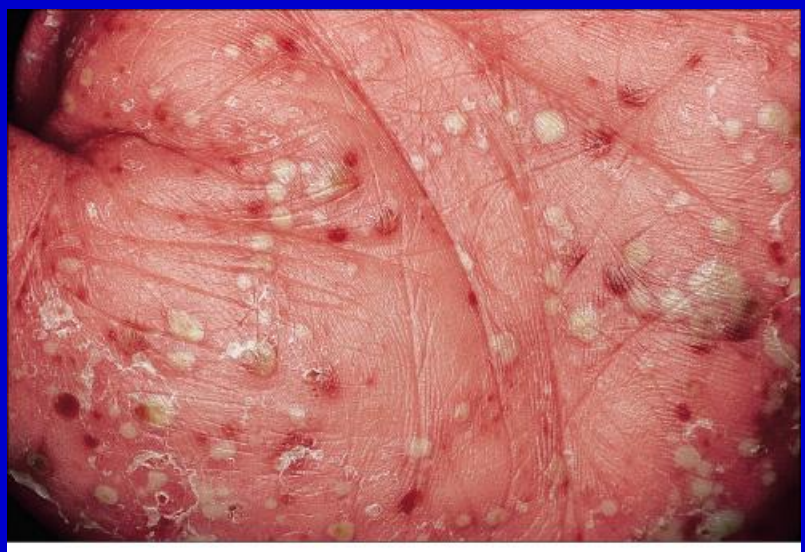
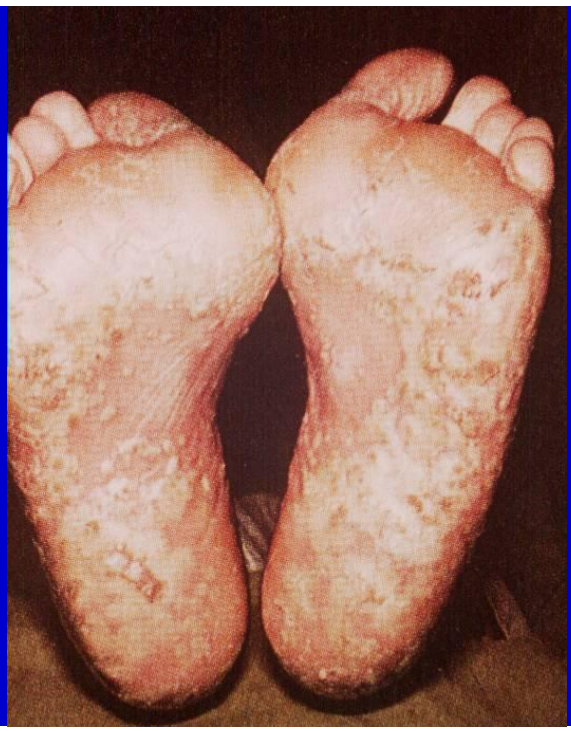

multiple superficial fine pustules
Pustular psoriasis






Pustular psoriasis
dried discolored pustules
pustules
background erythema
scale





Guttate Ps.

- Follows a streptococcal throat infection
- Rapid evolution
- Very small red papules, with thick white scales
- Mainly on trunk and limbs
- Resolution within four months

Guttate Psoriasis

- Characterized by numerous 0.5 to 1.5 cm papules and plaques
- Early age of onset
- Most common form in children
- Streptococcal throat infection often a trigger
- Spontaneous remissions in children
- Often chronic in adults

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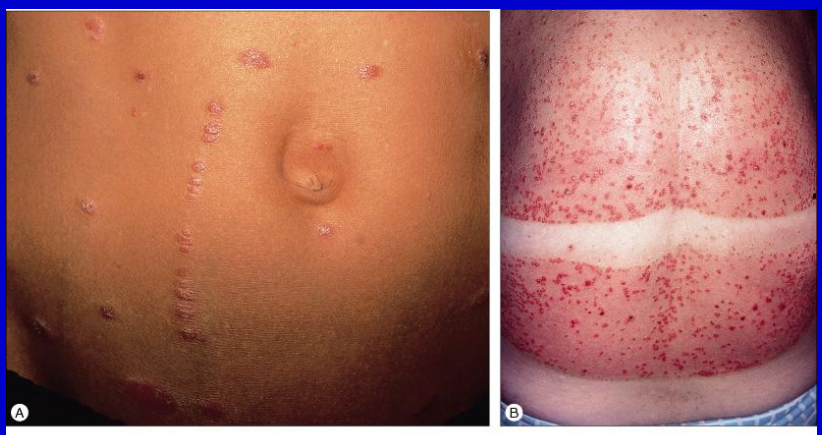
Guttate Psoriasis



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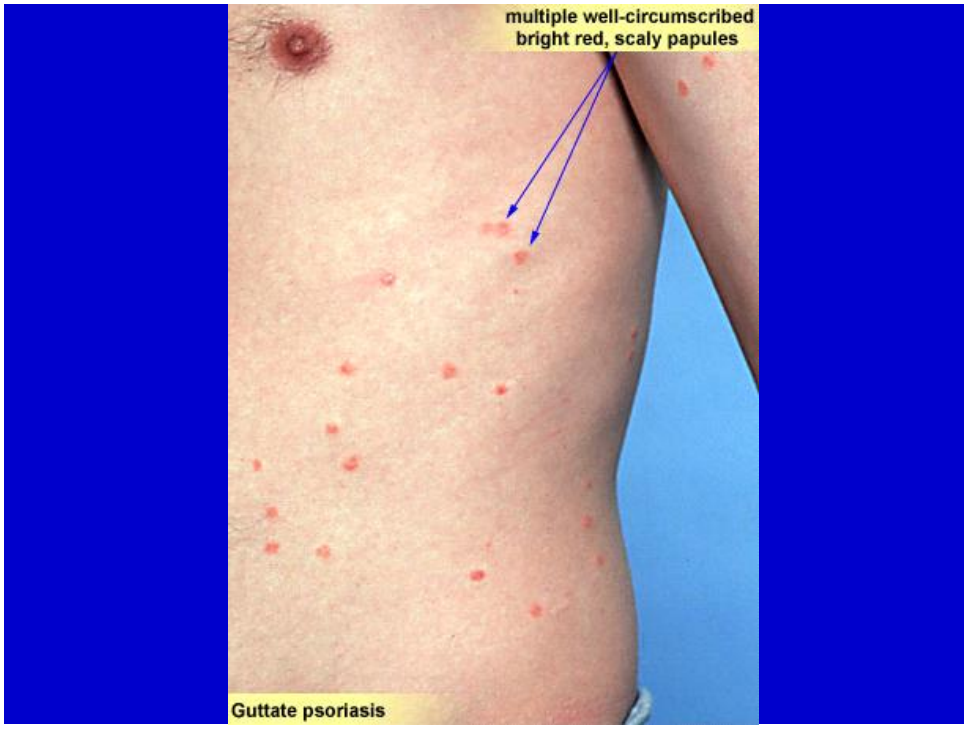
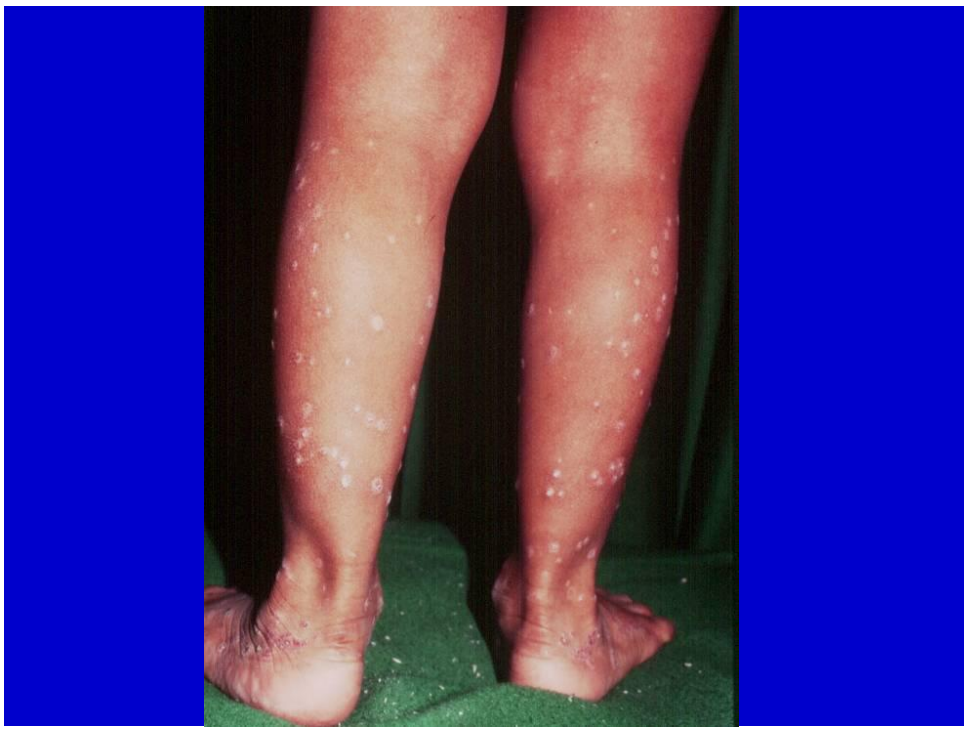
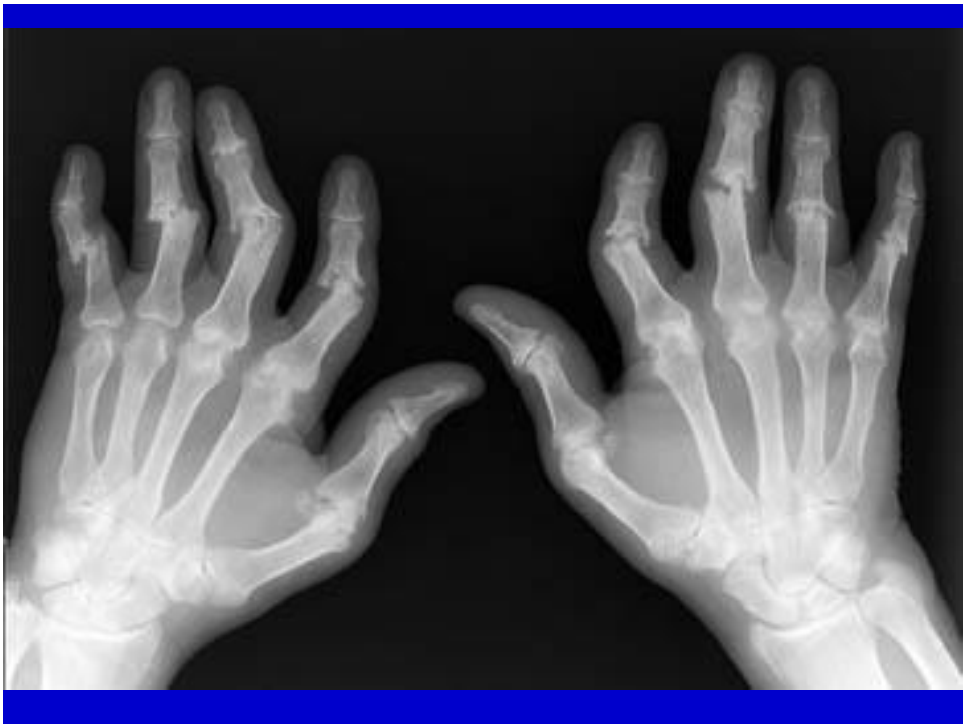

multiple well-circumscribed
bright red, scaly papules
Guttate psoriasis



Psoriatic Arthropathy

- Asymmetric distal interphalangeal joints
- Artheritis mutilans with osteolysis of phalanges and metacarpals
- Symmetric polyarthritis like, Rh. Arth. with claw hands
- Ankylosing spondylitis



