Papulosquamous diseases

Objectives:

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

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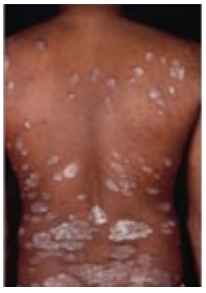
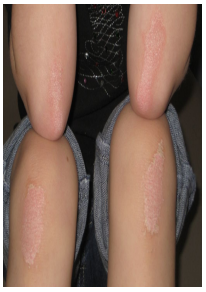
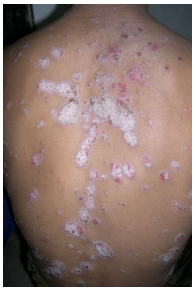
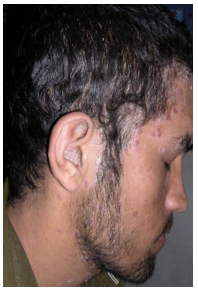
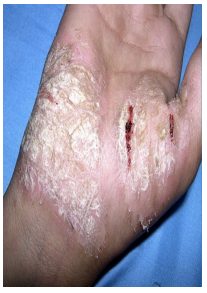
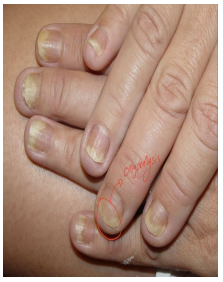
[Color index: Important | Notes | Extra]

Papulosquamous diseases

- The term squamous refers to scaling that represents thick stratum implies an **abnormal keratinization process.**
- **Papulosquamous diseases** are group of disorders characterized by scaly papules and plaques:
  1) Psoriasis.
  2) Lichen planus.
  3) Pityriasis rosea.
  4) Pityriasis rubra pilaris.
  5) Secondary syphilis.

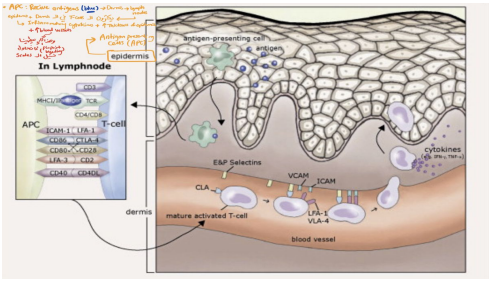
1) Psoriasis: الصدفية

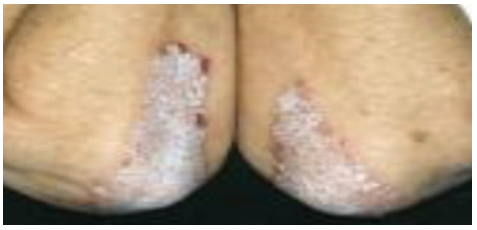
- Chronic common **non contagious** relapsing **inflammatory** disorder. inflammation,not infection!
- Genetic predisposition.
- Skin of elbow, knees, scalp, lumbosacral areas, intergluteal clefts and glans penis. trauma sites
- Joints also affected in up to **30%** of patients. in Hx: you have to ask about joint pain
- Frequency:
  - Between 2% and 2.6% of the US population.
  - Race: more common in Caucasians.
  - Sex: slightly more common in women than men.
  - Age: 2 onsets; 10-15% of new cases begin in children < 10 years. The first peak occurs in persons aged 16-22 years (type 1 psoriasis), and the 2nd in persons aged 57-60 years (type 2 psoriasis).

					
Scaly, slivery, adherent plaques.	Bilateral and very well defined. إذا بتعطين مثال على well defined قولي الصدفية لأنها مرسومة بدقة	slivery, adherent plaques.	Involvement of the scalp (scalp psoriasis)	Thick adherent scales + fissures	Pitting of nails, onycholysis (separation of nails from the nail bed)

Pathophysiology:

