Papulosquamous diseases

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Learning Objectives:

-Define the papulosquamous disease.
-know the pathogenesis of papulosquamous diseases.
-Discuss the clinical features of papulosquamous diseases.
-Highlight on the papulosquamous diseases treatment

The term squamous refers to scaling that represents thick stratum corneum and thus implies an abnormal keratinization process

Papulosquamous diseases:

group of disorders characterized by scaly papules and plaques :

Psoriasis

Lichen planus

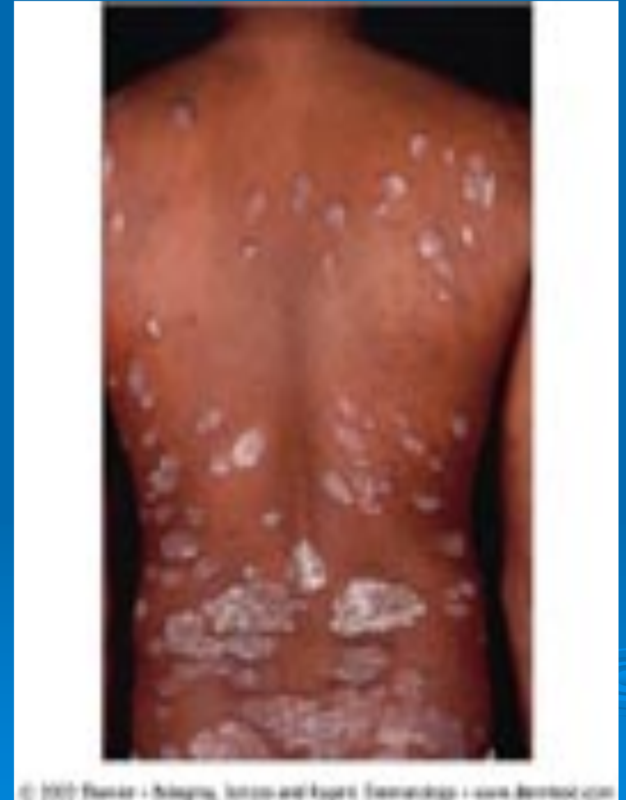
Pityriasis rosea

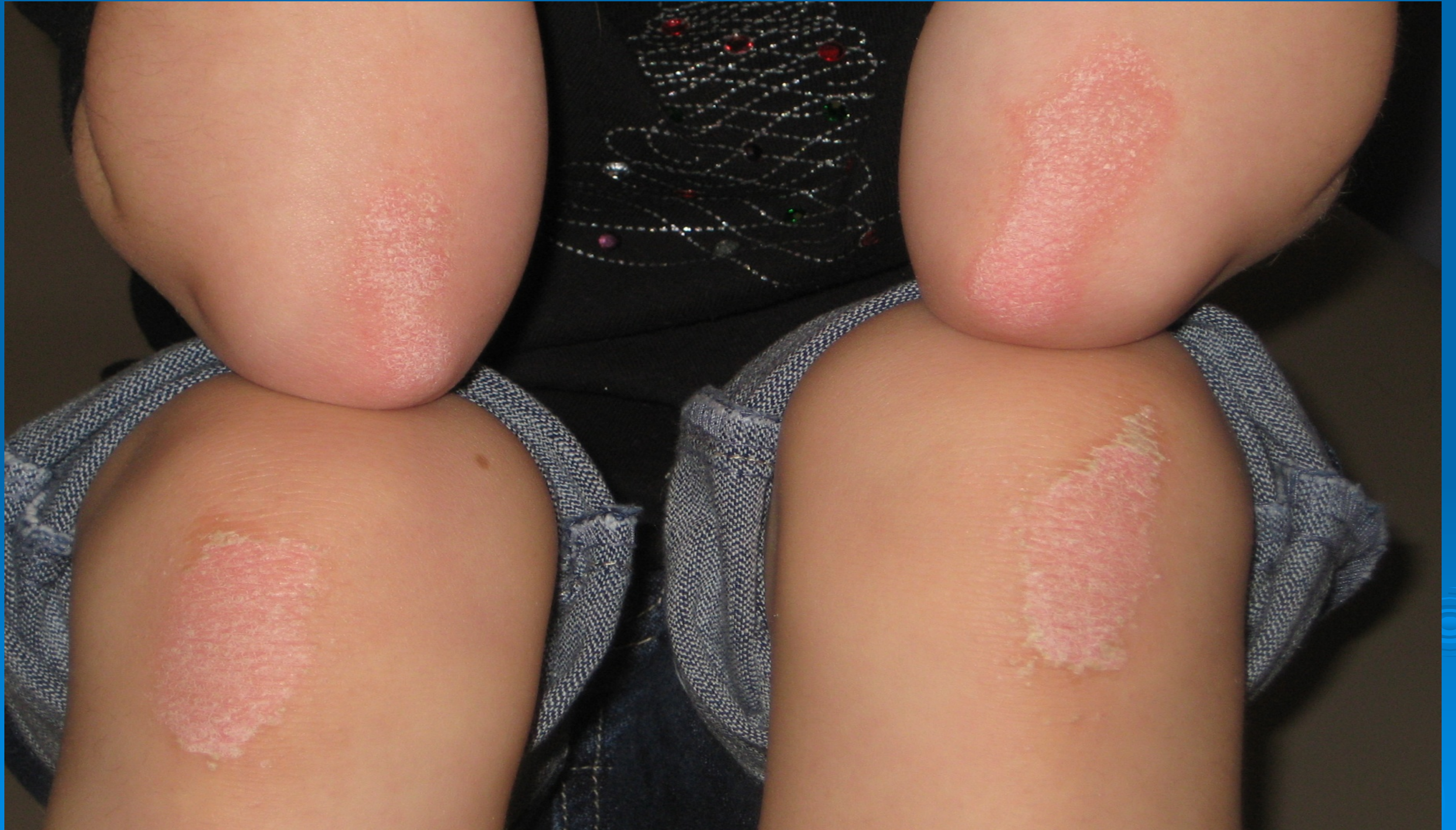
Pityriasis rubra pilaris

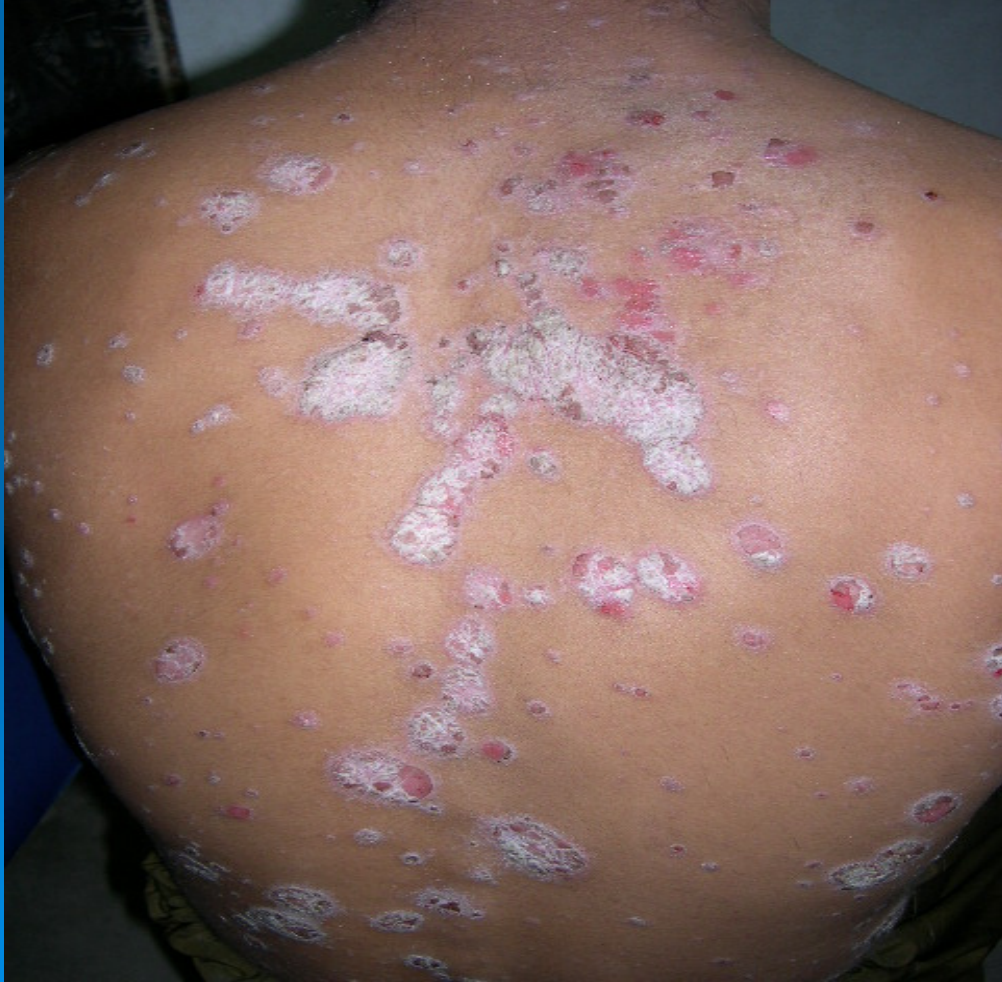
Secondary syphilis

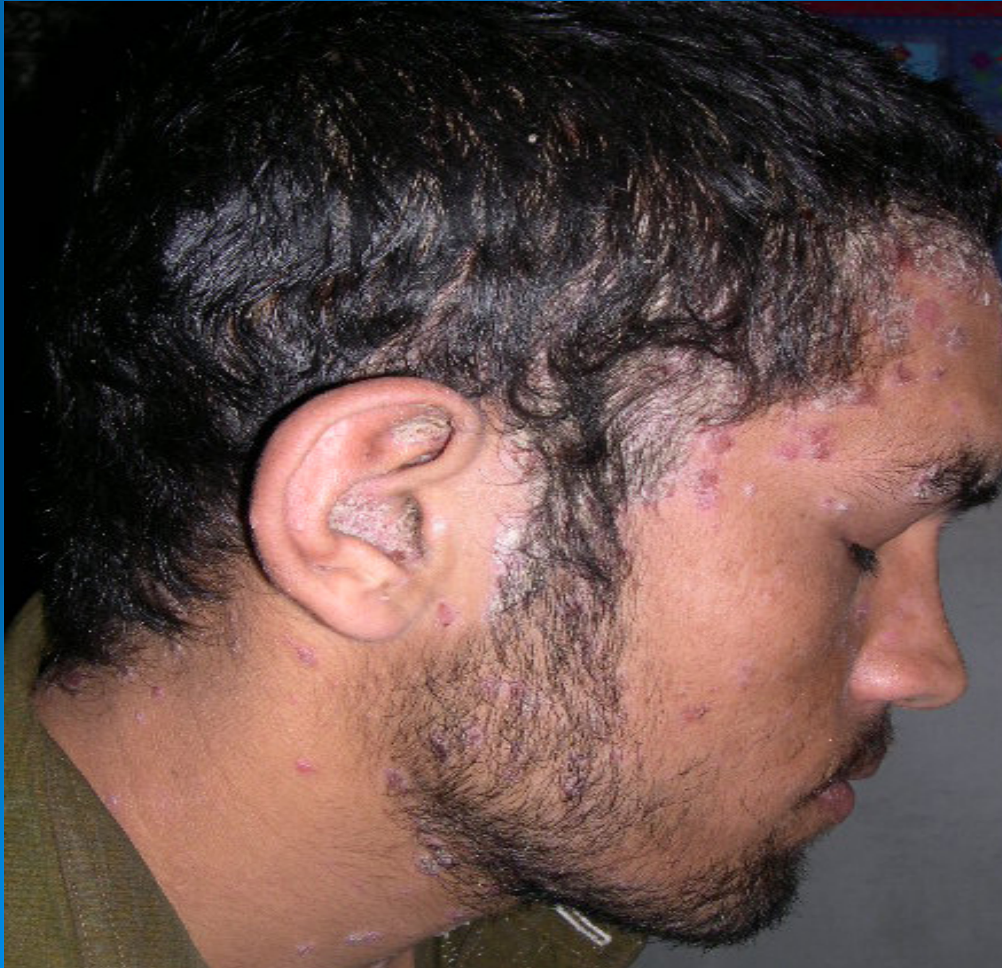
Psoriasis

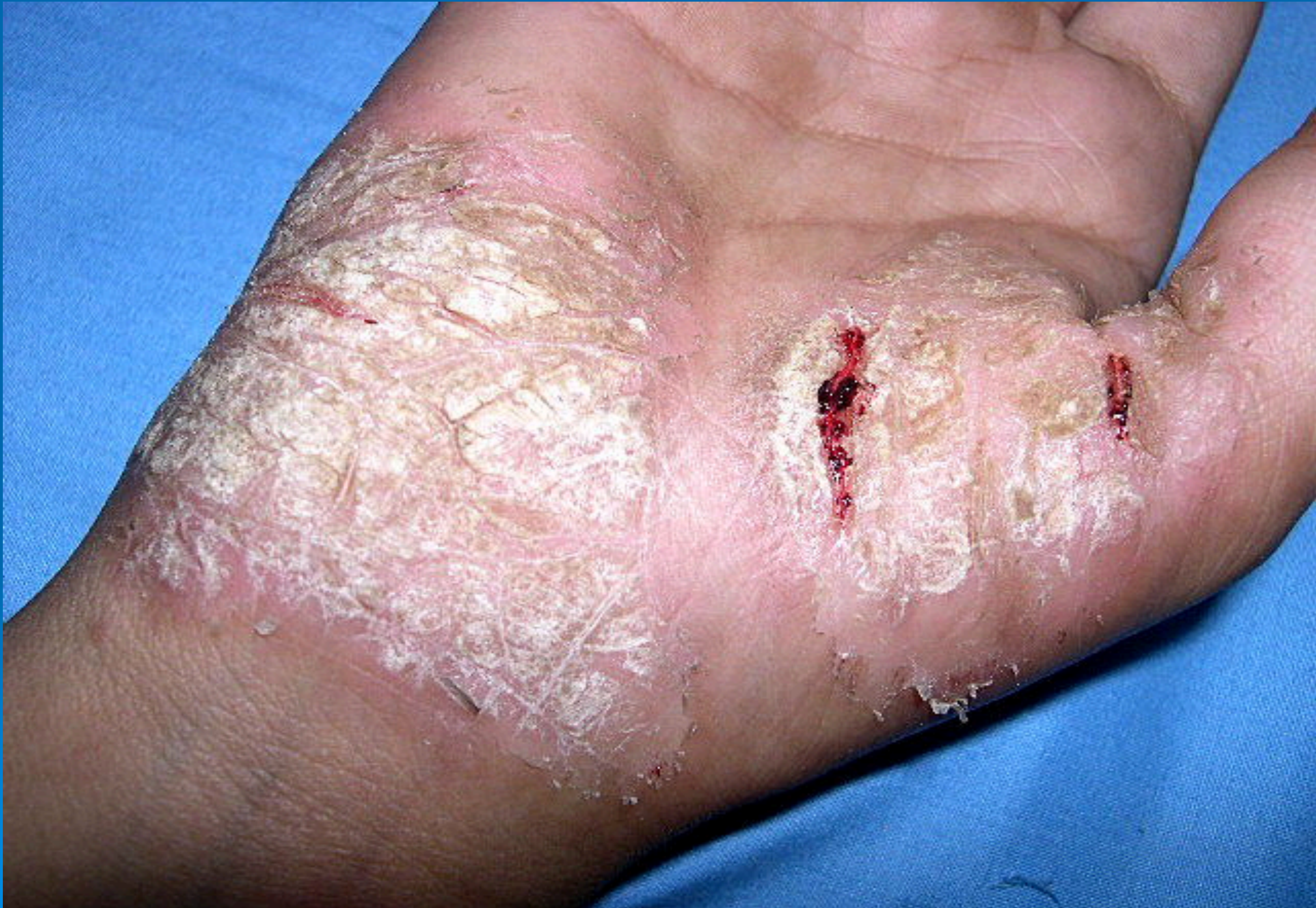
- Chronic common noncontagious relapsing inflammatory disorder.
- genetic predisposition
- skin of the elbows, knees, scalp, lumbosacral areas, intergluteal clefts, and glands penis.
- joints also affected in up to 30% of pts











Frequency

United States

- Between 2% and 2.6% of the US population.
- **Race**
- more common in Caucasians.