- **Drug-induced Ps.:**
  May be induced by many drugs e.g. Beta blockers Lithium, antimalarials & non-steroidal anti-inflammatory agents
- **DD:**
  - Seb. dermatitis
  - Pityriasis rosea
  - Lichen planus
  - Eczema
  - Syphilis
  - SLE
- **Treatment:**
  - The lesions may disappear spontaneously or as a result of therapy
  - Treatment methods will vary according to the site, severity, duration, previous therapy and the age of the patient

Laboratory findings

- Elevated uric acid
- Mild anemia
- Negative nitrogen balance
- Increase sedimentation rate
- Increase alpha-2-microglobulin
- Incresae IgA and IgA immune complex

Differential diagnosis

Erythroderma
- Atopic dermatitis
- Sezary syndrome
- Drug eruption
- Generalized contact dermatitis

Intertrigenous psoriasis
- Candidiasis
- Contact dermatitis
- Darier's disease

Differential diagnosis

Psoriasis vulgaris
- Nummular eczema
- Mycosis fungoides, plaque stage
- Tinea corporis

Guttate psoriasis
- Pityriasis rosea
- Pityriasis lichenoides et varioliformis
- Syphillis
- Tinea corporis

Differential diagnosis

Nail psoriasis
- Tinea ungium
- Dyskeratosis : secondary to injury

Scalp and face
- Seborrheic dermatitis

Genitalia
- In situ squamous cell CA

Current Treatment Approaches

Treatment of Psoriasis

- What influences therapy choice?
  - Clinical type and severity of psoriasis (eg, mild vs moderate-to-severe), assessed by Psoriasis Area and Severity Index (PASI)
  - Response to previous treatment
  - Therapeutic options
  - Patient preference
- The "1-2-3" step approach is no longer generally accepted for disease more than mild in severity
  - Level 1: Topical agents—do not work
  - Level 2: "Phototherapy"—difficult; not always available
  - Level 3: Systemic therapy
- Risk in relation to benefit must be evaluated

Camisa C. *Cleve Clin J Med*. 2000;67:105-119.

Topical Agents

- Initial therapeutic choice for mild-to-moderate psoriasis
  - Emollients
  - Keratolytics (salicylic acid, lactic acid, urea)
  - Coal tar
  - Anthralin
  - Vitamin D_3 analogues (calcipotriene)
  - Corticosteroids
  - Retinoids (tazarotene, acitretin)
- Compliance can be difficult due to amount of time required to apply topicals 2 to 4 times/day

Camisa C. *Cleve Clin J Med*. 2000;67:105-119.

Phototherapy

- Used to treat moderate-to-severe psoriasis
- Phototherapy causes death of T cells in the skin
  - Natural sunlight
  - Ultraviolet (UV) B light
  - UVB light + coal tar (Goeckerman treatment)
    - Best therapeutic index for moderate-to-severe disease
  - UVB light + anthralin + coal tar (Ingram regimen)
  - Usually 3 treatments/week for 2 to 3 months is needed
  - Accessibility to a light box facility and compliance necessary

Camisa C. *Cleve Clin J Med*. 2000;67:105-119.

Systemic Therapy

- Systemic therapy should be reserved for patients with disabling psoriasis despite topical therapy
  - Psoralen + UVA light
  - Oral retinoids: acitretin (+/- phototherapy)
  - Methotrexate
  - Cyclosporine

Camisa C. *Cleve Clin J Med*. 2000;67:105-119.

UVA Light With Psoralen (PUVA)

- Psoralen is a drug that causes a toxic reaction to skin lymphocytes when it is activated by UVA light
- Psoralen can be given systemically or topically
- Effective treatment—longest remissions of any treatment available
- Adverse effects
  - Nausea, burning, pruritus
  - Risk of cancer with cumulative use—both squamous cell carcinoma and melanoma
    - >160 cumulative treatments

Greaves MW, et al. *N Engl J Med*. 1995;332:581-588.

Methotrexate

- Folic acid metabolite
  - Blocks deoxyribonucleic acid synthesis, inhibits cell proliferation
- Dose
  - Start at about 15 mg/week; maximum 30 mg/week
  - Can also be given intramuscularly
- Adverse effects
  - Headache, nausea, bone marrow suppression
  - Cumulative dose predictive of liver toxicity
    - Prospectively identify risk factors for liver disease
    - Guidelines recommend liver biopsy after 1.5 g
    - Teratogenic in men and women

Greaves MW, et al. *N Engl J Med*. 1995;332:581-588.

Acitretin: Oral Retinoid

- Frequently used in combination with topical agents, systemic therapies, and UV light
- Less effective as monotherapy for plaque psoriasis
- Plaque psoriasis dose
  - Start at 10 to 25 mg/day
- Adverse effects (fewest dose-related adverse effects)
  - Peeling/dry skin, alopecia, muscle pain
  - Lipid abnormalities
- Teratogenic: avoid pregnancy

Greaves MW, et al. *N Engl J Med*. 1995;332:581-588.

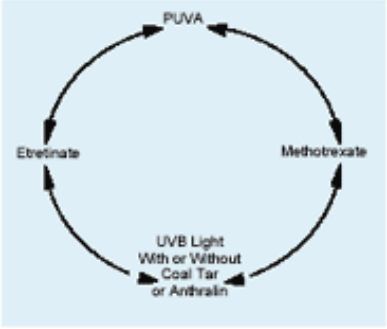
Cyclosporine

- Reserved for severe, recalcitrant disease
- Inhibits the proliferation of activated T cells
- Dose: 4 mg/kg/day, not to exceed 5 mg/kg/day
  - Tapering slowly may improve remission
- Use not recommended for >1 year
  - Renal toxicity
- Patients relapse 2 to 4 months after discontinuing
- Adverse effects
  - Immunosuppression: infections, possible malignancy
  - Hirsutism, gingival hyperplasia, muscle pain, infection
  - Serious: hypertension, renal failure

Lebwohl M, et al. *J Am Acad Dermatol*. 1998;39:464-475.

Rotational/Sequential Treatment

- Therapeutic strategy for moderate-to-severe disease
  - Switch to alternative therapy before early evidence of toxicity

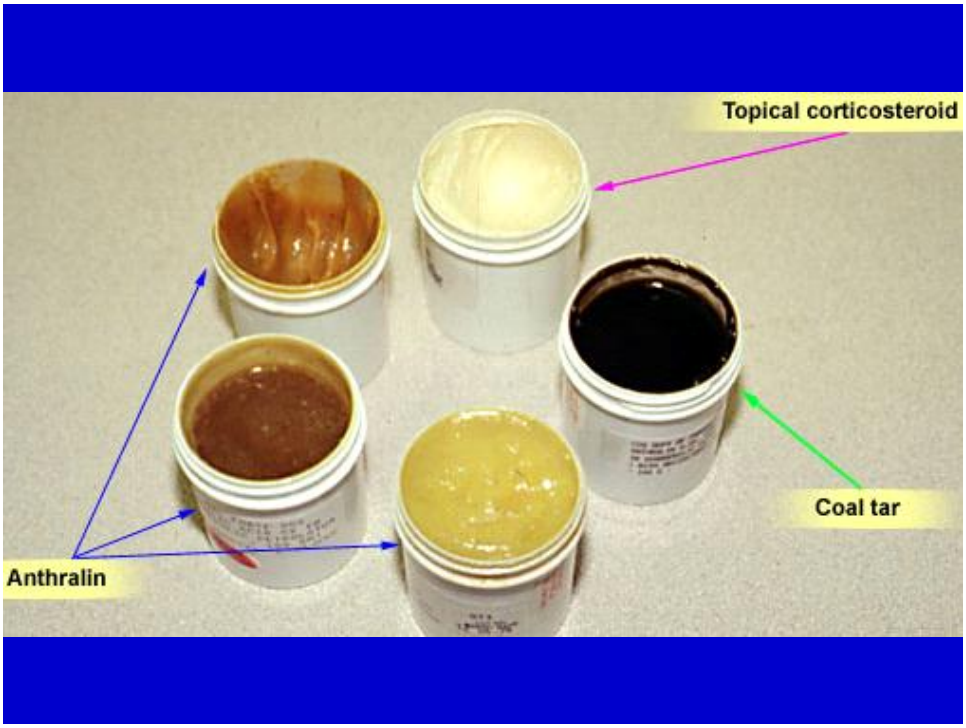


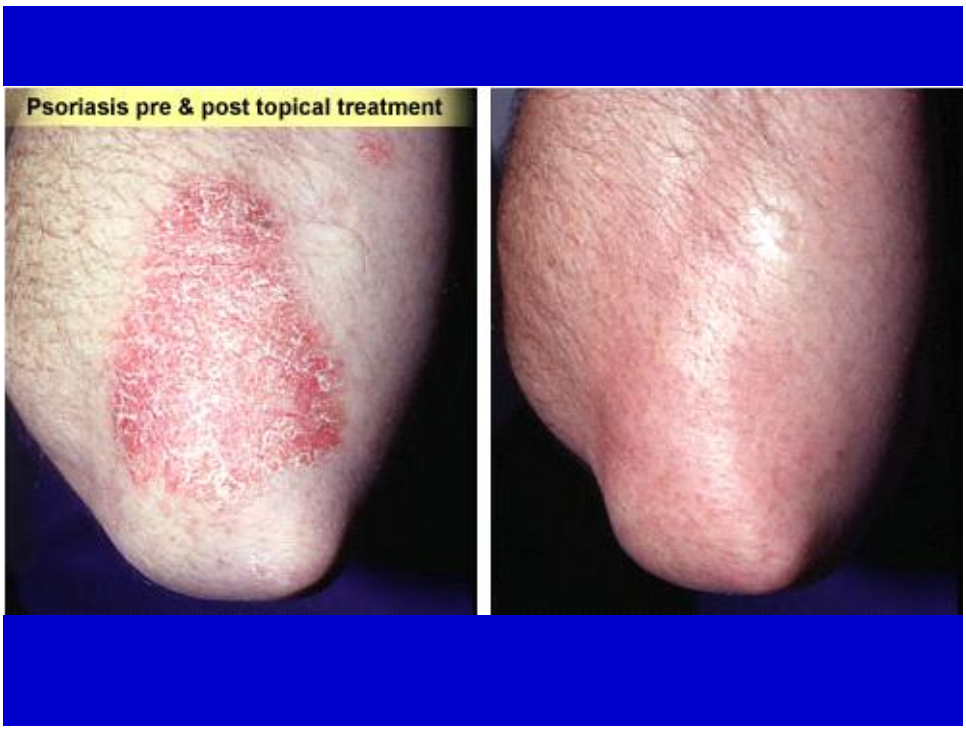
Greaves MW, et al. *N Engl J Med*. 1995;332:581-588.

