AUTOIMMUNEVESICULO-BULLOUS DEMATOSIS

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AUTOIMMUNE VESICULO - BULLOUS DERMATOSIS

A group of diseases results from an autoimmune response to intercellular molecules in desmosomes or the basement membrane zone.

Therefore they are classified generally into intraepidermal and subepidermal autoimmune vesiculobullous dermatosis

Immunobullous Dermatosis (IBD)

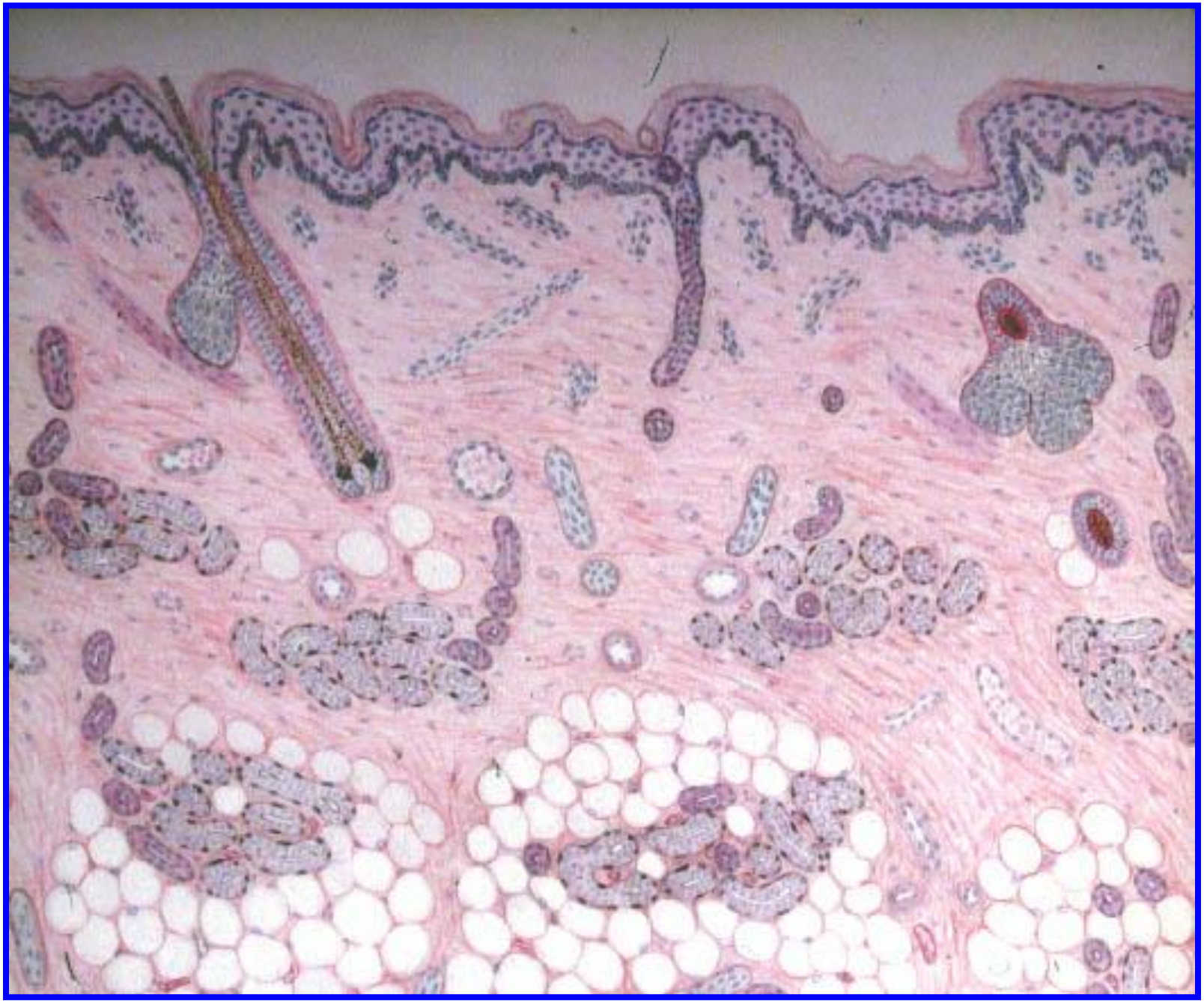
