Blistering Disorders

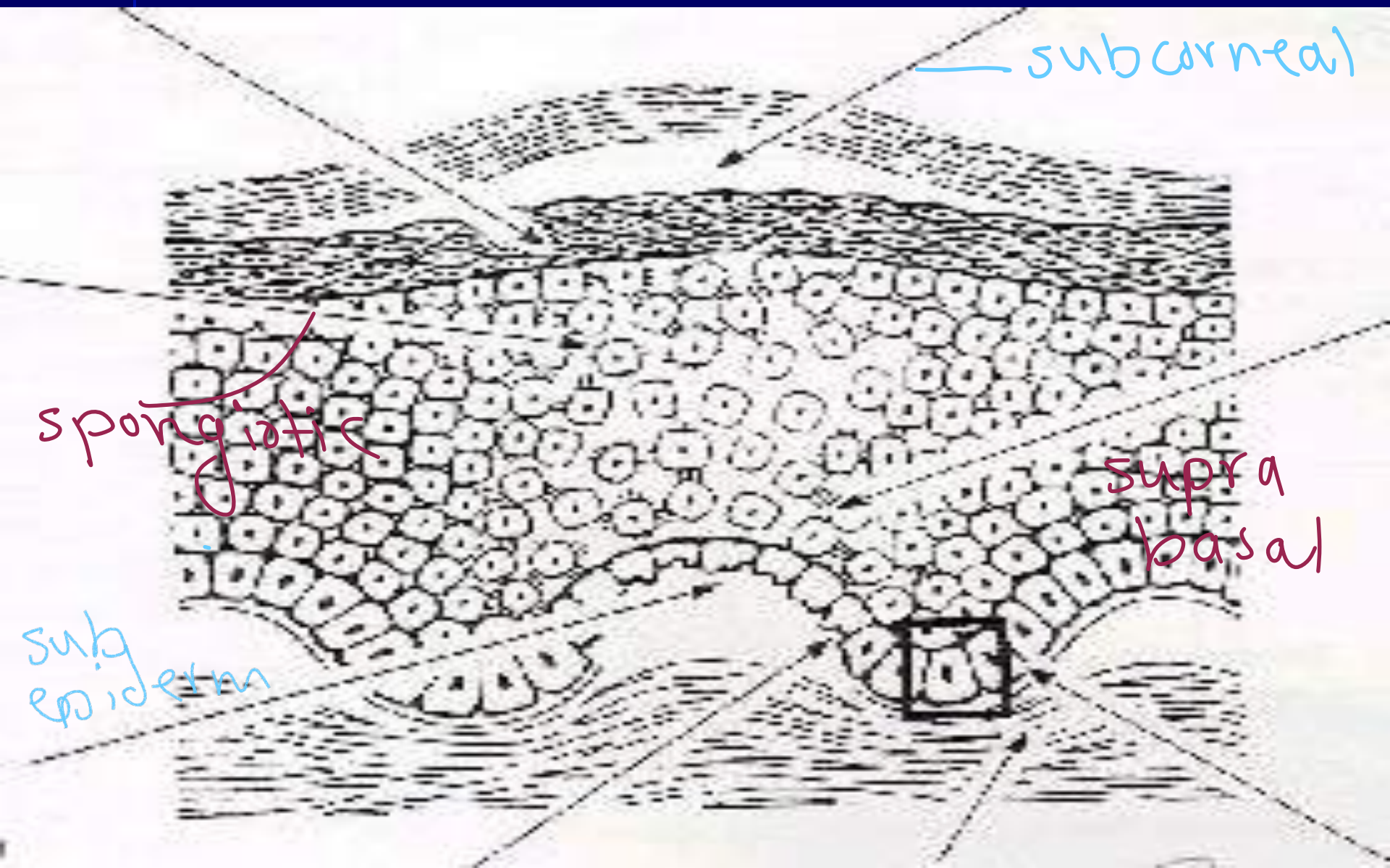
Blistering Disorders

Blisters :-are accumulation of fluid within or under the epidermis .

The appearance of blister is determined by the level at which it form .

1. Intra epidermal blisters :- appear within the prickle cell layer of the epidermis , & so have thin roof & rupture easily to leave an oozing denuded surface ; this tendency is even more marked with sub corneal blisters .
2. Sub epidermal blisters :- occur between the dermis & epidermis , their roof are relatively thick , & so they tend to be tense & intact .

Classification of bullous diseases according to the location of blister in the skin :-



Classification according to their location

Subcorneal blister:-

1-bullous impetigo.

2-candidiasis.

3-miliria crystallina.

4-SSSS.

Intraepidermal blister:-

1-acute dermatitis.

2-viral vesicle.

3-pemphigus vulgaris.

4-Scabies.

5-friction blister.