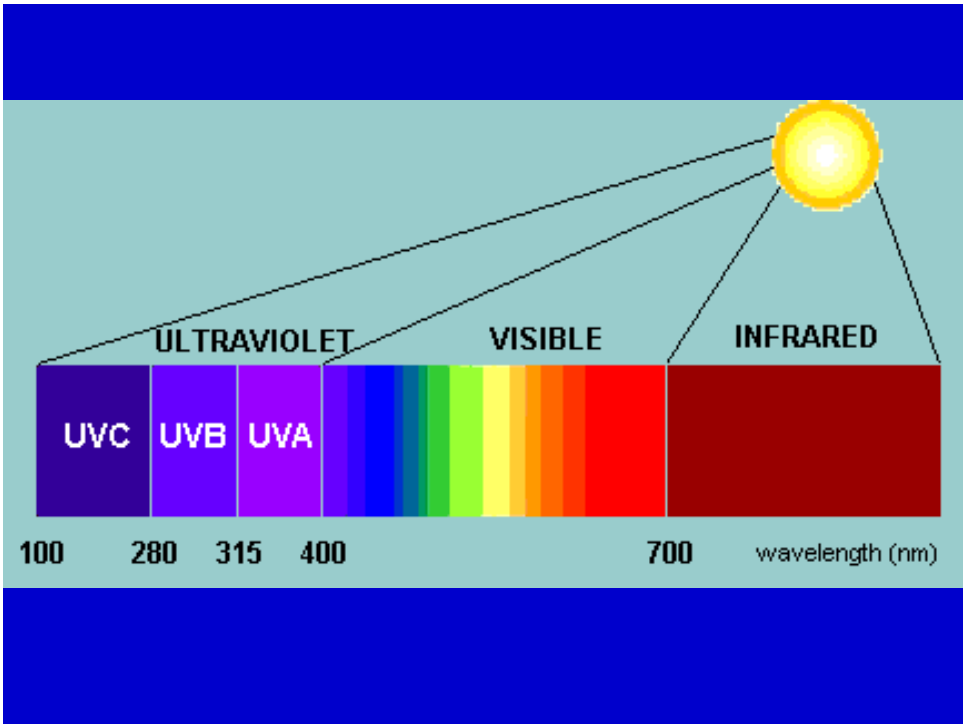
New Treatments

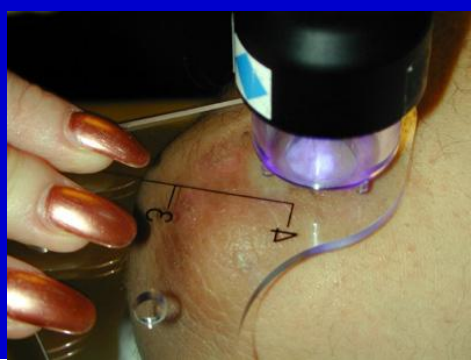
Biologic Therapies Currently Approved for the Treatment of Psoriasis

- Alefacept
- Efalizumab
- Etanercept


Topical corticosteroid
Coal tar
Anthralin


Psoriasis pre & post topical treatment


ULTRAVIOLET
VISIBLE
INFRARED
UVC
UVB
UVA
100
280
315
400
700
wavelength (nm)



Pityriasis Rosea

Acute, self-limiting, mild inflammatory exanthem of unknown origin.

Etiology:

- Unknown
- A virus infection is most frequently suggested?
  - The formation of herald patch
  - The self-limited course
  - The seasonal preponderance & rare recurrence
- The Pit. rosea-like may occur as a reaction to:
  - Captopril
  - Gold
  - Clonidine
  - Barbiturates
  - Arsenicals
  - Bismuth
  - Methoxypromazine

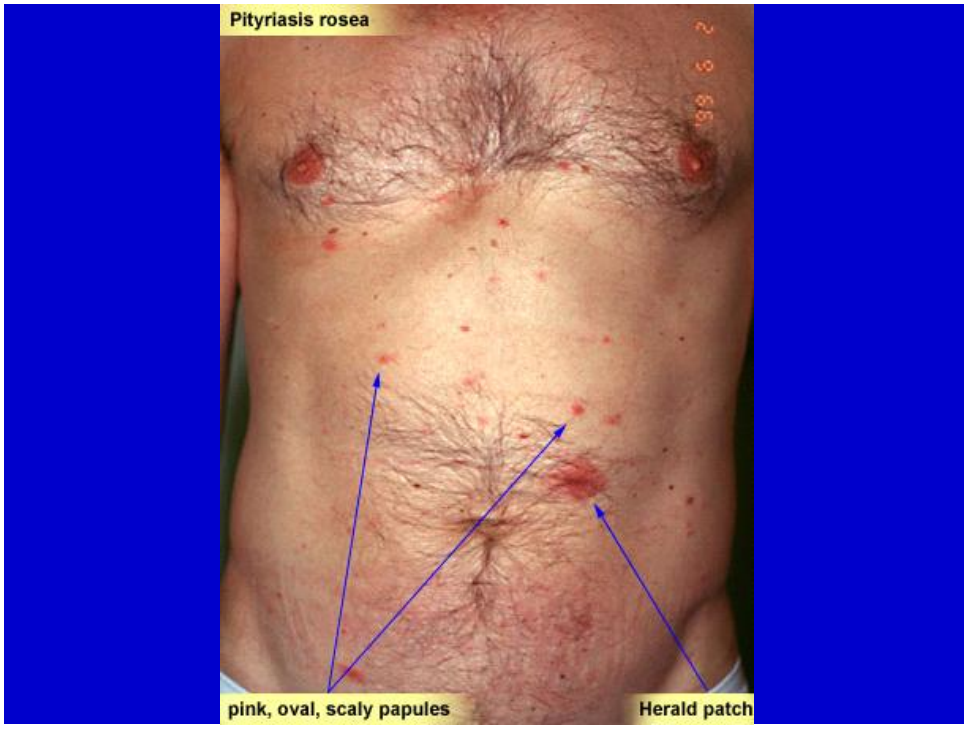
Pityriasis Rosea

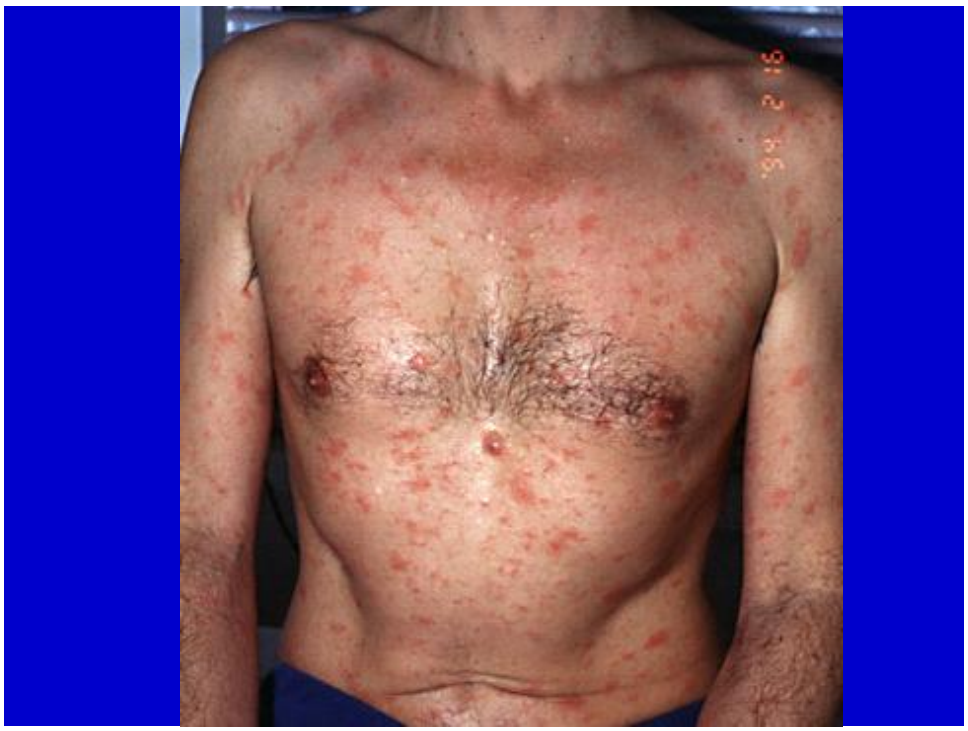
Clinical features

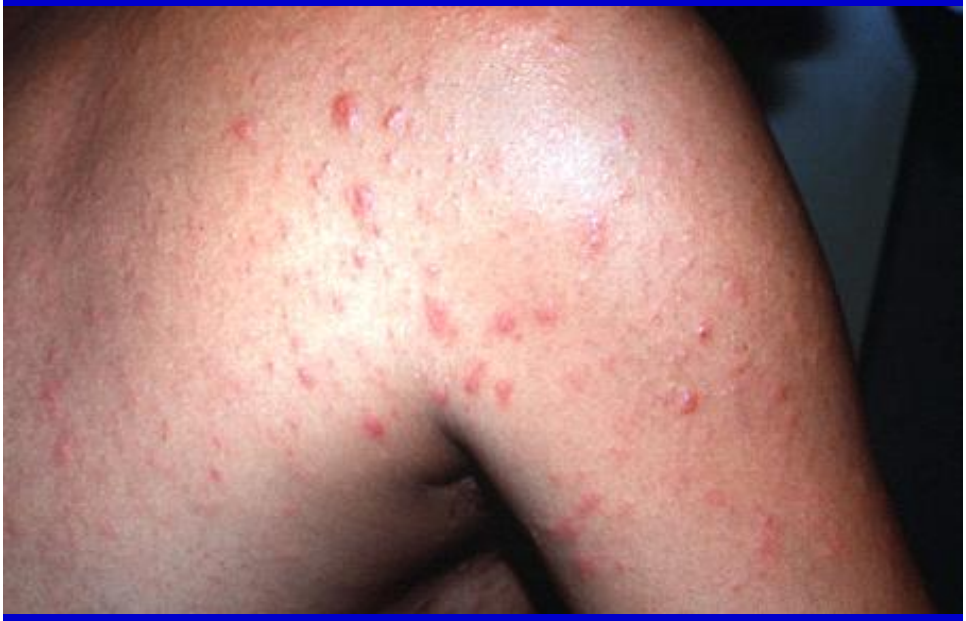
- Salmon-colored papular & macular lesions
- Oval patches or circinate covered with finely crinkled, dry epidermis ➡ often desquamates