Intraepidermal IBD

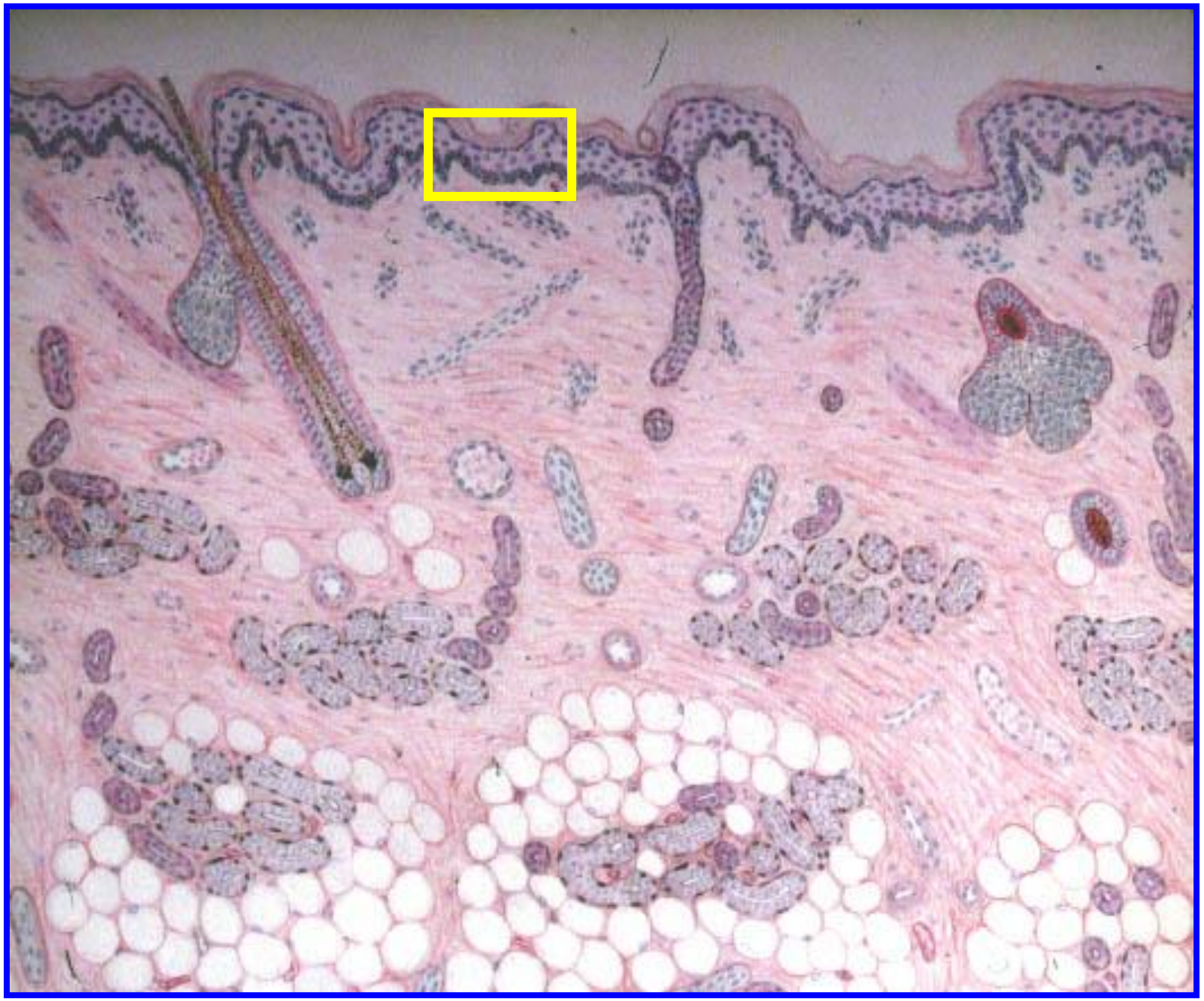
Pemphigus

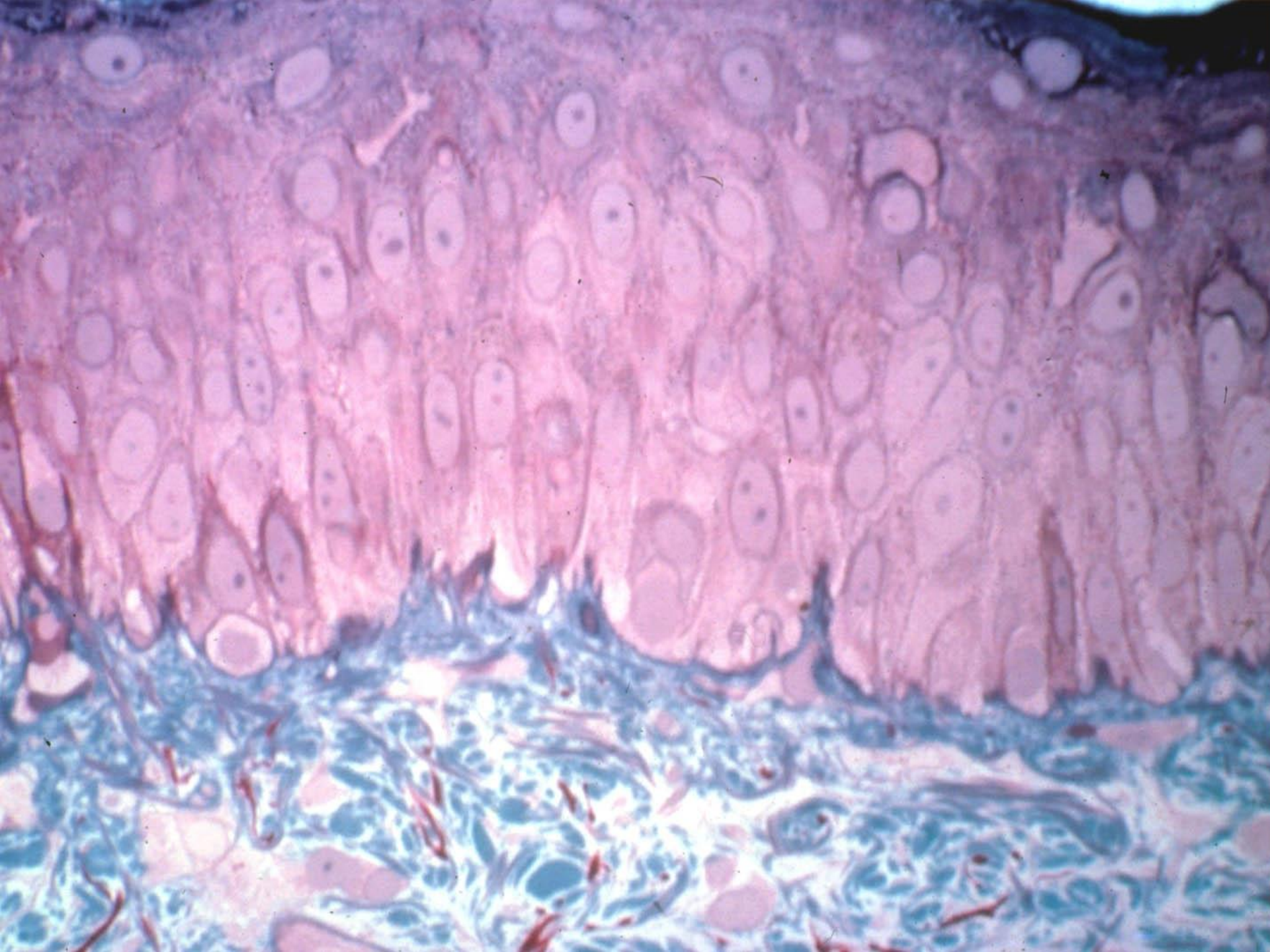
Subepidermal IBD

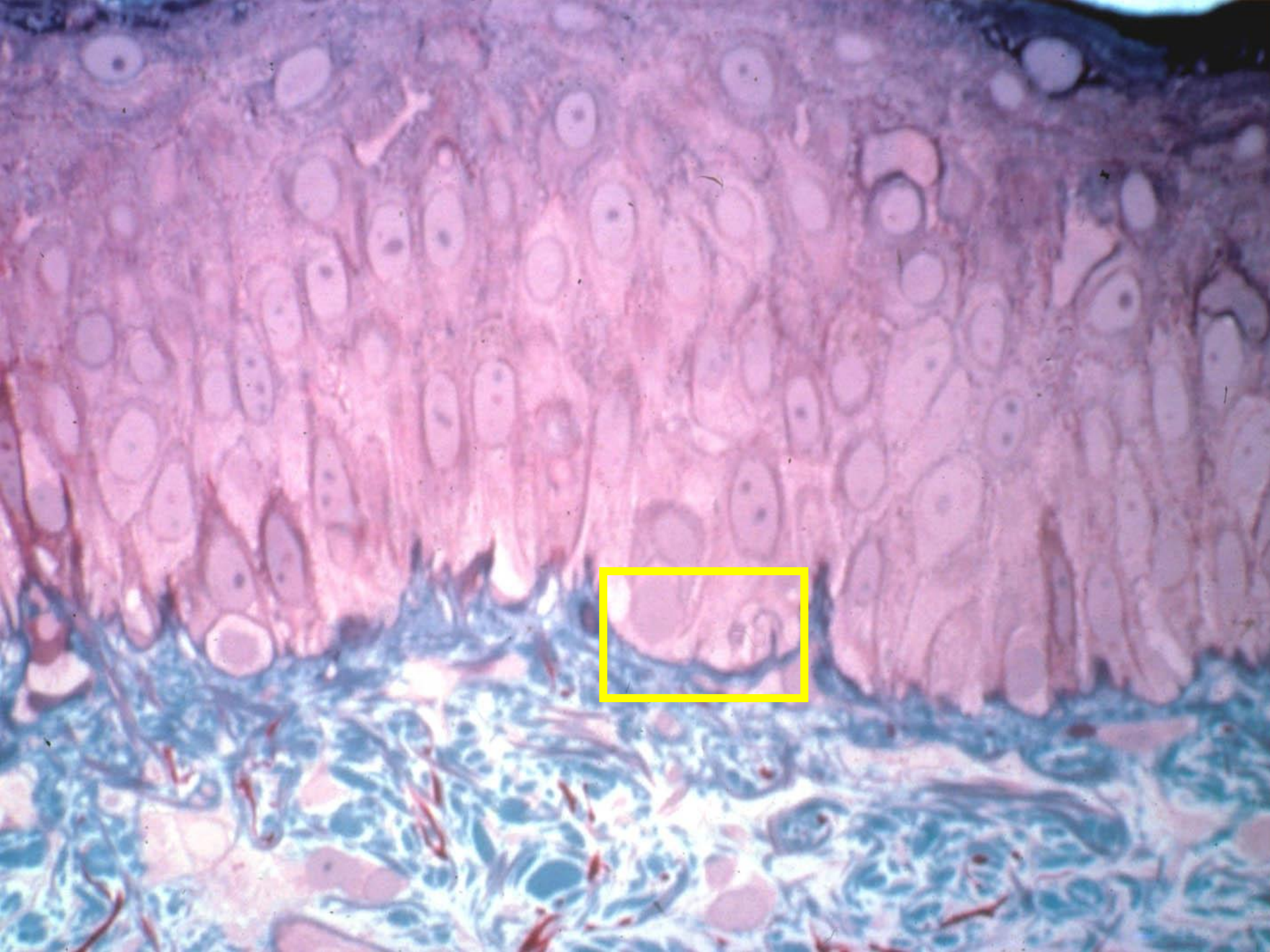
Bullous pemphigoid

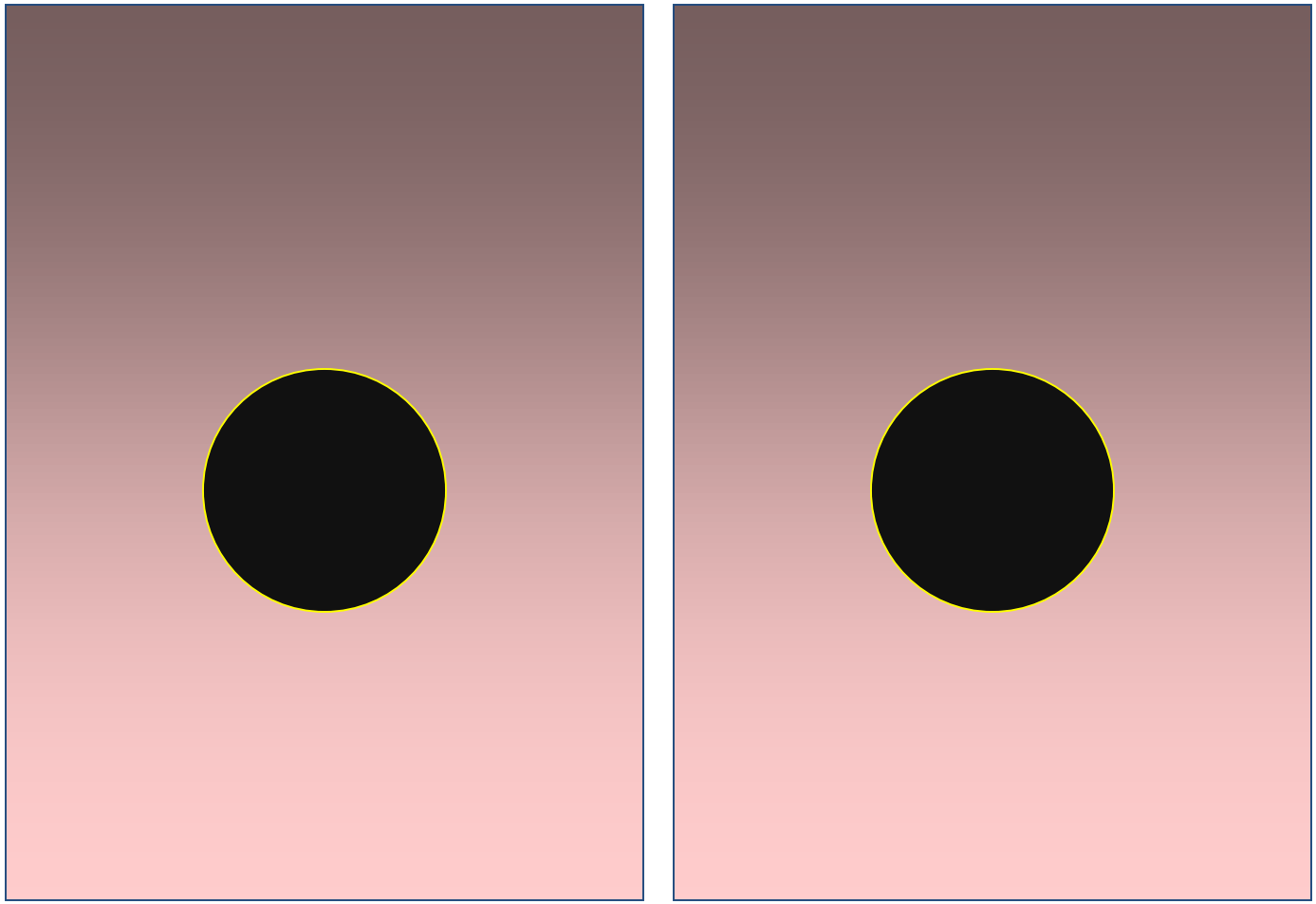
Dermatits Herpitiformis



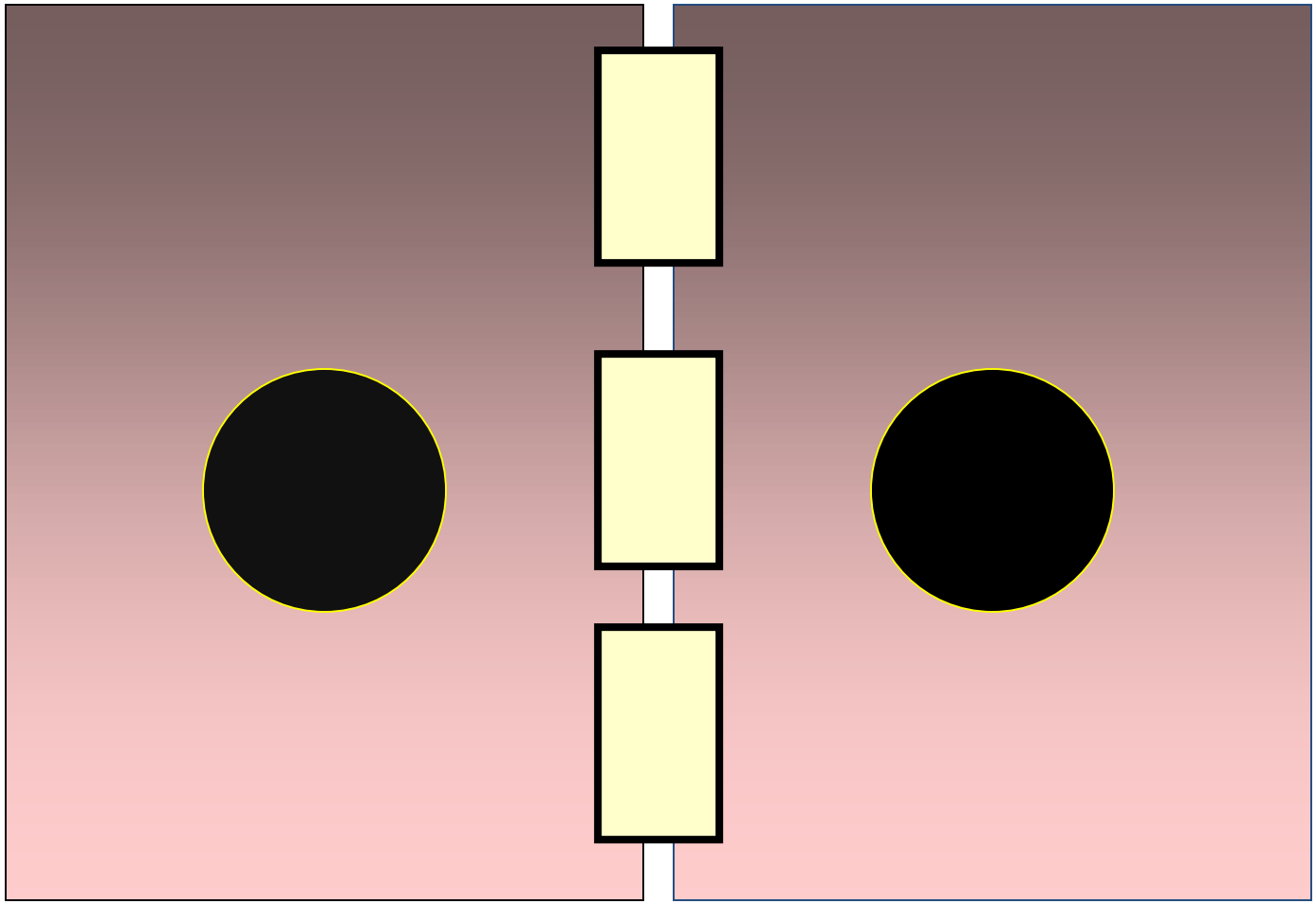


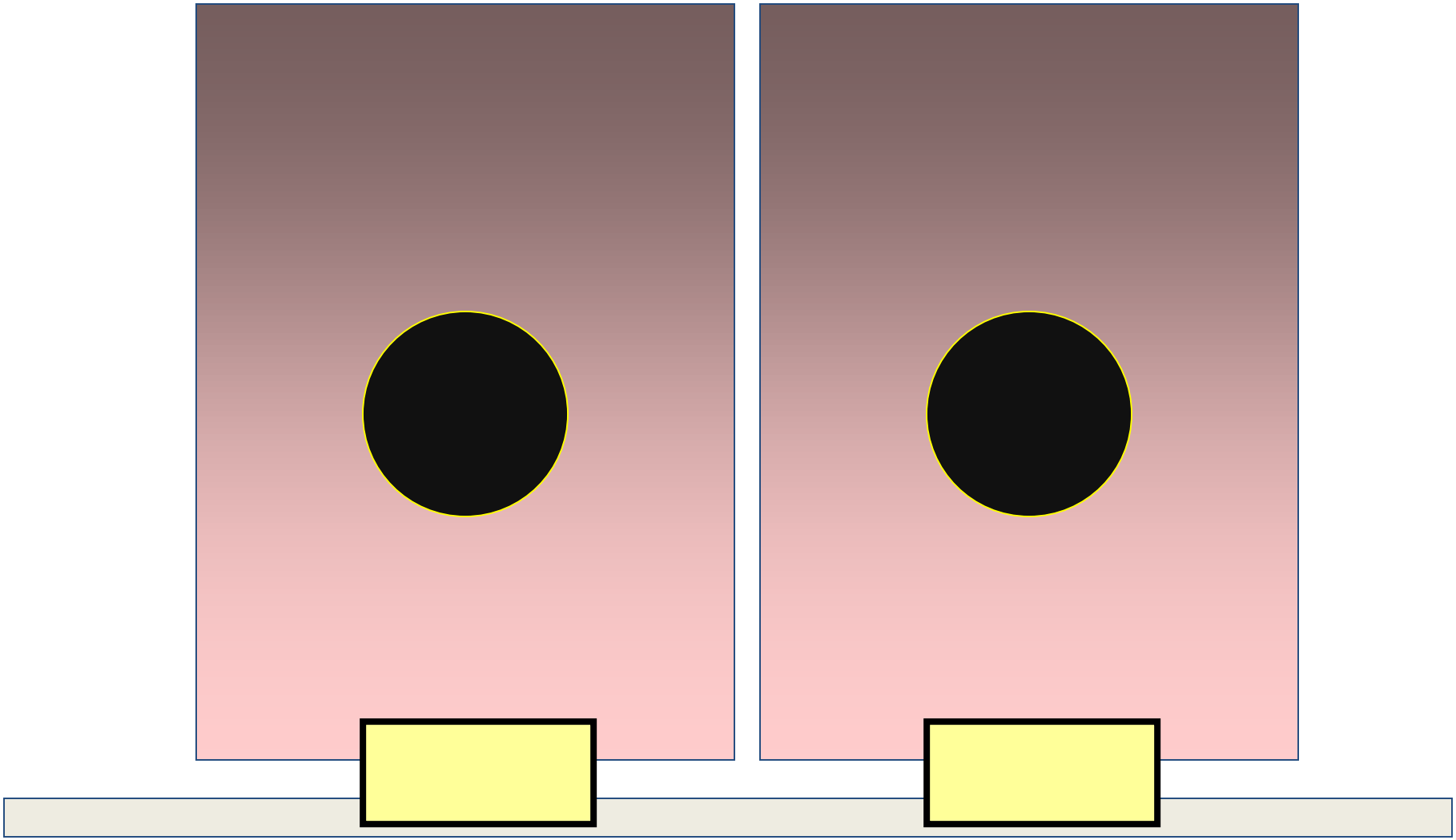






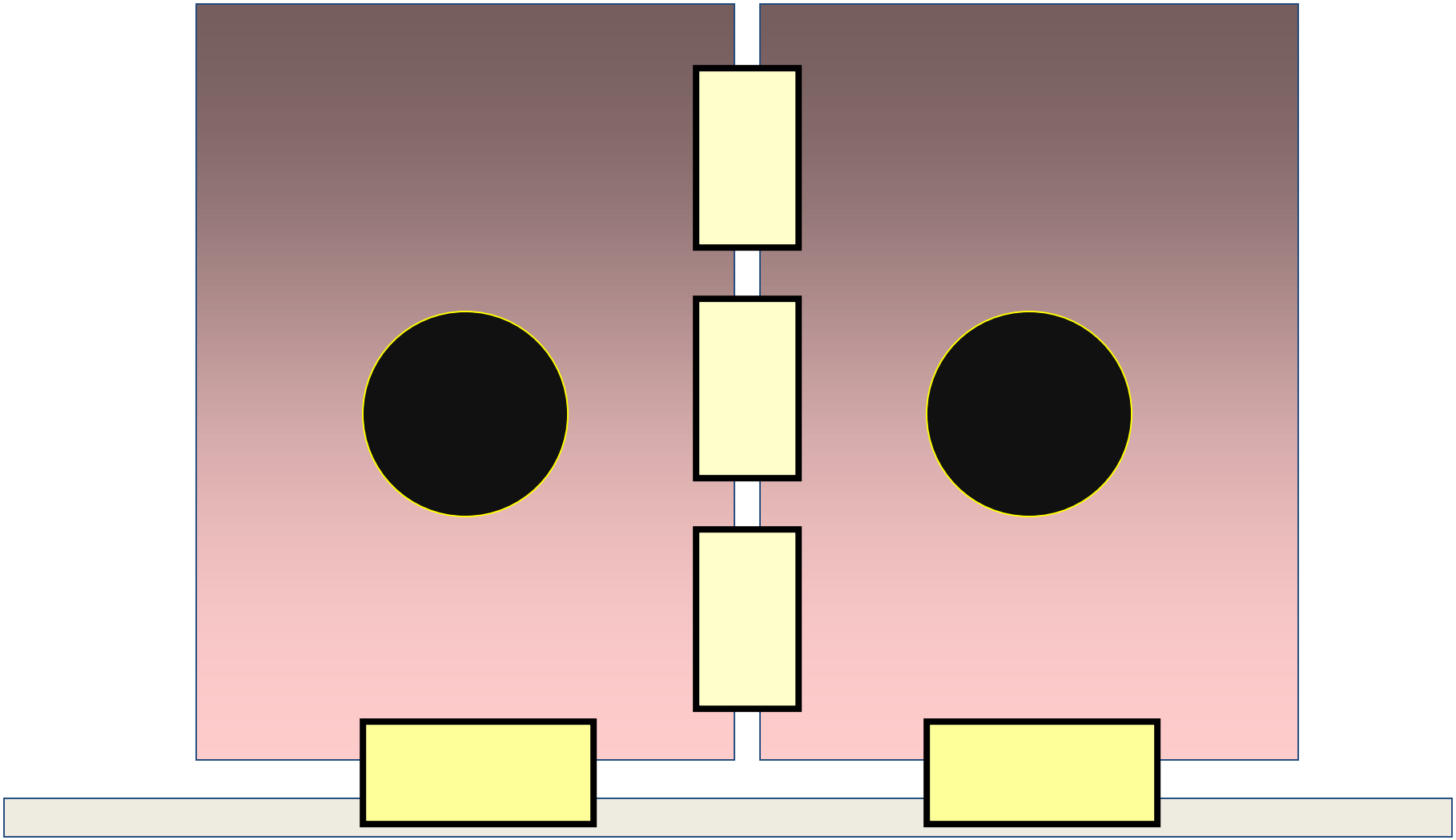
Desmosome





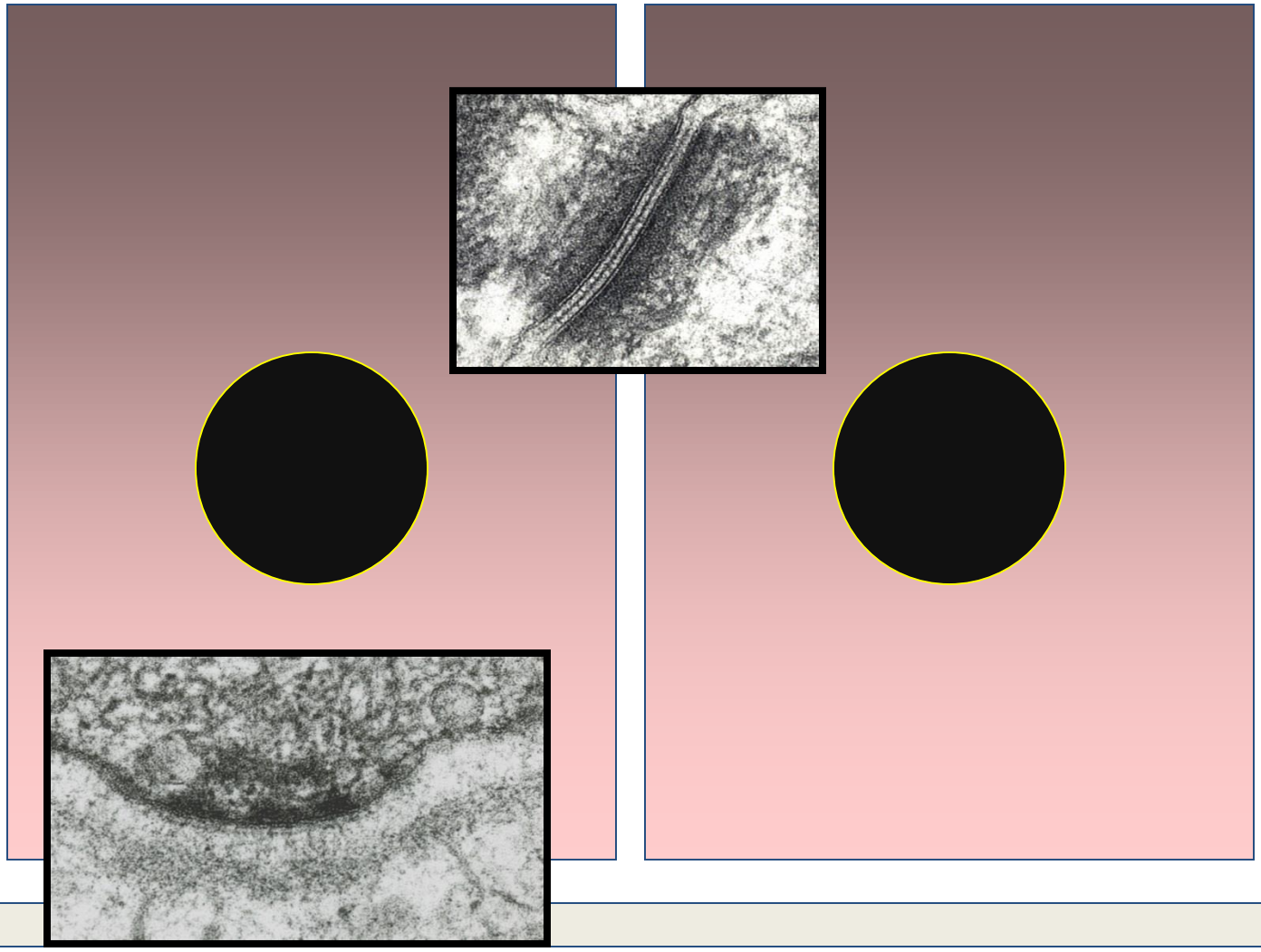
Hemidesmosome