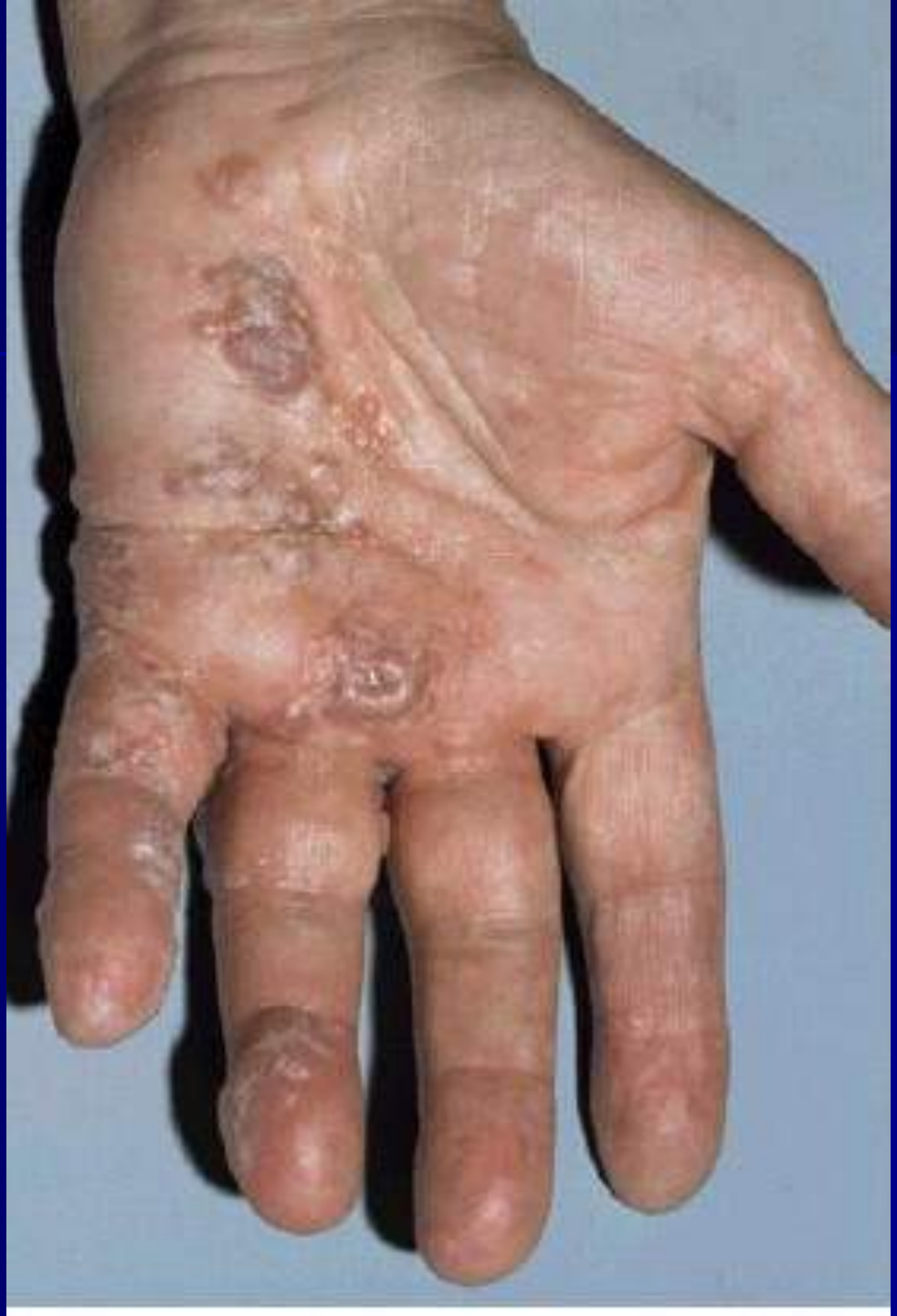
Subepidermal blister:-

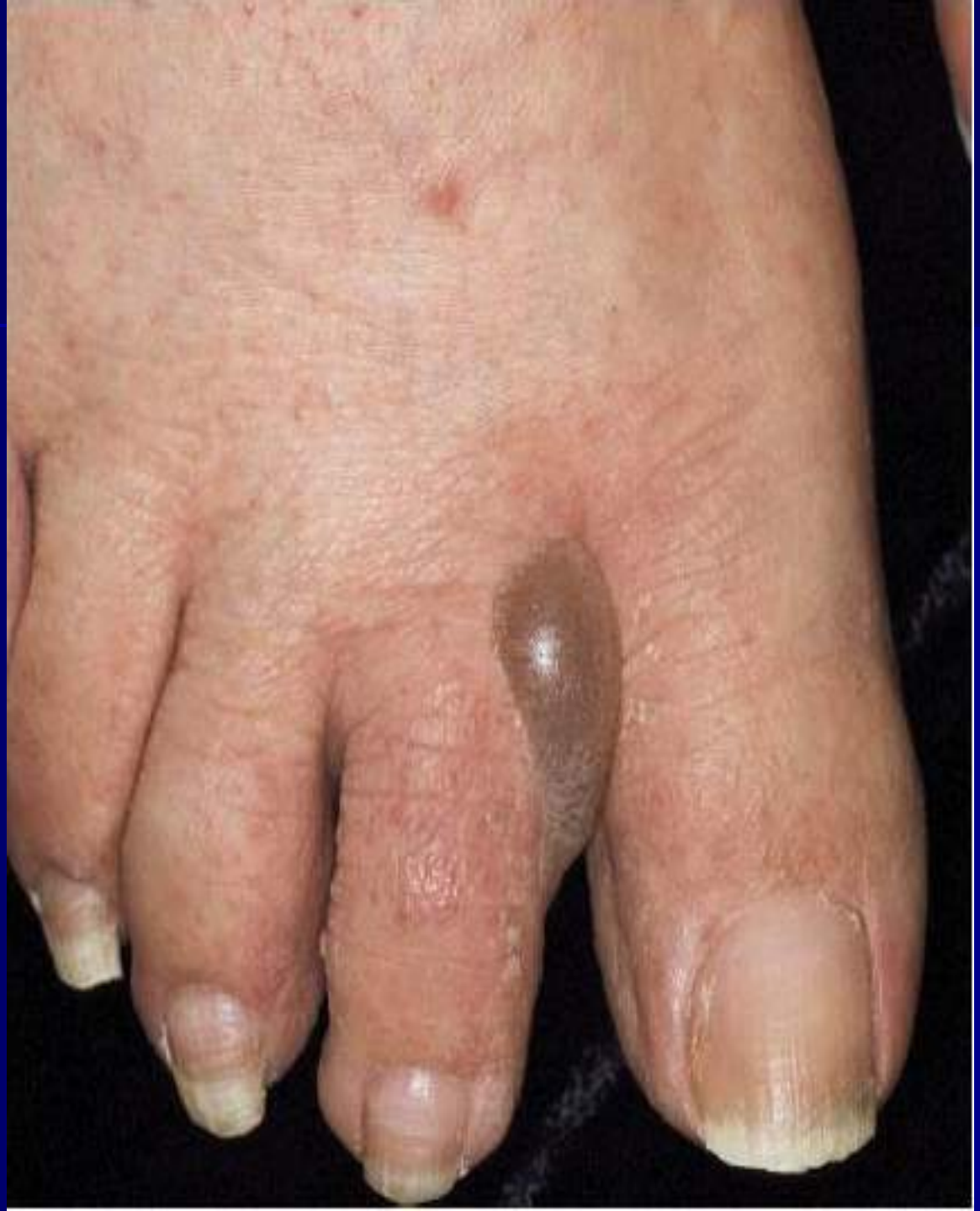
1-bullous LE.

2- bullous lichen planus.

3-TEN.

4- erythema multiforme.





Blistering Disorder

Classification according to cause :-

1. Infection :- bullous impetigo , herpes simplex or zoster , SSSS , & candidiasis .
2. Auto immune :- pemphigus , bullous LE , & bullous lichen planus .
3. Hereditary :- epidermolysis bullosa .
4. Unknown :- toxic epidermal necrolysis .
5. Miscellaneous :- dermatitis , erythema multiforme , friction blister .

Autoimmune Blistering Disorder

These are disorders by which the damage is created by auto antibodies directed at molecules that normally bind the skin . These diseases tend to be chronic & many are associated with tissue bound or circulating Ab. . These include :-

1. Pemphigus .
2. Bullous LE .
3. Bullous lichen planus .

Blistering Disorders

Diagnosis of blistering disorder :-

1. History :- sometimes history helps to reach the diagnosis , as in cold or thermal injury , or in an acute contact dermatitis .
2. Examination :- the morphology or distribution of a bullous eruption gives the diagnosis , as in herpes simplex or zoster .
3. Investigation :- when the cause is not obvious (skin biopsy , direct immunoflouresence , indirect immunoflouresence) .

Pemphigus

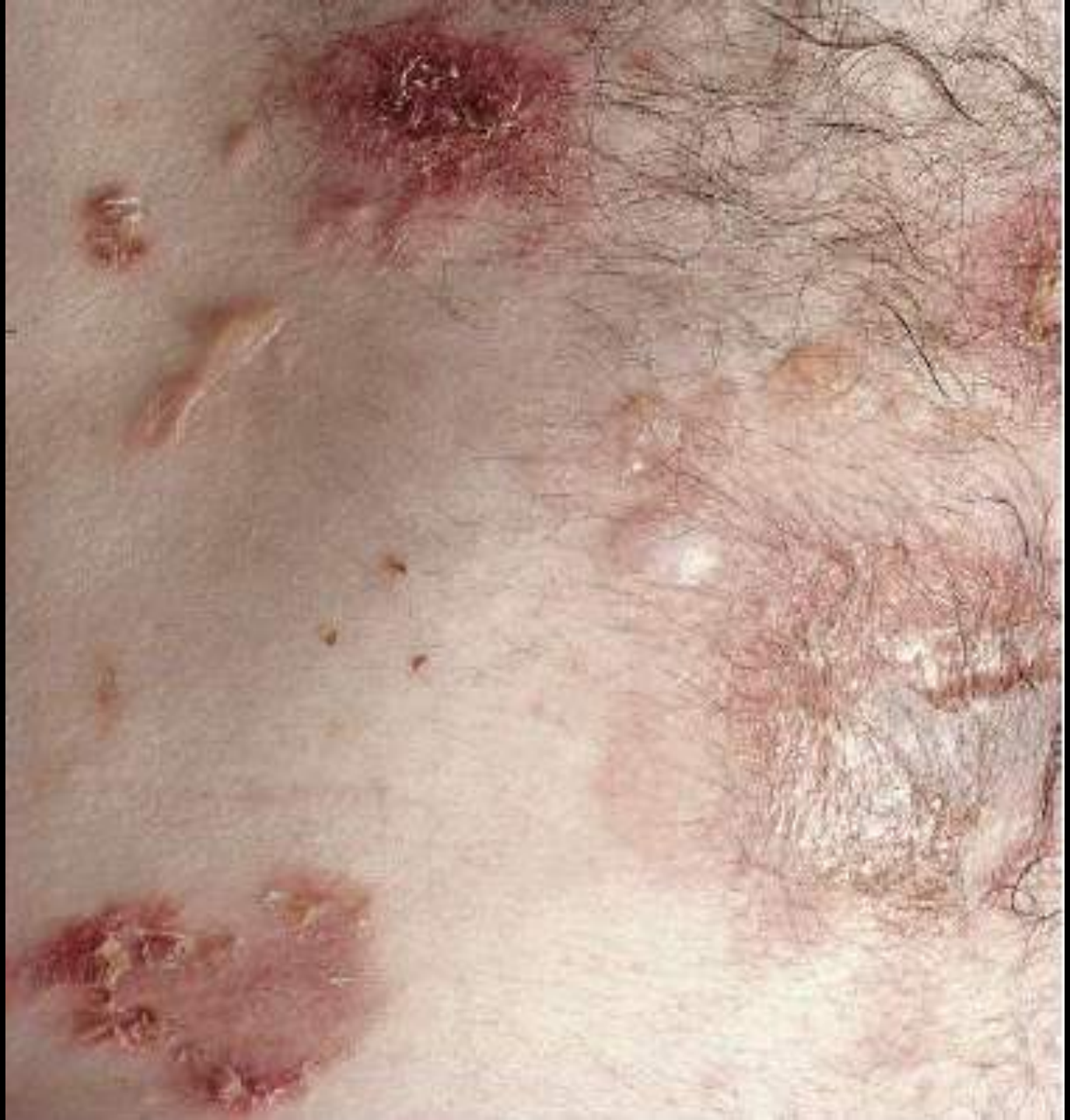
Pemphigus :- is a chronic lethal autoimmune intra epidermal blistering disease involving the skin & mucous membrane because of circulating IgG antibodies directed against the cell surface of keratinocytes , destroying the adhesion between epidermal cells producing blisters .