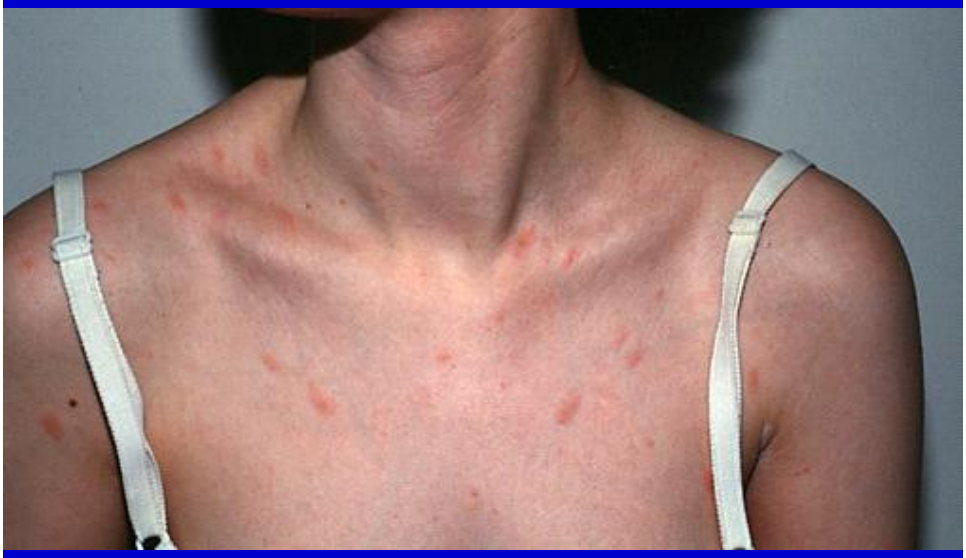
oval

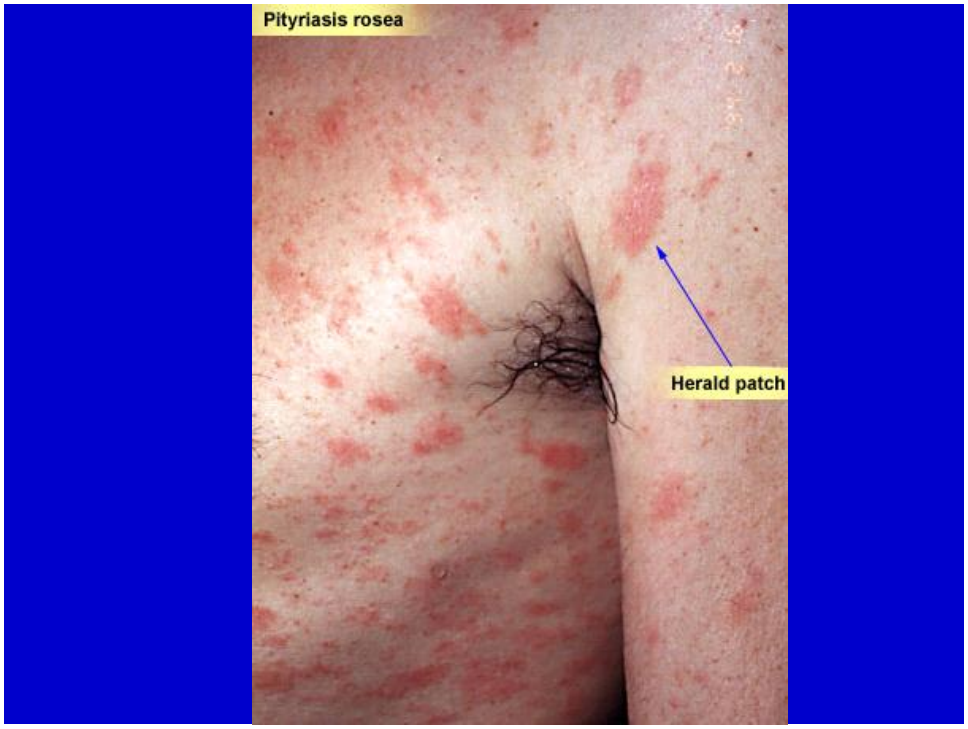
- Usually begins with a single - herald or mother patch
- The new lesions spread rapidly
- Arranged - runs parallel to the lines of cleavage
- Generalized, affecting the trunk & sparing the sun-exposed surfaces
- Moderate pruritus may be present
- Variations in the mode of onset, course and clinical manifestations are common (papular Pit. Rosea)