Desmosome



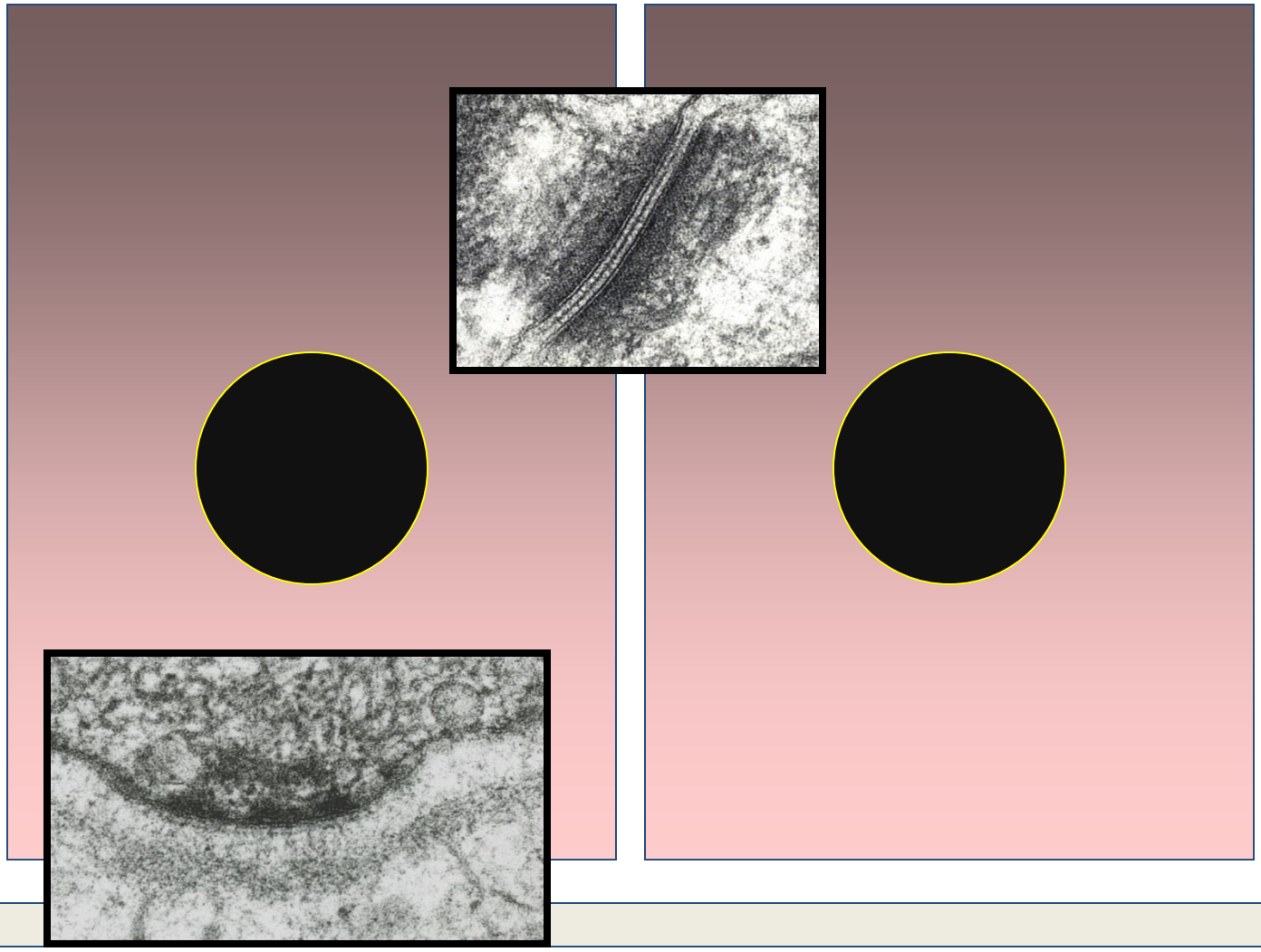
Hemidesmosome

Desmosome



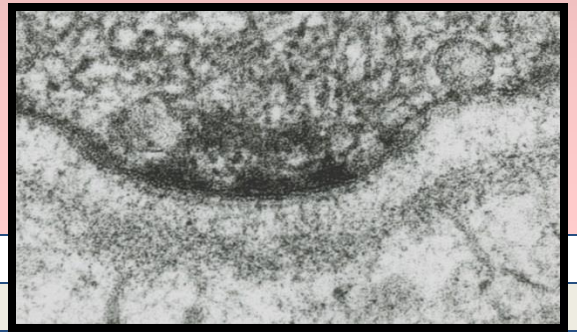
Hemidesmosome

IgG → Desmosome → Pemphigus



Hemidesmosome

Desmosome



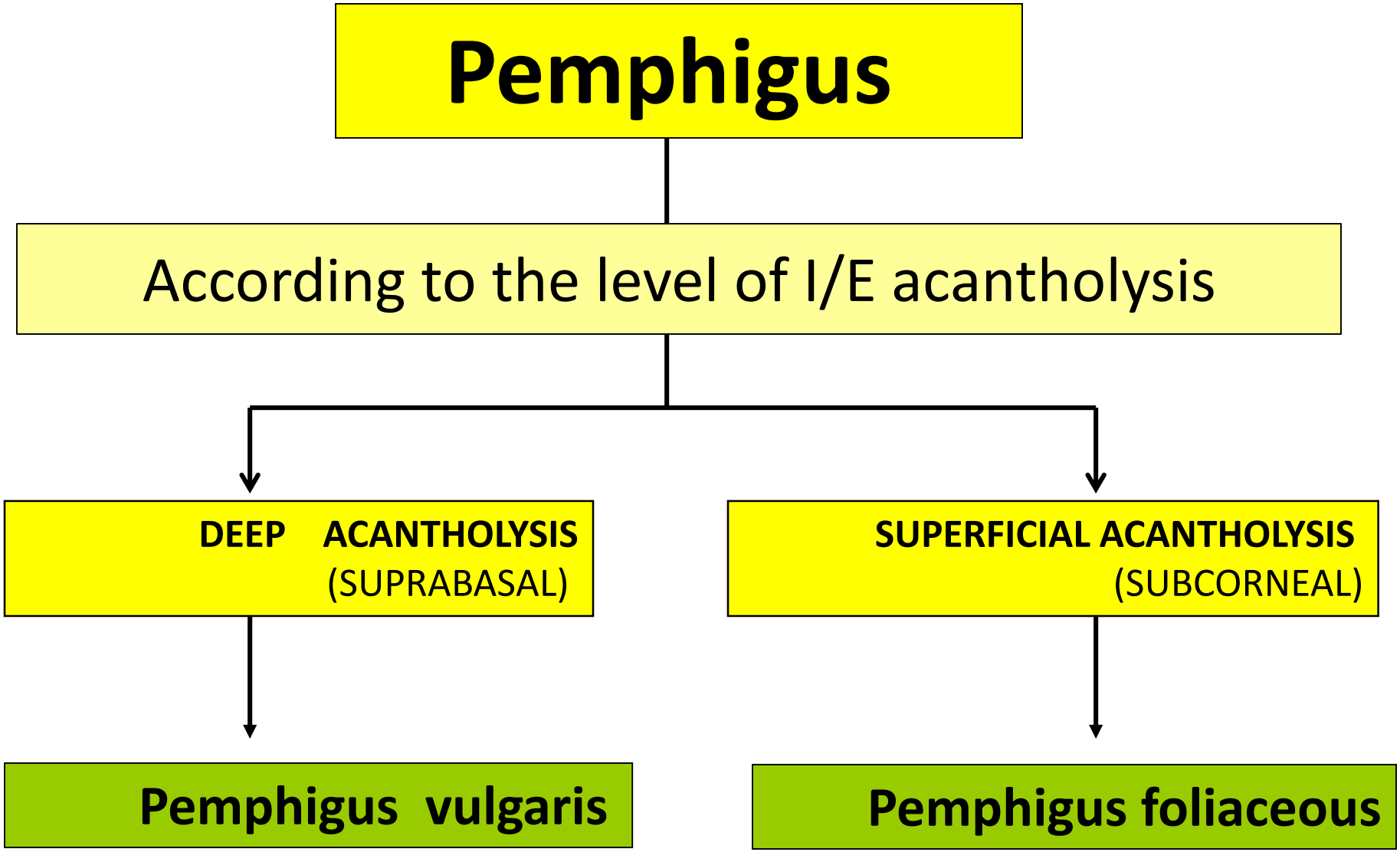
IgG

Hemidesmosome

Pemphigoid

Pemphigus

Pemphigus " *Greek, pemphigus = blister* " is a sever or even life-threatening intraepidermal blistering diseases characterized by acantholysis of the epidermal cells.