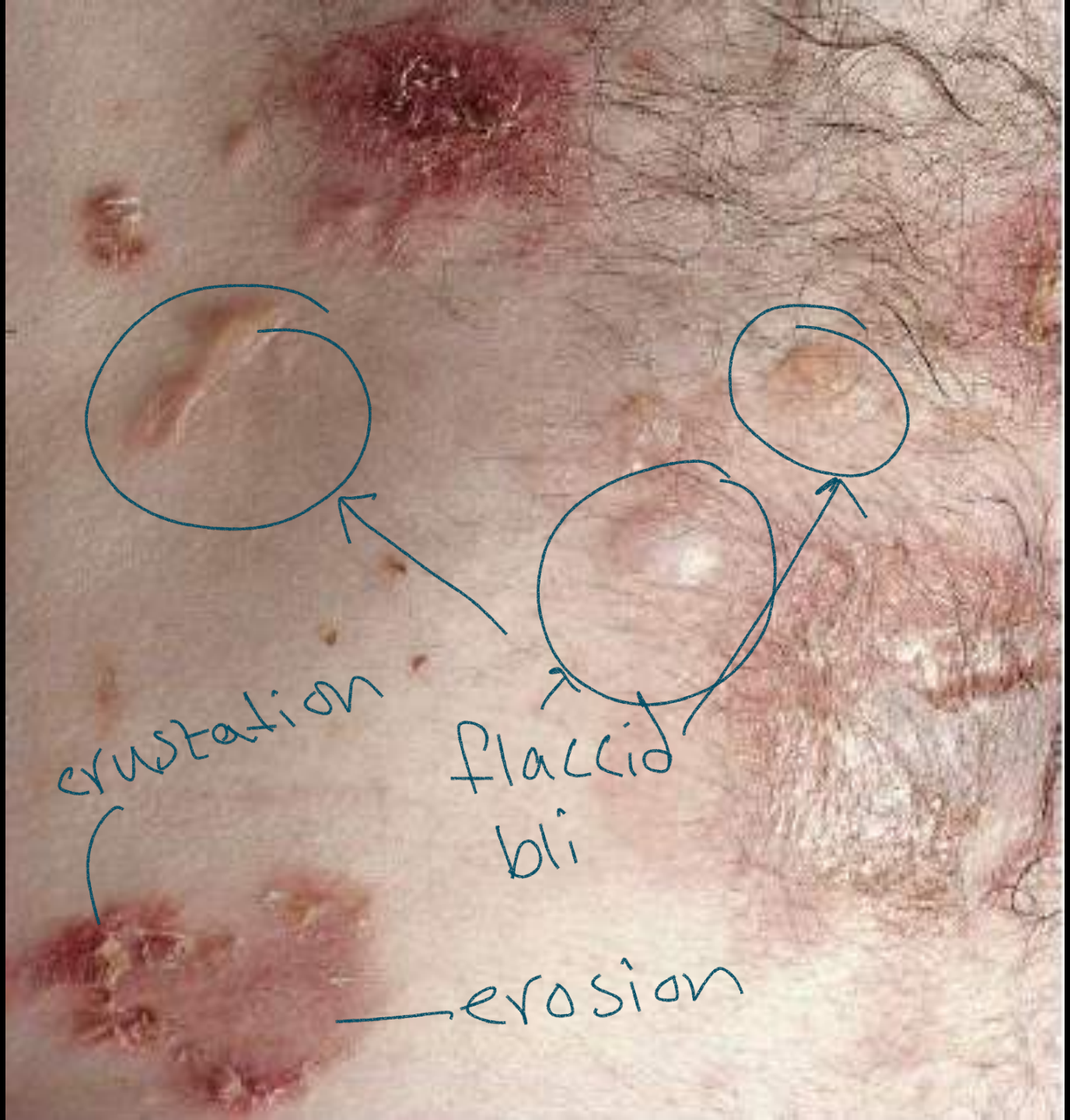
Course :- of all form of pemphigus is prolonged even with treatment & mortality rate of pemphigus is still at least 15% .

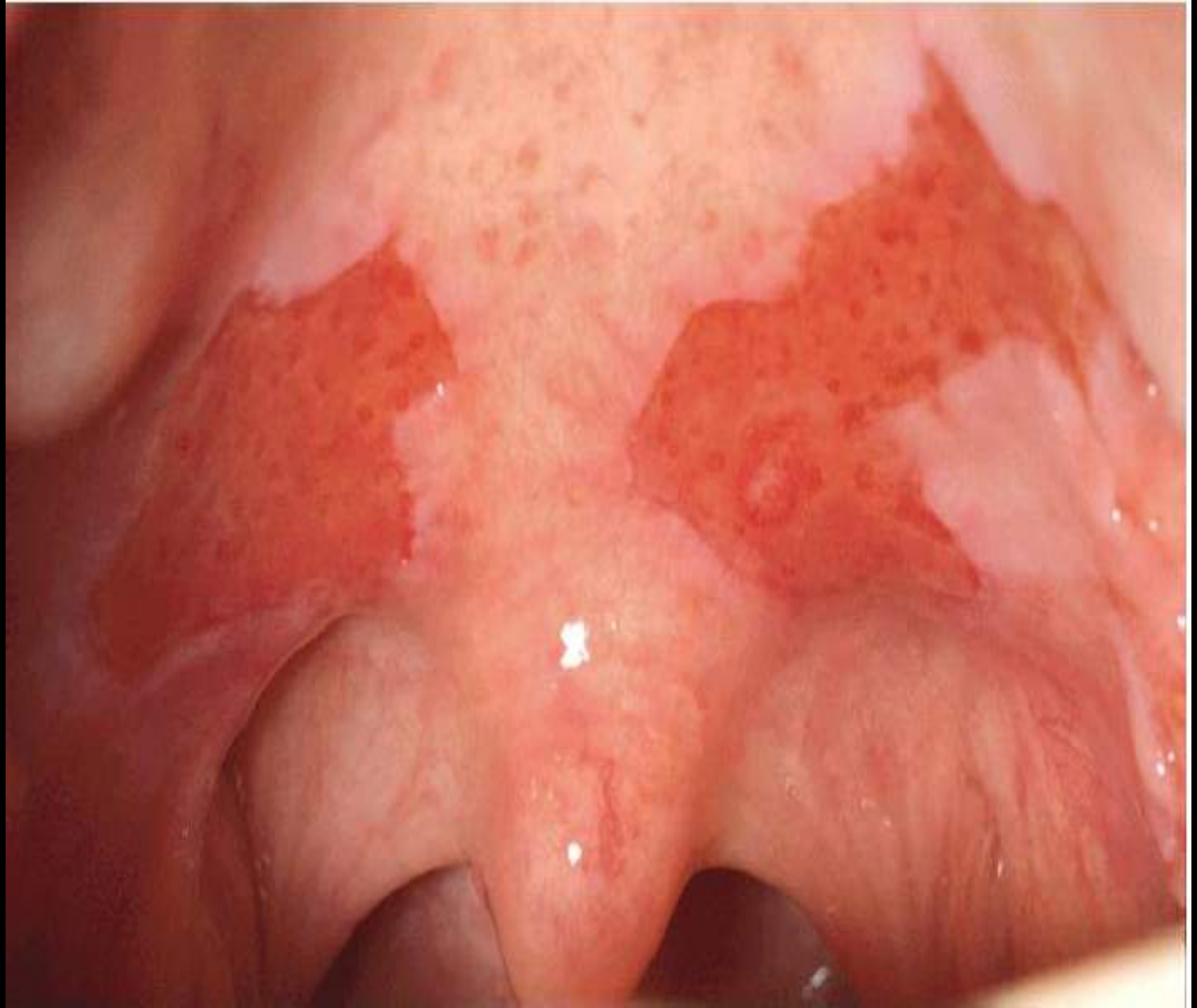
Pemphigus Vulgaris

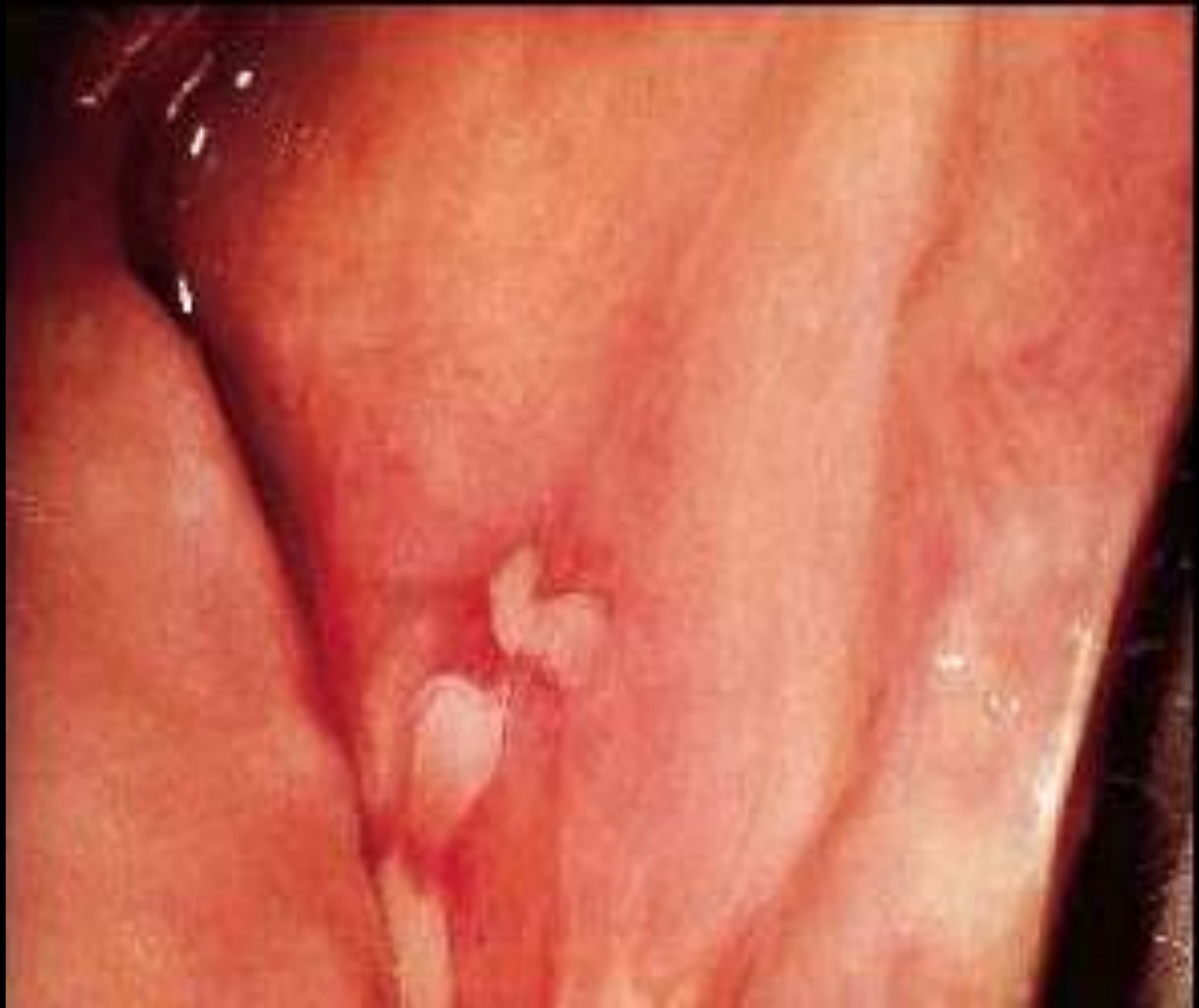
The most common & severest form of pemphigus , charact. by thin walled flaccid , easily ruptured bullae that appear on either apparently normal skin & mucous membrane or on erythematous base .the bullae soon rupture to form erosions , covered with crusts that have little or no tendency to heal . The healed lesions often leave post inflammatory hyper- pigmentation .