Sex

- slightly more common in women > men.

Age

- = 10-15% of new cases begin in children < 10 y.
- The1st peak occurs in persons aged 16-22 y, and the 2nd in persons aged 57-60 y
- (type 1 and 2 psoriasis)

Pathophysiology

- Complex multifactorial disease influenced by genetic and immune-mediated components.
- not completely understood
- Genetic :
- Genetic predisposition for(HLA-B13, -B17, B27,DR7 and -Cw6)
- There are two inheritance mode:
- A-one has onset in younger age with family history of ps.
  B-the other has onset in late adulthood without family history of ps

- A child with one affected parent…………..16%
  If child with both parents affected 50%

Immunological factors

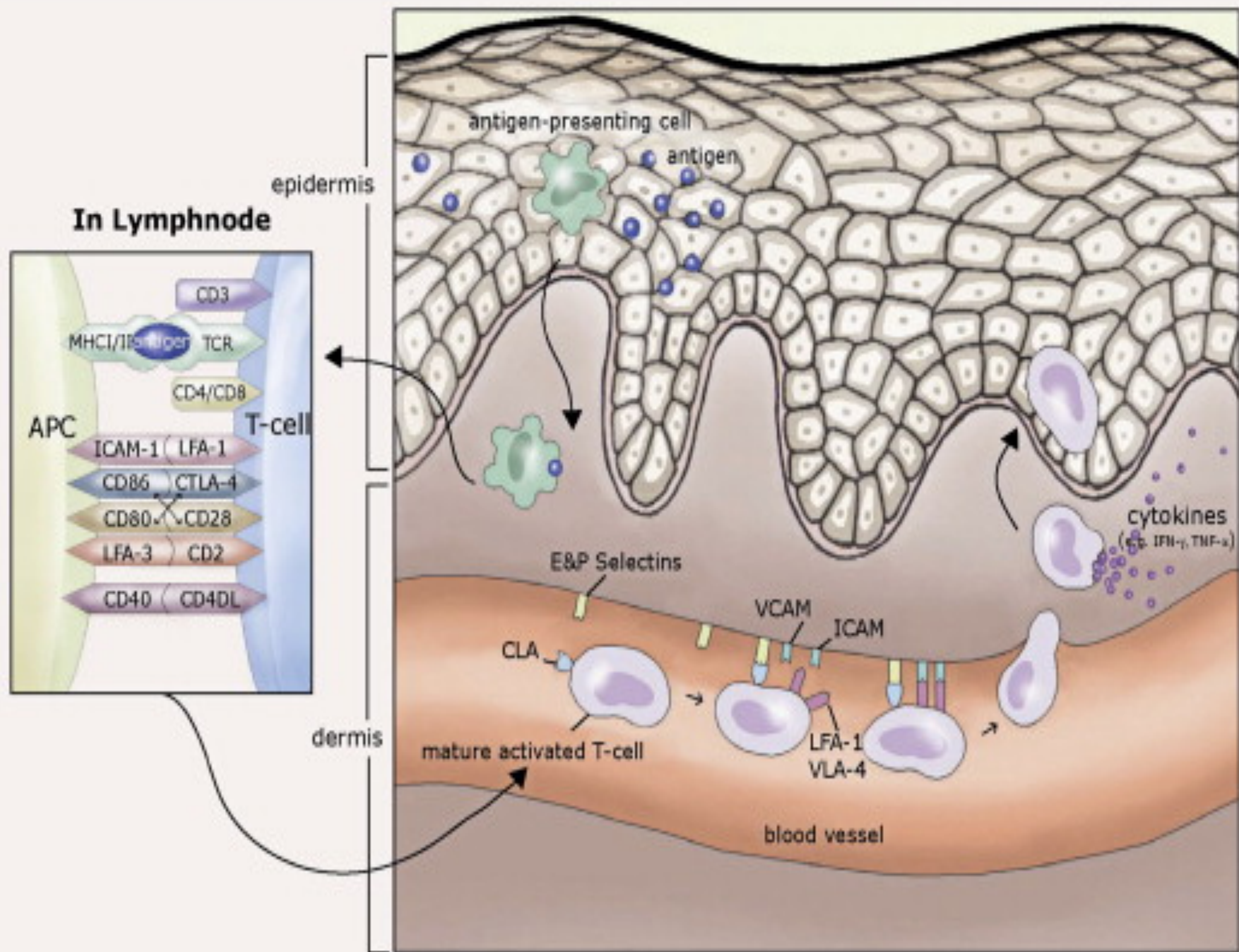
- Studies show high levels of dermal and circulating TNF-alpha.
- TNF receptors are upregulated .
- Rx with TNF-alpha inhibitors is often successful.
- Increase level of interferon gamma .
- Increase level of interleukin 2 and 12 as well as increase in IL-23/IL-17.
- Increased activity of T cells of psoriatic lesions

Environmental factors

- Multiple theories regarding triggers of disease :
- Stress & smoking & UV,truma and alcohol exacerbate psoriasis.
- Infection:
- - Pharyngeal strept & guttate psoriasis .
- - HIV
- Drugs (NSAIDs,lithium, anti- malarials , beta-blockers and withdrawal from systemic corticosteroids)
- Association with obesity .
- In many pts, no obvious trigger exists at all

Epidermal cell kinetics

- The growth fraction of basal cells is increased to almost 100% compared with 30% in normal skin.
- The epidermal turnover time is shortened to less than 10 days compared with 30 to 60 days in normal skin



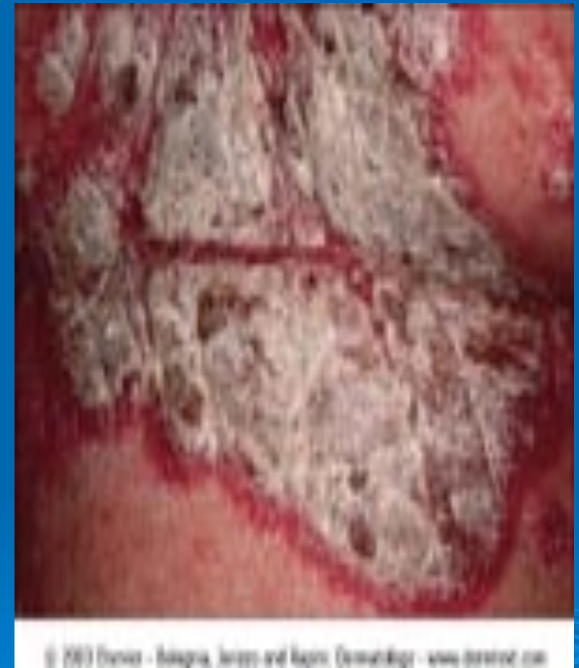
In Lymphnode
CD3
MHCI/II
TCR
CD4/CD8
APC
T-cell
ICAM-1
LFA-1
CD86
CTLA-4
CD80
CD28
LFA-3
CD2
CD40
CD4DL
epidermis
antigen-presenting cell
antigen
cytokines
(e.g. IFN-γ, TNF-α)
E&P Selectins
VCAM
ICAM
CLA
LFA-1
VLA-4
mature activated T-cell
dermis
blood vessel

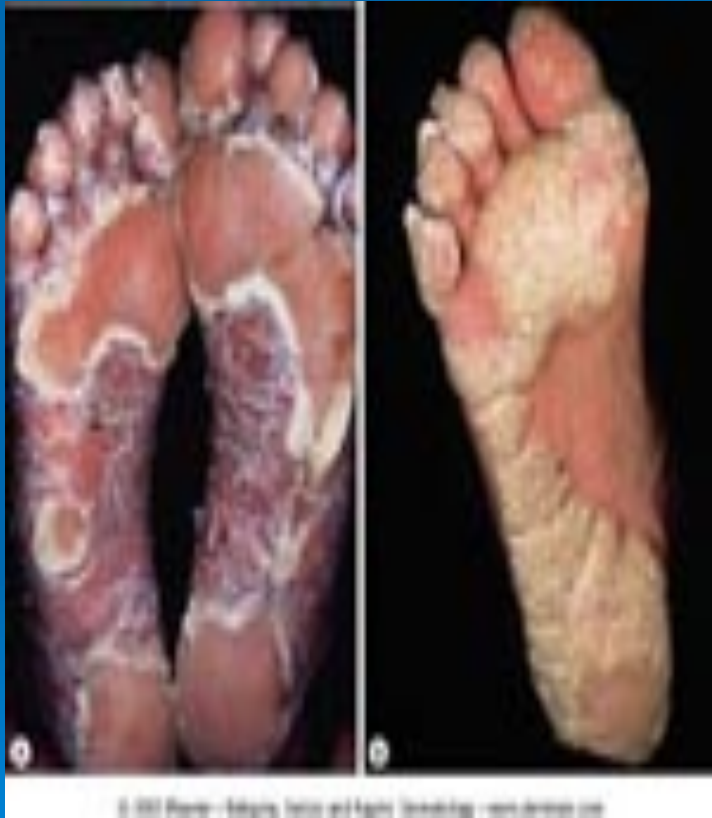
- Increase in the turnover rate of epidermal cells from 23 to 3-5 days→ dead skin cells layer as silver scales.
- At sites of trauma to the skin→ new lesions appear (**Koebner phenomenon)**

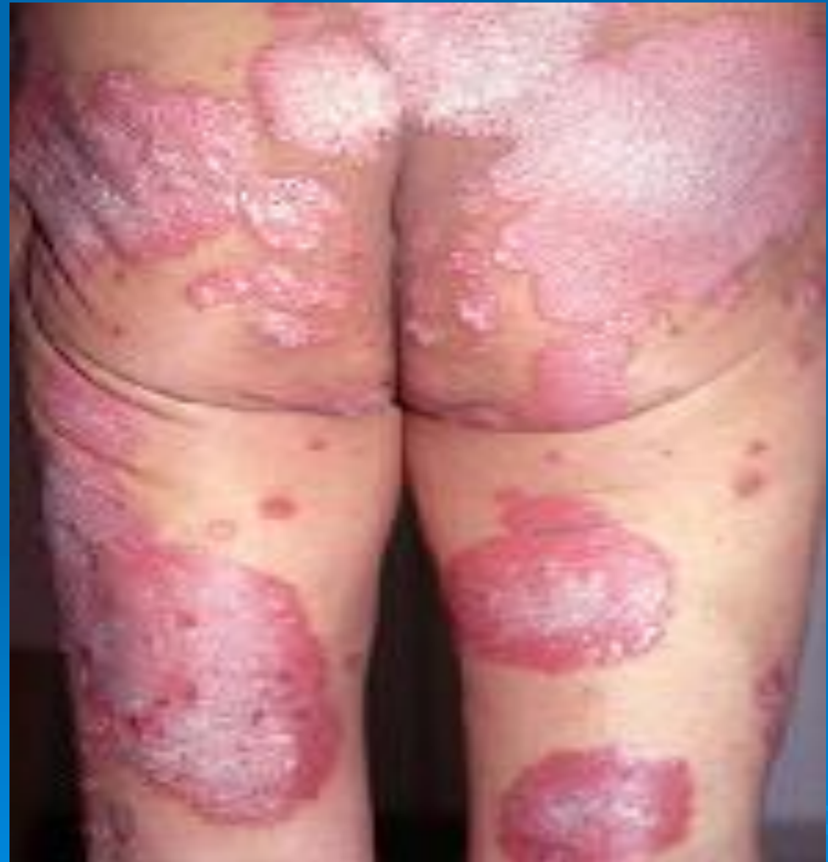
Clinical Features

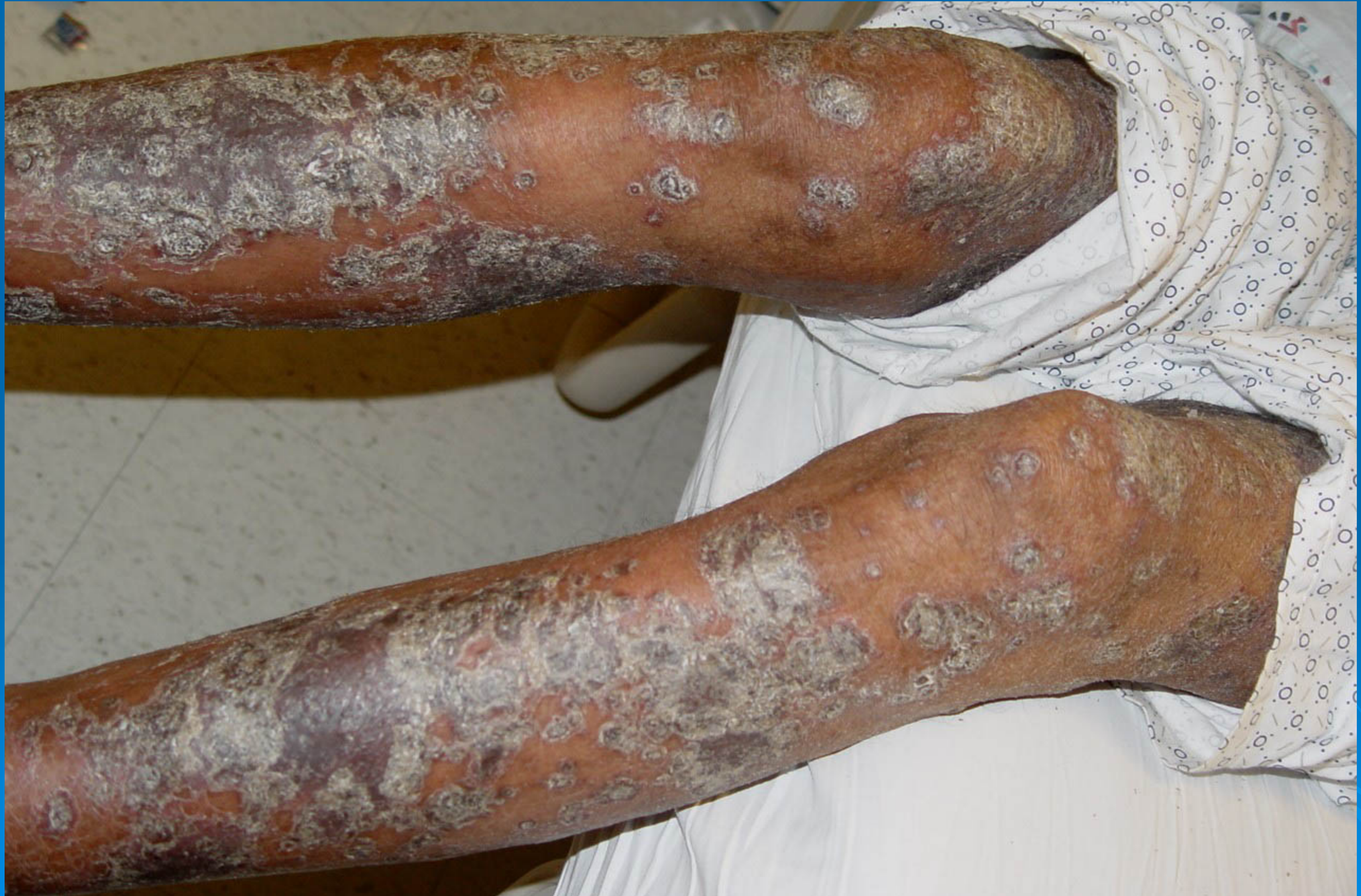
plaque psoriasis (most common)