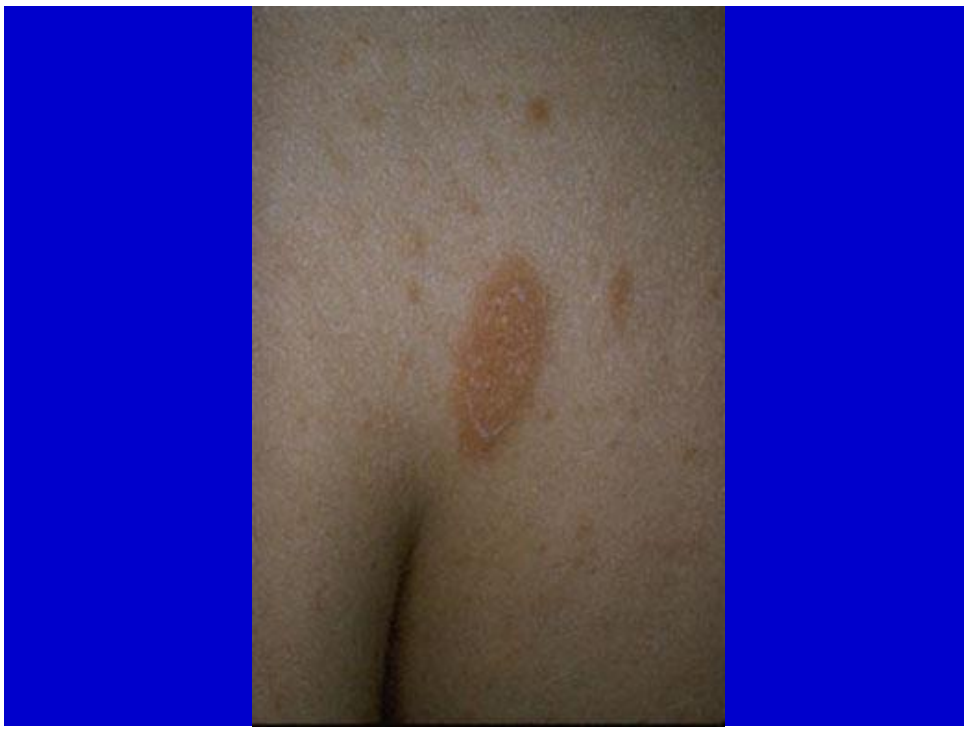

Pityriasis rosea
pink, oval, scaly papules
Herald patch

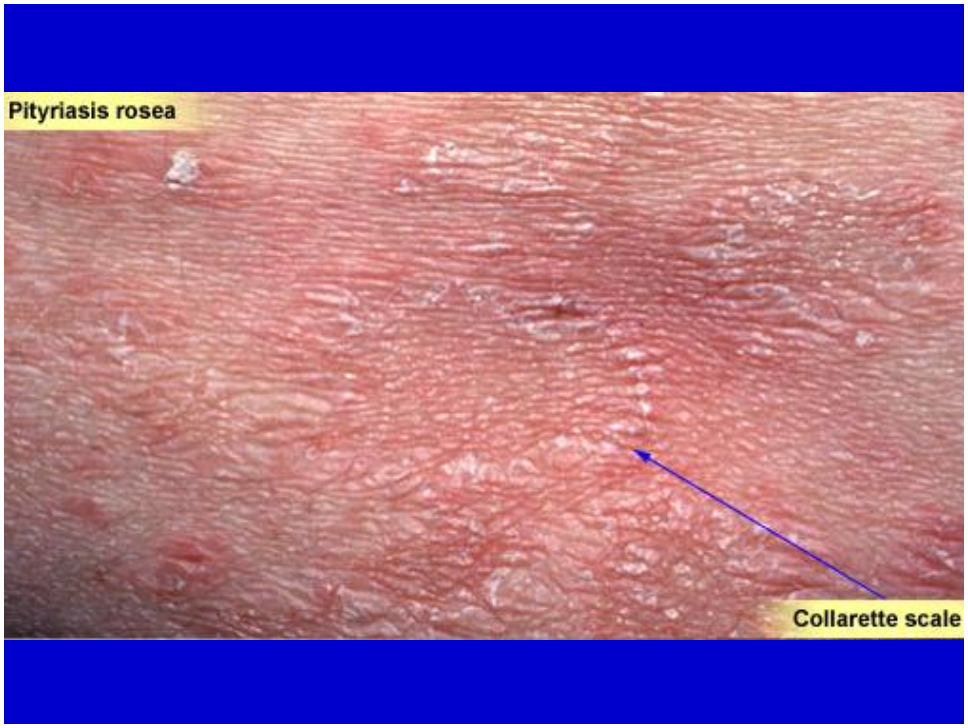


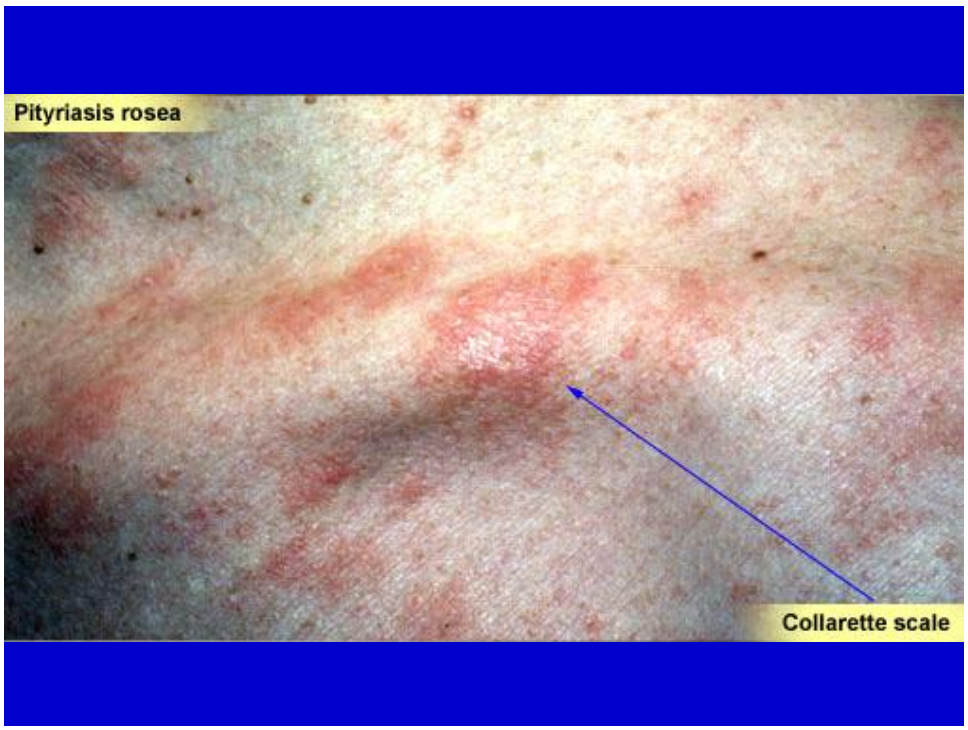


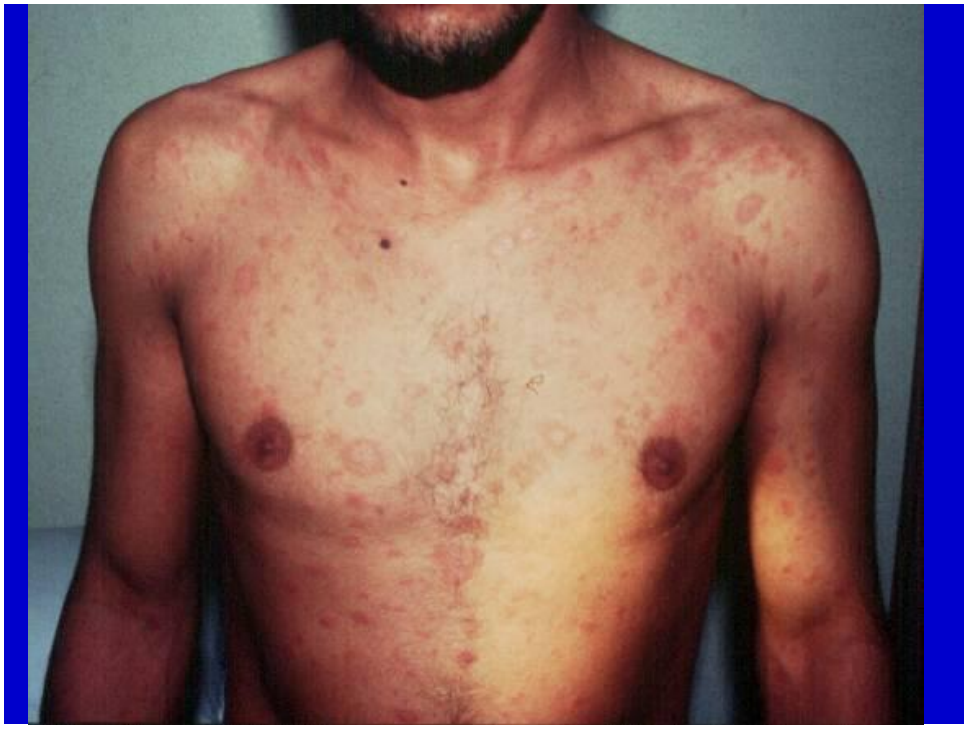


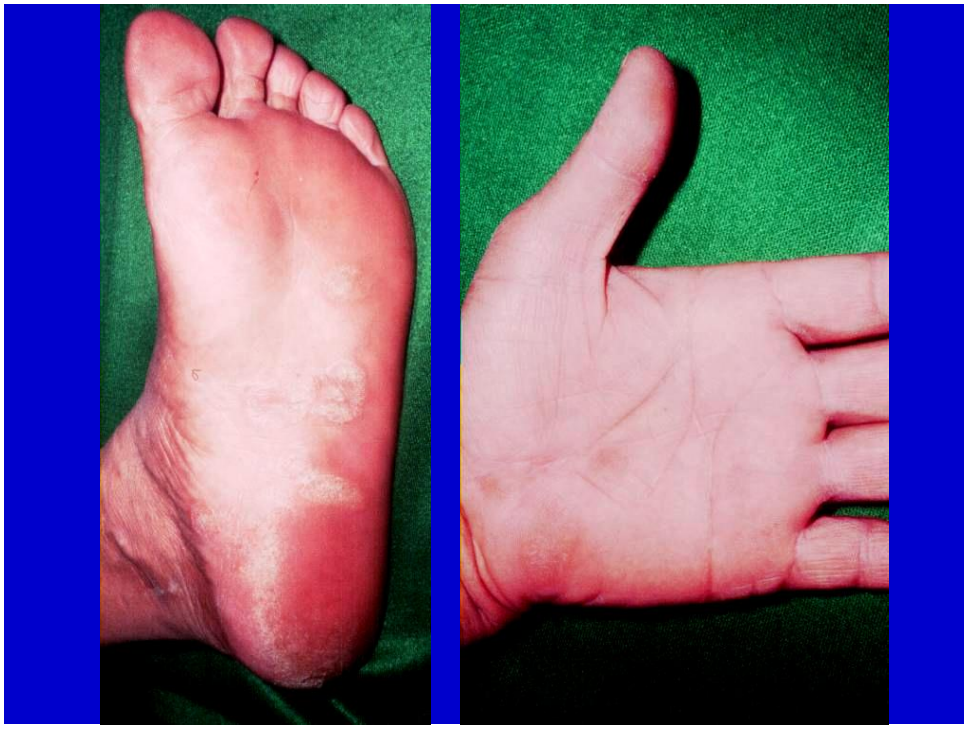

Pityriasis rosea
Herald patch




Pityriasis rosea
Collarette scale


Pityriasis rosea
Collarette scale





Pityriasis Rosea

Differential diagnosis

- Seborrheic dermatitis
- Tinea Circinata (T.C.)
- Macular Syphilid (secondary syphilis)
- Drug eruption
- Viral exanthema
- Psoriasis

Pityriasis Rosea

Treatment

- Prevent irritable hot baths & soaps and woolen clothes
- Symptomatic
- Emollients
- Corticocosteroid (Topical, Oral, IM)
- UVB

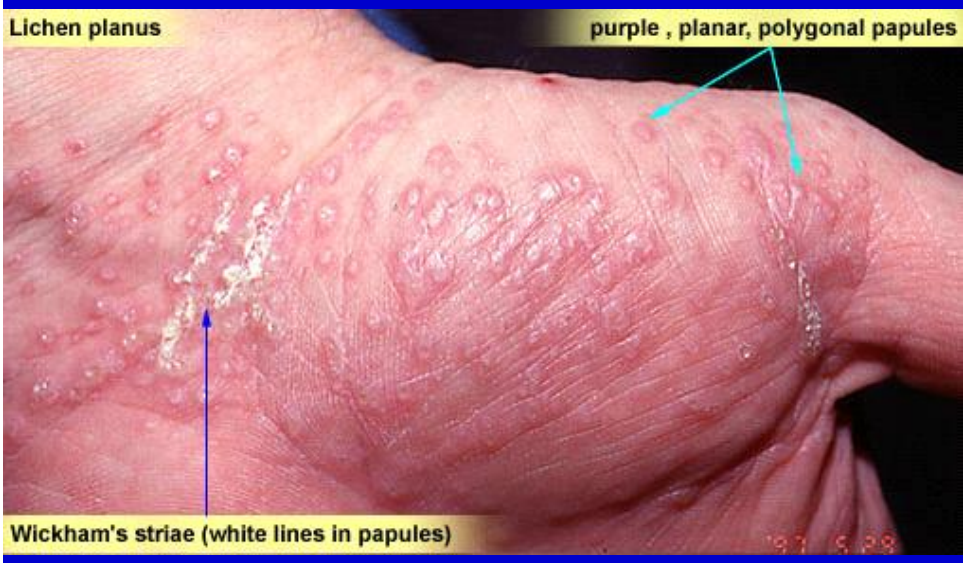
Lichen Planus & Lichenoid Eruption

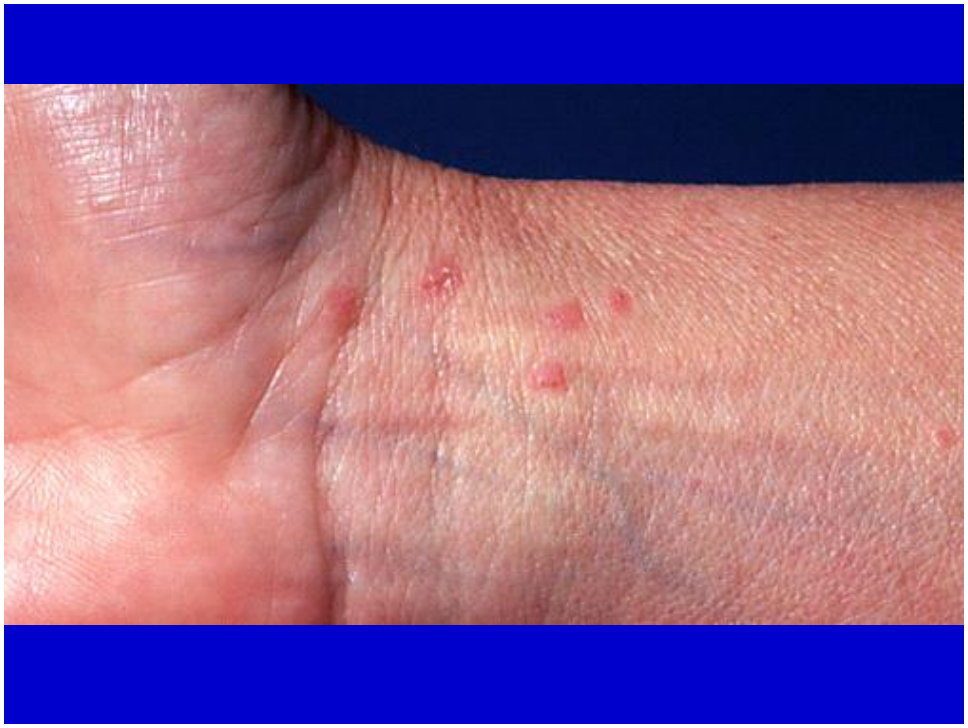
- Inflammatory pruritic disease of the skin and mucous membranes
- Rare in children
- Etiology:
  - The cause of LP remains unknown
  - ?? an alteration of epidermal cell antigens induce a cell mediated immune response
  - ?? may be familial (early age & chronic)
  - Drugs ➡ may induce lichenoid reactions (e.g. antimalarials, thiazide derivatives, propranolol..)
  - Viral infection? Symmetrically associated with viral hepatitis
  - A psychogenic origin? Severe psychic trauma
  - An auto-immune phenomenon?