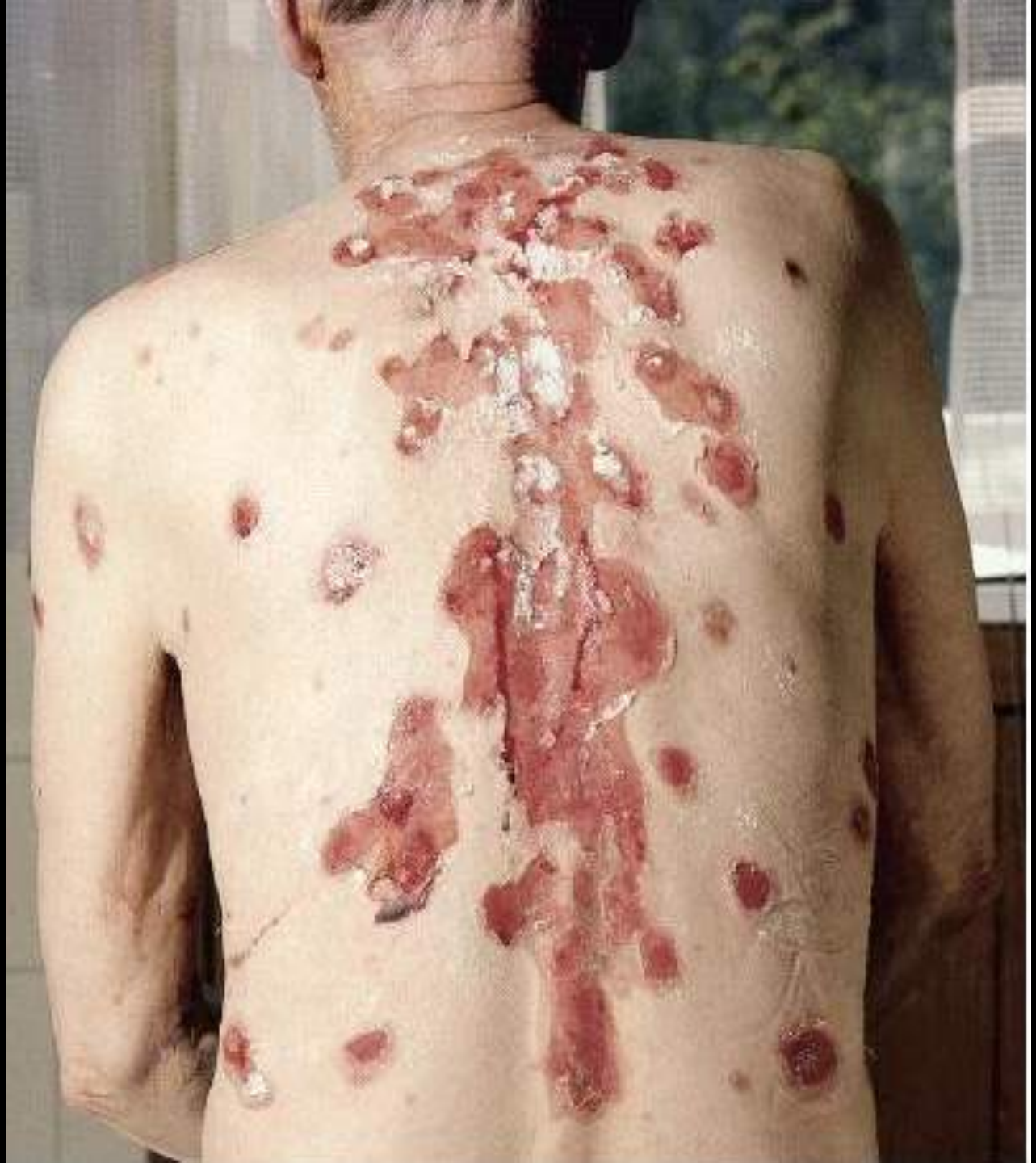
- It may begin in many ways , but most commonly the lesions appear first in the mouth in up to 60% .
- During the course of illness , oral mucosal involvement occur in 100% in the form of short lived bullae that quickly rupture in to painful erosions , lips are fissured & crusted .