- Well-circumscribed red plaques covered with a silvery white thick scale .
- if scale scraped away→ reveal inflamed skin beneath with pin point bleeding (Auspitz sign)
- Symmetrical on extensor surfaces of knees, elbows, scalp, and sacral area.
- up to 10-20% of patients with plaque psoriasis may evolve into more severe disease, such as pustular or erythrodermic psoriasis



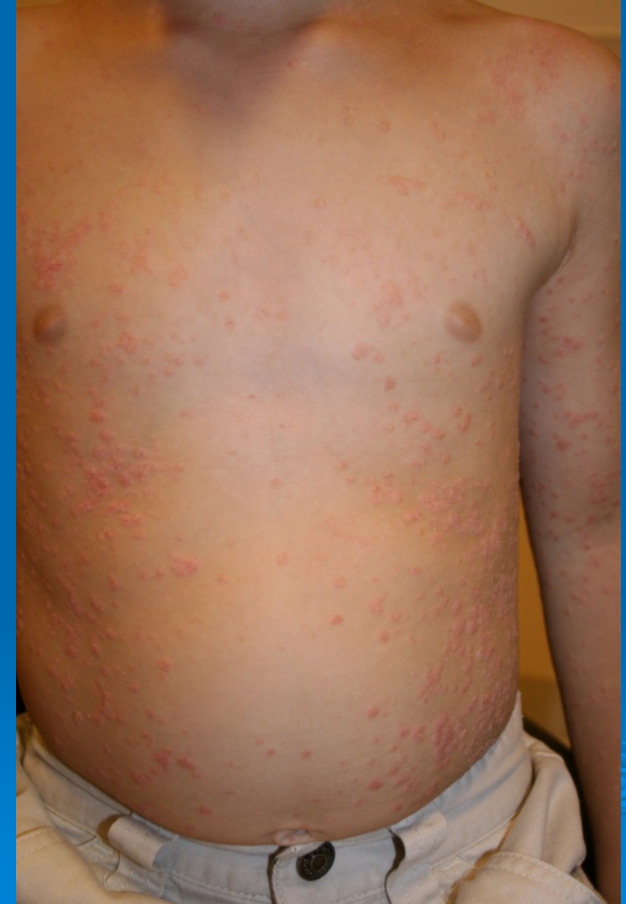


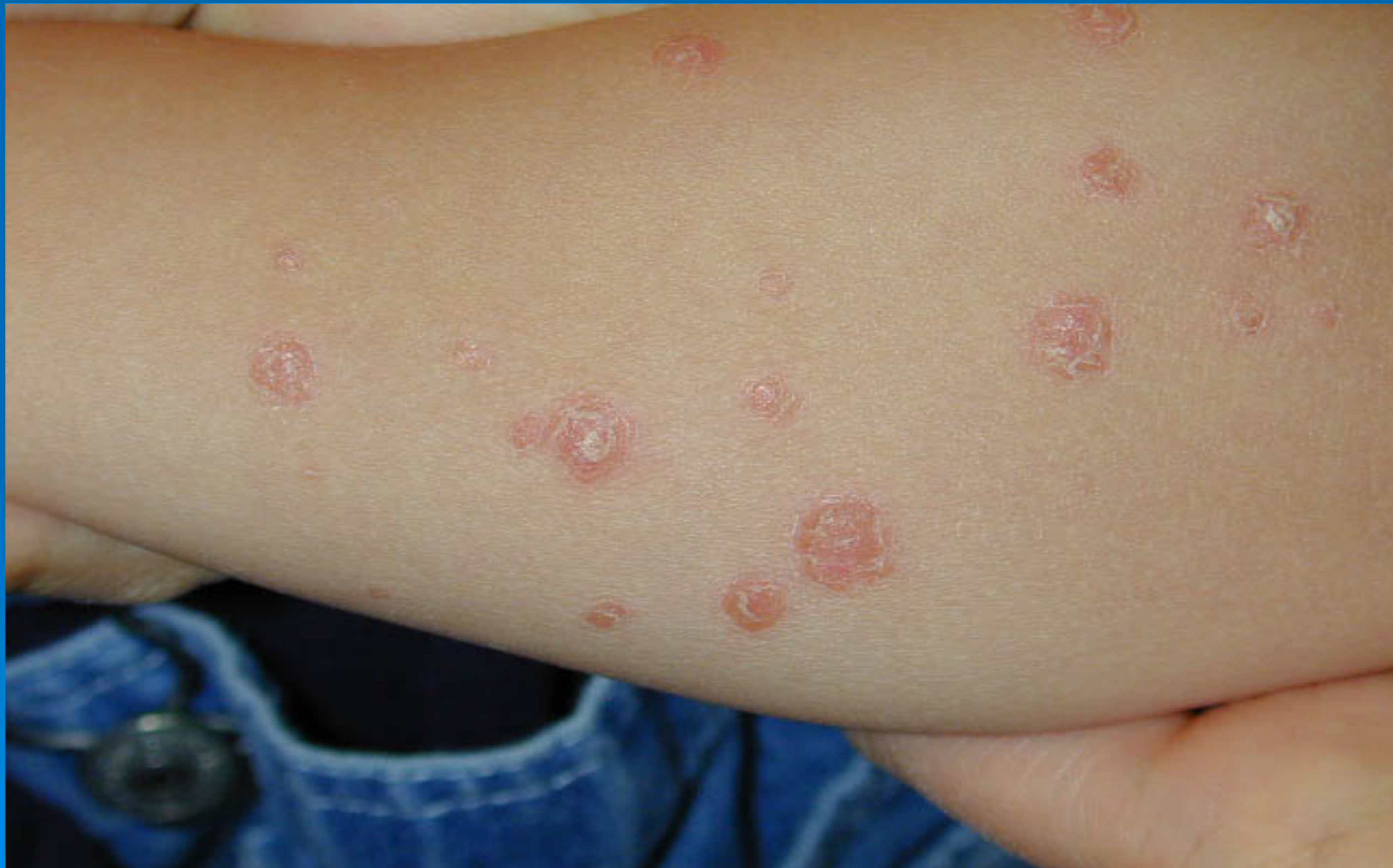




Guttate psoriasis

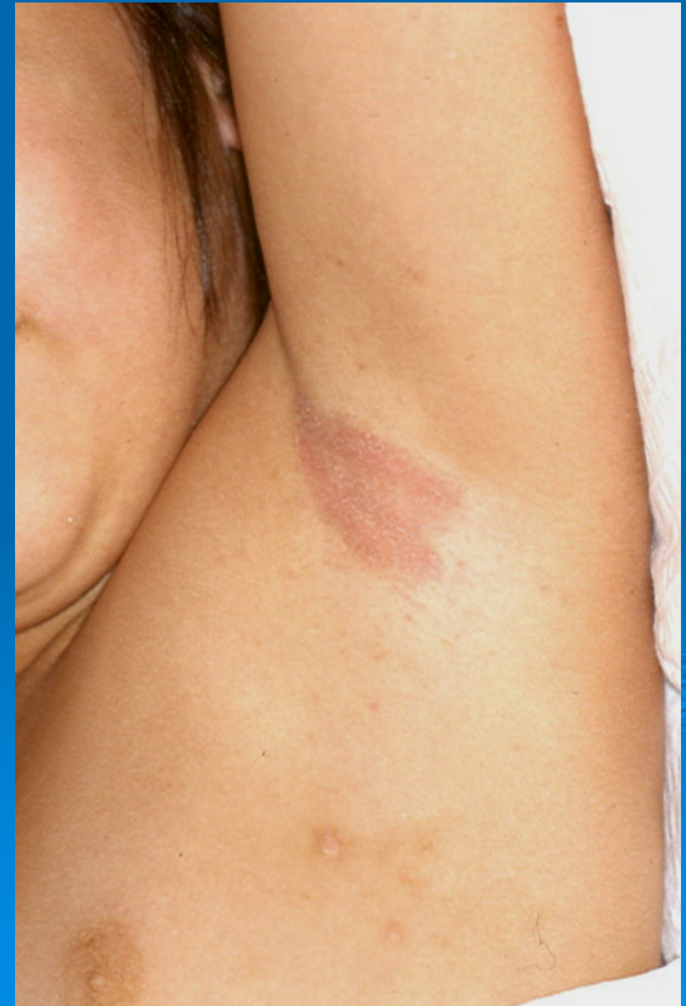
- Children> adult
- Presents as small droplike salmon-pink scaly papules, 1-10 mm in diameter
- On the trunk and the proximal extremities .
- Suddenly, 2-3 weeks after URI with group A beta-hemolytic streptococci
- HLA- Cw6
- Resolution within few months.

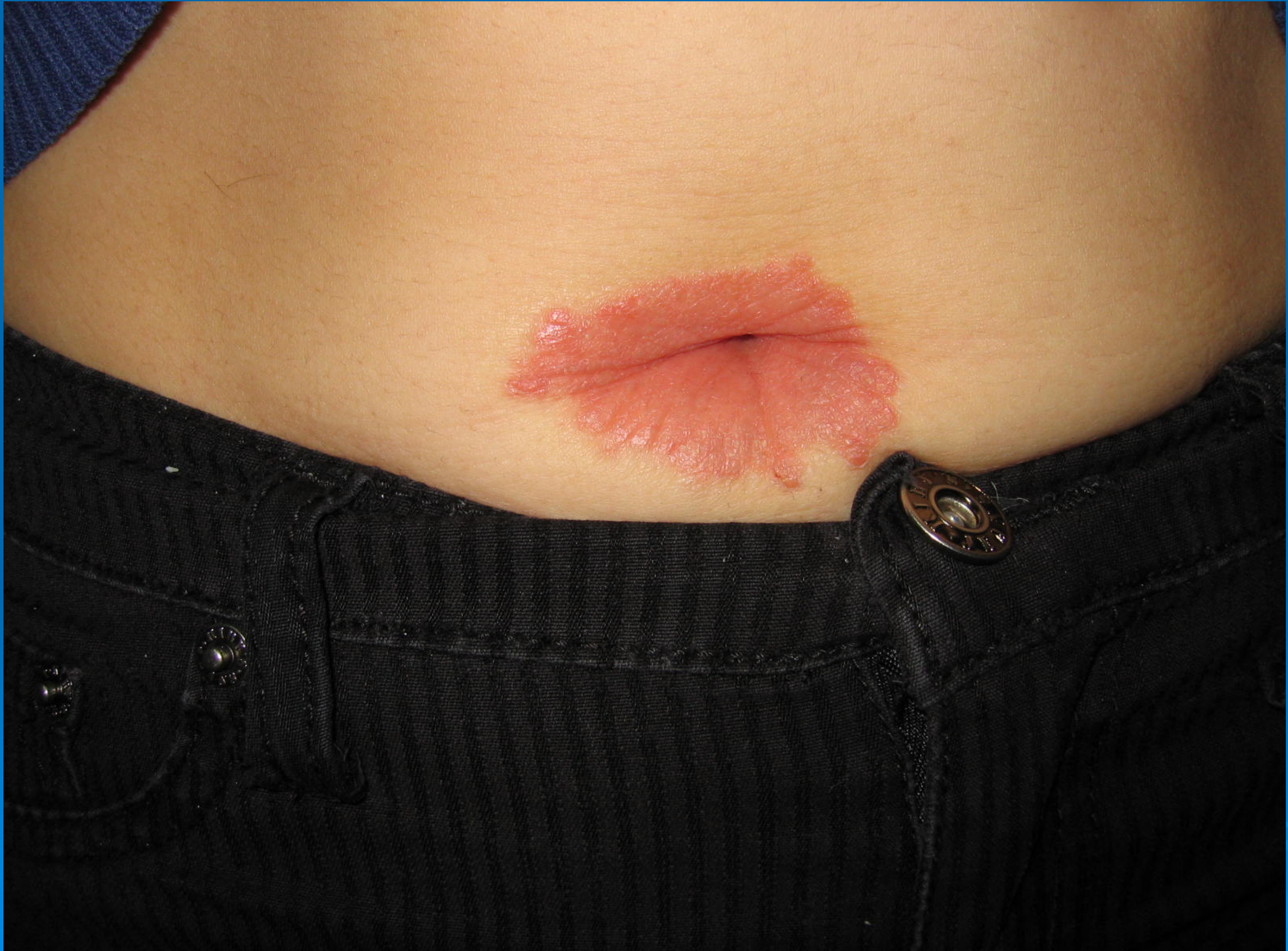




Inverse psoriasis

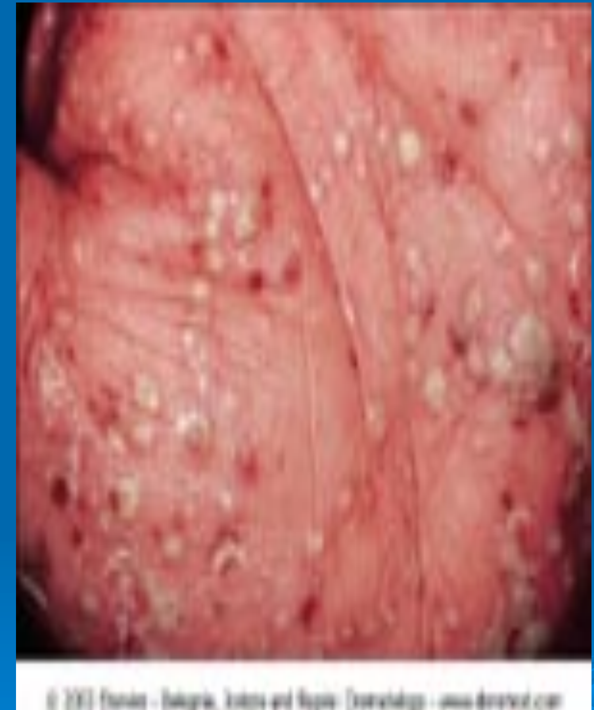
- occurs on the flexural surfaces, armpit, groin, under the breast, and in the skin folds.
- It is characterized by smooth, inflamed lesions without scaling due to the moist nature of the area