Pemphigus
According to the level of I/E acantholysis
DEEP ACANTHOLYSIS (SUPRABASAL)
SUPERFICIAL ACANTHOLYSIS (SUBCORNEAL)
Pemphigus vulgaris
Pemphigus foliaceous

Pemphigus vulgaris.

P.V is a chronic , potentially fatal , autoimmune blistering disease of the skin and mucous membranes characterized by acantholysis in the suprabasal layer of the epidermis.

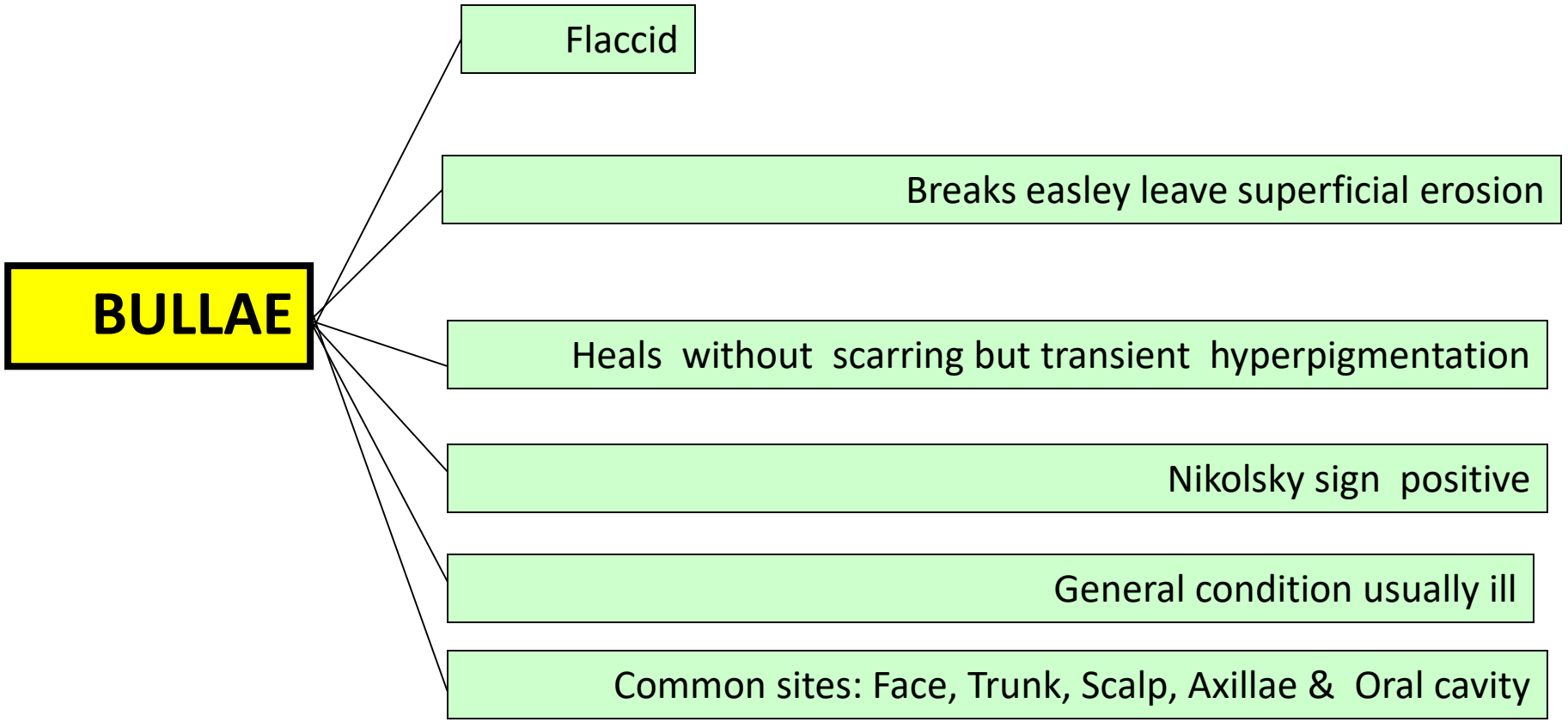
Epidemiology

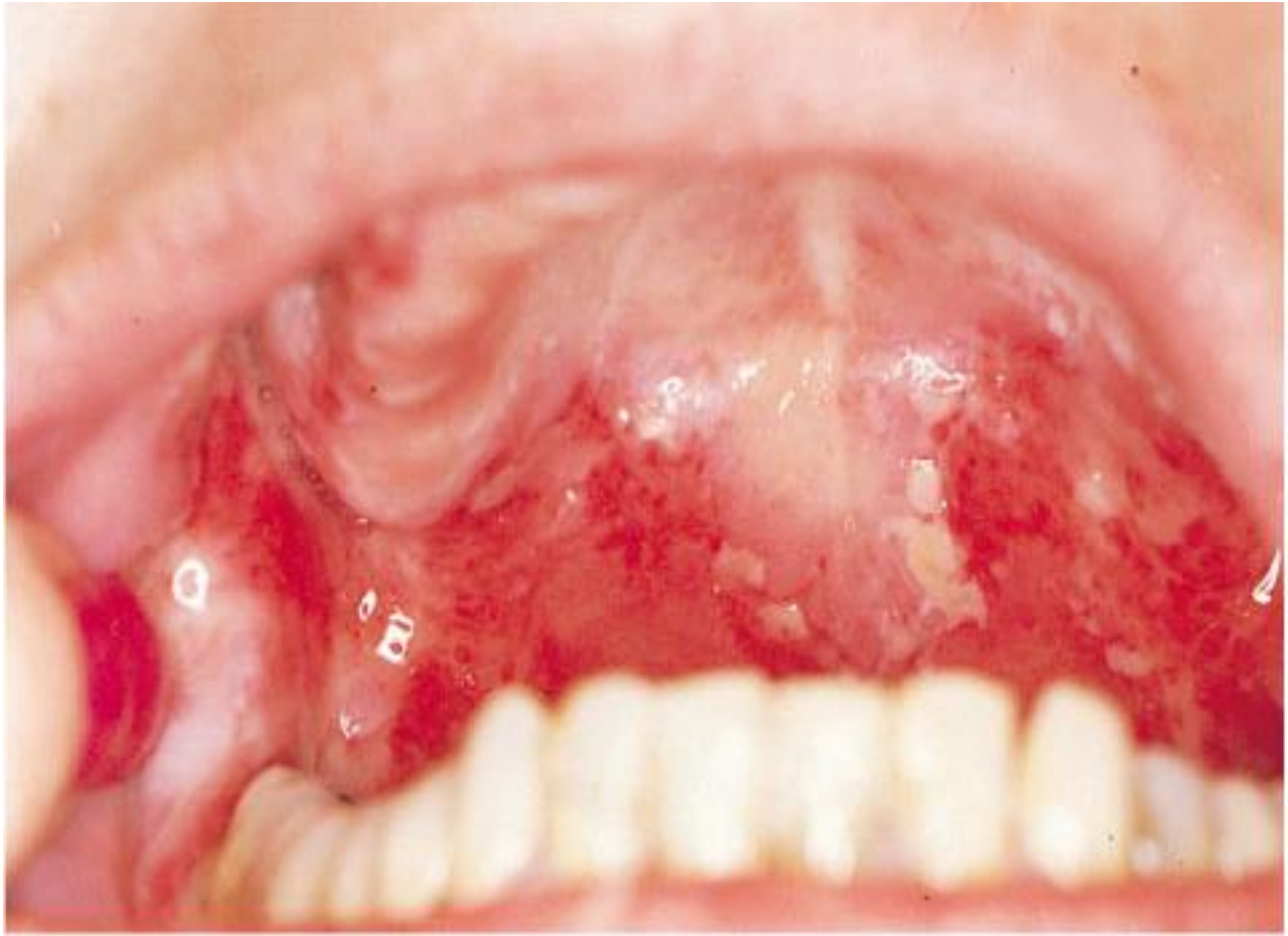
- P.V accounts 70% of all types of pemphigus.
- It affects the middle age group (50 – 60 years).
- Almost equal in both sexes.

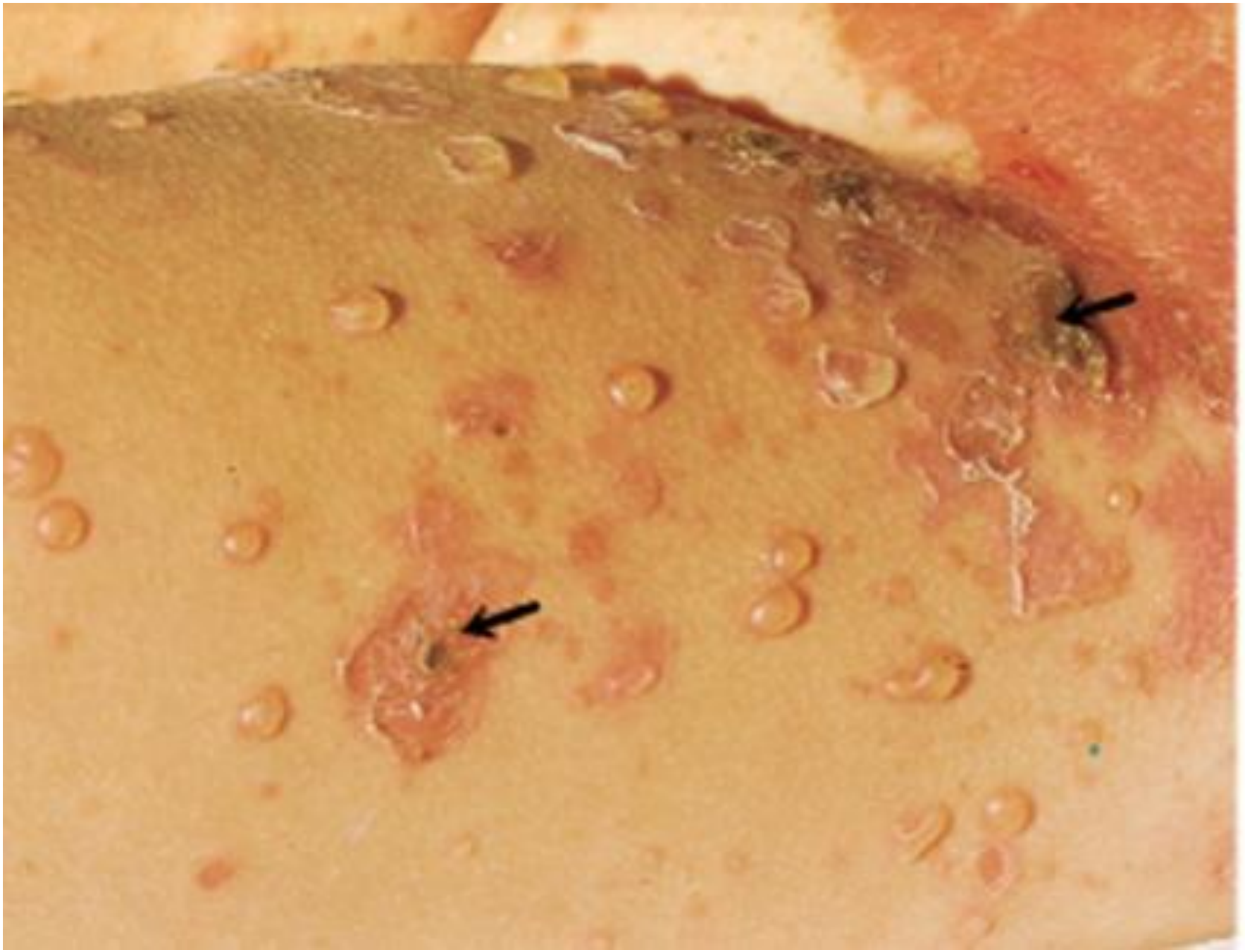
Clinical features

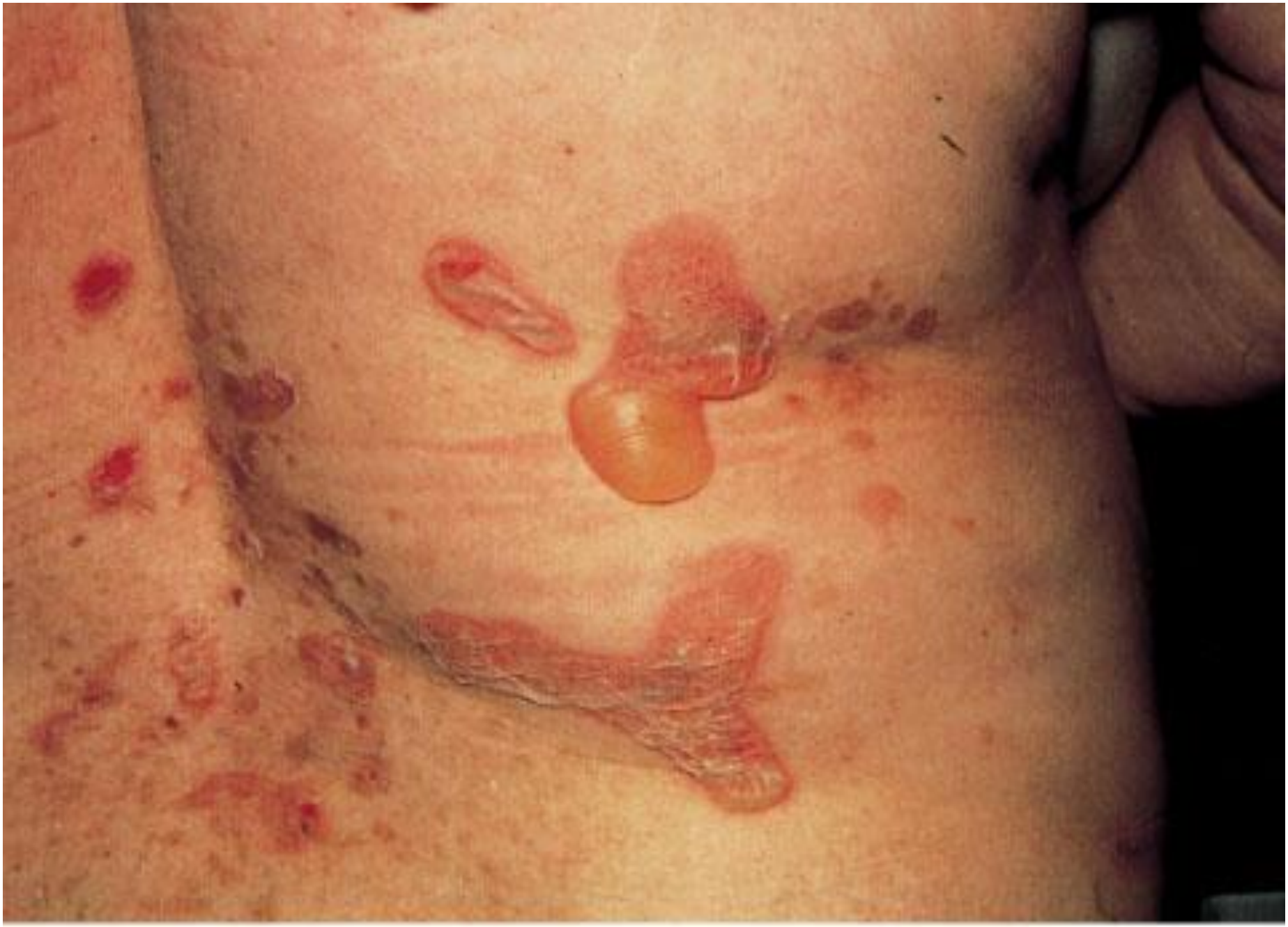
In more than 50 % of patients the disease starts with lesions in the oral cavity, but almost all patients have oral lesions at some time during the course of the disease.