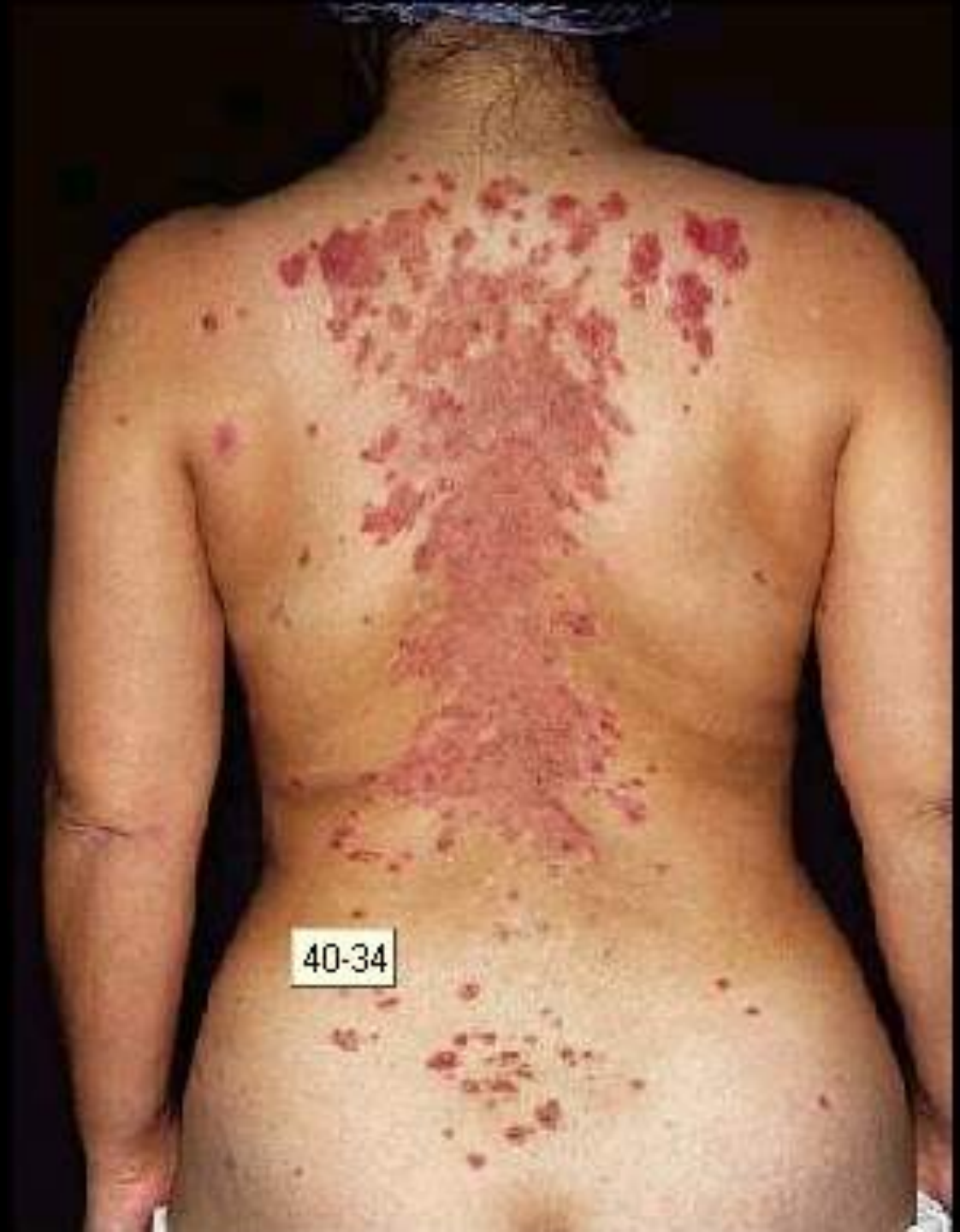


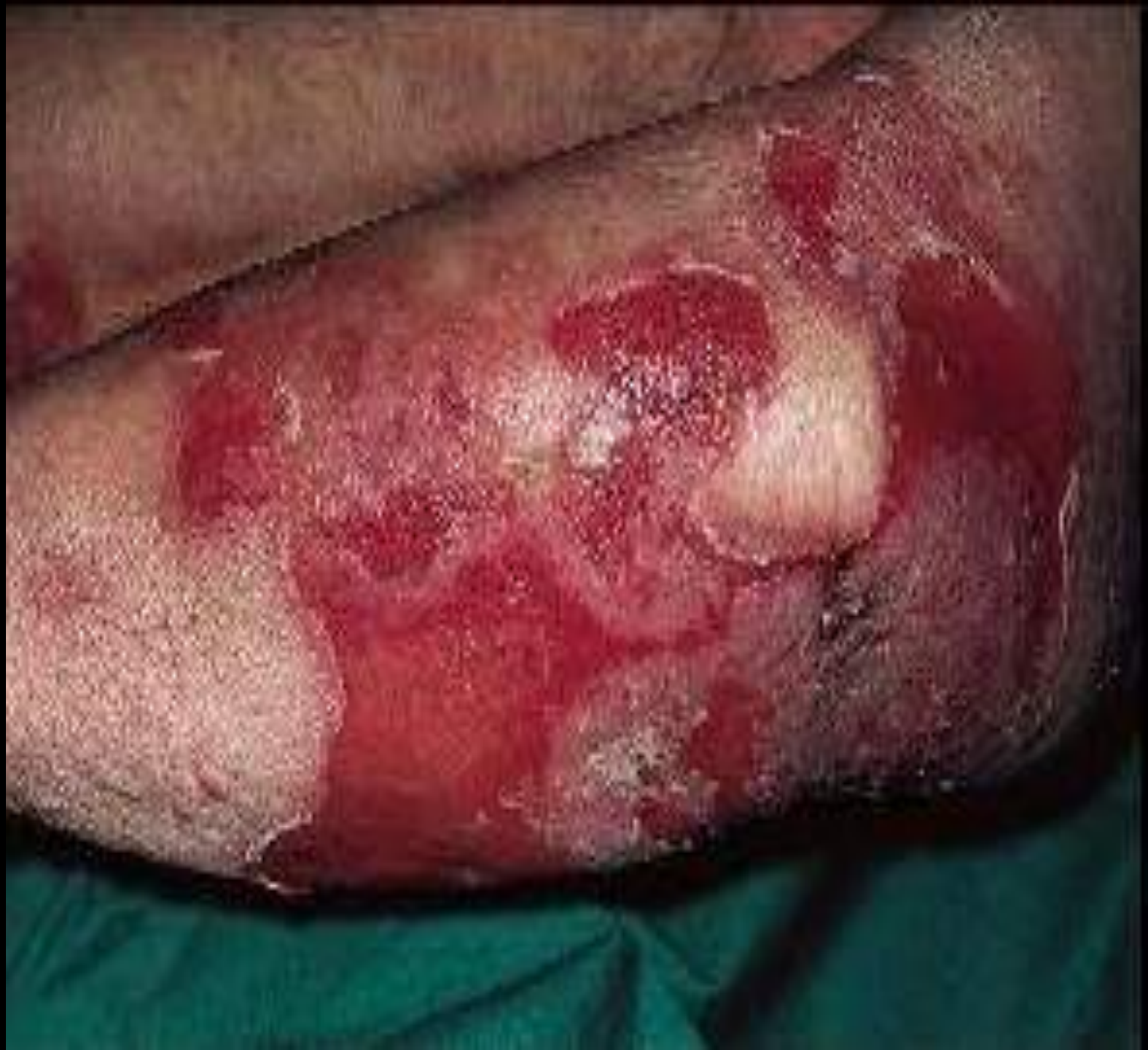

crustation
flaccid
bli
erosion








40-34



Pemphigus Vulgaris

- Also there is involvement of the throat , esophagus , vagina , & penis .
- The next most common sites are the groin , scalp , face , neck , axillae or genitalia .
- Nikolsky sign is +ve (there is an absence of cohesion in the epidermis , so the upper layers are easily made to slip laterally by slight pressure or rubbing .
- male = female .
- 5^{th} – 6^{th} decades .
- Mediterraneous descent .

Pemphigus Vulgaris

Diagnosis :-

1. Skin biopsy for light microscopy :- small , early vesicles or skin adjacent to a blister shows an intra-epidermal bulla , supra basal acantholysis , mild - moderate infiltrate of eosinophils .
2. Direct immunoflouresence :- shows deposition of IgG & in most instances C_3 in the intercellular space of the epidermis .
3. Indirect immunoflouresence :- detecting serum IgG which is present in all forms of pemphigus reflecting disease activity .

Pemphigus Vulgaris

Complication :-

1. Due to high dose of systemic steroid & immunosuppressive drugs .
2. Infections of all types are common . The large areas of denuded skin may become infected .
3. Sever oral ulcers make eating painful .
4. Fluid & electrolyte imbalance (fluid loss & painful eating) .

Pemphigus Vulgaris

Treatment :-

1. Admission to hospital .
2. Very high dose of systemic steroid such as prednisolon 80-320 mg\day , the dose is dropped only when new blister stop appearing & taper gradually to maintenance level .
3. Immunosuppresive agents as azathioprine , cyclophosphamide , & methotraxate are often used as steroid sparing agent .
4. Plasmapheresis & intravenous immunoglobulin .
5. Biologic therapy in the form of ritoximab which is CD20 inhibitor.

Pemphigus

Drug induced pemphigus :- mostly due to pencillamine , captopril , pencilline , IL2 , rifampicin . There is less mucosal involvement , most of the cases resolve after discontinuation of treatment .

Para neoplastic pemphigus :- rare type , associated with thymoma or underlying carcinoma , chara. by unusually sever mucosal lesion .

Erythema Multiforme

acute often recurrent inflamm. Disease charac.by target shaped urticarial plaques which are arranged symmetrcally on the palms, back of the hand & feet.

Pathogenesis:- immune complex mediated in which IgM &C3

deposition around upper dermal blood vessel.

Etiology:- 50% idiopathic

1- viral infection:- herpes, hepatitisA,B,C, AIDs, ORF, polio, mycoplasma pneumonia.

2- bacterial infection:- TB, brucilla, typhoid.

3- fungal infection:- histoplasmosis.

4- Protozoal infection:- malaria, trichomoniasis.

5- Radiotherapy.

6- Drugs:- sulfonamide, pencilline, contraceptive, phenytoin phenothiazine.

7- polyarteritis nodosa, lupus erythematosus.

8- carcinoma, lymphoma, leukemia

9-Sarcoidos

Erythema Multiforme

Charac. Clinically by dusky red maculopapular lesions or vesicular or bullous lesions, charactristic lesion is target lesion.

Site back of hand and feet, palm, extensor of forearm & legs, but could be generalized.

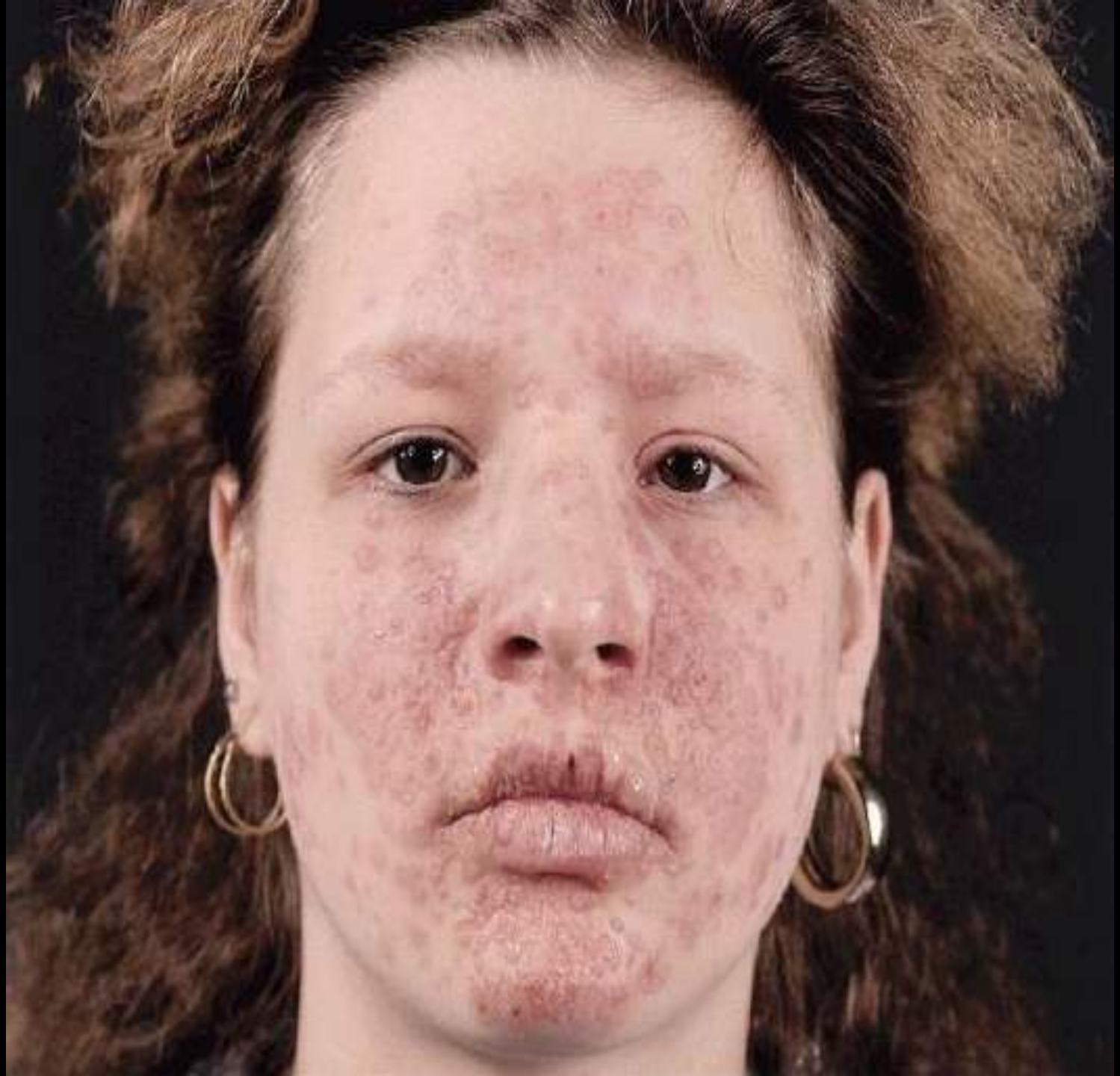
Mucus membrane could be affected.