Pustular psoriasis

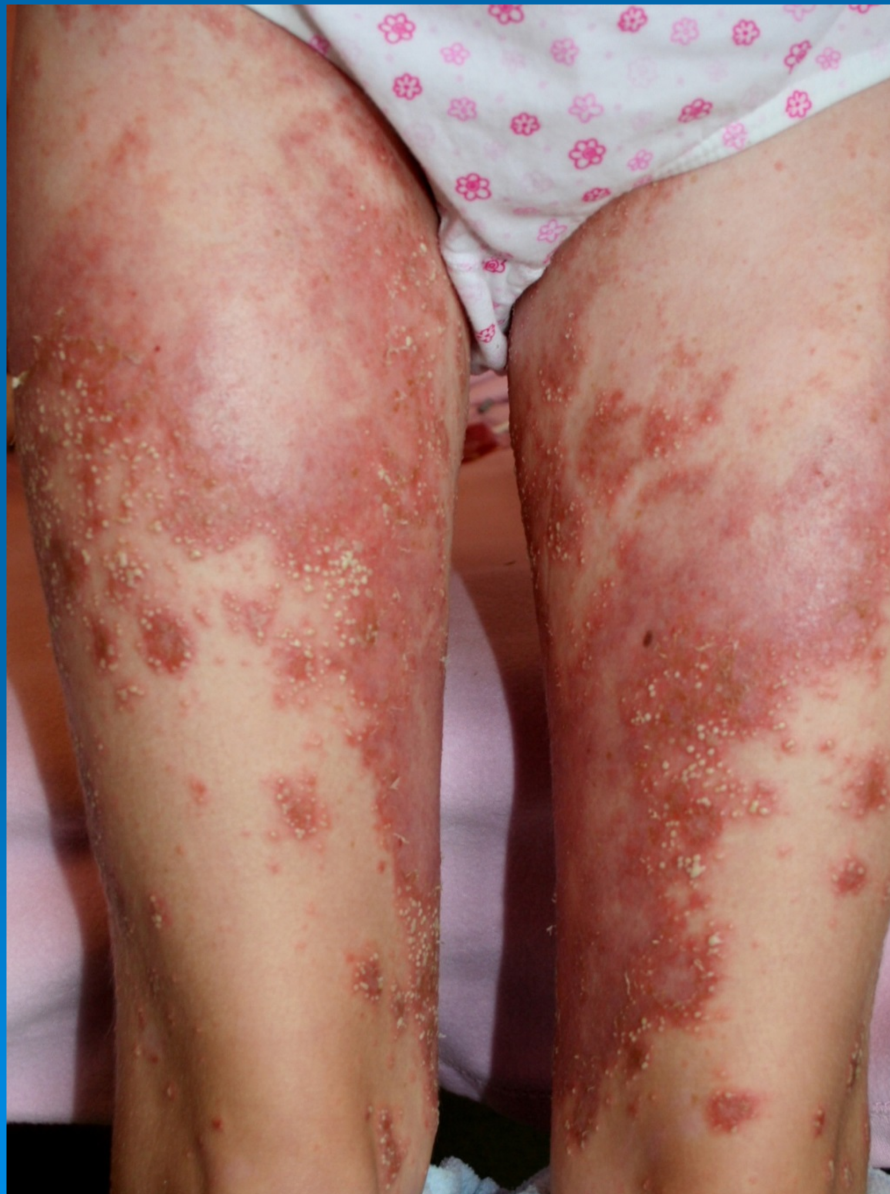
- Uncommon form of psoriasis.
- Sterile pustules on palms & soles or diffusely over the body.
- Pustular psoriasis -->erythema then scaling.
- Psoriasis vulgaris may be present before, during, or after.

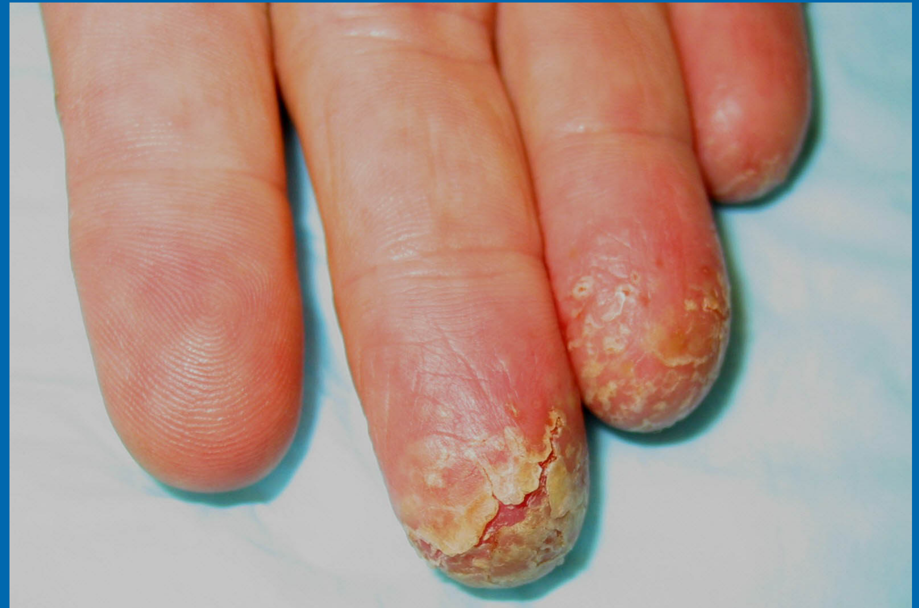
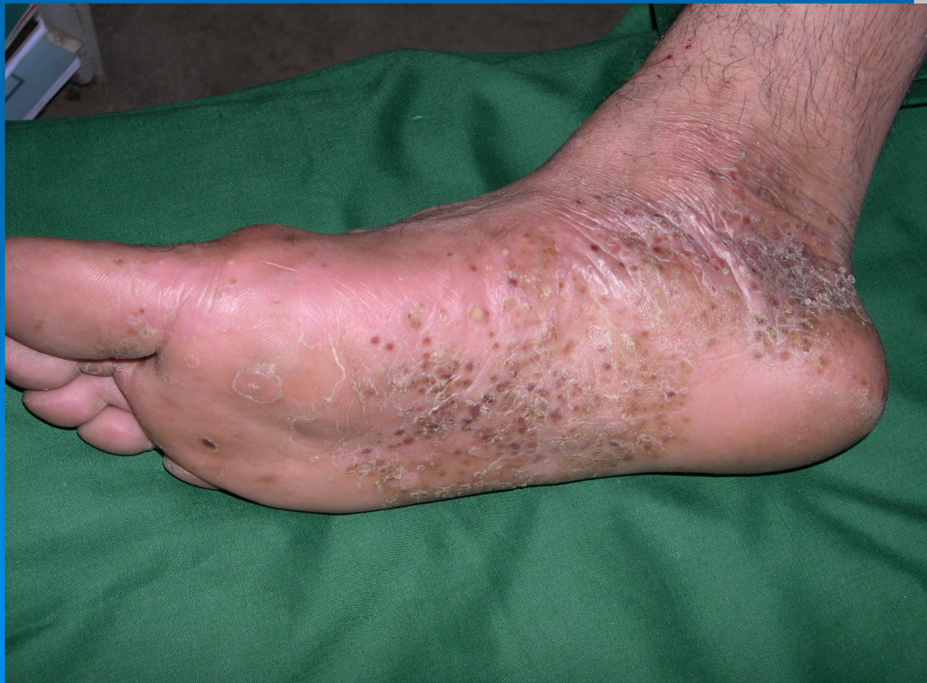


- pustular psoriasis may be classified into several types:
- 1-generalized type(von Zumbusch variant):
- Generalized erythema studded with interfolecular pustules
- Fever, intense ill feeling , tachypneic, tachycardic
- Absolute lymphopenia with polymorph nuclear leukocytosis up to 40,000/µL
- 2-Localized form (palms and soles)

Causes of pustular psoriasis

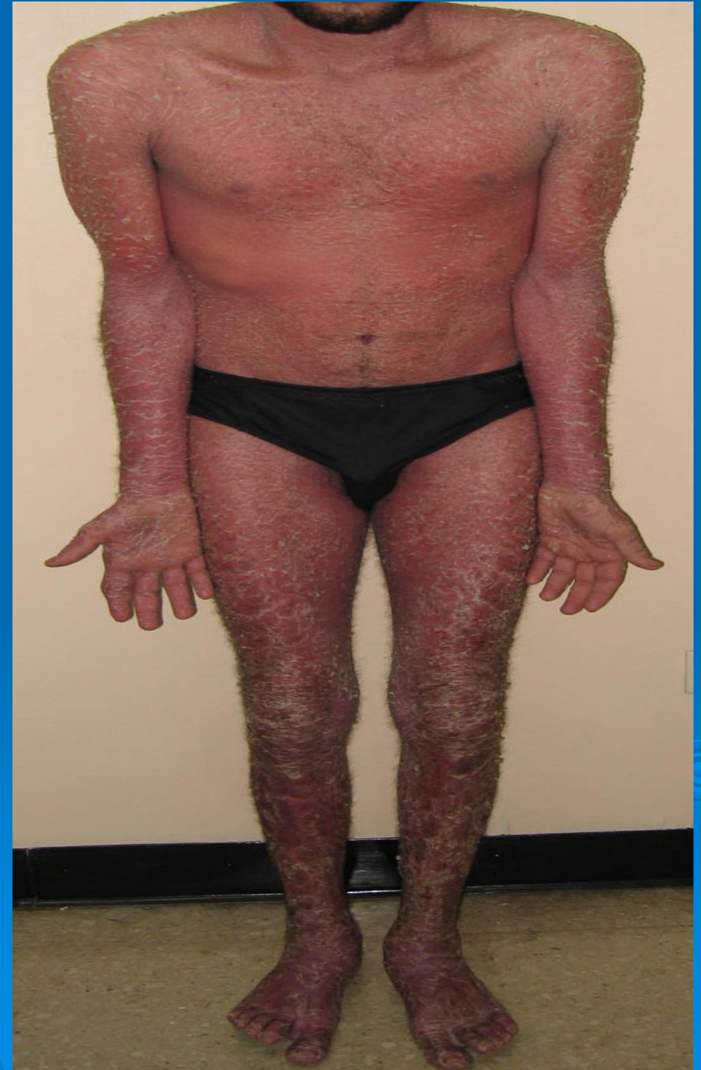
- Withdrawal of systemic steroids.
- Drugs, including salicylates, lithium, phenylbutazone,, hydroxychloroquine, interferon.
- Strong, irritating topicals, including tar, anthralin, steroids under occlusion, and zinc pyrithione in shampoo
- Infections .
- Sunlight or phototherapy.
- Cholestatic jaundice.
- Hypocalcemia.
- Idiopathic in many patients.



Erythrodermic psoriasis

- Generalized painful scaly erythematous lesions, involving 90% or more of the cutaneous surface
- Hair may shed; nails may become ridged and thickened
- Few typical psoriatic plaques
- Unwell ,fever, chills, hypothermia, and dehydration secondary to the large BSA involvement

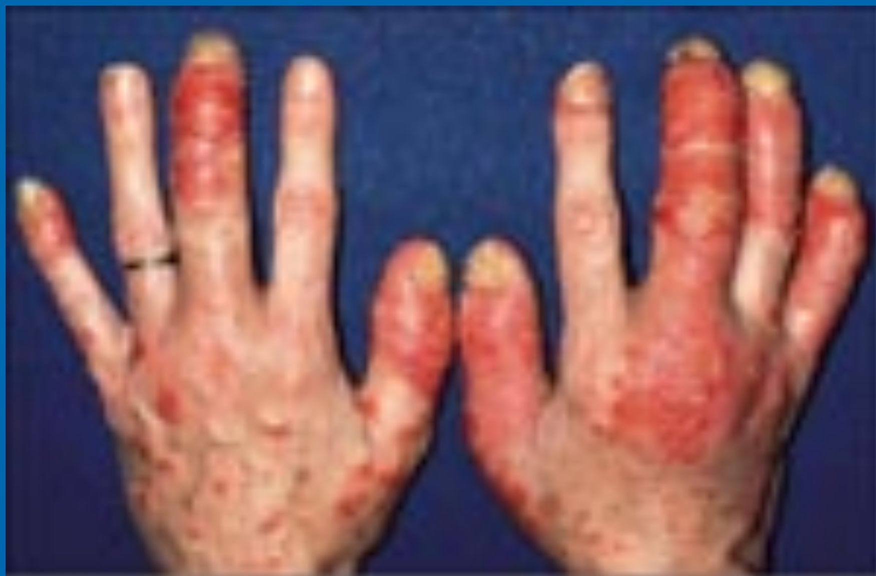


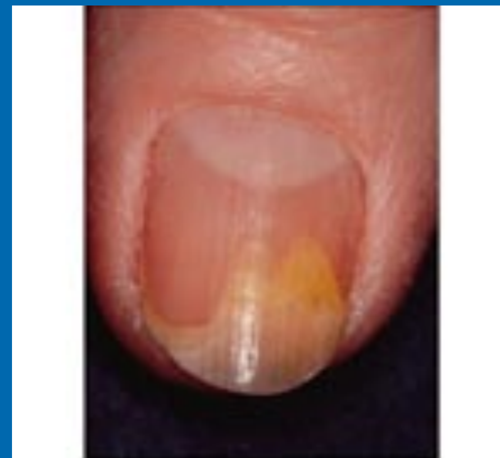
Psoriatic arthritis

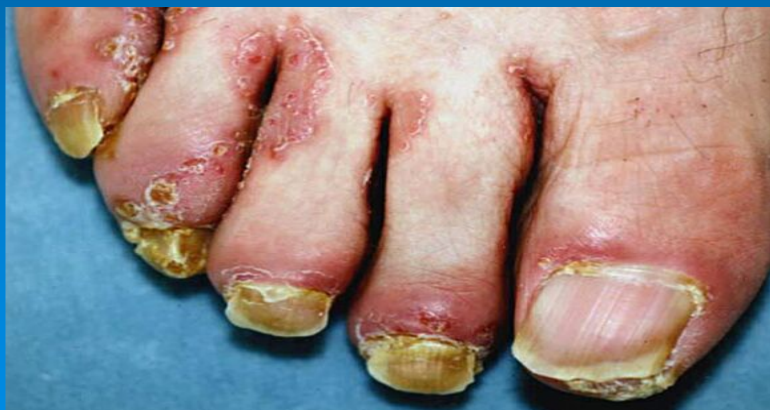
- Is a chronic inflammatory arthritis that is commonly associated with psoriasis
- One in five patients with psoriasis has psoriatic arthritis .
- Psoriasis before psoriatic arthritis in 60-80% of patients.
- In 15-20 % of patients, arthritis appears before psoriasis .
- Most commonly a seronegative oligoarthritis
- Asymmetric oligoarthritis occurs in as many as 70% of patients with psoriatic arthritis
- DIP joint involvement occurs in approximately 5-10% of patients with psoriatic arthritis
- Arthritis mutilans is a rare form of psoriatic arthritis occurring in 5% of patients with psoriatic arthritis
- Spondylitis occurs in about 5% of patients with psoriatic arthritis and is often asymptomatic