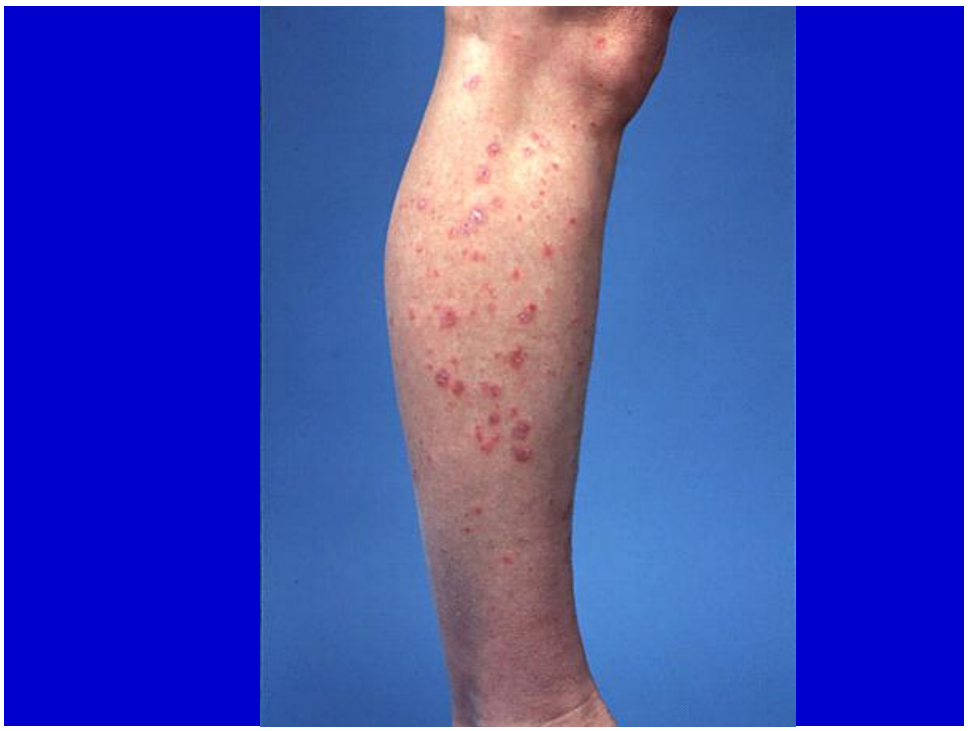
Clinical Features

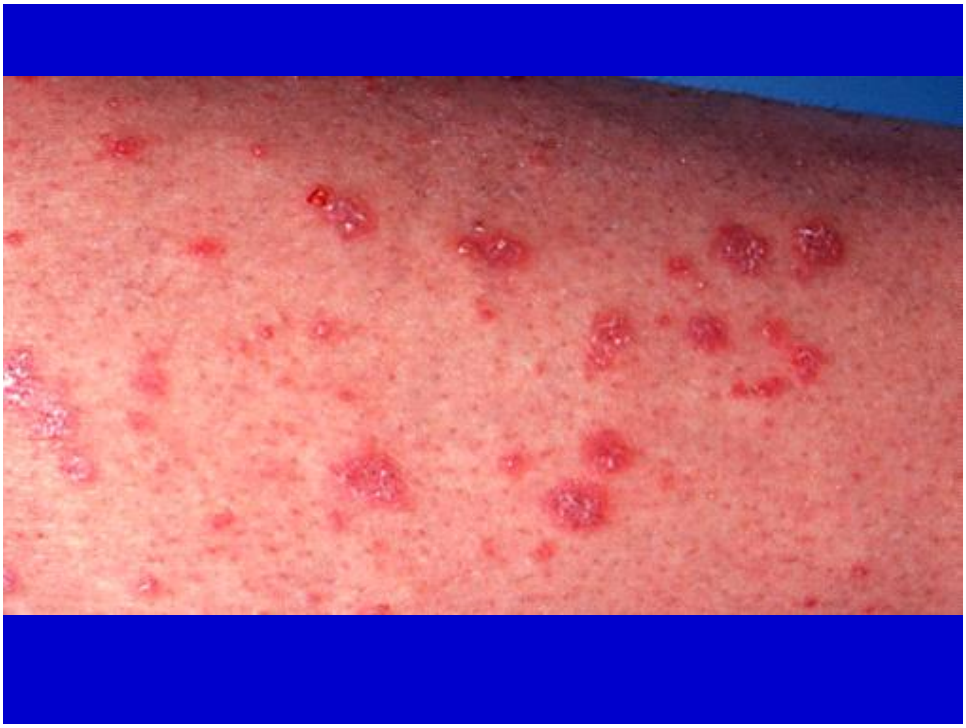
- Initial lesions: Flat topped dry with scanty adherent scales shiny, polygonal, violaceous (violet) itchy papular eruption, sometimes centrally umblicated
- Occurs especially on:
  - The volar aspects of the wrists
  - Medial sides of the thighs
  - Shins
  - Back of the hands
  - The glans penis
  - The disease may appear on any part of the body but rarely on the face (except the upper eyelids and lips
- After the LP lesions have disappeared ➡ deep pigmentation may persist for several months

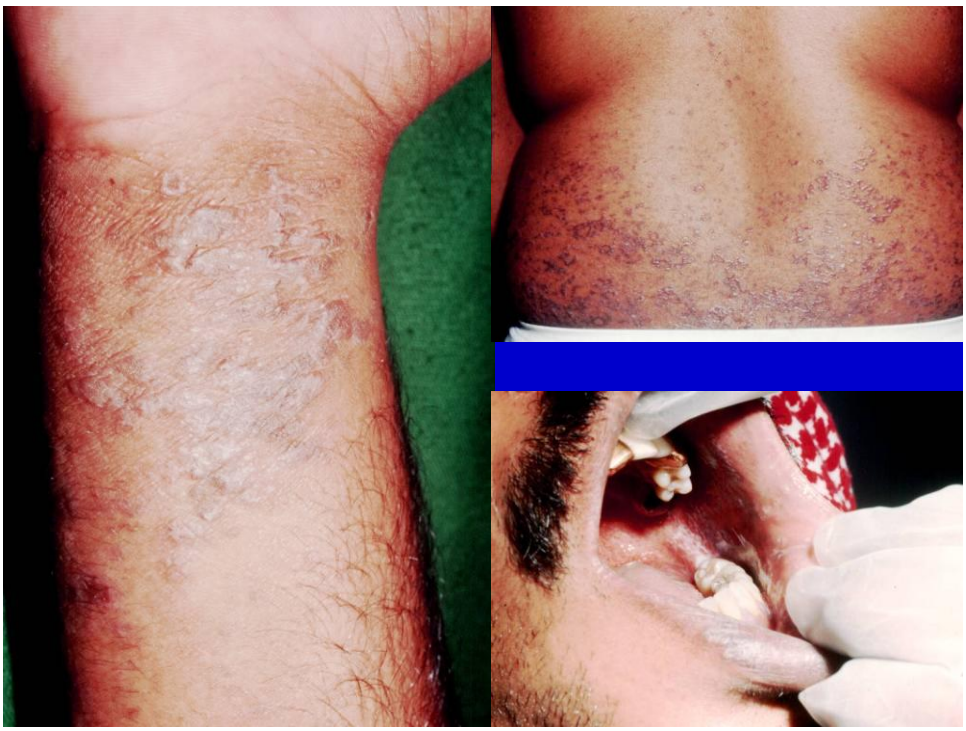
- **Wickham's striae:**
  Grayish puncta or streaks which form a network on the surface of the papules (focal increase in thickness of granular layer & infiltrate)
- **Koebner's isomorphic phenomenon:**
  As in psoriasis by physical trauma (scratching) skin lesions are produced in the scratch marks identical to those already on the pat. skin.
- **Pruritus:**
  - It is intolerable in acute cases
  - Most pat. react by rubbing rather than scratching