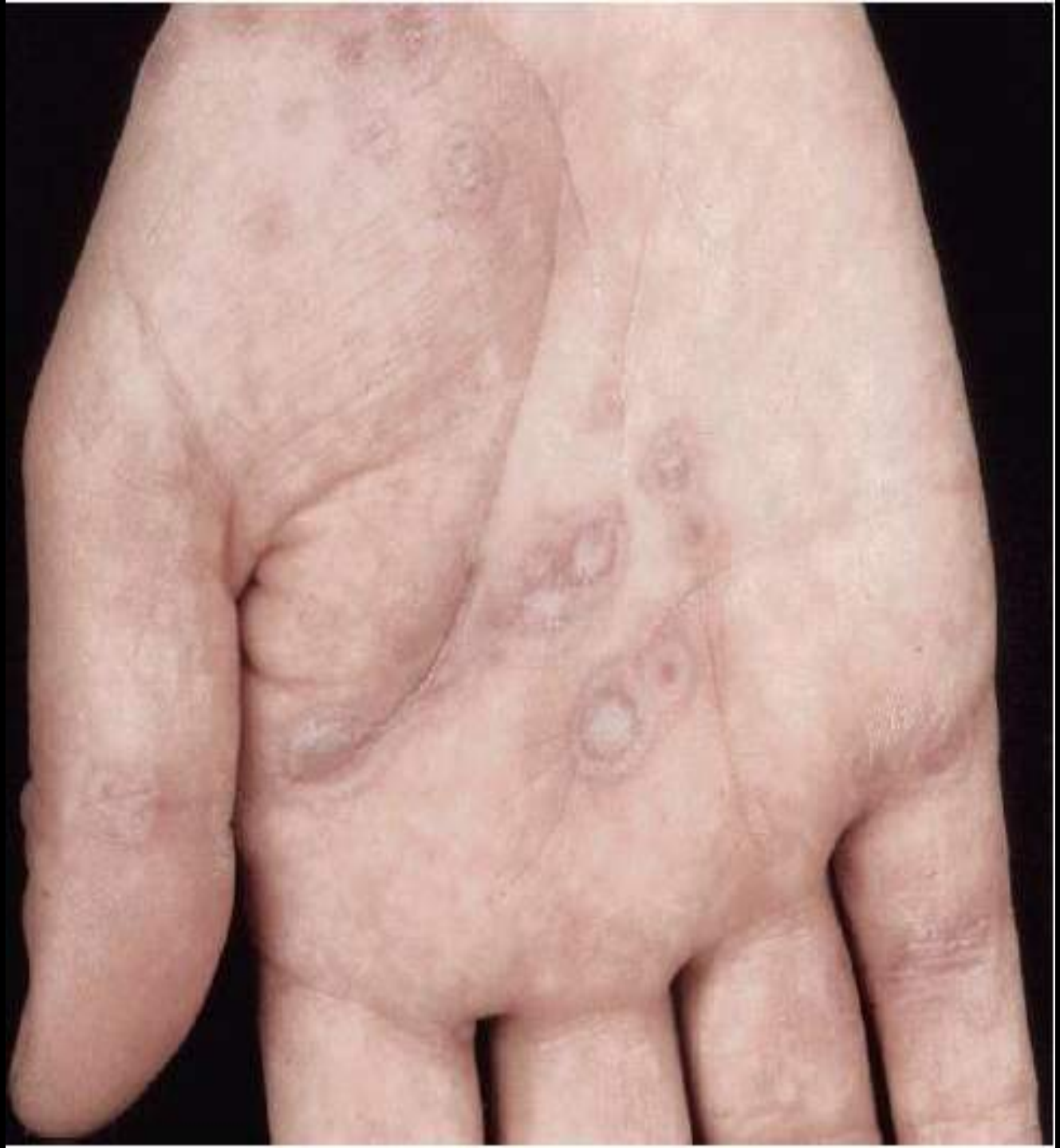
Not associated with prodromal symptoms.