The primary skin lesion in pemphigus is blisters (Bullae).





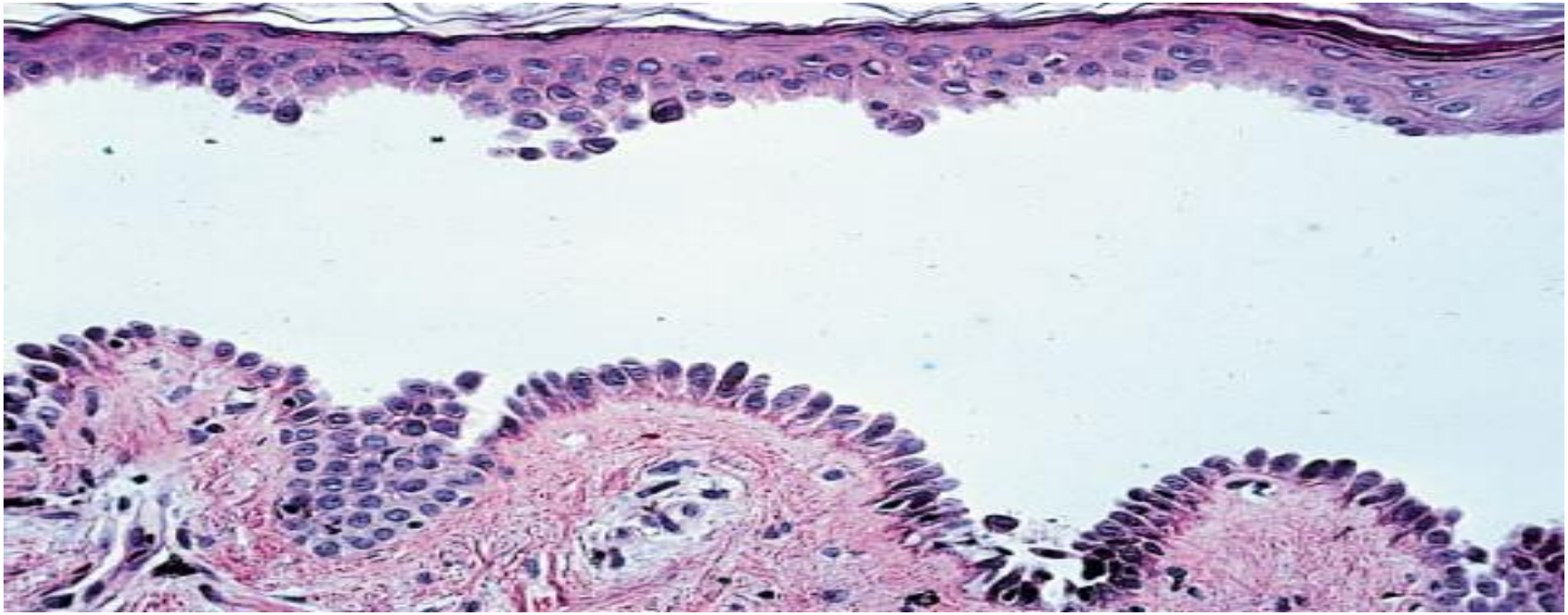




Nikolsky sign : **Firm sliding pressure on normally appearing skin leads to avulsion of the outermost layer of the skin.**

Histopathology

The major histopathological feature of pemphigue is acantholysis (seperation of keratinocytes), that leads to intraepidermal bullea, which in pemphigus vulgaris in suprabasal layer of the epidermis

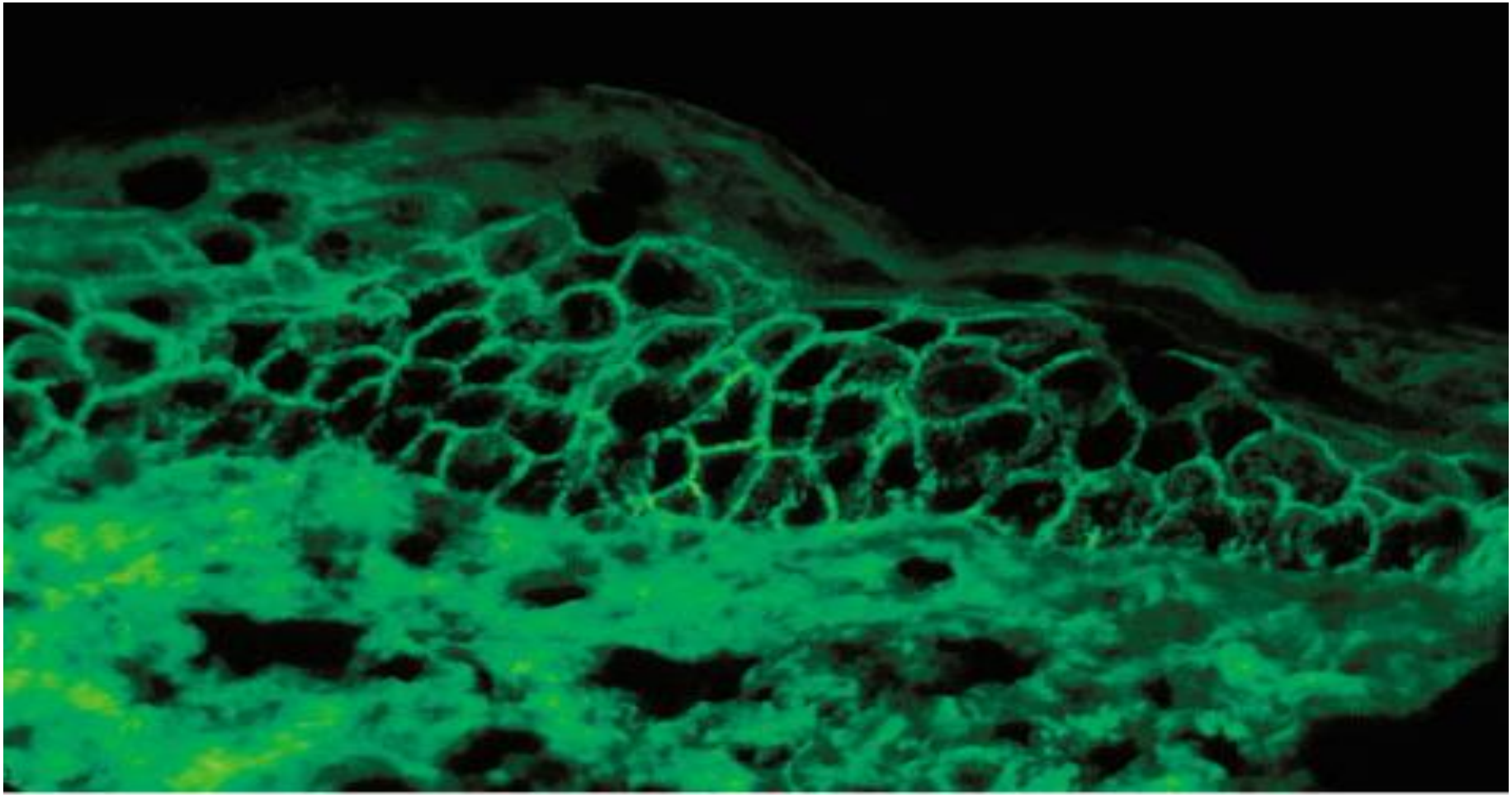


The basal cells remains attached to the basement membrane but separated from one another and stand like a row of tombstones.

IMMUNOFLUORESCENCE

DIF of pemphigus skin shows intercellular deposition of IgG in almost 100% of patients.

IIF : circulating Ig G against the skin desmosomes.



TREATMENT

Systemic Corticosteroids

- **Most patients are treated by systemic corticosteroids.**
- **A dose of 60 -120 mg/day for 6 -8 weeks or until cessation of new blisters.**

Pemphigus Foliaceous

PF, is a mild form of pemphigus in which blistering occur high in the epidermis, just beneath the stratum croneum (subcorneal acantholysis).

Epidemiology

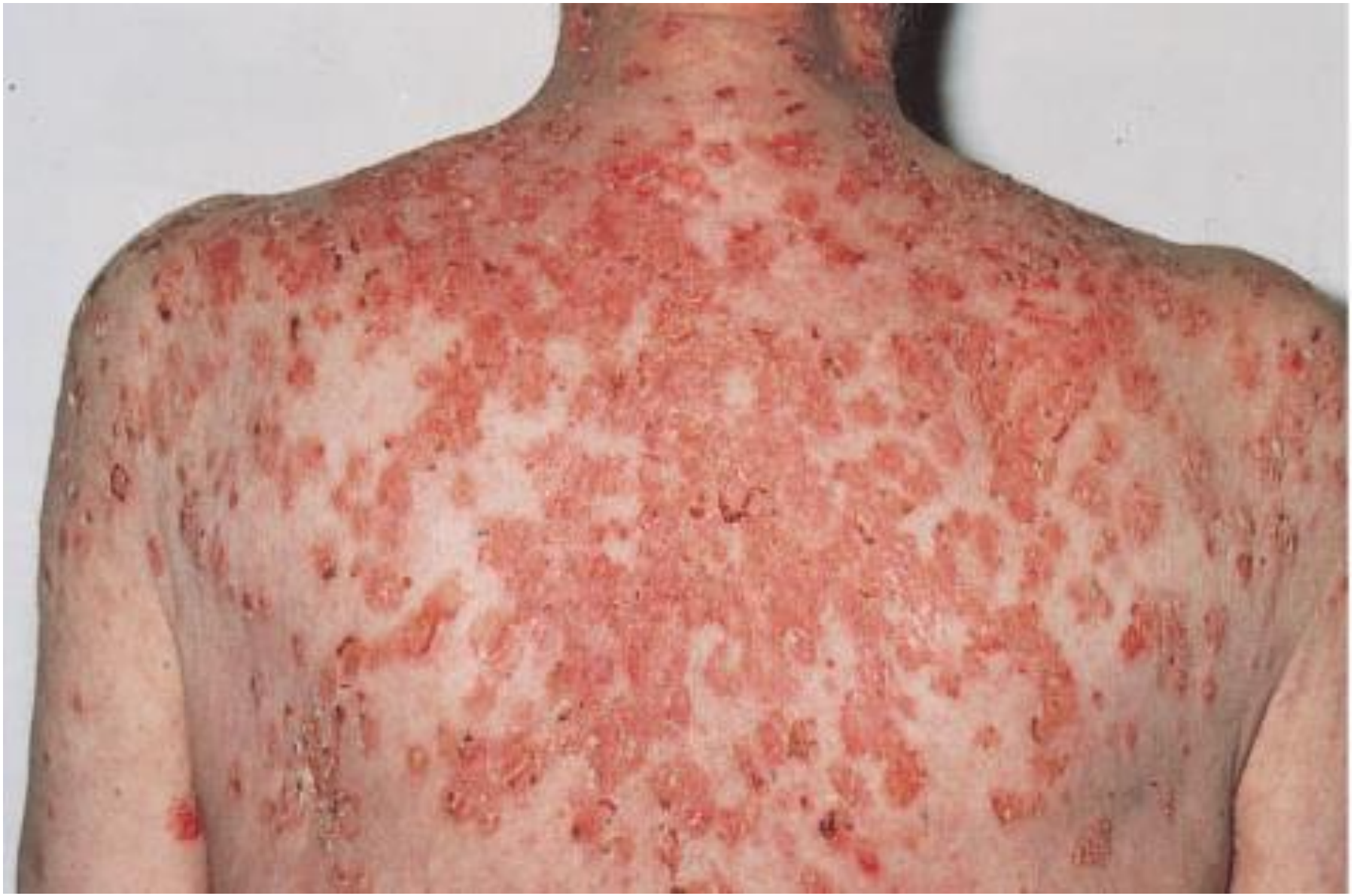
- PF less common than PV.
- It accounts only 10% - 20% of all cases of pemphigus.
- The mean age of onset of PF is about 50 – 60 years.
- Both sexes were equally affected.