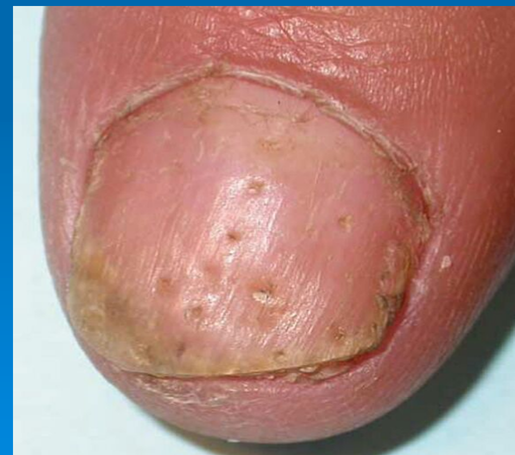
Psoriatic nail

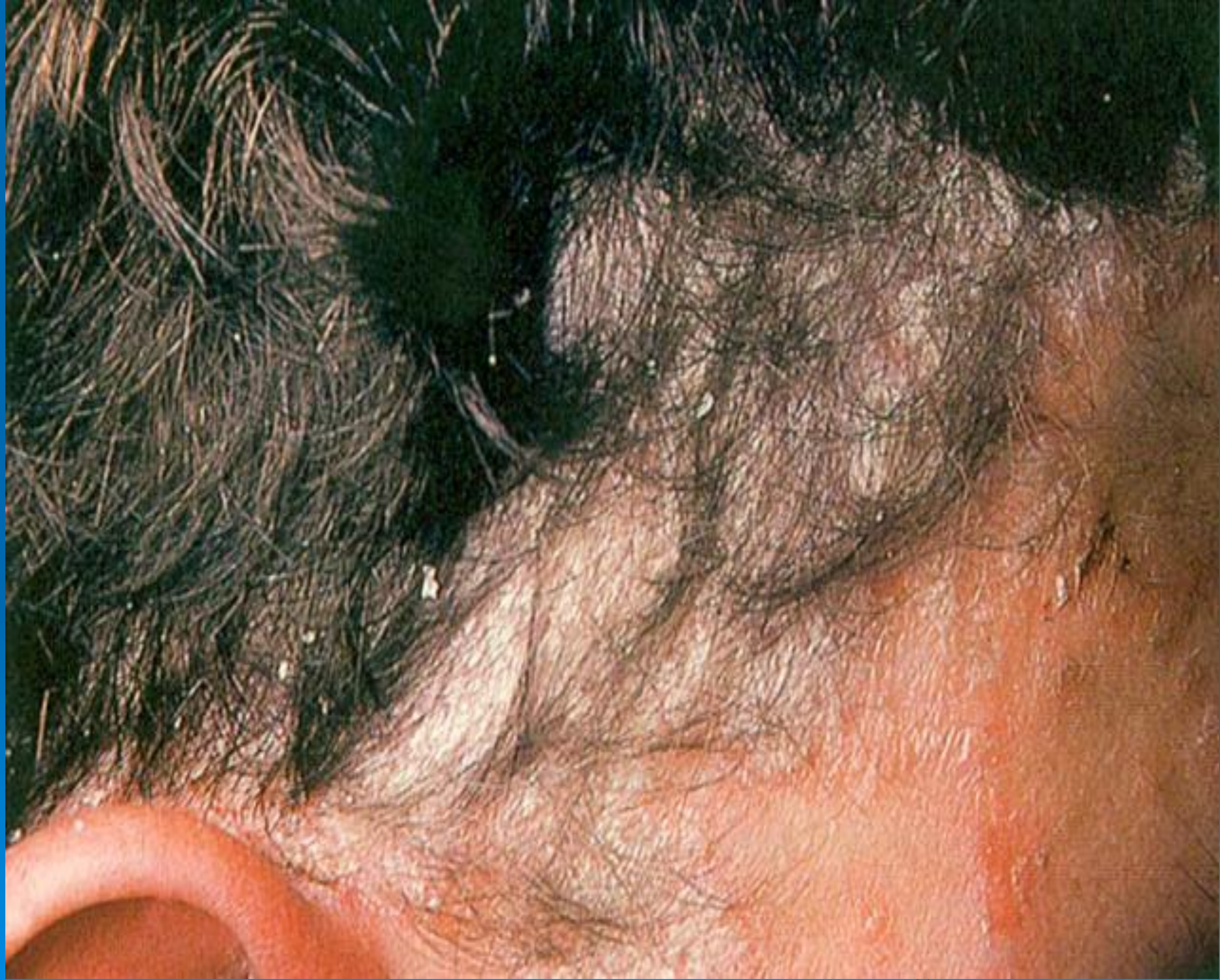
- Psoriatic nail disease in 10-55% of all pts with psoriasis
- Less than 5% of psoriatic nail disease cases occur in patients without other cutaneous findings
- Nail changes are seen in 53-86% of pts with PA
- Oil drop or salmon patch/nail bed
- Pitting
- Subungual hyperkeratosis
- Onycholysis
- Beau lines
- Scalp psoriasis
- 50% of patients with psoriasis .
- Erythematous raised plaques with silvery white scales











Differential Diagnoses

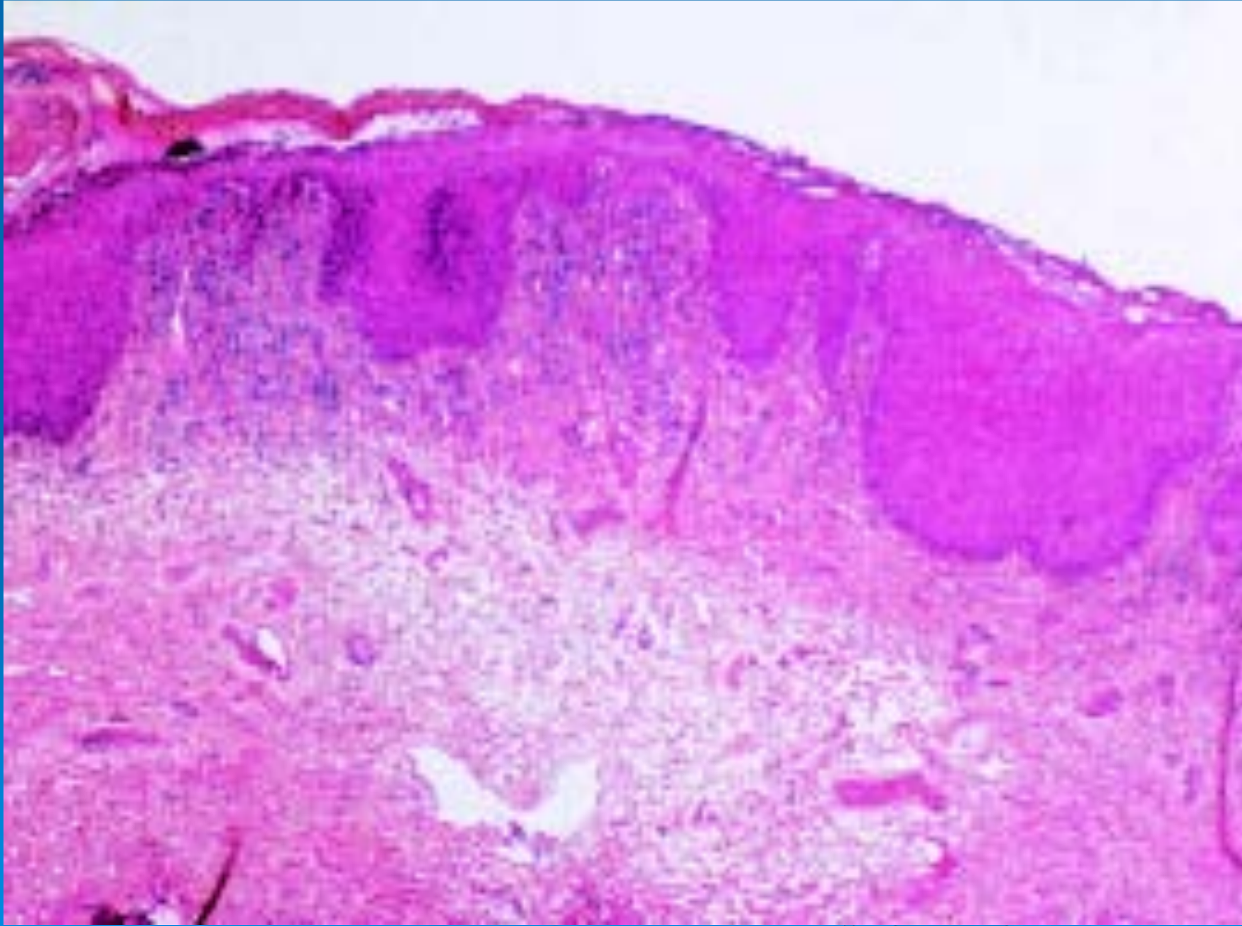
Seborrheic dermatitis
Nummular eczema
Lichen planus
Pityriasis Rosea
Drug eruptions
Reiter's disease
Syphilis
Tine Corporis
Nail : Onychomycosis

INVESTIGATIONS

- Skin biopsy
- Others

Histopathology

- Parakeratosis(nuclei retained in the horny layer)
  Irregular thickening of the epidermis oever the rete ridges but thinning over dermal papillae.
- Epidermal polymorphonuclear leucocyte infiltrates (munro abscesses).
- Dilated capillary loops in the dermal papillae.
- T-lymph infiltrate in the upper dermis



Treatment

Prevention

- Avoid injury to skin(sunburn and other physical trauma)
- Avoid drugs known to worsen the problem
- Treatment regimens must be individualized according to age, sex, occupation, personal motivation, severity, other health conditions, and available resources.
- Rx :

(topical agents, phototherapy, and systemic agents, including biologic therapies)

Topical corticosteroids

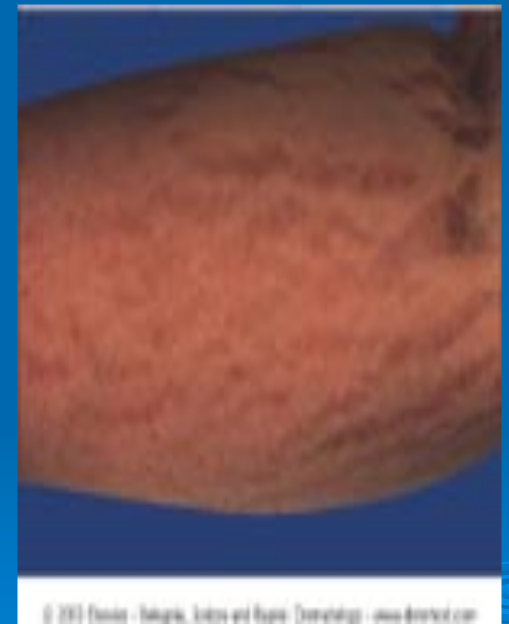
- anti-inflammatory effects
- modify body's immune response to diverse stimuli.

Systemic SE:(rare):

- HPA axis suppression, Cushing syndrome

local cutaneous SE:(common)

- Atrophy of the epidermis and dermis & striae
- Purpura, Telangiectases
- tachyphylaxis
- **Betamethasone dipropionate (Diprolene) 0.05% cream**



Coal tar

- Antipruritic and antibacterial that inhibits deregulated epidermal proliferation
- In shampoos or lotions
- Useful in hair-bearing areas
- SE: messy, ? carcinogenicity

Vitamin D-3 analogs

- **Calcipotriene (Dovonex)**
- Regulates skin cell production & development
- SE: irritation, transiently but reversibly elevate serum calcium level

Keratolytic agents

- to remove scale, to smooth the skin

Anthralin 0.1-1%

- **Short-contact**
- **SE: irritation ,staining**

salicylic acid

- **Scalp, palms and soles**
- **SE: Salicylicism if high con**

Phototherapy:

Psoralen plus UVA(PUVA)

- Ingestion of 8-methoxypsoralen (8-MOP) then UVA
- 2 or 3 times per week
- long-term remissions
- SE: nausea, phototoxicity, lentigines
- If > 260 individual PUVA sessions, 11-fold increase in SCC (Male genitalia) ? malignant melanoma

Narrowband UVB

- Range around 311 nm
- Not as effective as PUVA
- less carcinogenic
- Safer > PUVA

Retinoids

- Stimulate cell differentiation
- Can be used in combination with UV phototherapy
- SE: Teratogenicity, hyperlipidemia

e.g. Acitretin

Antimetabolites

Methotrexate:

- interferes with DNA synthesis, repair, and cellular replication
- 2.5-7.5 mg PO q12h for 3 doses/wk
- give with folic acid 1 mg/d
- SE: Teratogenicity, Liver , BM & Renal

Immunosuppresive

Cyclosporine

Remission is rapid

skin lesions tend to recur after Rx is stopped

SE: Risk of renal damage

Other medication:

Mycophenolate mofetil

Hydoxyurea

Biologic Therapies Currently Approved for the treatment of psoriasis

Alefacept

Adalimumab (Humira):
Infliximab (Remicade):
Etanercept

Ustekinumab (Stelara)
Secukinumab

New treatment

Biologic therapies:

Alefacept

Efalizumab

Tumor Necrosis Factor Inhibitors:

= Infliximab

= Etanercept

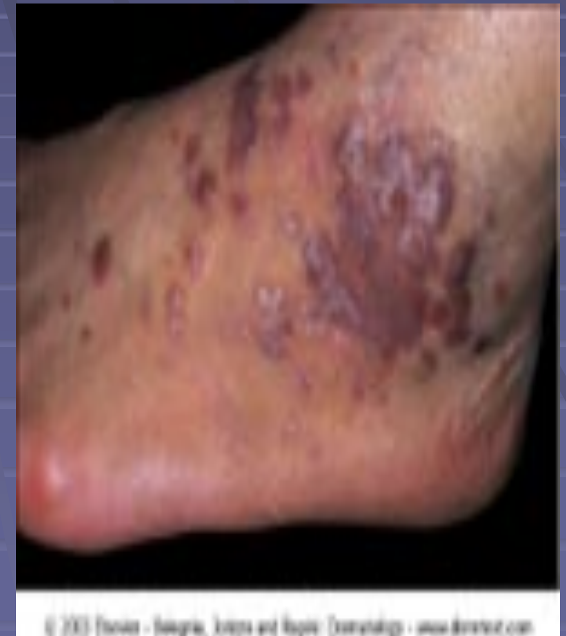
= Adalimumab

Prognosis

- The course of plaque psoriasis is **unpredictable**.
- relapses occurring in **most** patients.
- **early** onset and a **family** history of disease are considered **poor** prognostic indicators.