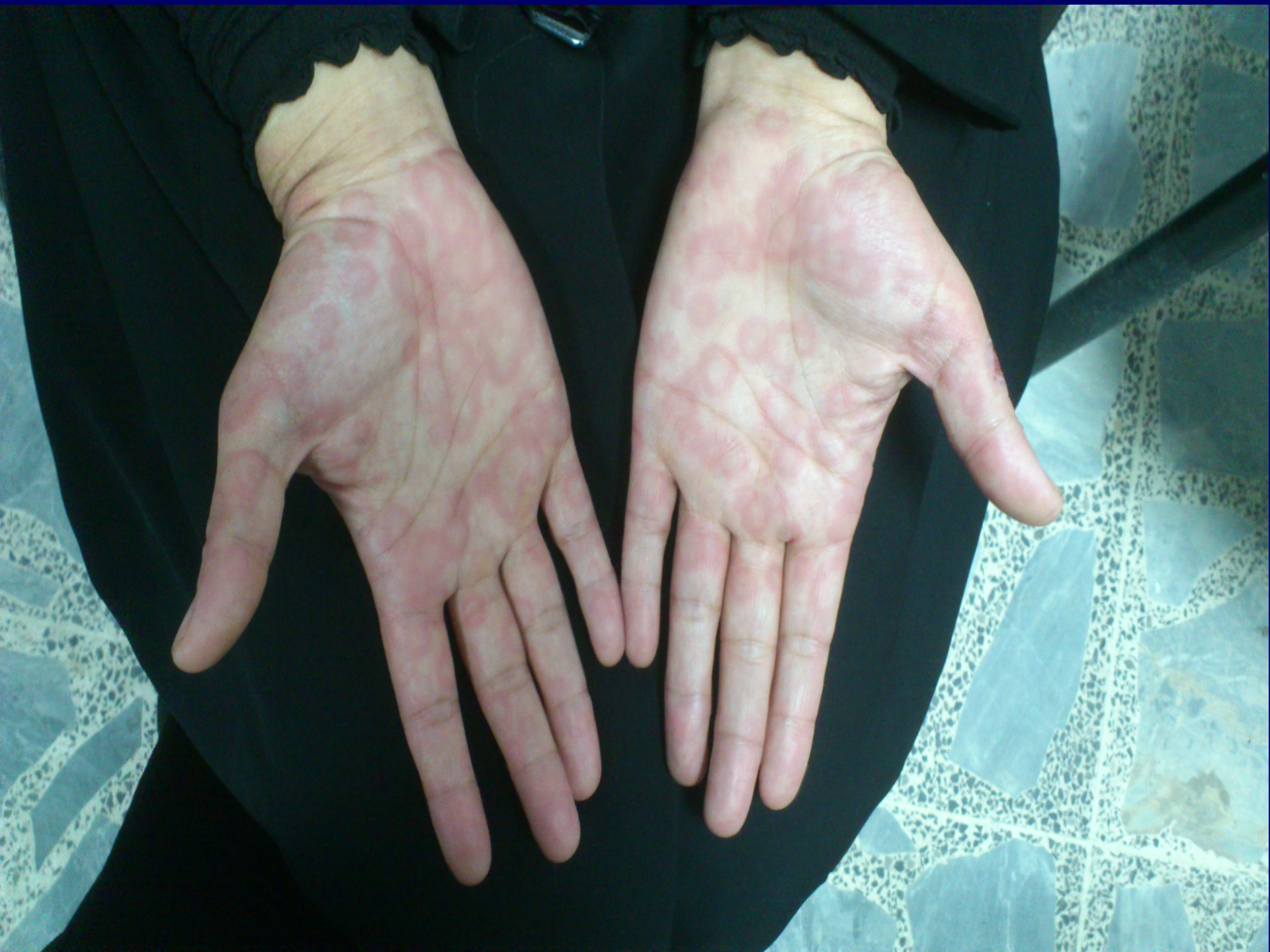
Treatment :-

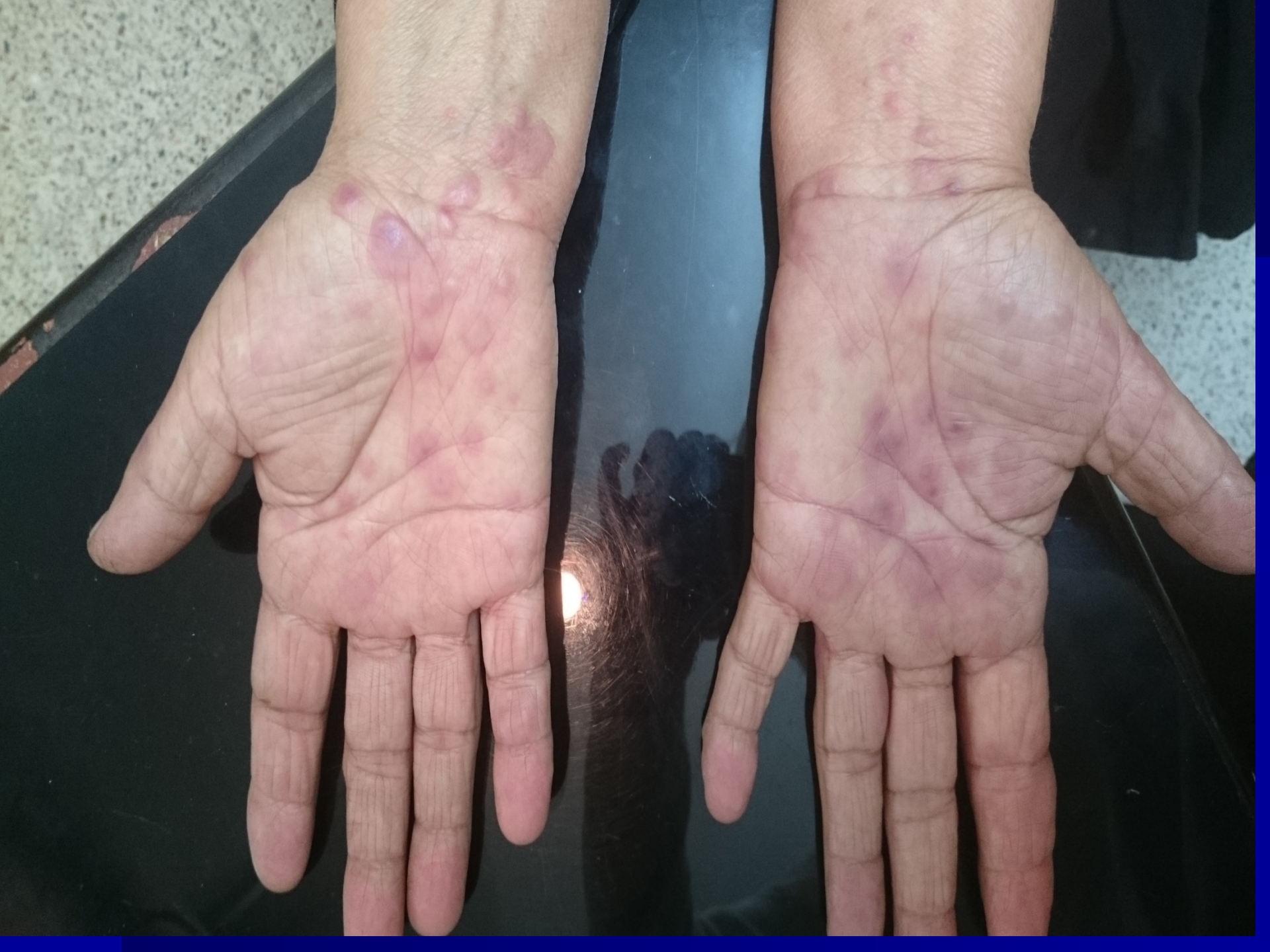
Self limited, sever systemic steroid 40-80mg 1-3 weeks.