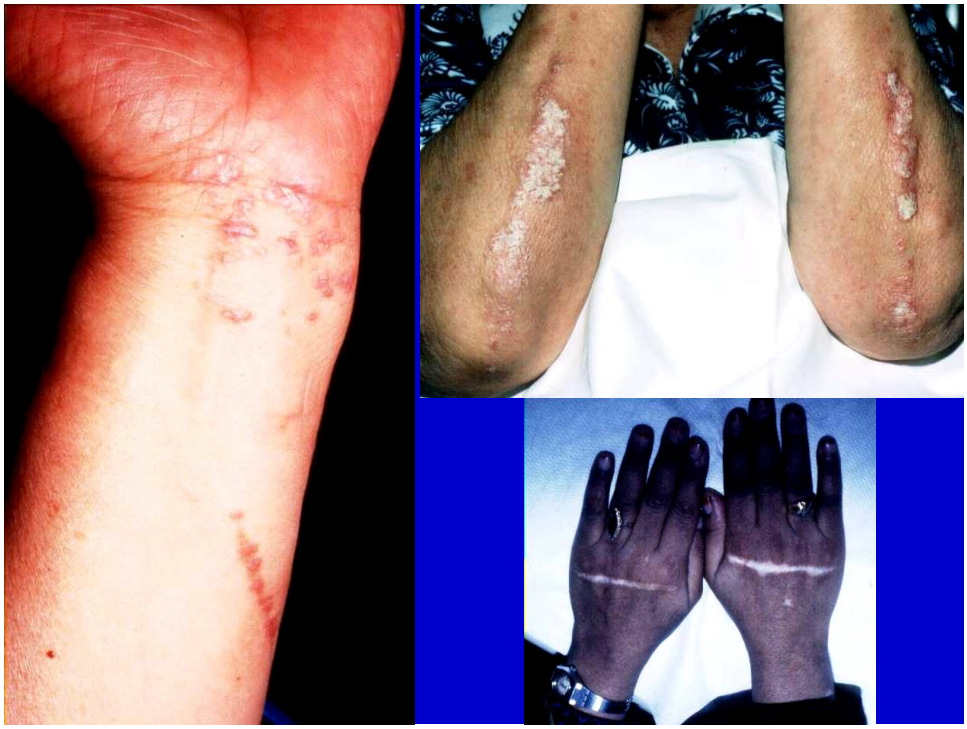


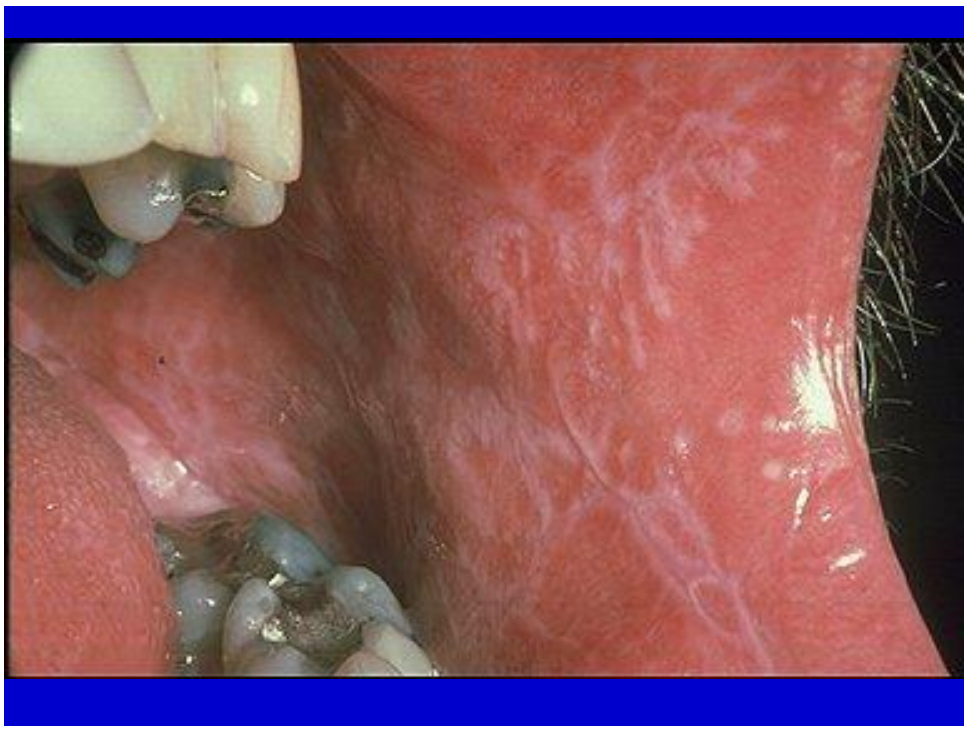


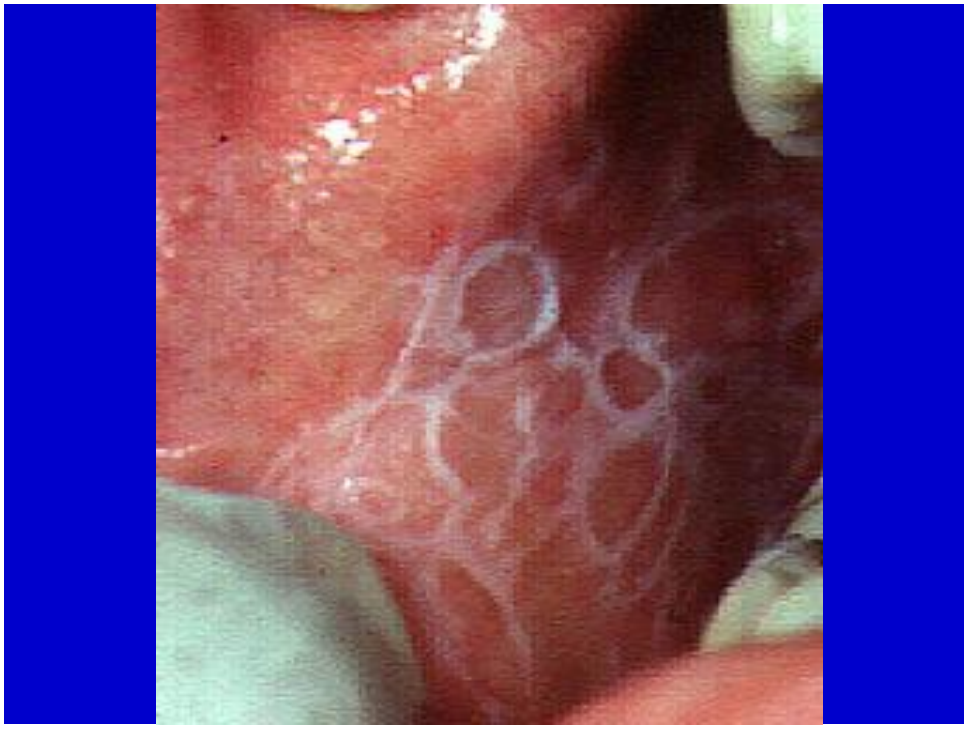


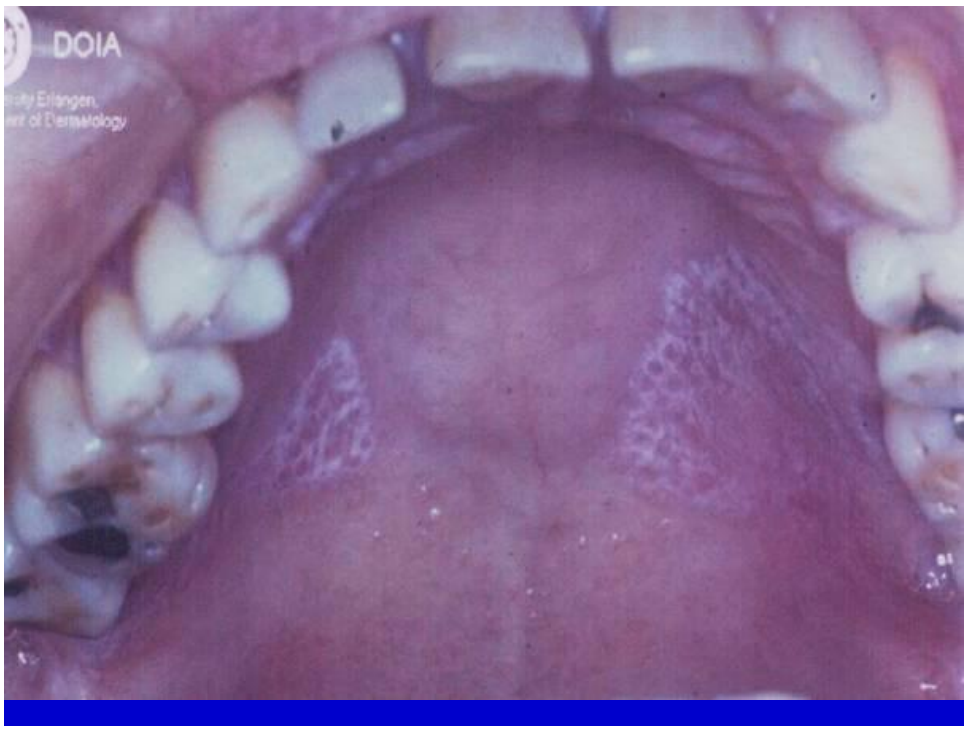








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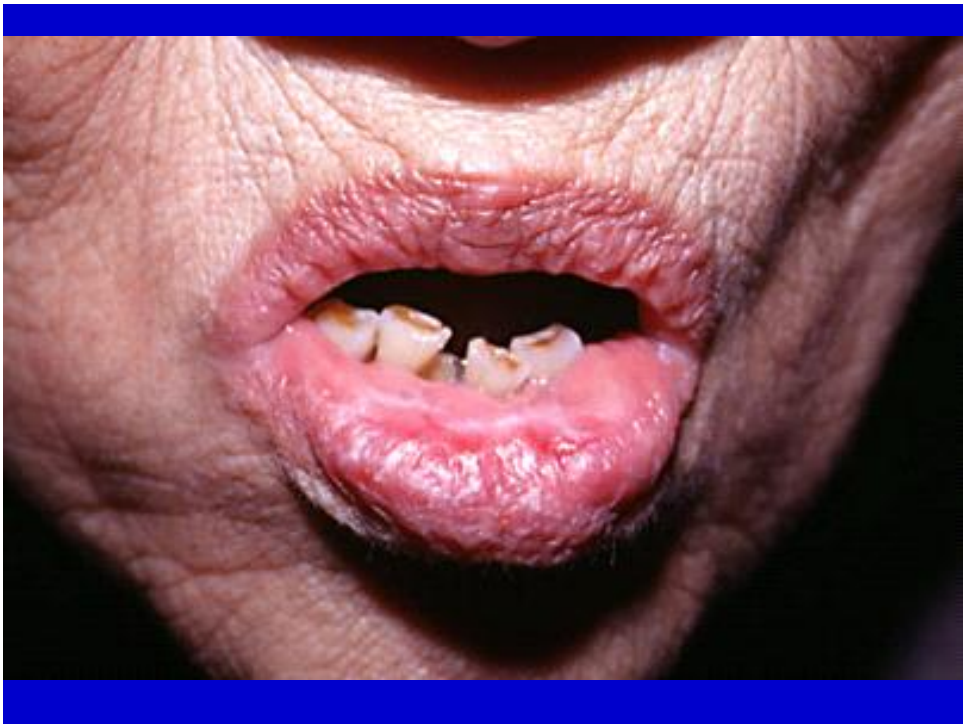