Lichen planus

- Is a pruritic, papular eruption characterized by its violaceous color; polygonal shape; and, sometimes, fine scale.
- on the flexor surfaces of upper extremities, genitalia & on the mucous membranes



Frequency

- LP is reported in approximately 1% of all new pts seen at health care clinics in US.

F=M

Age

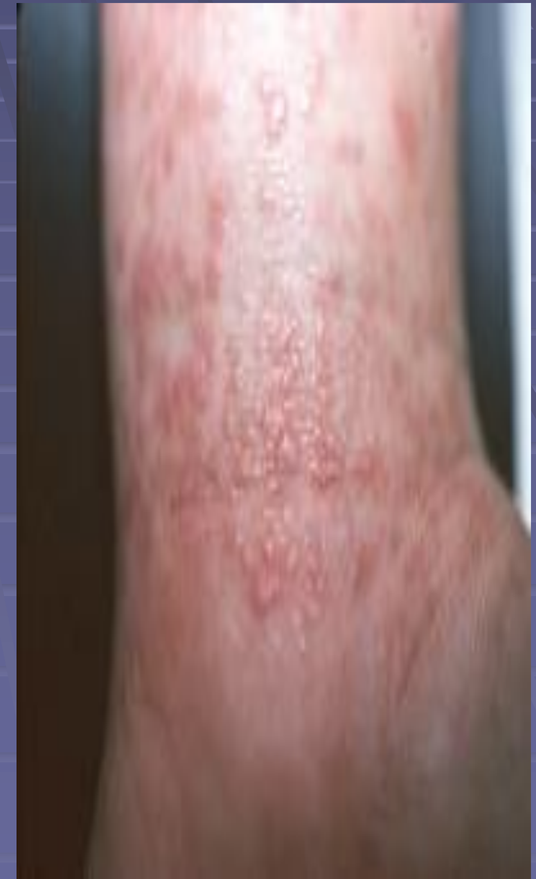
- Rare in children
- > two thirds of pts are aged 30-60 y; however, can occur at any age

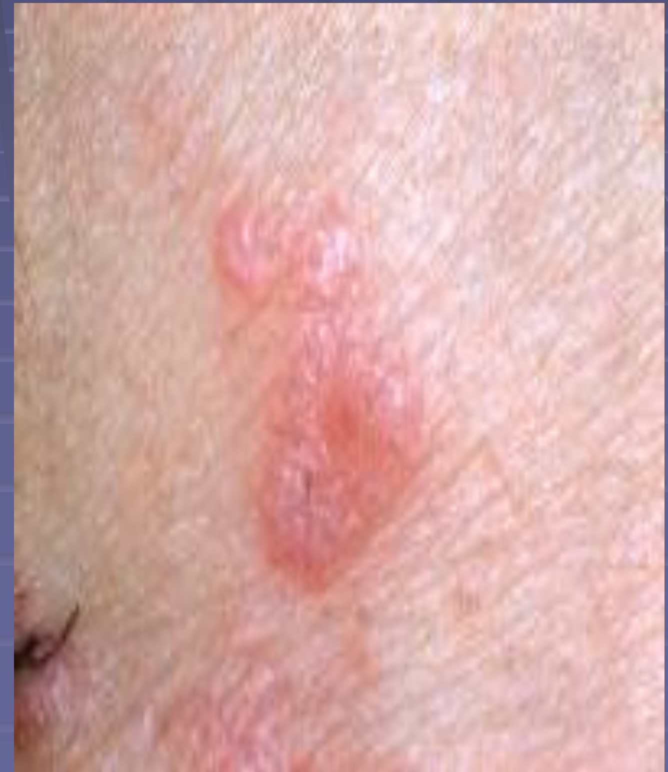
causes

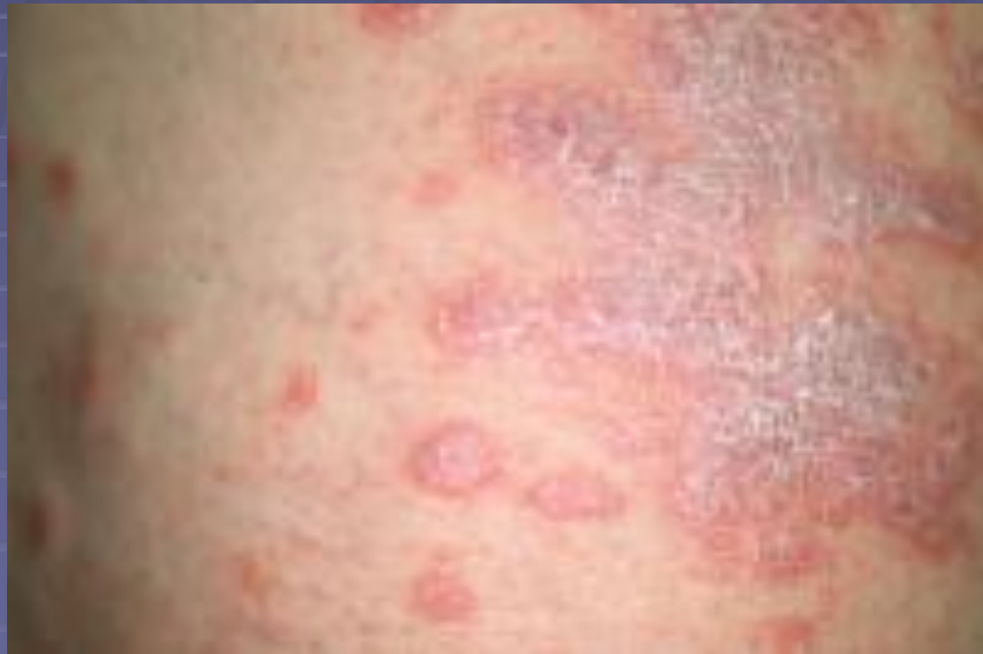
- Is a cell-mediated immune response of unknown origin.
- LP may be found with other diseases of altered immunity(UC, alopecia areata, vitiligo, DM, morphea)
- An association between LP and hepatitis C virus AND primary biliary cirrhosis
- ? genetic predisposition/ Familial cases.
- Onset or exacerbation of LP has been linked to stressful events
- Drugs induce lichenoid reaction like thiazide,antimalarials,propranolol

Clinical Features

- The papules are violaceous, shiny,flat- topped and polygonal; varying in size.
- They can be discrete or arranged in groups of lines or circles.
- Characteristic fine, white lines on the papules (Wickham stria)
- Sites: flexor of wrists and legs.



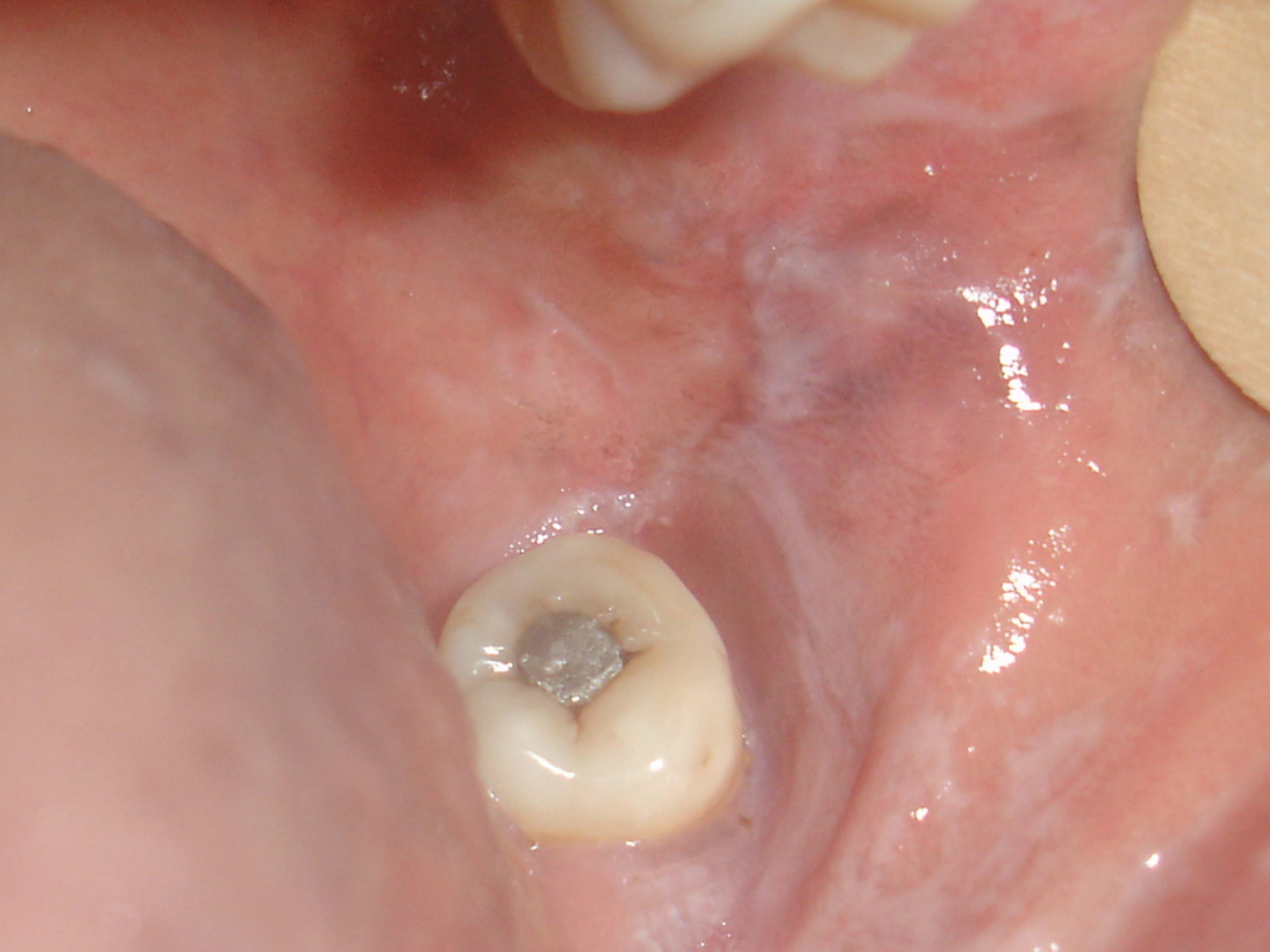




Mucous membrane involvement :

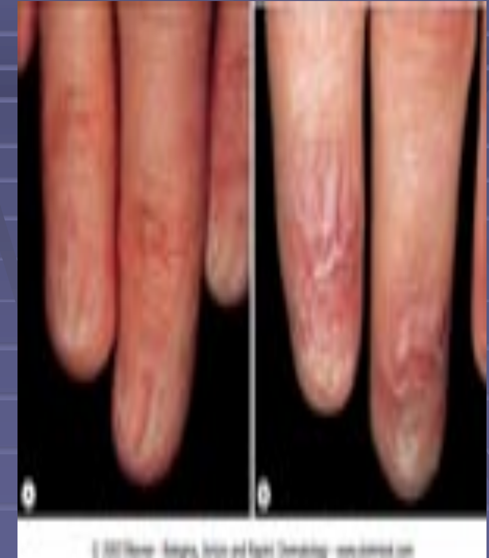
- common and may be found without skin.
- Asymptomatic
- on the tongue and buccal mucosa
- characterized by white or gray streaks forming a linear or reticular pattern on a violaceous background).
- Oral lesions are **classified** as:

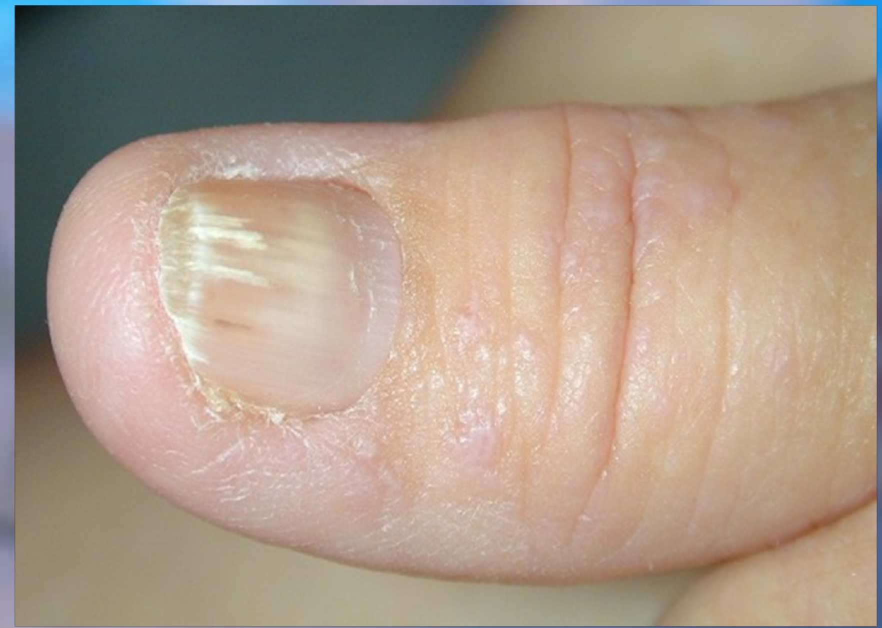
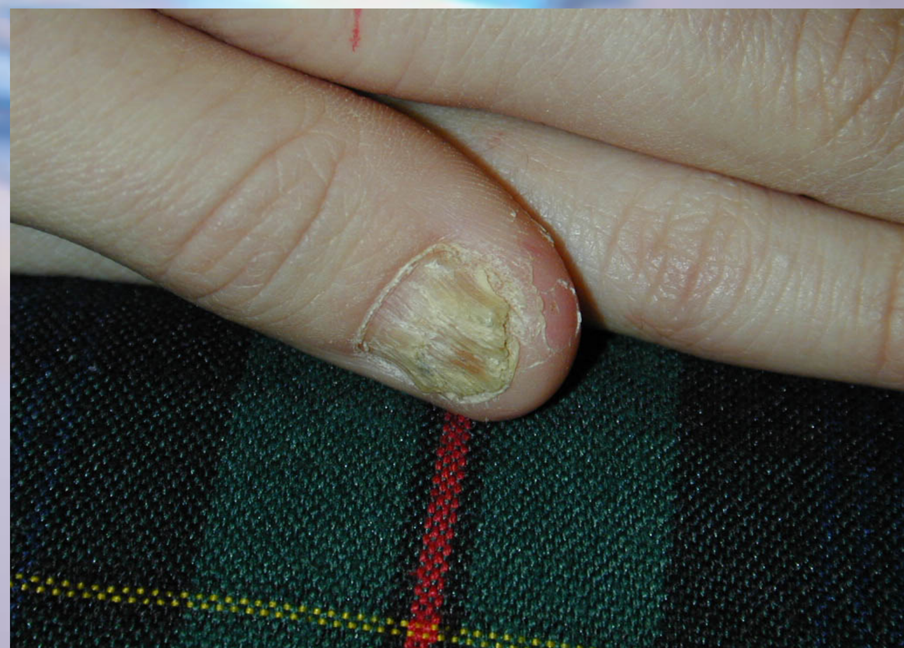
  reticular, plaquelike, atrophic, papular, erosive, and bullous.
- Lesions may also be found on genitalia & GIT