Clinical Features

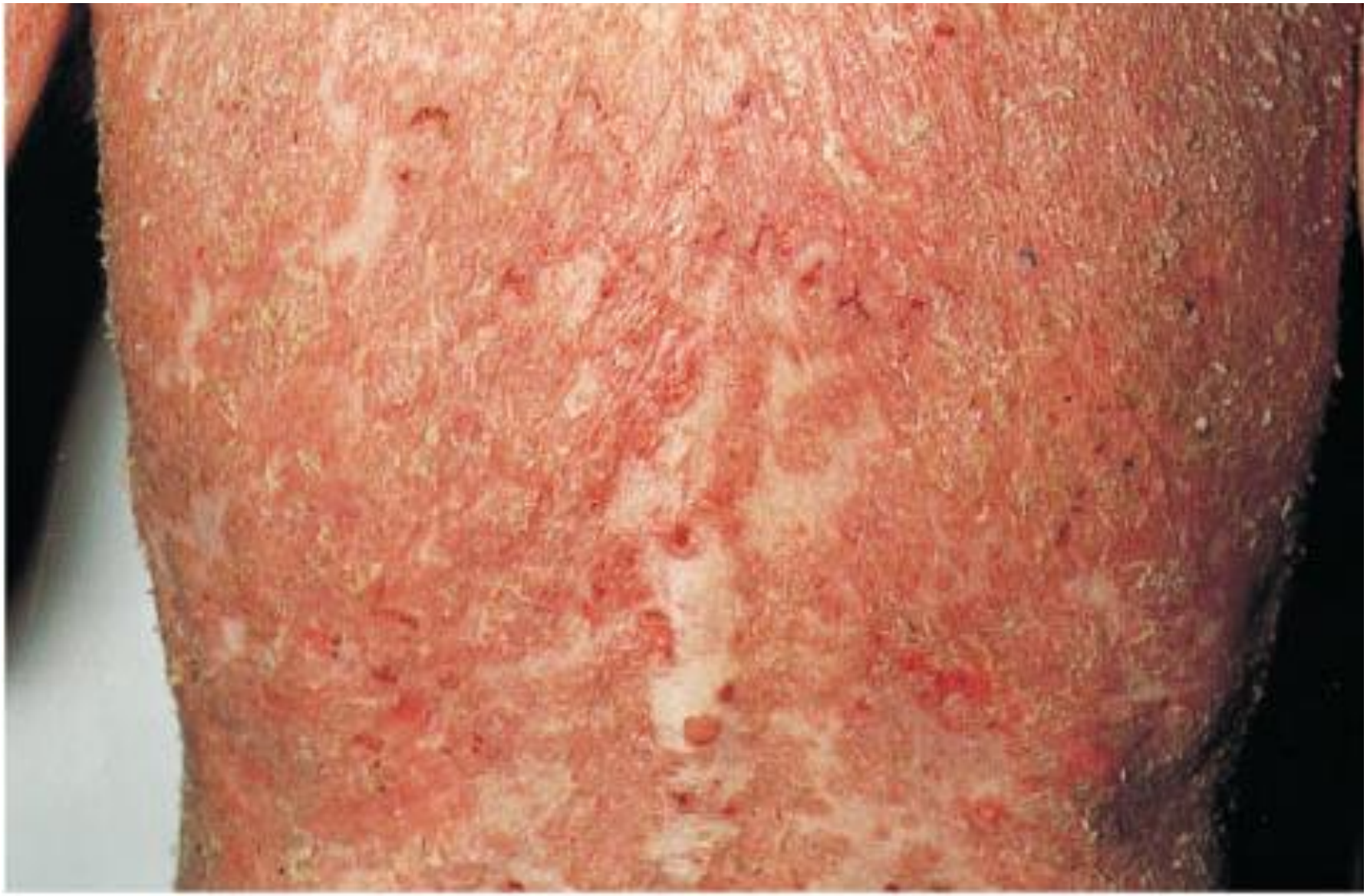
- PF, is generally less sever than pemphigus vulgaris .
- The mucous membranes usually not involved.
- The 1ry skin lesion is flaccid bullae which difficult to fined.
- Because they ruptured easley and transformed into erosion.
- Affected sites includes face, scalp, and upper trunk.
- Occasionally the erosion becomes generalize erythrodermic.

Treatment

In mild localised cases potent topical coricosteroids may be sufficient.
In sever extensive cases predinisolone 20 – 40 mg/ day.



Flaccid bullae that ruptured easley & transformed into crusted erosions & scales



Generalized crusted erosions and scales that becomes erythrodermic

Immunobullous Dermatosis (IBD)

Intraepidermal IBD

Pemphigus

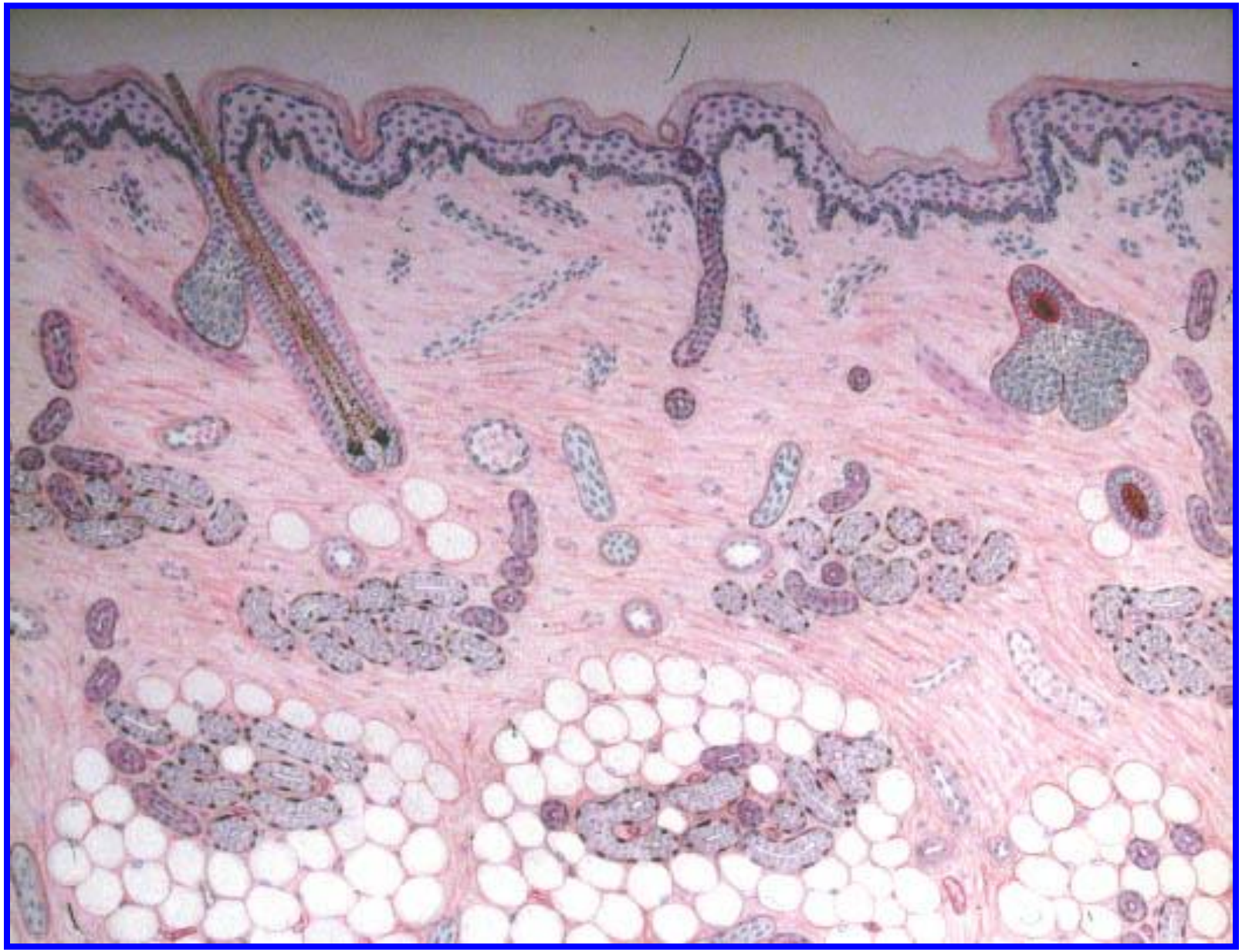
Subepidermal IBD

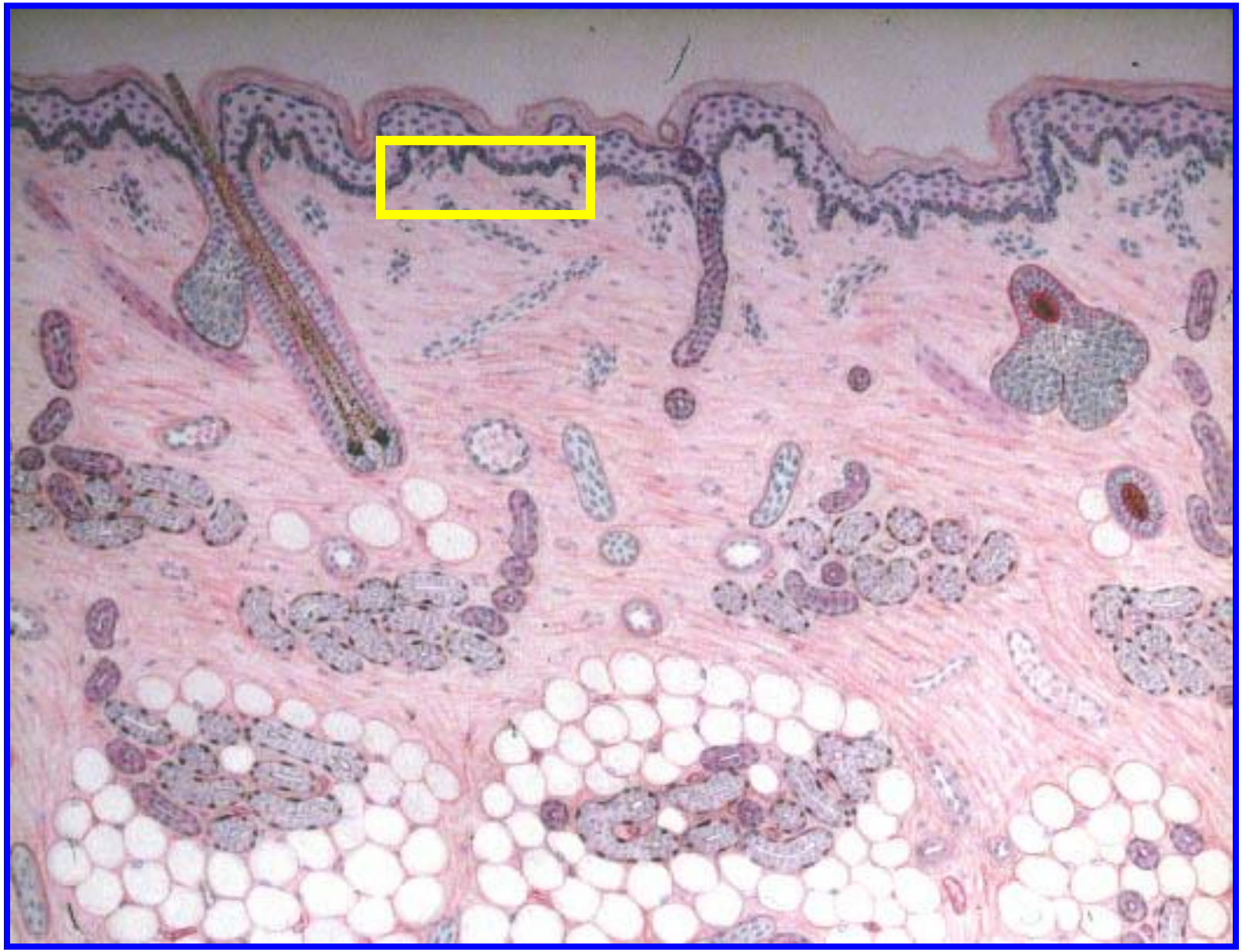
Bullous pemphigoid

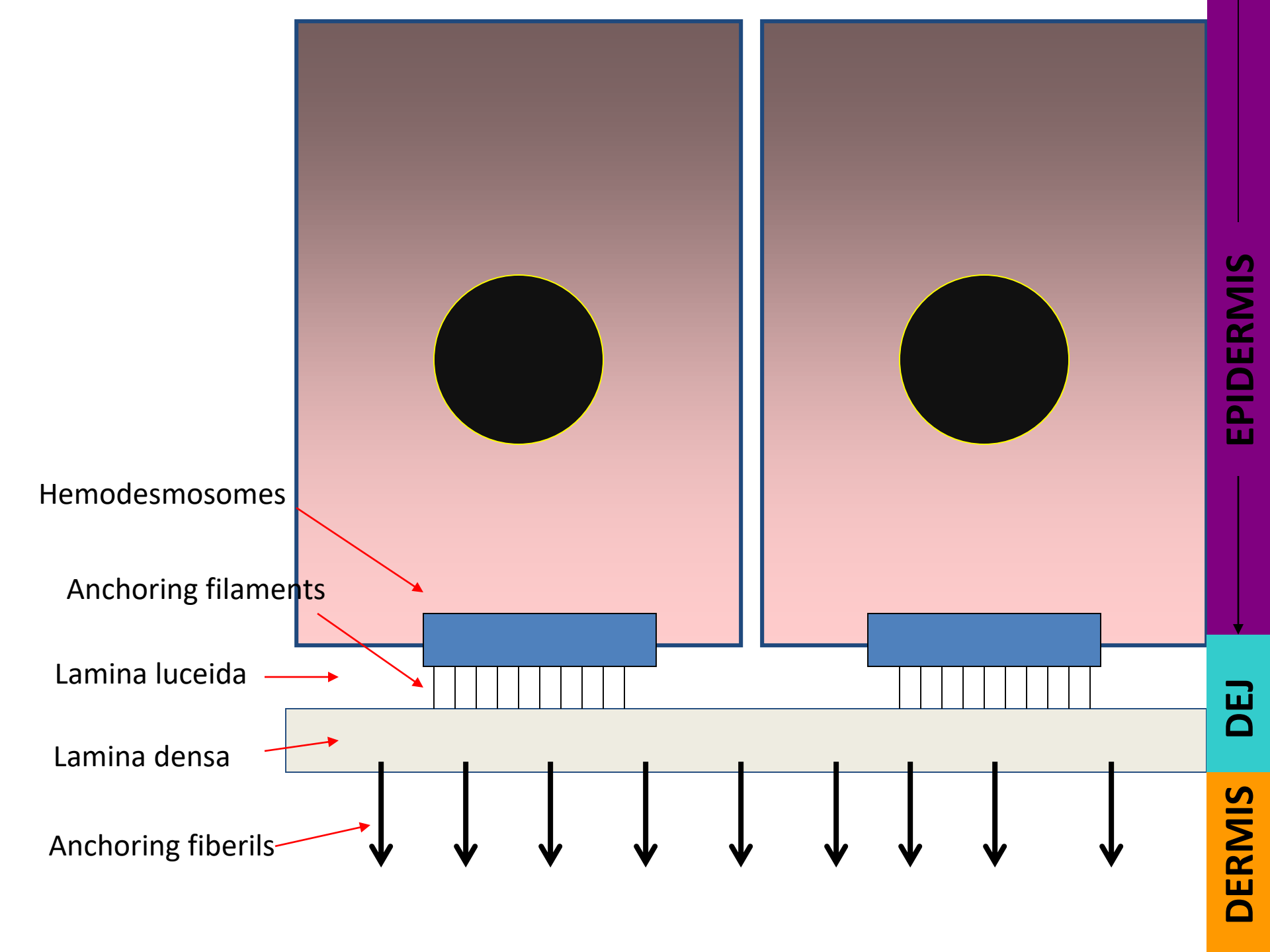
Dermatits Herpitiformis

Subepidermal Autoimmune Vesiculo-bullous Dermatosis

1. Bullous Pemphigoid.
2. Dermatitis Herpitiformis.






Hemodesmosomes
Anchoring filaments
Lamina luceida
Lamina densa
Anchoring fiberils
EPIDERMIS
DEJ
DERMIS