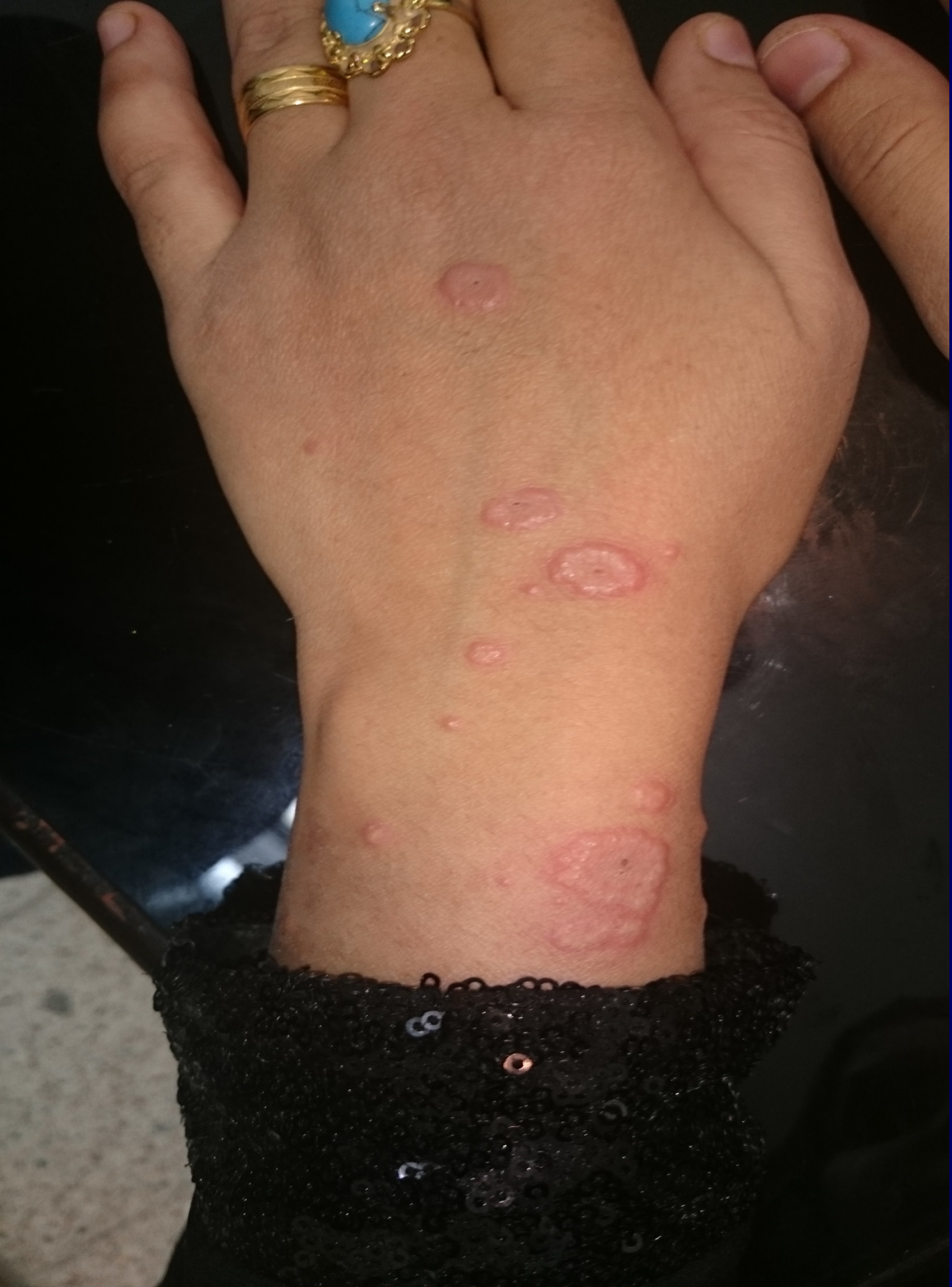
Oral acyclovir for recurrent EM





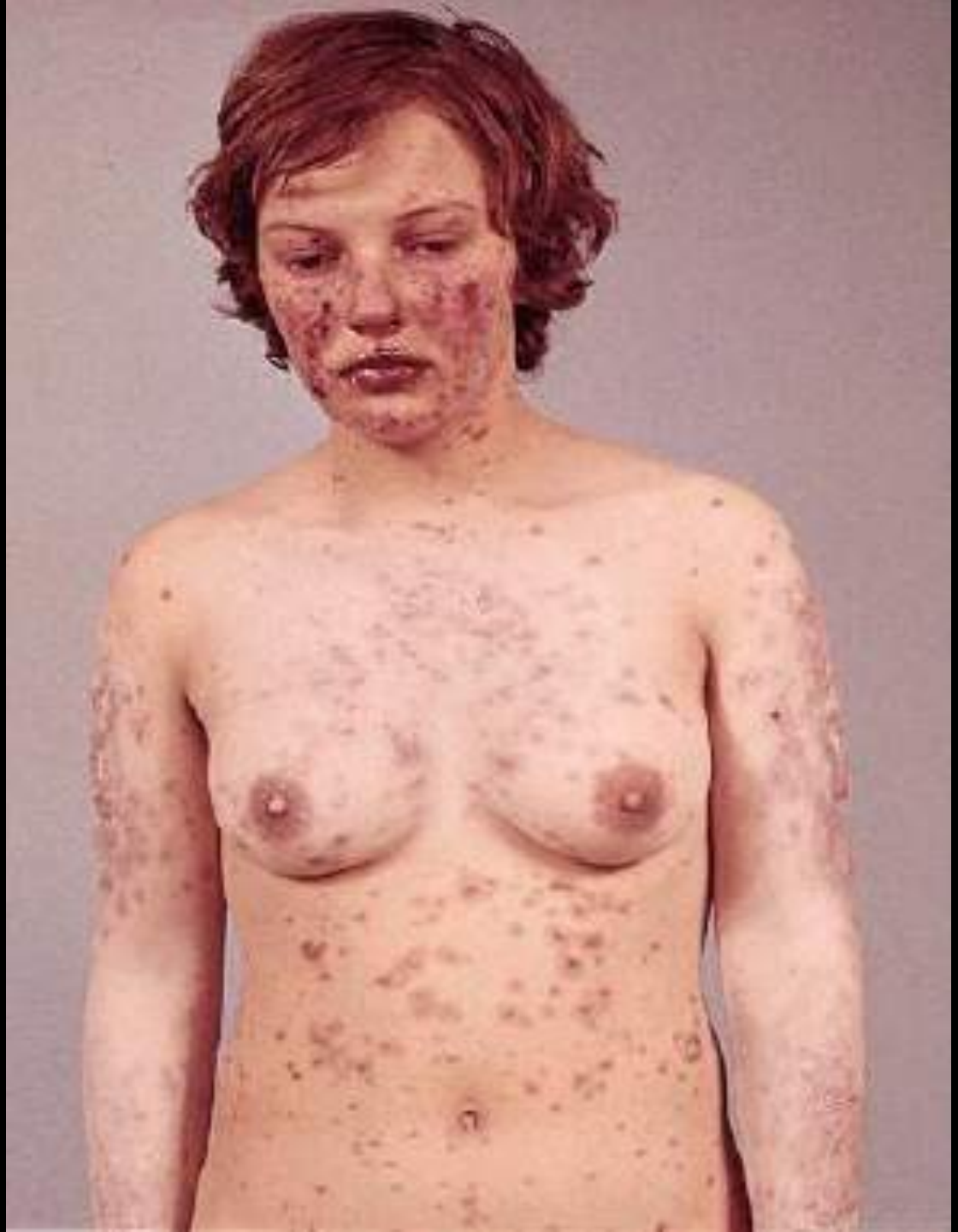


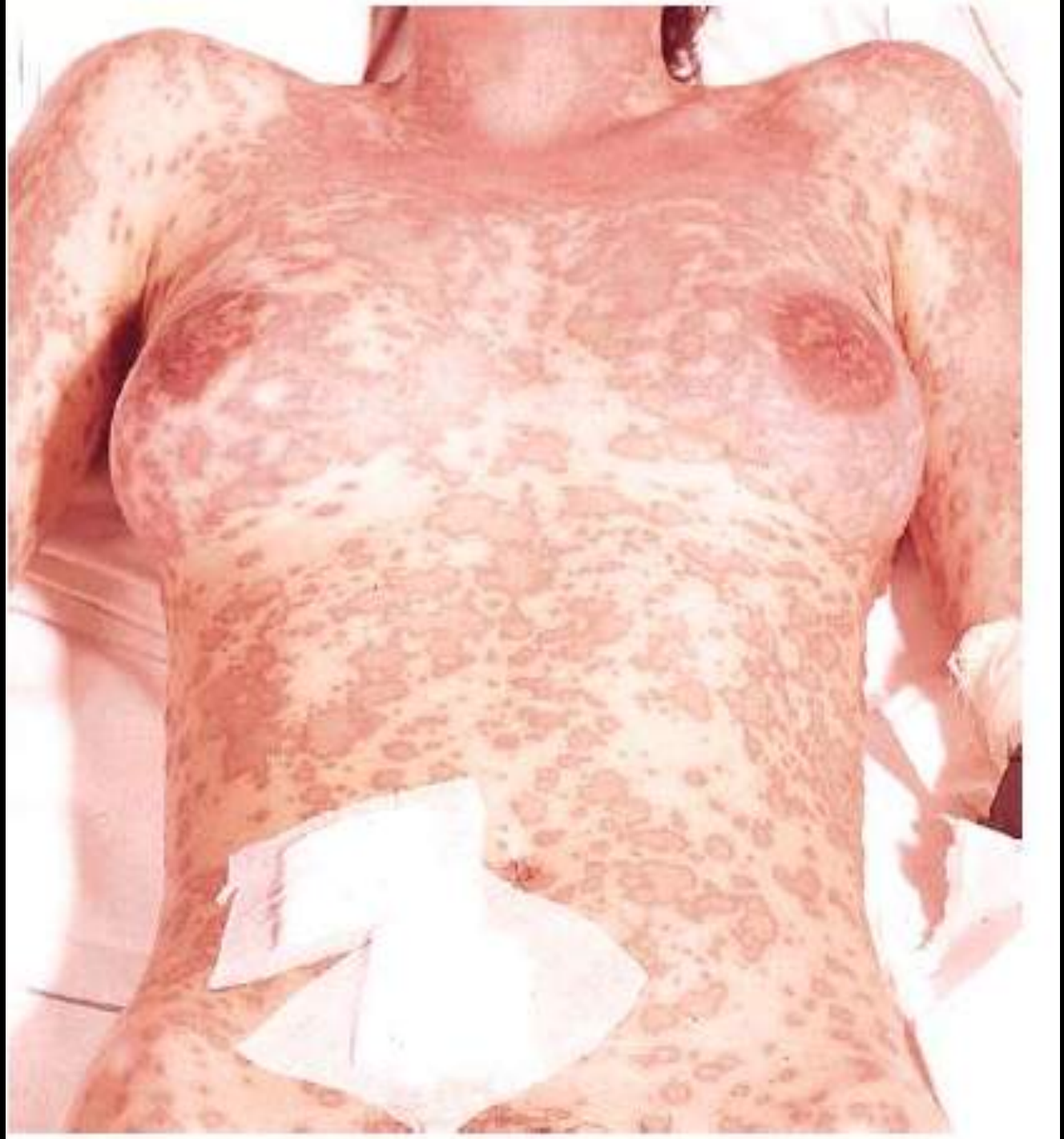


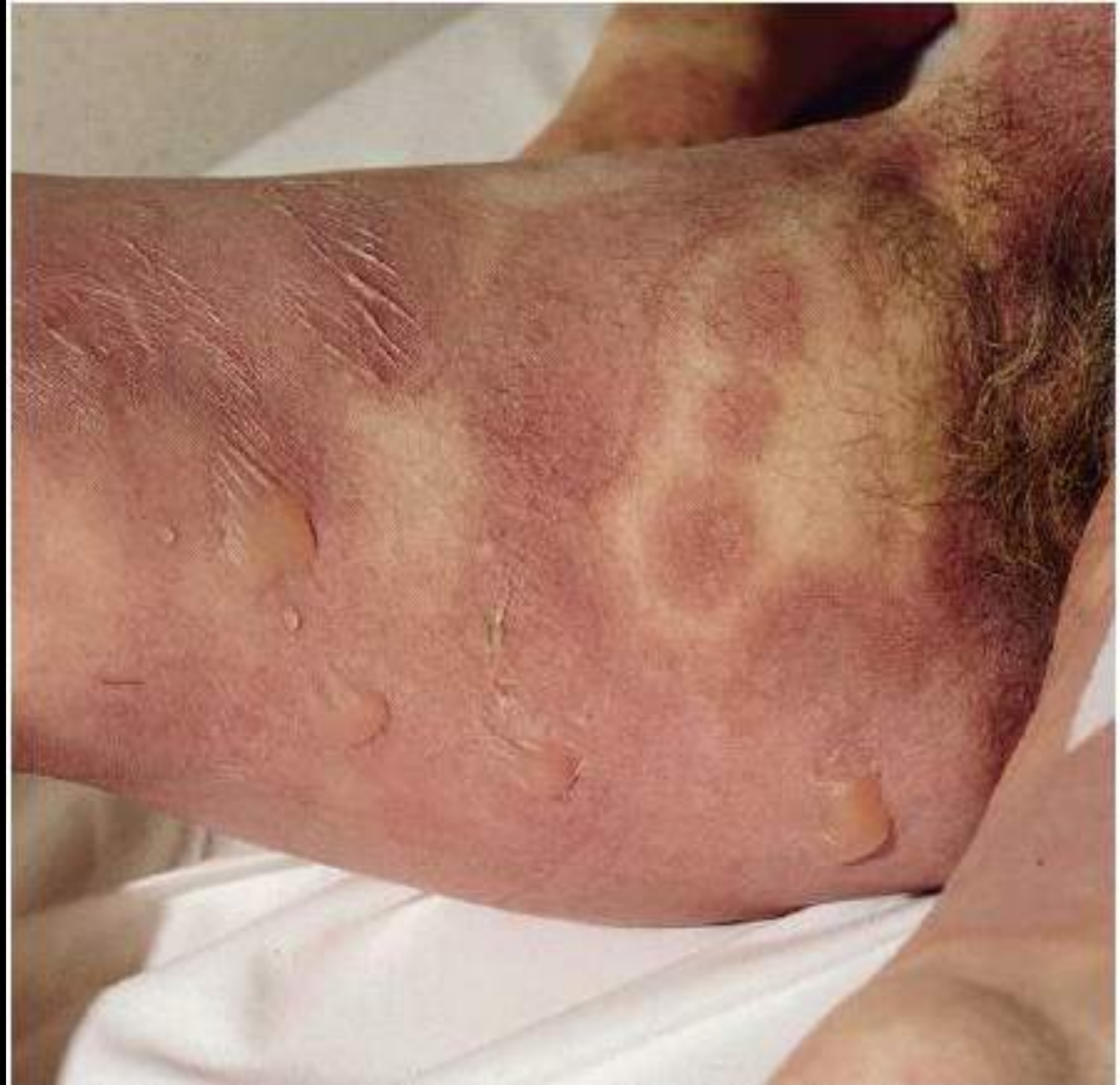


Stevens-johnson syndrome

- SJS is considered the most sever form of EM .
- characterized by widespread blisters and purpuric macules, is usually a sever drug-induced reaction with high morbidity and a poor prognosis with the mortality reaching 10%.
- Cutaneous eruption is preceded by fever and symptoms of an upper respiratory infection.
- Cutaneous lesions are flat atypical target or purpuric macules widespread.
- mucosal lesions: bullea appearing suddenly 1-14 days after prodromal symptoms, affecting conjunctivae, nose, mouth, anorectal junction, vulvovaginal, urethral.







Toxic epidermal necrolysis

A life threatening full thickness loss of epidermis with a high mortality reaching 35-40%. Usually is a sever drug reaction.

Chracterized by sever prodromal symptoms , the skin hot and red and painful, and slide laterally with slight pressure(nikolsky's sign). With sever mucosal involvement.