- Complex **multifactorial** disease influenced by genetic and immune-mediated components.
- Not completely understood.
- **Genetic predisposition for:** HLA-B13, HLA-B17, HLA-B27, DR-7, and CW6).
- **There are 2 inheritance modes:**
  - ❖ One has onset in younger age with family history of psoriasis.
  - ❖ The other has onset in late adulthood without family history of psoriasis.
- A child with one affected parent : **16%** risk.
- A child with both parents affected: **50%** risk.
- **Immunological factors:** T-cell mediated + inflammatory Th1
  - ❖ Studies shown high levels of dermal and circulating **TNF-alpha.** cytokine (from Th1)
  - ❖ TNF receptors are upregulated; Rx with TNF-alpha inhibitors is often successful.
  - ❖ Increased levels of **interferon gamma.**
  - ❖ Increased levels of interleukin 2 & 12 as well as IL-23 and IL-17.
  - ❖ Increased activity of T cells of psoriatic lesions.
- **Environmental factors:** multiple theories regarding <u>triggers</u> of disease:
  - ❖ Stress, smoking, UV, trauma and alcohol exacerbate psoriasis.
  - ❖ Infections: pharyngeal streptococcal & guttate psoriasis, HIV.
  - ❖ Drugs: NSAIDs, lithium, antimalarials, beta-blockers, and withdrawal from systemic corticosteroids. in genetically susceptible pts.
  - ❖ Association with obesity.
  - ❖ In many patients, no obvious trigger exist at all. Idiopathic
- **Epidermal cell kinetics:**
  - ❖ The growth fraction of basal cells is increased to almost 100% compared with 30% in normal skin. المصنع سريع جداً، الجلد إعادة يتبدل كل ٢٨ يوم لكن اللي يصير هنا إنه يتبدل كل ٣-٥ أيام
  - ❖ The epidermal turnover time is shortened to less than 10 days compared with 30 to 60 days in normal skin. المشكلة في استبدال خلايا الجلد ketatinaization
  - ❖ 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** not specific



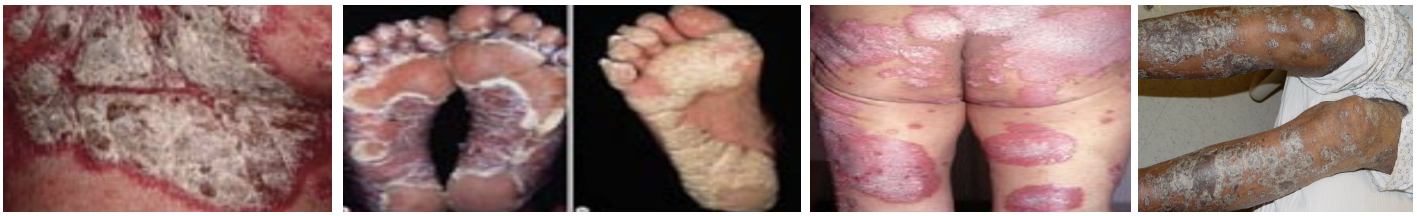


Clinical features: (types)

1) Plaque psoriasis (psoriasis vulgaris) most common
2) Guttate psoriasis.
3) Inverse psoriasis.
4) Pustular psoriasis.
5) Erythrodermic psoriasis.
6) Psoriatic arthritis.
7) Psoriatic nail.
8) Scalp psoriasis.

Plaque psoriasis

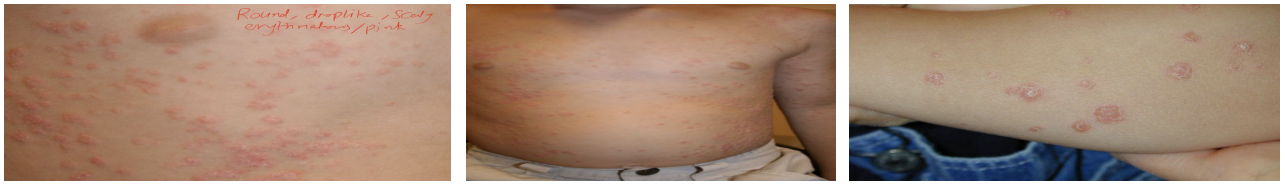
- Well circumscribed red plaques covered with a silvery white thick scale.
- If scale scraped away, it will reveal inflamed skin beneath with pinpoint bleeding **(Auspitz sign).**
- **Symmetrical on extensor surfaces of knees, elbows, scalp, and sacral area.**
- Up to 10-20% of patients with plaque psoriasis may evolve into more severe disease such as pustular or erythrodermic psoriasis. it's a chronic disease that could transform into acute one



*well defined bilateral scaly erythematous plaques.
*Buttock area examination is very important in psoriasis.
*First pic on the Rt: typical well defined bilateral silvery thick plaques on extensors.