Nail involvement :

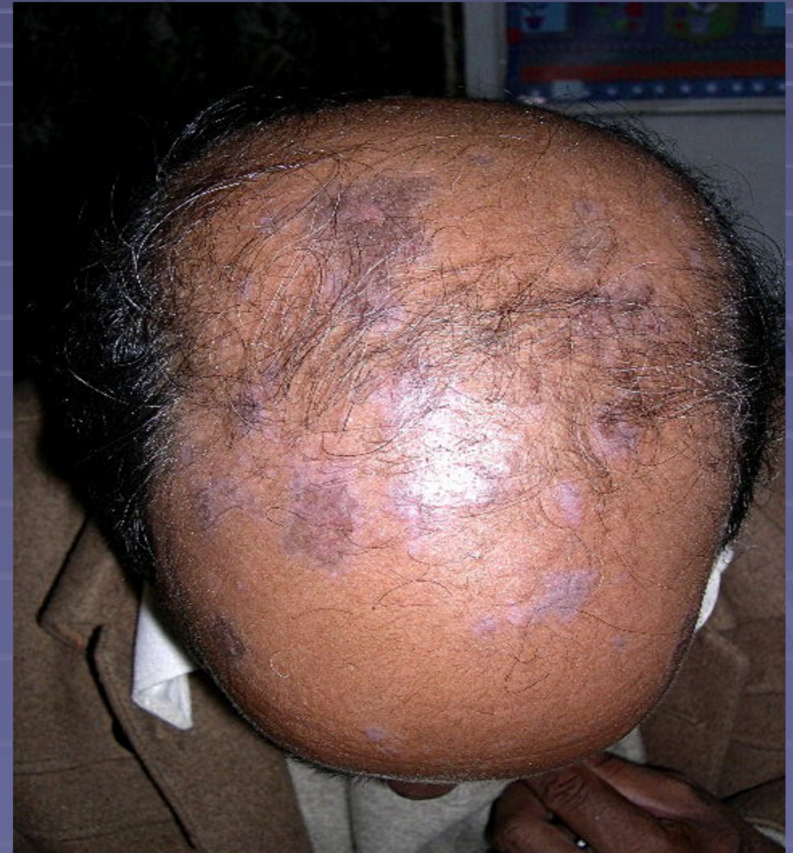
- In 10% of pts
- Commonly, nail plate thinning causes longitudinal grooving and ridging.
- Subungual hyperkeratosis, onycholysis
- Rarely, the matrix permanently destroyed with prominent pterygium formation.
- Twenty-nail dystrophy



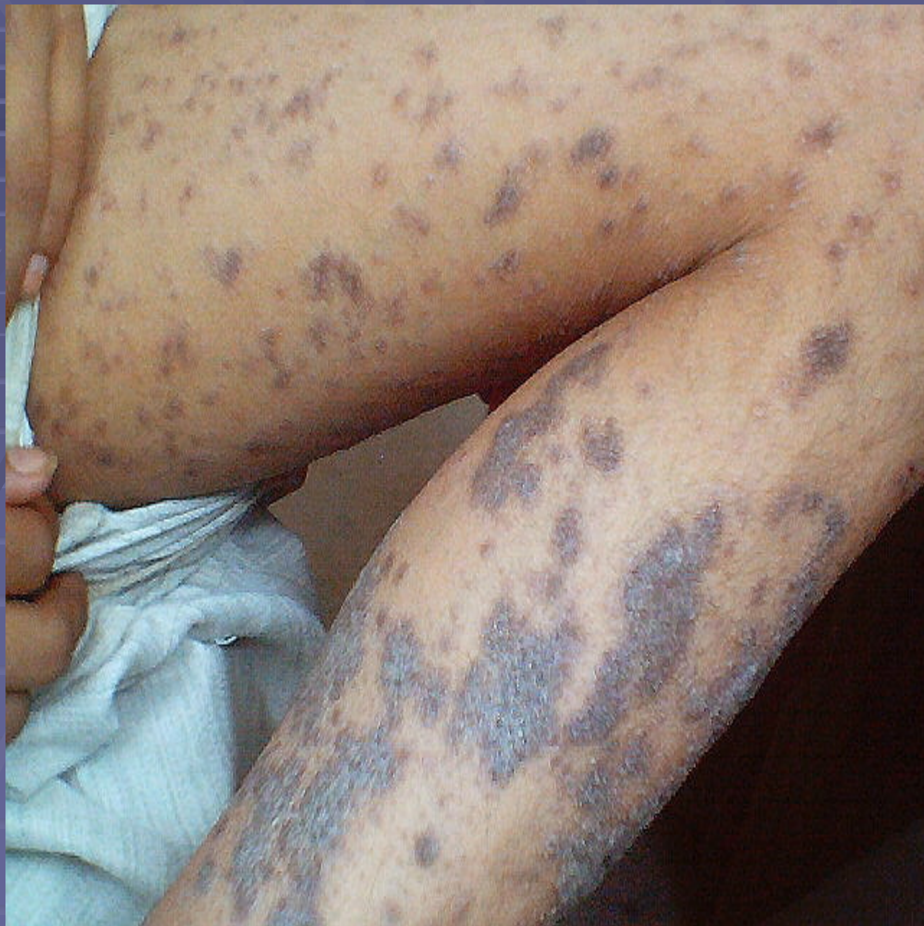
Scalp involvement:

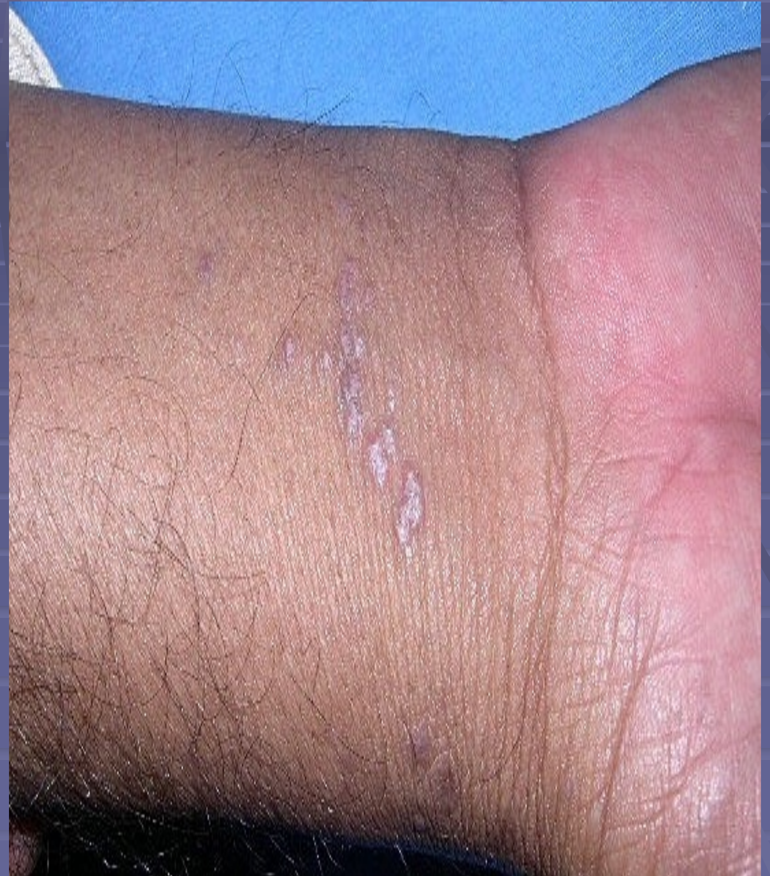
- Follicular and perifollicular violaceous, scaly, pruritic papules
- Can progress to scaring alopecia



Variations in LP :

- Hypertrophic LP
- Atrophic LP
- Erosive LP
- Follicular LP (Lichen planopilaris)
- Annular LP
- Linear LP
- Vesicular and bullous LP
- Actinic LP
- LP pigmentosus
- LP pemphigoides





Differential Diagnoses

Psoriasis

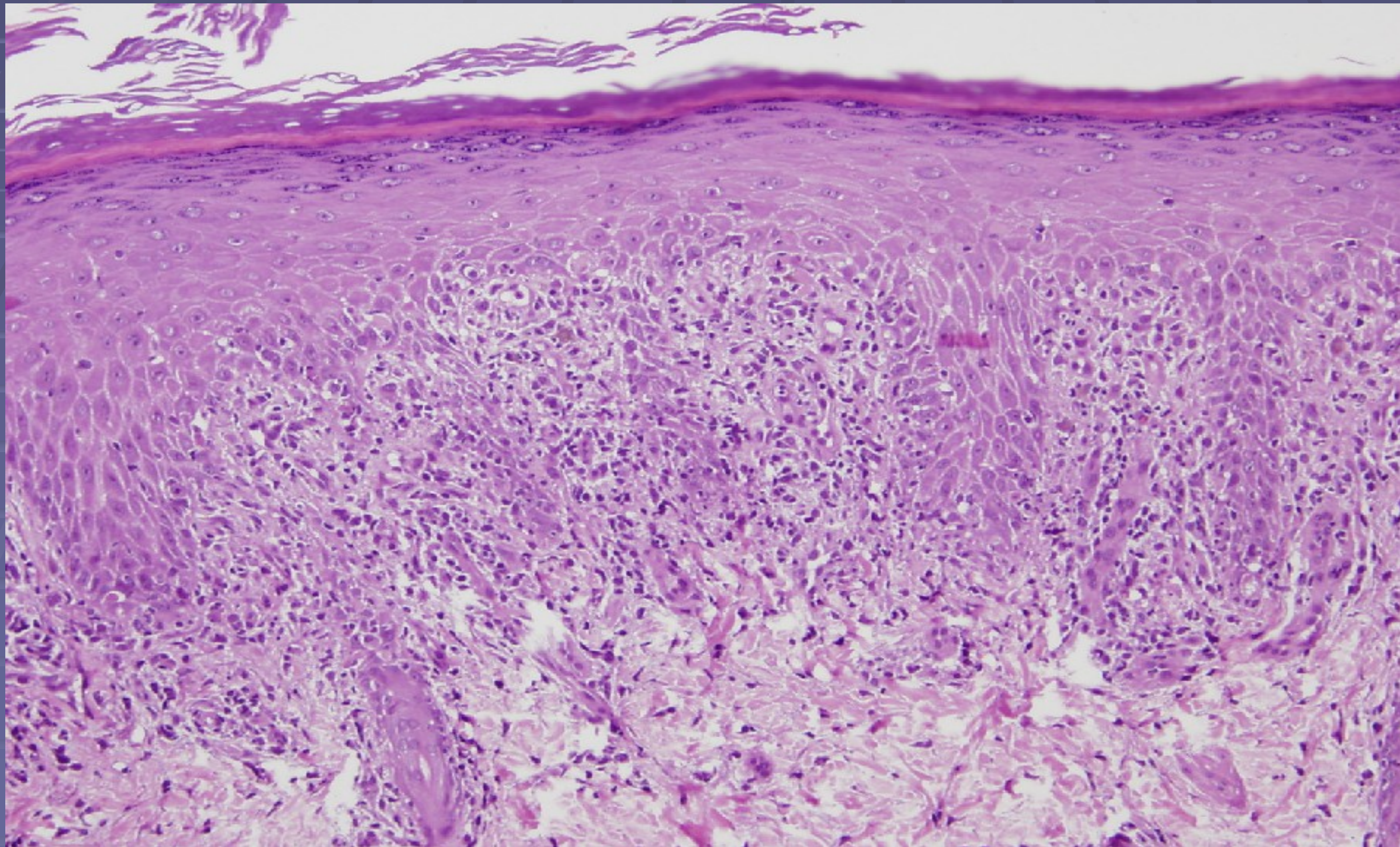
Lichenoid drug eruption

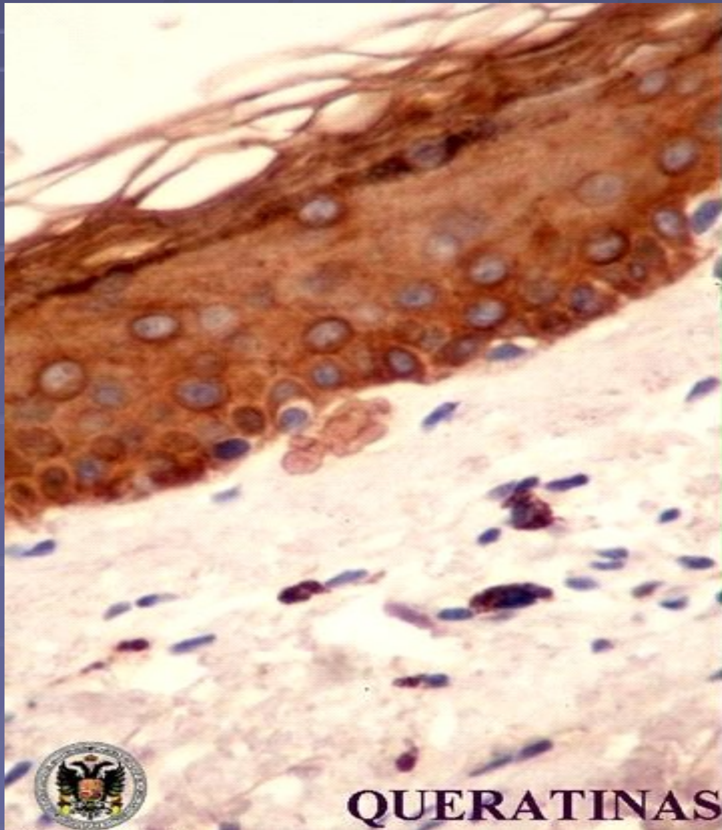
Syphilis

Tine Corporis

Histopathology

- The inflammatory reaction pattern is characteristic (lichenoid tissue reaction):
- destruction of the basal layer..
- Degenerative keratinocytes, known as colloid or Civatte bodies, are found in the lower epidermis.
- The upper dermis has a bandlike infiltrate of lymphocytic (primarily helper T) and histiocytic cells, The infiltrate is very close to the epidermis and often disrupts the dermal-epidermal junction
- IF study reveals globular deposits of IgM and complement mixed with apoptotic keratinocytes

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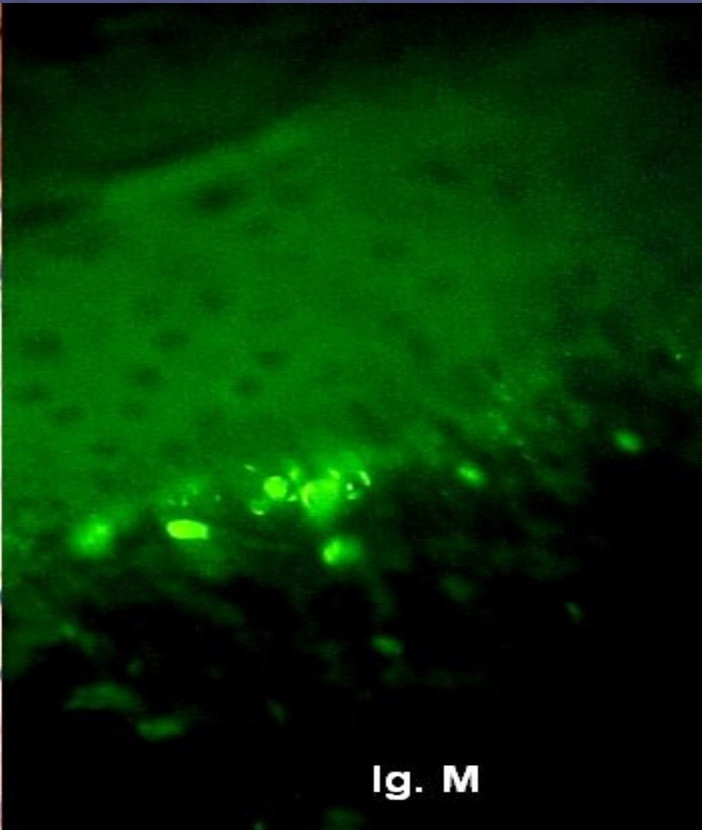

Ig. M

Treatment

- Self-limited disease tusually resolves within 8-12 months
- Sedative antihistamines for itching
- Topical steroids, particularly class I or II ointments.
- Intralesional steroid inj (hypertrophic LP)
- Systemic steroids (short course)

widespread LP:

- NBUV-B therapy OR   PUVA
- Oral Retinoids

LP of the oral mucosa:

- Topical steroids
- Topical and systemic cyclosporin
- Newer topical calcineurin inhibitors have replaced topical cyclosporin
- Oral or topical retinoids.