BOULOUS PEMPHIGOID

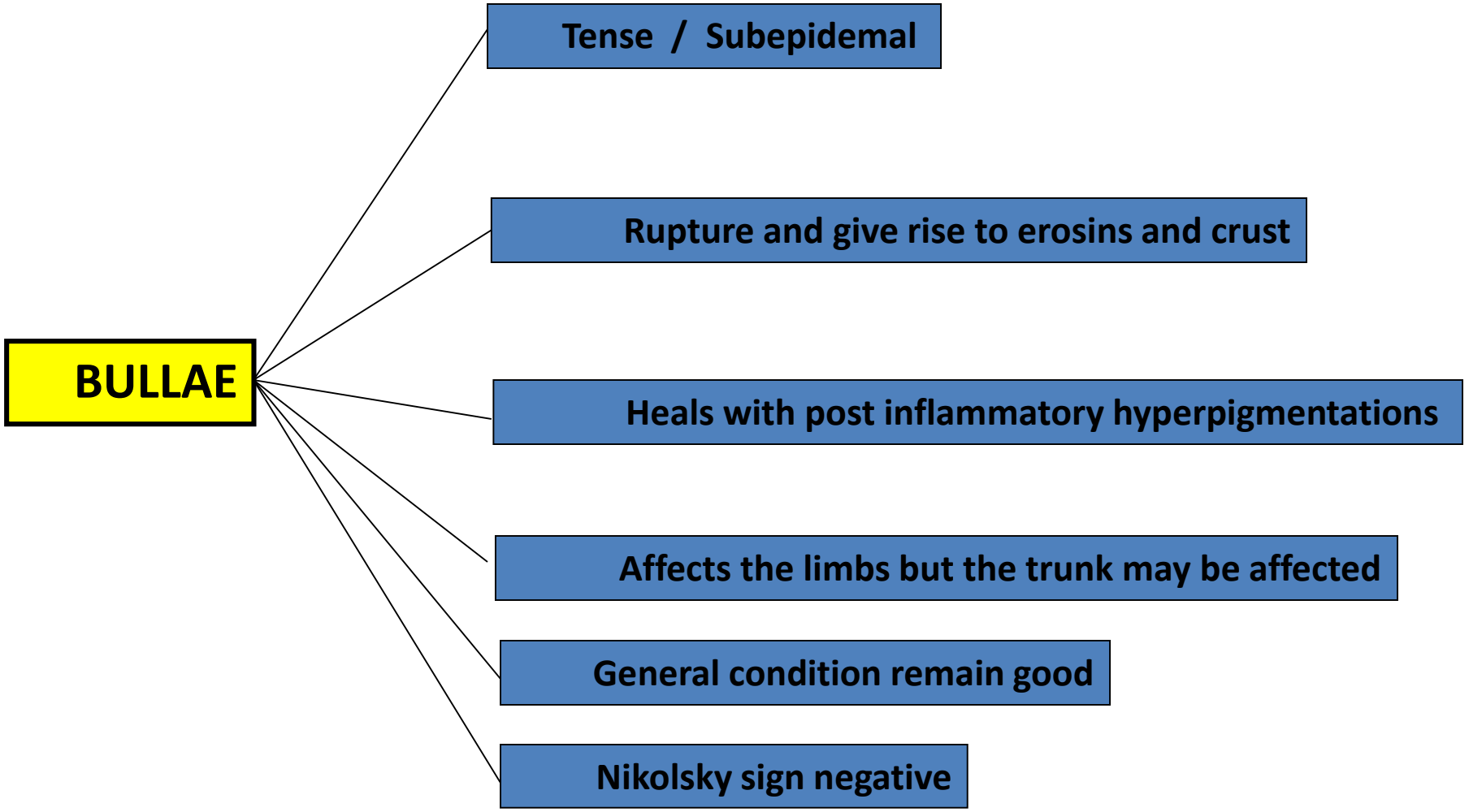
BP, is a chronic benign subepidermal blistering disease characterized by self - limited course with remission and exacerbation and low mortality rate.

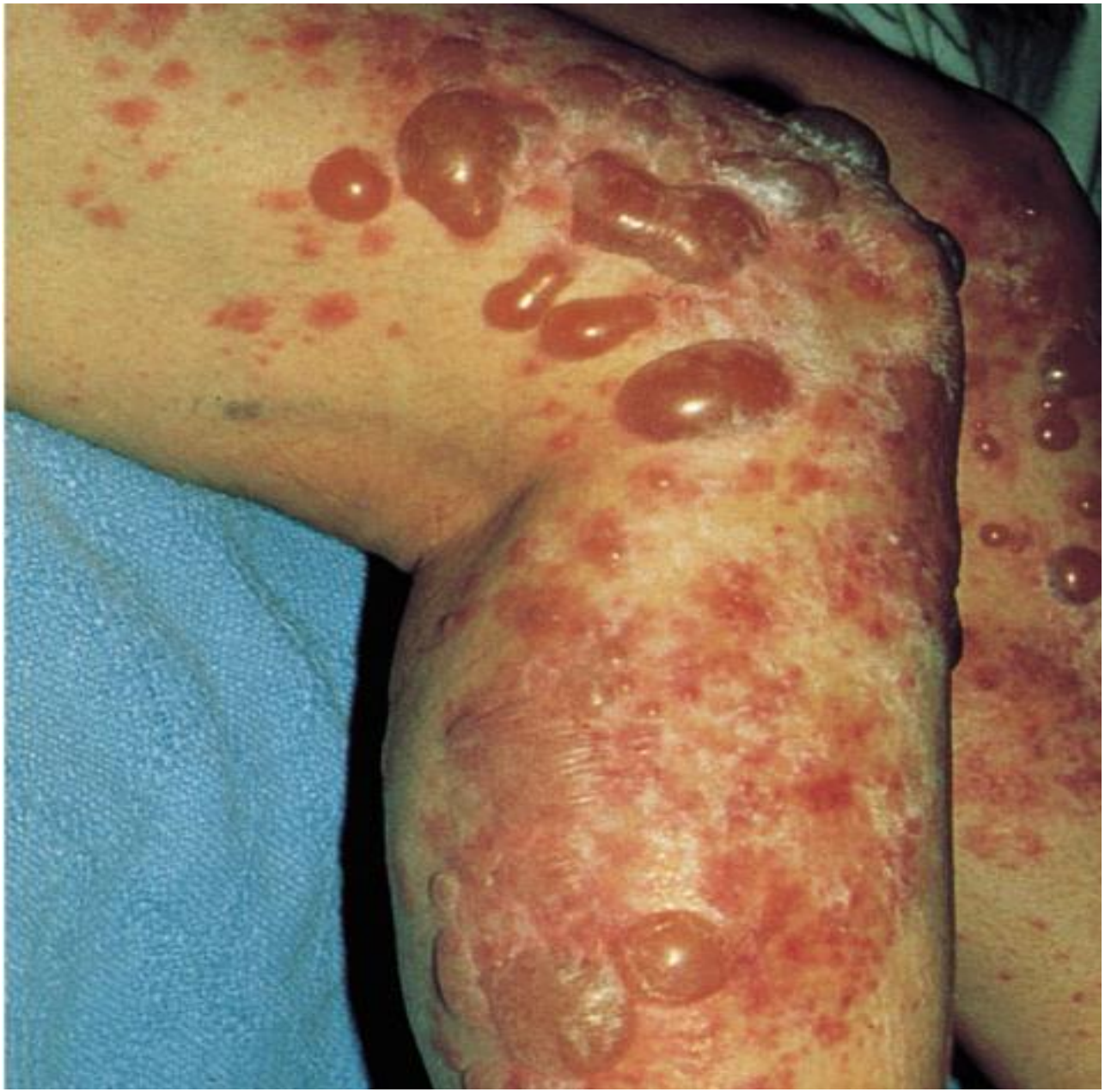
Epidemiology

- BP is a disease of elderly (> 60 years).
- No racial or sex predilection.

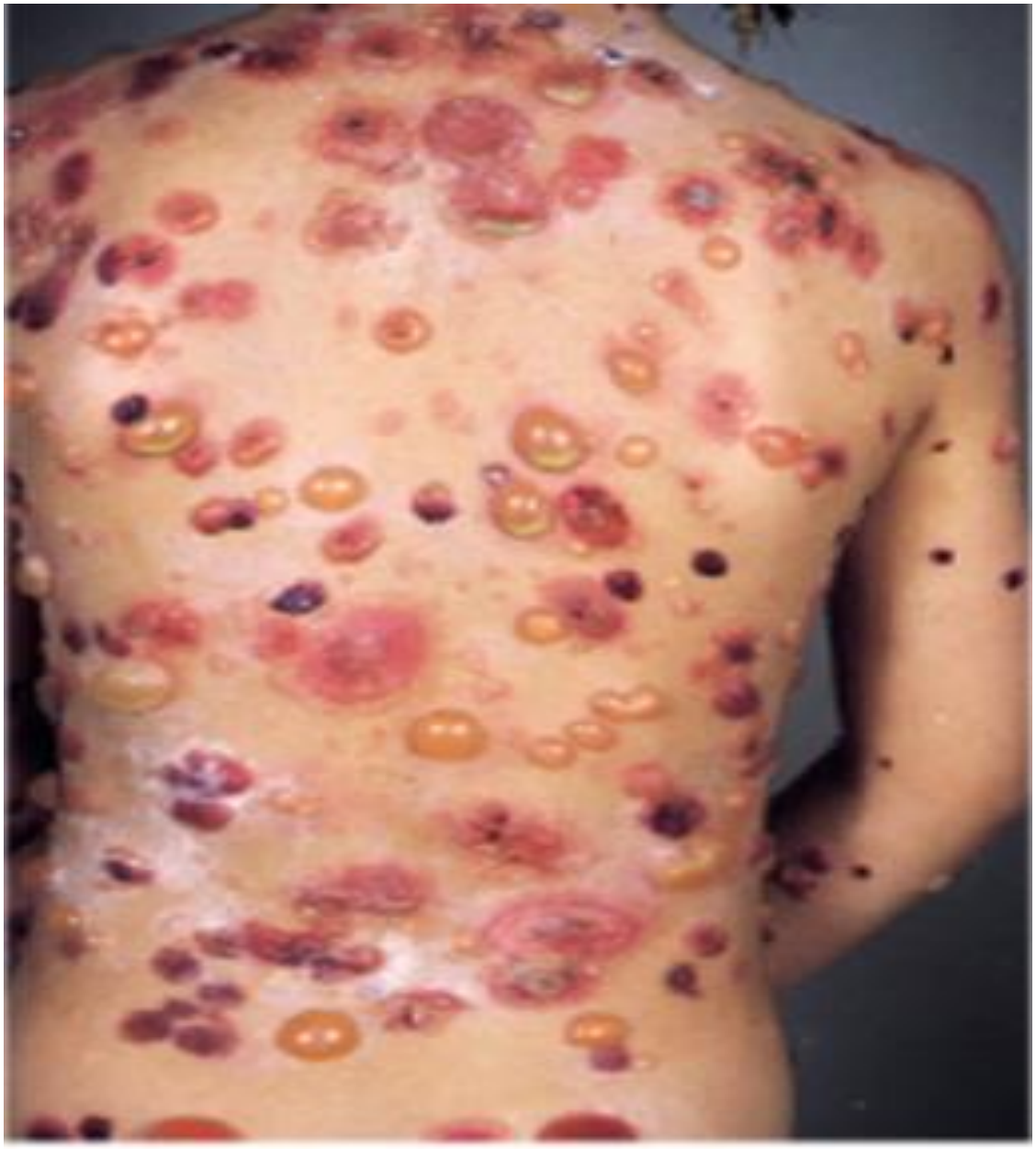
Clinical features

- The primary lesion in bullous pemphigoid is bulla.
- Mucous membranes were not affected.