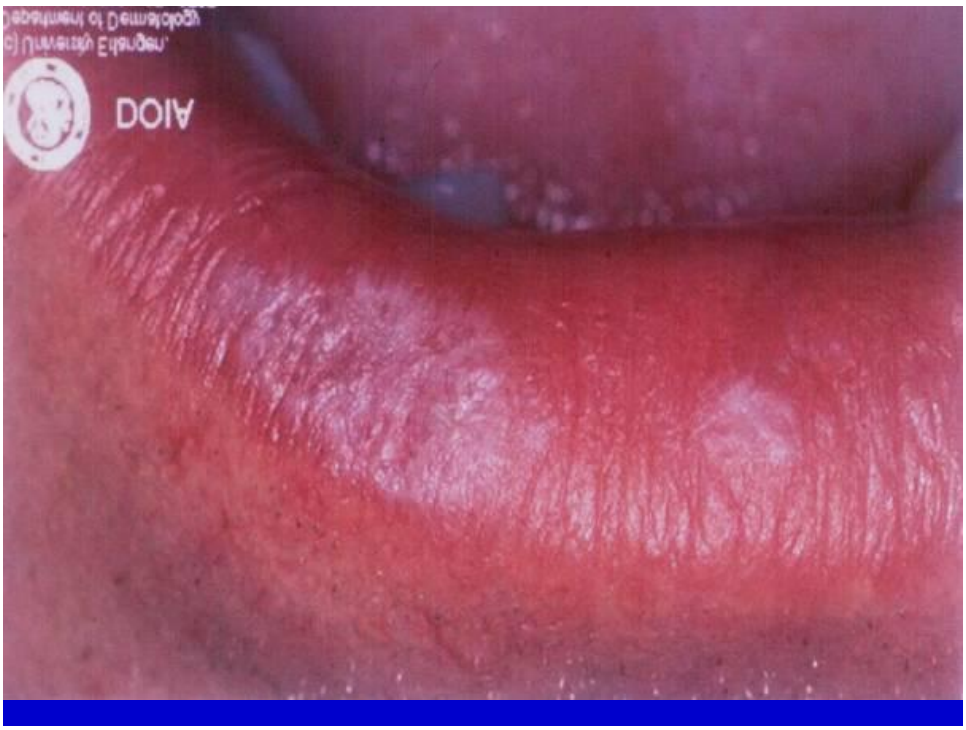

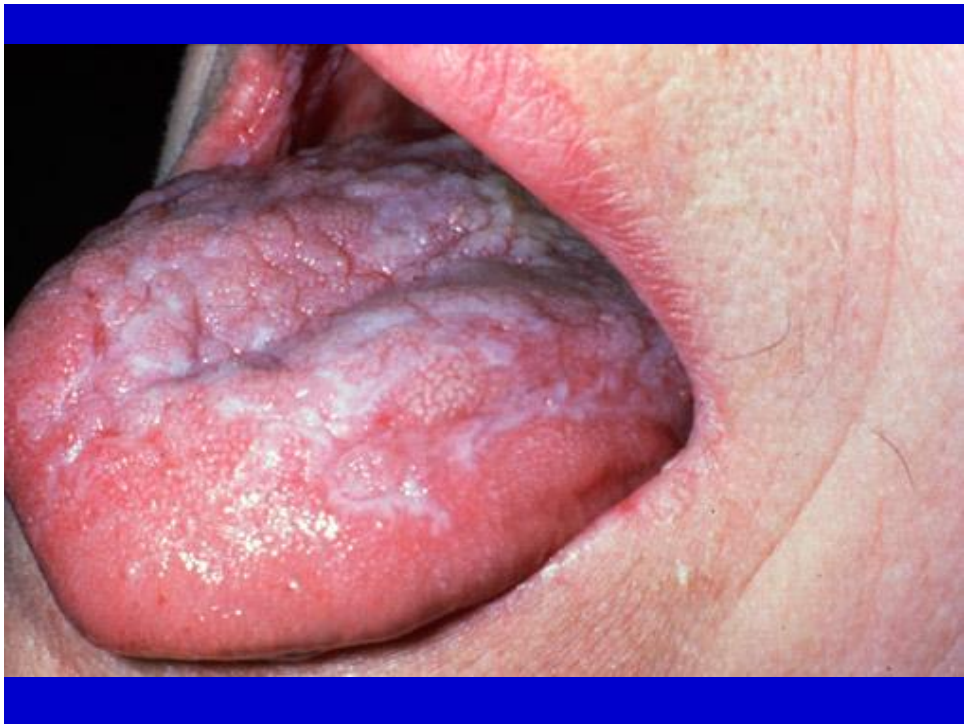

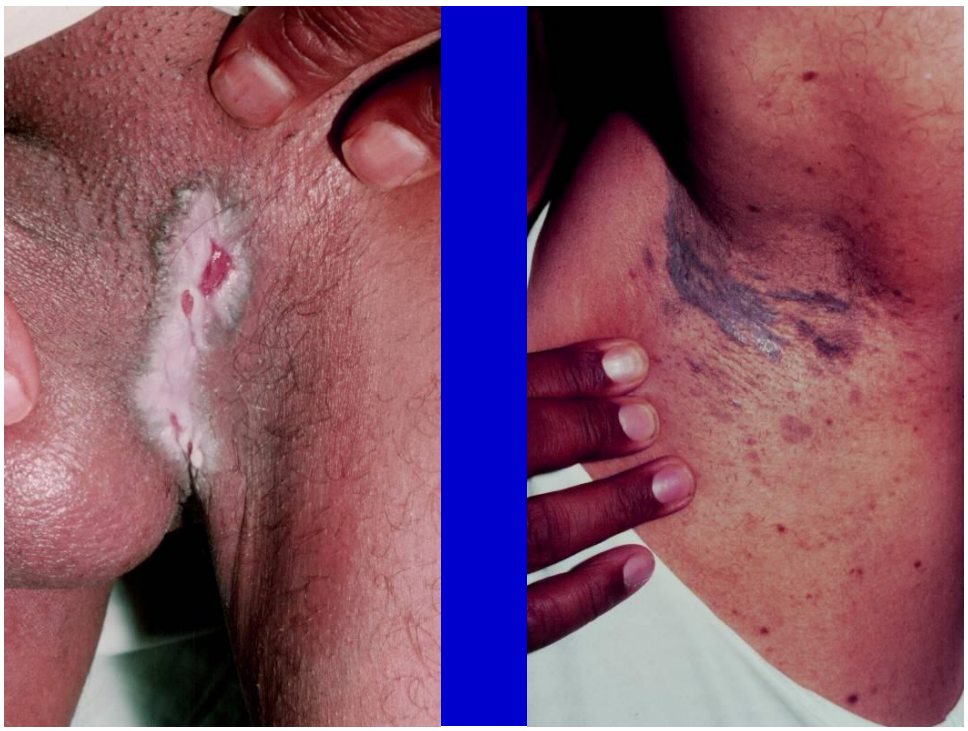

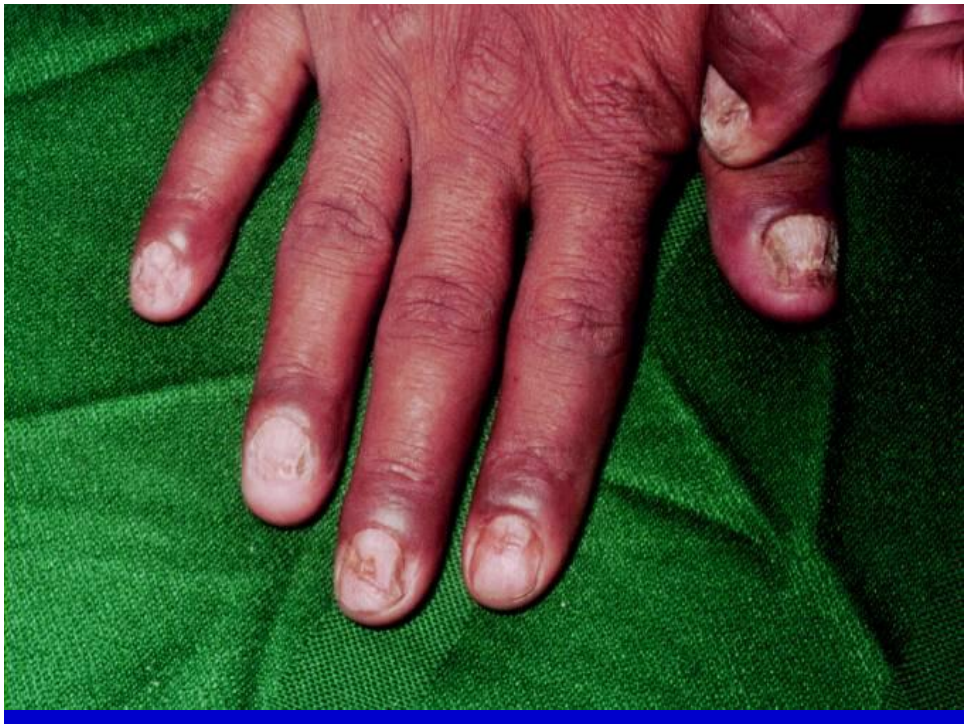

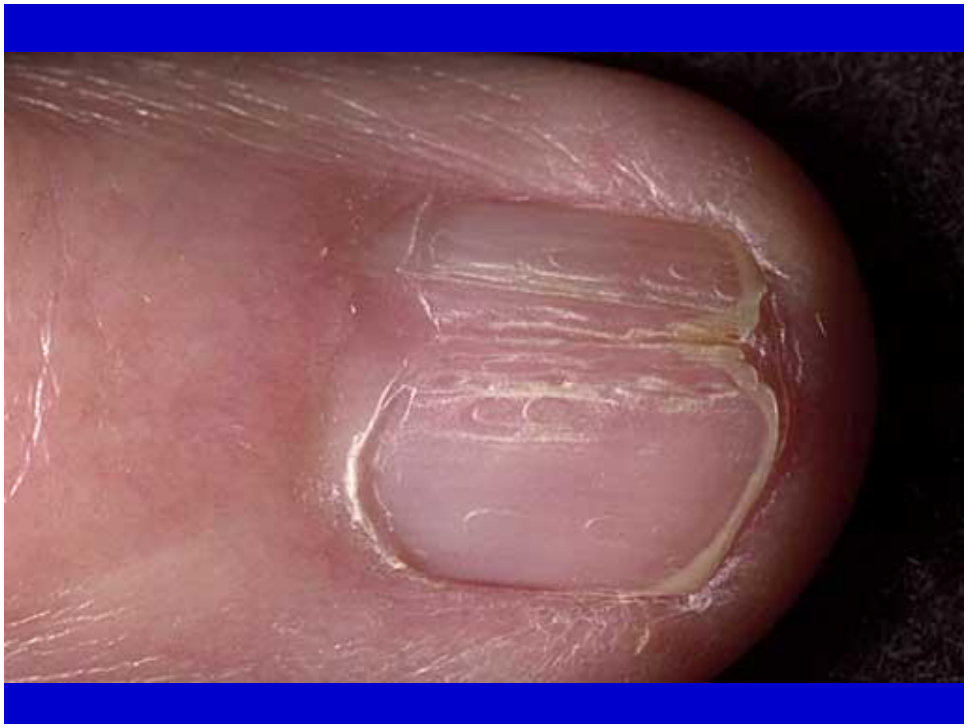

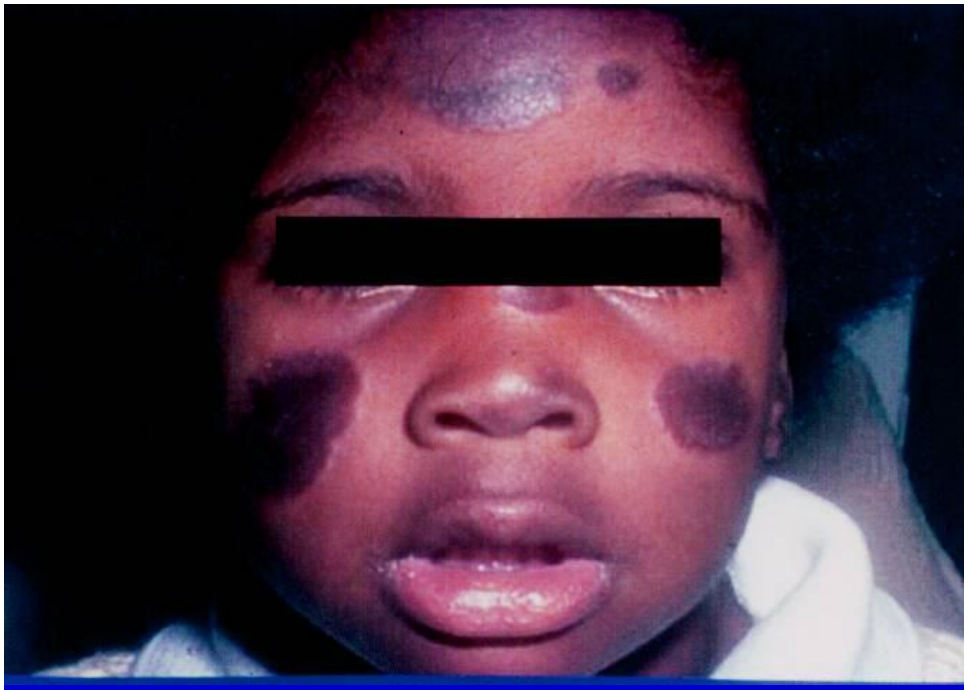

Diagnostic Features (L.P.)

- Very itchy
- Purple, flat-topped, shiny, polygonal papules
- May occur anywhere (wrists, ankles, arms, legs, genitalia)
- Lace-like patterning on the buccal mucosa
- Koebner phenomenon
- Leaves hyperpigmentation as it heels

A Skin Manifestations May Occur in LP

- Acute widespread LP
- Chronic localized LP
- Hypertrophic LP (verrucosus)
- LP atrophicus
- Errosive & Bullous oral lesions of LP
- Ulcerative LP
- Hepatitis - associated LP

A Skin Manifestations May Occur in LP (Cont...)

- LP - LE overlap
- LP - erythematosus
- Follicular LP (Lichen Planopilaris)
- Graham - Little - Piccardi - Lassueur Syndrome
- Annular LP
- LP - Tropicus (subtropicus)
- LP - Pemphigoids

Differential diagnosis

- Papular syphilis
- Guttate psoriasis
- Lichenoid forms of (eczema, scabies)
- Pityriasis rosea
- Leukoplakia (mucous m.)

Management

- The condition is neither serious nor infectious
- The average patient is free of it within few months
- Topical steroids - under polythene (occlusion)
- Systemic steroids - 30 - 35 mg, Prenisolone ➡ Decrease 5 mg/wk.
- Antihistamines - sedative antihistamine
- Rx of specific variants (hypertrophic, oral lesions)