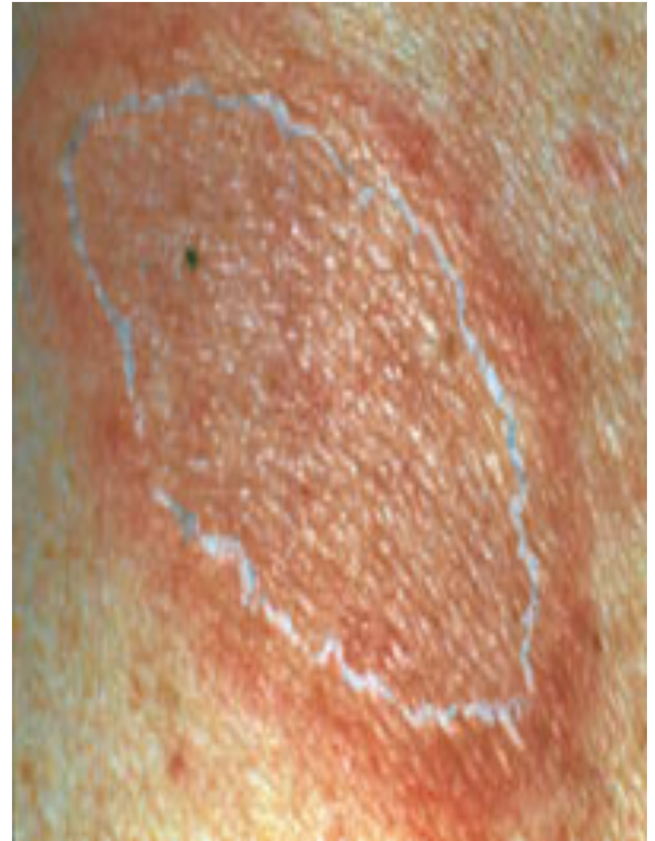
Prognosis

- Good
- In more than 50% of patients with cutaneous disease, the lesions resolve within 6 months but most cases regress within 18 months.
- Some cases recur
- Oral ulcerations in men have the potential to become malignant.
- Alopecia is often permanent

Pityriasis Rosea

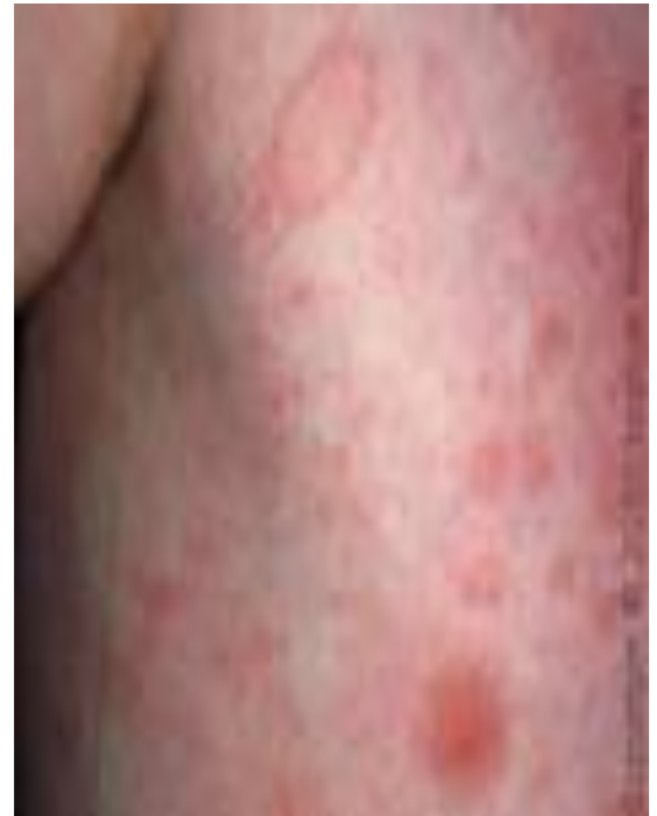
- common Acute self-limited
- Usually asymptomatic
- > 75% of pts: 10 - 35 y of age.
- Increased incidence in spring and autum
- Many pts report a mild prodromal symptoms (eg, malaise, nausea, anorexia, fever, joint pain, LN swelling, headache) or URTI within a month of onset.

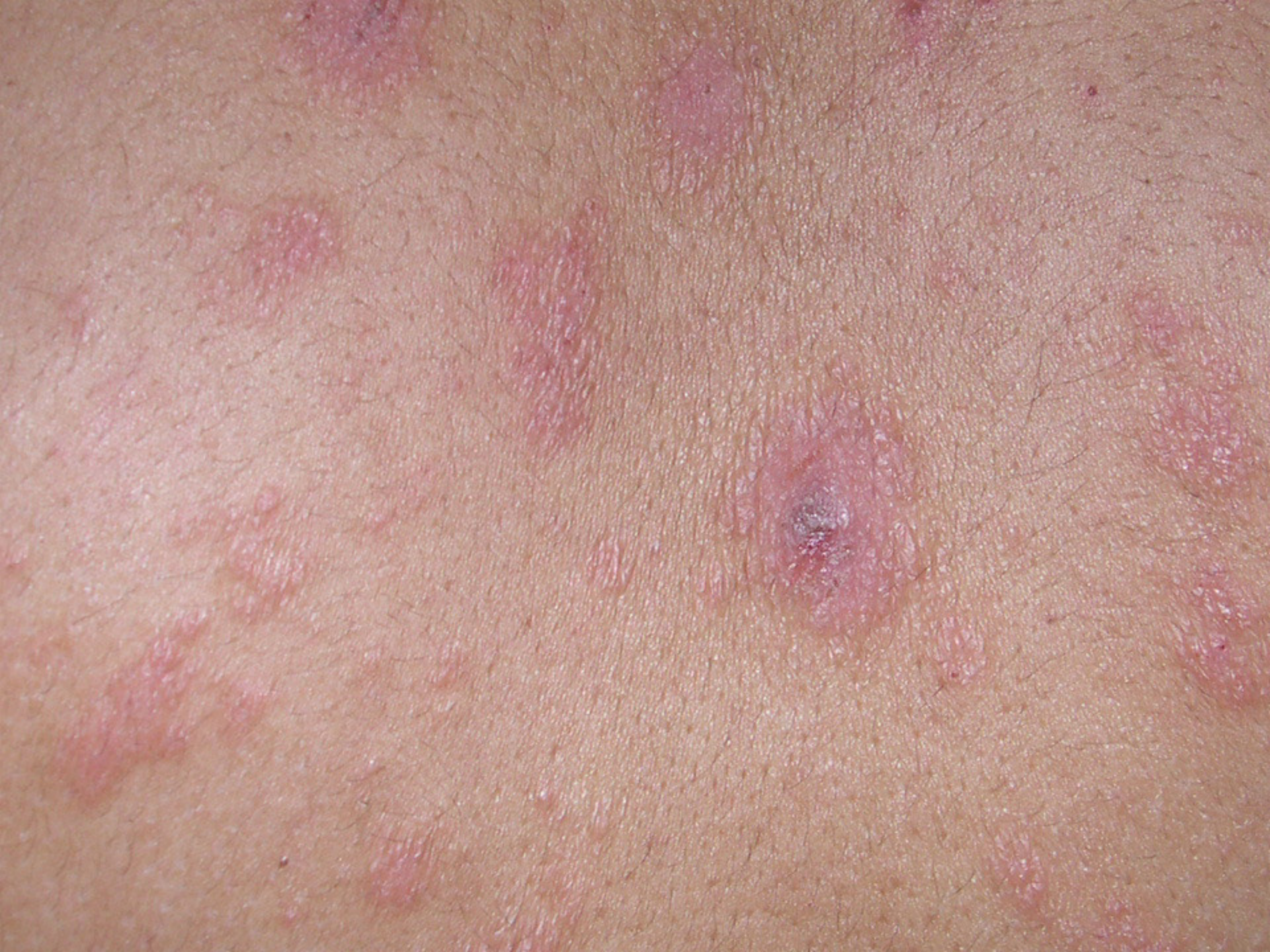
- herald patch (on the trunk).
- The lesion is1-2 cm in diameter oval or round patch with a central, wrinkled, salmon-colored area and a dark red peripheral zone. The areas are separated by a collarette of fine scales

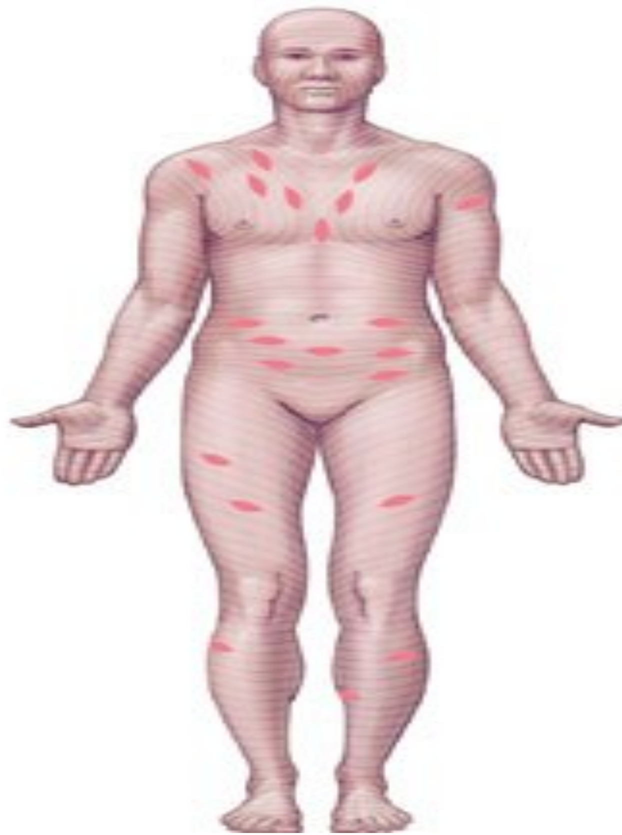
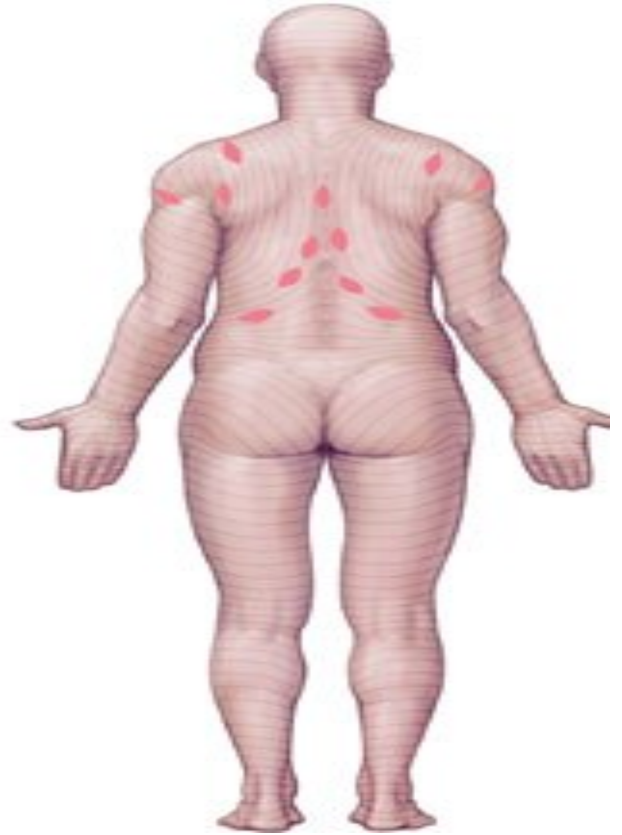


The secondary eruption

- appears at its maximum = 10 days
- **symmetric (trunk ,neck and extremities).**
- **appear as the primary patch, with the two red zones separated by the scaling ring.**
- **distributed in a Christmas tree pattern with their long axes following the lines of cleavage of the skin.**
- **Hypo and hyperpigmentary skin changes may follow the inflammatory stage**





atypical PR :

- herald patch may be missing or confluent
- The distribution of rash may be peripheral, & facial involvement may be seen in children.
- Involvement of axilla and groin (inverse variant)
- The lesions of PR may be large, urticarial ,vesicular, pustular, purpuric, and erythema multiforme–like

Causes

? Infectious exanthems:

- In clusters among contacts
- Self-limited course
- Seasonal (spring & autumn) , rare recurrence
- ? HHV-6 and 7

Drugs

bismuth, barbiturates, captopril, gold, organic mercurials, methoxypromazine, metronidazole, D-penicillamine, isotretinoin

Differential Diagnoses

Guttate psoriasis

Nummular eczema

Pityriasis versicolor

Drug eruptions

Secondary syphilis

Treatment

- In most cases, Rx is not necessary
- Avoid irritable hot baths and soap
- Symptomatic and emollients
- Topical or oral steroids If the disease is severe or widespread (e.g. vesicular PR)
- Erythromycin (pts > age 2 y)
- UVB
- Acyclovir

Prognosis

- Excellent
- **The secondary rash develops over 2 weeks, persists for another 2 weeks, and then fades over another 2 weeks .**
- **Some lesions have persisted for 3-4 months**

THANK YOU