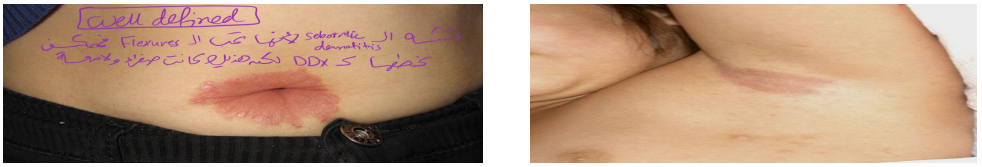
Guttate psoriasis

- Children > adult
- Presents as **small droplike salmon pink scaly papules**, 1-10 mm in diameter. well defined
- On the **trunk and the proximal extremities.**
- Suddenly, 2-3 weeks after URTI 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 occurs in flexural areas and does not have scales that's why it's called inverse.
- It is characterized by smooth, inflamed lesions without scaling due to the moist nature of the area.

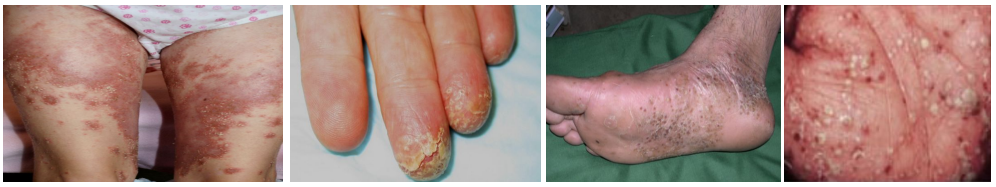


Pustular psoriasis

- Uncommon form of psoriasis.
- Sterile pustules on palms and soles or diffusely over the body.
- **Pustular psoriasis = erythema then scaling** erythema and on top of it there are pustules
- Psoriasis vulgaris may be present before, during or after.
- Could be divided into several types:

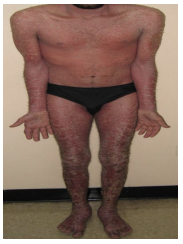
1) **Generalized type (Von Zumbusch variant):**
   - Generalized erythema studded with interfolecular pustules.
   - Fever, intense ill feeling, tachypneic, tachycardic. pts come to ER
   - Absolute lymphopenia with polymorphonuclear leukocytosis up to 40,000/μL.
2) **Localized form (palms and soles).**

- **Causes of pustular psoriasis:**
- Withdrawal of systemic steroids.
- Drugs, including salicylates, lithium, 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.** it's an emergency
- 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.**

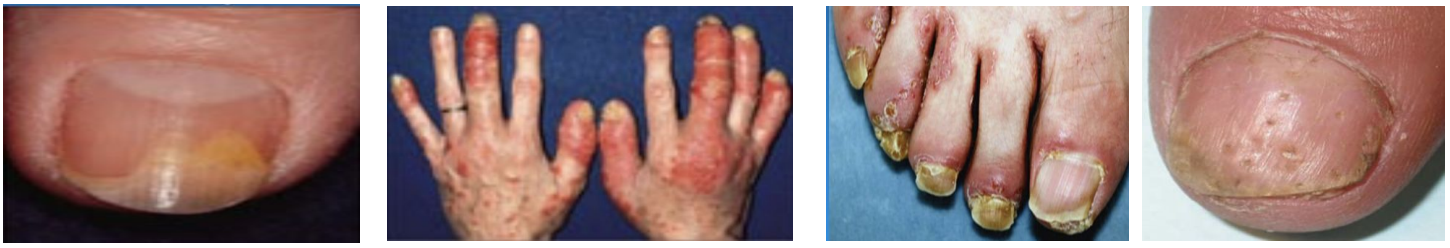


Psoriatic arthritis

- 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 appear before psoriasis.
- Most commonly a seronegative oligoarthritis. RF -Ve
- Asymmetrical 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 occuring in 5% of patients with psoriatic arthritis.
- Spondylitis occurs in about 5% of patients with psoriatic arthritis and is often asymptomatic.

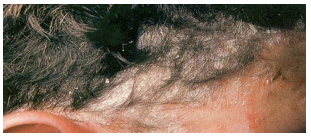
Psoriatic nail

- Psoriatic nail disease in 10-55% of all patients 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 patients with psoriatic arthritis.
- **Oil drop or salmon patch/nail bed** مميزة للصدفية
- Pitting, subungual hyperkeratosis, onycholysis, Beau lines.



Scalp psoriasis

- 50% of patients with psoriasis.
- **Erythematous raised plaques with silvery white scales.**
- Seborrheic dermatitis: yellowish greasy itchy (fungal)
- Psoriasis: Silvery and adherent



◄ Differential diagnosis of psoriasis:

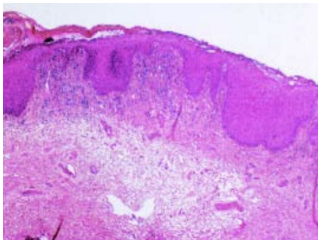
1) **Seborrheic dermatitis**
2) Nummular eczema
3) Lichen planus
4) Pityriasis rosea
5) Drug eruptions
6) Reiter's disease
7) Syphilis (mimicker)
8) Tinea corporis
9) Onychomycosis

◄ Investigations: mostly, it's a clinical dx

- Skin biopsy
- Others

◄ Histopathology:

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



◄ Prevention and treatment of psoriasis:

- Avoid injury to skin (sunburn and other physical trauma).
- Avoid drugs known to worsen the problem.
- Treatment regimens must be INDIVIDUALISED according to the sex, age, occupation, severity, other health conditions and available resources.
- Rx: topical agents, phototherapy, and systemic agents including biologic therapies.

1) Topical corticosteroids:

- Anti- inflammatory effects.
- **Betamethasone dipropionate** (Diprolene) 0.05% cream
- Modify body's immune response to diverse stimuli.
- Systemic side effects (rare): HPA axis suppression, cushing syndrome.
- Local/cutaneous side effects (common): atrophy of the epidermis and dermis, striae, purpura, telangiectasia, tachyphylaxis.

2) Coal tar: القطران

- Antipruritic and antibacterial that inhibits deregulated epidermal proliferation.
- In shampoos or lotions. for scalp psoriasis
- Useful in hair bearing areas.
- S/E: messy, carcinogenicity?

3) Vitamin D3 analogs:

- Calcipotriene (Dovonex).
- Regulates skin cell production and development.
- S/E: irritation, transient but reversibly elevate serum calcium level. caution in cardiac pts

4) Keratolytic agents:

- To remove scale, to smoothen the skin.
- **Anthralin 0.1-1% :** short contact. S/E: irritation, staining.
- **Salicylic acid:** scalp, palms and soles. S/E: salicylicism if high concentration. ماينفع للأطفال والحوامل

5) Phototherapy:

- **Psoralen plus UVA (PUVA):**
  - Ingestion of 8-methoxypsoralen (8-MOP) then UVA.
  - 2 or 3 times per week.
  - Long-term remession.
  - SE: nausea, phototoxicity, lentigines.
  - If > 260 individual PUVA sessions, **11 fold increase in SCC (male genitalia) + malignant melanoma.**

- **Narrowband UVB:** هذا اللي يستخدمونه بمستشفانا
  - Range around 311 nm
  - Not as effective as PUVA
  - Less carcinogenic > safer than PUVA.

6) Retinoids:

- Stimulate cell differentiation.
- Can be used in combination with UV phototherapy.
- S/E: **Teratogenicity**, hyperlipidemia.
- Example: **Acitretin**

7) Antimetabolites:

- Methotrexate: interferes with DNA synthesis, repair and cellular replication.
- 2.5-7.5 mg PO q12h for 3 doses/week.
- Give with folic acid 1 mg/d
- S/E: **Teratogenicity**, liver, BM & renal.

8) Immunosuppressive:

- Cyclosporine: remission is rapid, skin lesions tend to recur after Rx is stopped.
  S/E: risk of renal damage + it increases the BP.
- Other medications: Mycophenolate mofetil, Hydroxyurea.

9) Biologic therapies (new treatment currently approved for the treatment of psoriasis):

- **Biologic therapies:**
  - Alefacept.
  - Efalizumab.
  - Secukinumab.
  - Ustekinumab (Stelara).
- **Tumor necrosis factor inhibitors:**
  - Infliximab (Remicade).
  - Etanercept.
  - Adalimumab (Humira).

◄ Prognosis:

- The course of plaque psoriasis is unpredictable.
- Relapses occurring in most patients.
- Early onset and a family history of disease are considered bad prognostic factors.

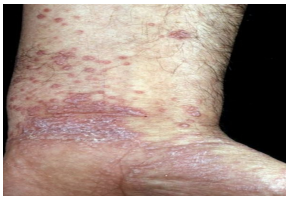
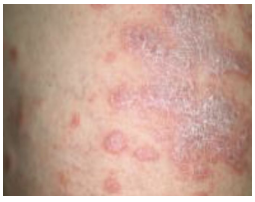
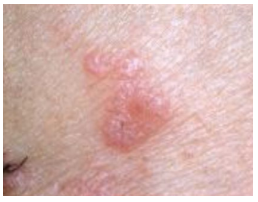
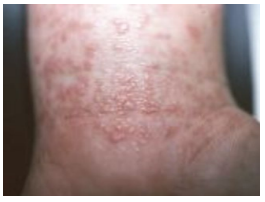
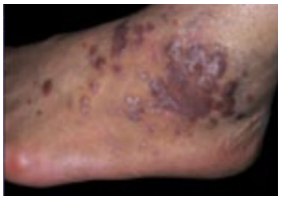
2) Lichen planus: الحزاز

- Is a **pruritic**, papular eruption characterized by its **violaceous color**, **polygonal shape** and sometimes, **fine scales** **on the flexor surfaces of upper extremities, genitalia and on the mucous membranes.** ITCHY/PRURITIC/PAPULAR
- **Frequency:** LP is reported in approximately 1% of all new patients seen at health care clinics in US. F=M.
- **Age:** rare in children, more than two thirds of patients are aged 30-60 years. However, can occur at any age.
- **Causes:**

  - Is a cell-mediated immune response of unknown origin. Hyper Reactive immune response
  - 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. اكتشفوا إن الحشوات المعدنية للأسنان ممكن تسبب الحزاز الفموي

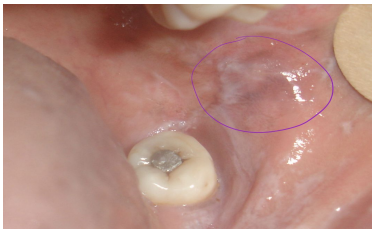
◄ Clinical features:

- 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 striae)**.
- Sites: flexors of wrist and legs.

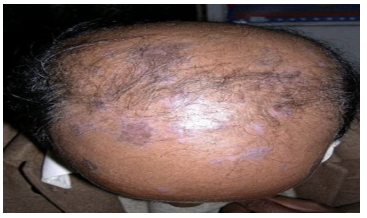
◄ Mucous membranes involvement:

- Common and may be found without skin involvement. مهم تفحصين الفم حتى لو ما اشتكى المريض
- 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, plaque like,atrophic, papular,erosive "the worst, has increased risk for SCC",and bullous.
- Lesions may also be found on genetalia and GIT.



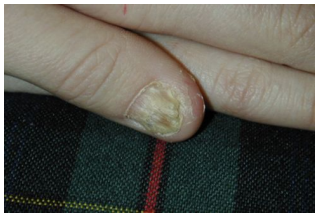
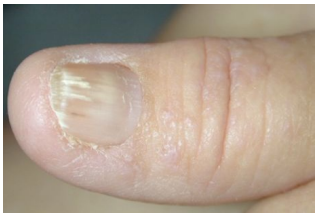
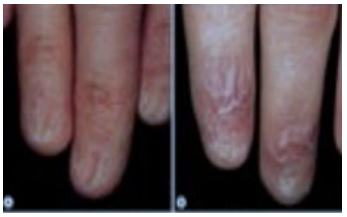
◄ Scalp involvement:

- Follicular and perifollicular violaceous, scaly, pruritic papules.
- Can progress to **scarring alopecia**.



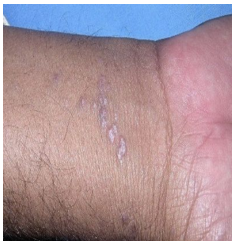
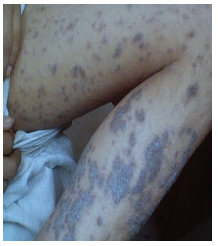
◄ Nail involvement:

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

◄ Variations in LP:

- Hypertrophic LP.
- Atrophic LP.
- Erosive LP.
- Follicular LP (lichen planopilaris)
- Annular LP
- Linear LP
- Vesicular and bullous LP
- Actinic LP
- LP pigmentosum
- LP pemphigoides

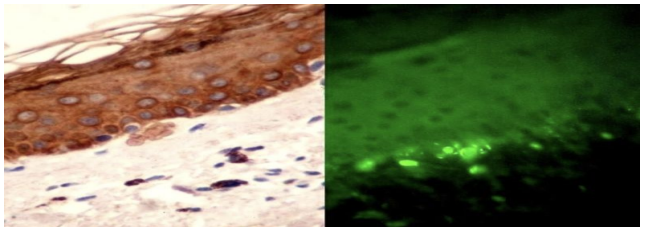
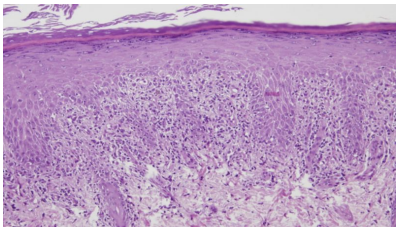
◄ Differential diagnosis:

- Psoriasis
- Lichenoid drug eruption
- Syphilis
- Tinea 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 band-like 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 (immunofluorescence) study reveals globular deposits of **IgM and complement** mixed with apoptotic keratinocytes.

◄ Treatment:

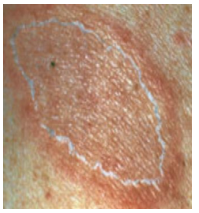
- Self-limited disease usually resolves within 8-12 months.
- Sedative antihistamine for itching.
- Topical steroids particularly class 1 or 2 ointments.
- Intralesional steroid injection (hypertrophic LP).
- Systemic steroids (short course).
- **Widespread LP:** NBUV-B therapy or PUVA, oral Retinoids.
- **LP of oral mucosa:** topical steroids, topical and systemic cyclosporine, newer topical calcineurin inhibitors have replaced cyclosporine, oral or topical Retinoids.

◄ Prognosis:

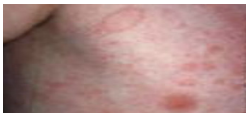
- Good, in more than 50% of patients with cutaneous disease, the lesions resolve within 6 months but most cases regress within 18 months.
- Some cases recur.
- Oral ulcerations in men have the potential to become malignant.
- Alopecia is often permanent.

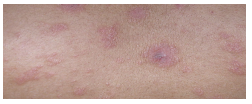
3) Pityriasis Rosea: النخالية الوردية

- Common Acute self-limited
- Usually asymptomatic
- > 75% of pts: 10 - 35 y of age.
- Increased incidence in spring and autumn
- 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 is 1-2 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





Differential Diagnosis	Treatment	Prognosis
• Guttate psoriasis • Nummular eczema • Pityriasis versicolor • Drug eruptions • Secondary syphilis	• 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	• 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