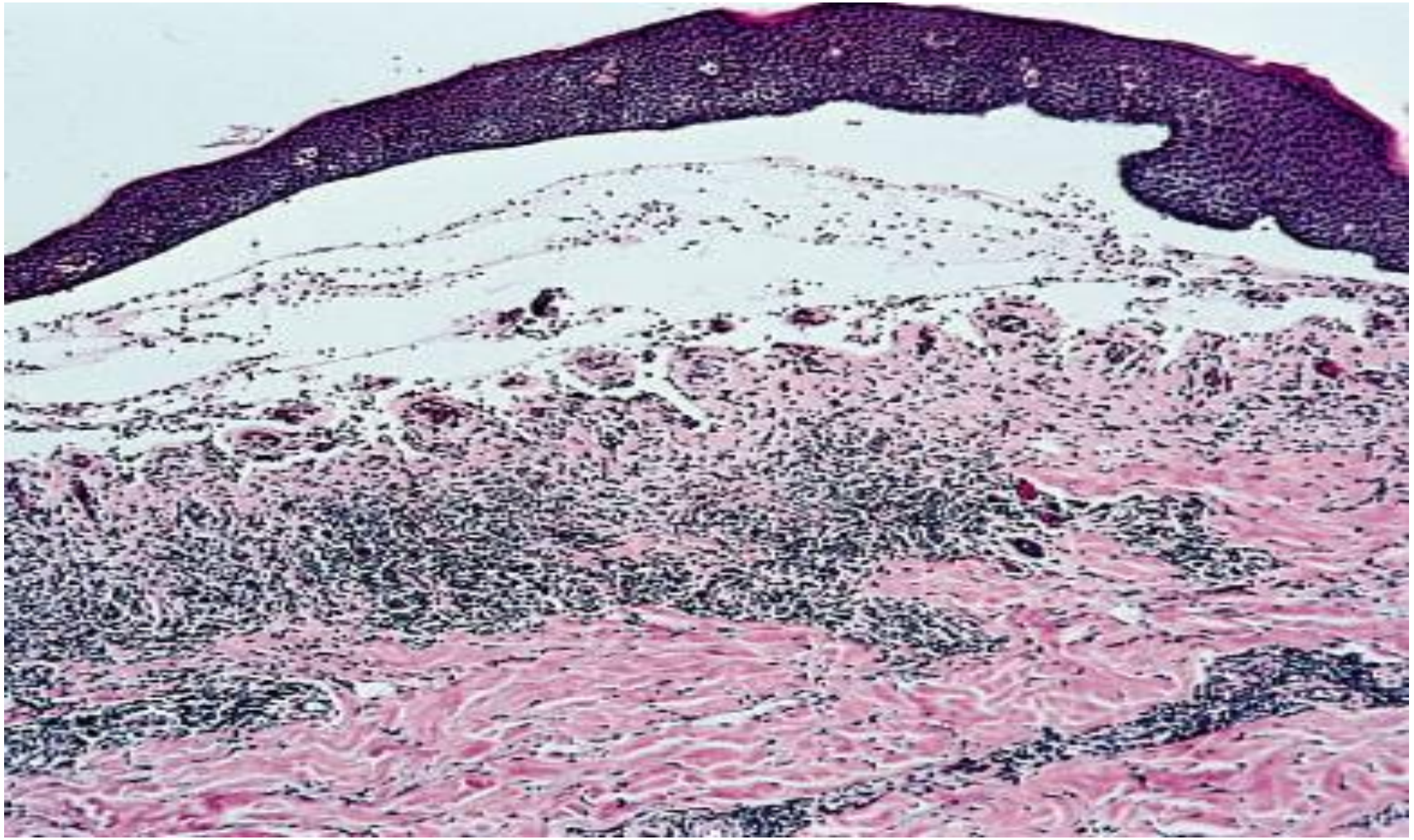


BP, tense clear, turbed and heamrrhagic bullea on an erythemotous base



BP, the trunk may be involved by tense bullea , erosions and crust

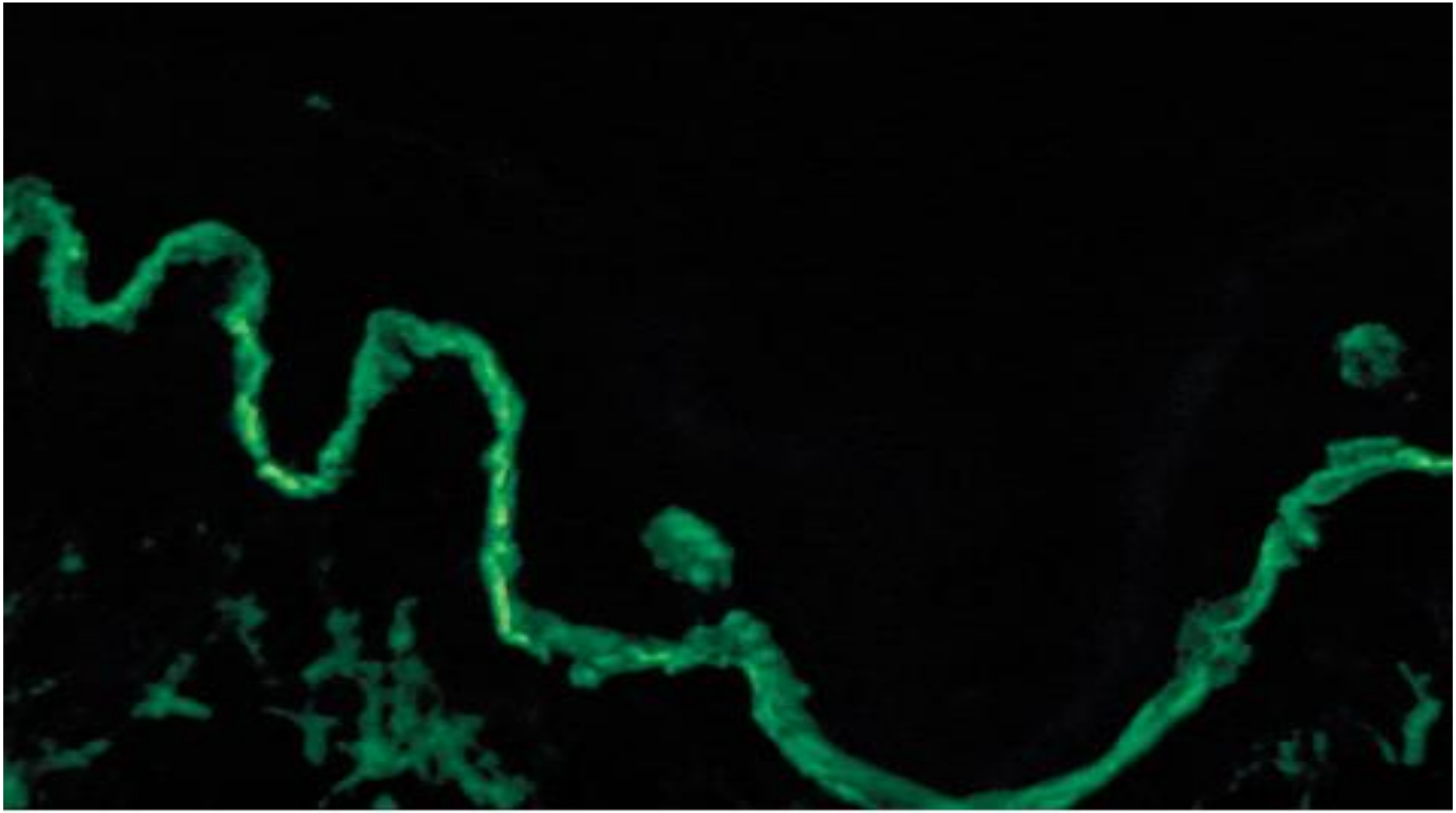
Histopathology



BP , Subepidermal bulla with eosinophilic infiltations

Immunopathology

DIF : Linear deposition of IgG & C3 at the DEJ.
II F : Circulating IgG autoantibodies against BP Ag.



Treatment

- Systemic corticosteroids in a dose of 40 – 60 mg/day.
- In patients with corticosteroids C / I.
  - Dapsone.
  - Azathioprin.

Dermatitis Herpetiformis (DH)

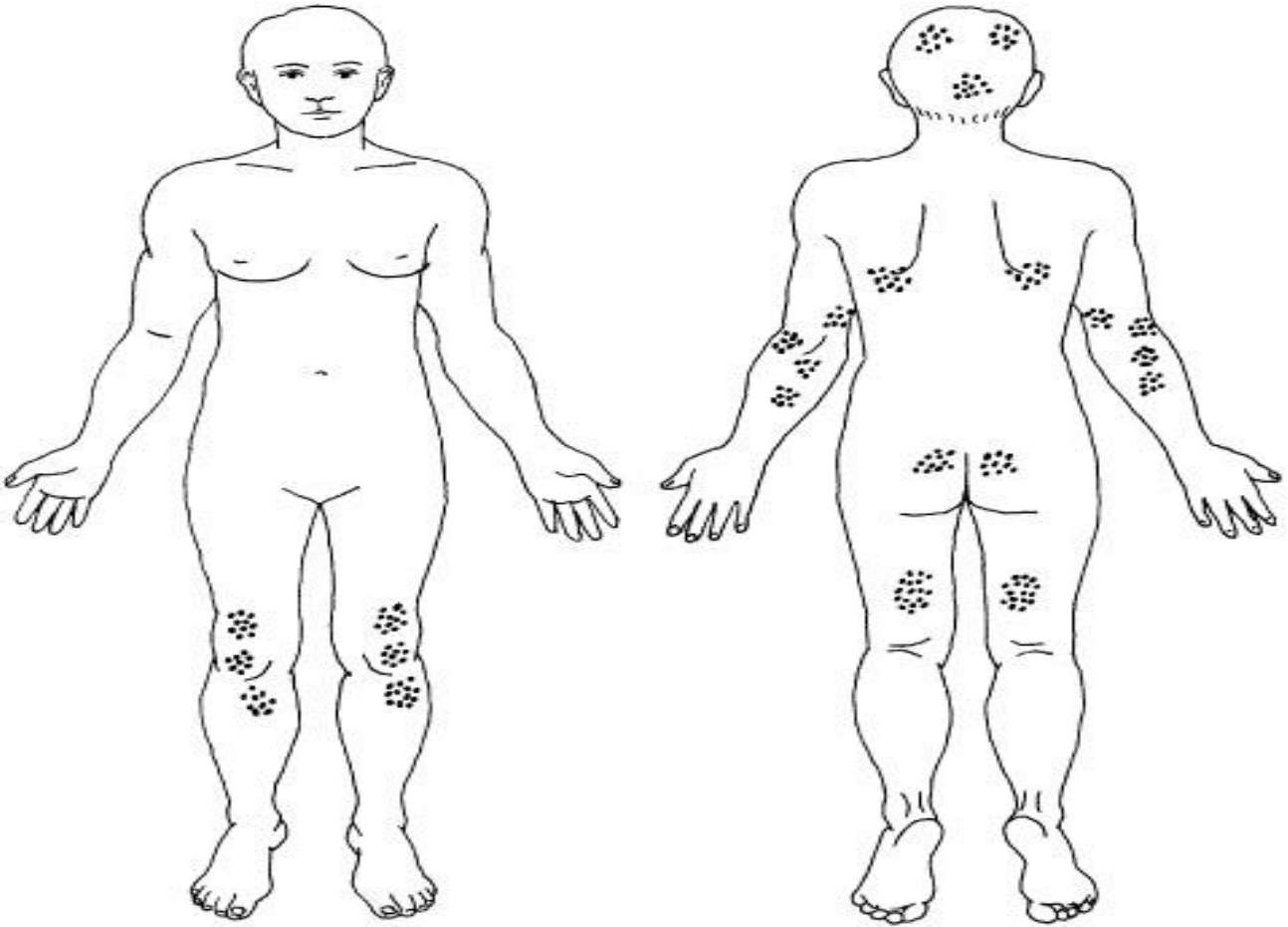
DH, is a chronic autoimmune vesicular dermatosis characterized by severely itchy grouped vesicles most frequently located on extensor surfaces and associated with gluten-sensitive entheropathy.

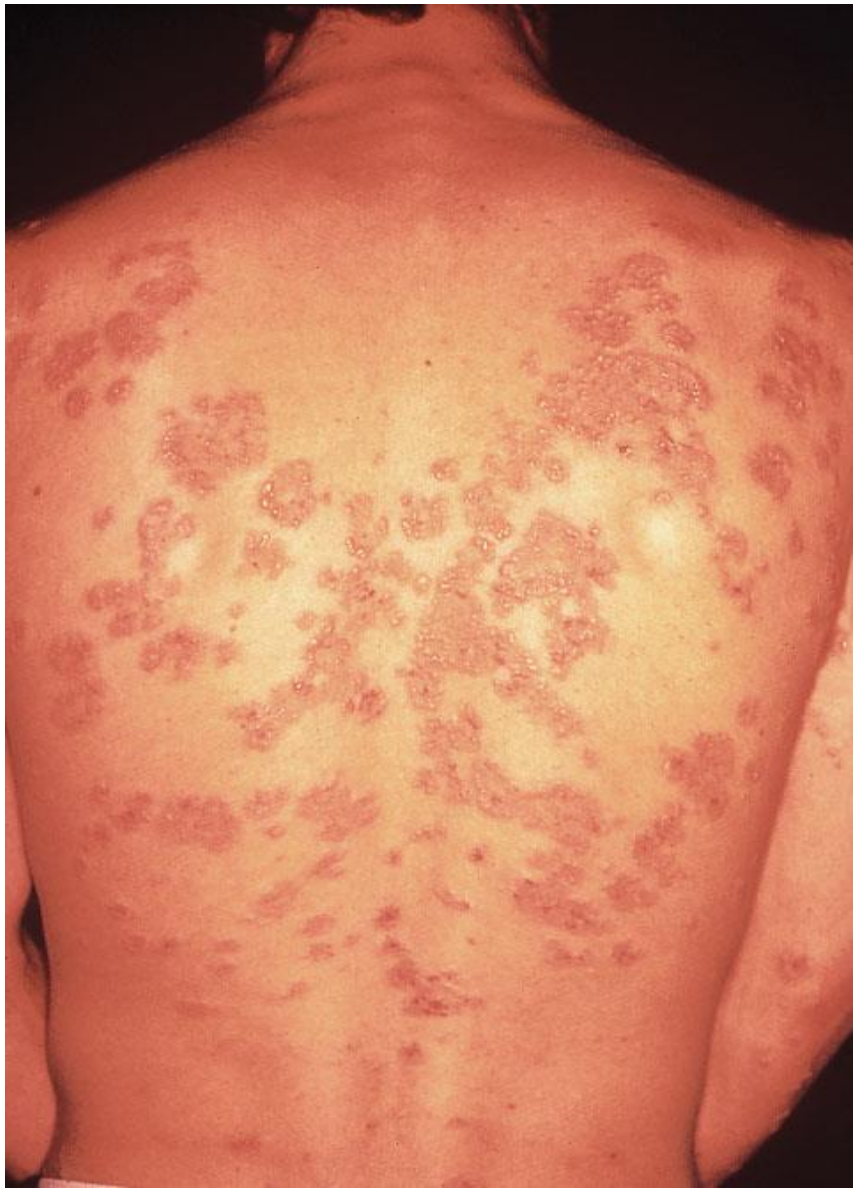
Epidemiology

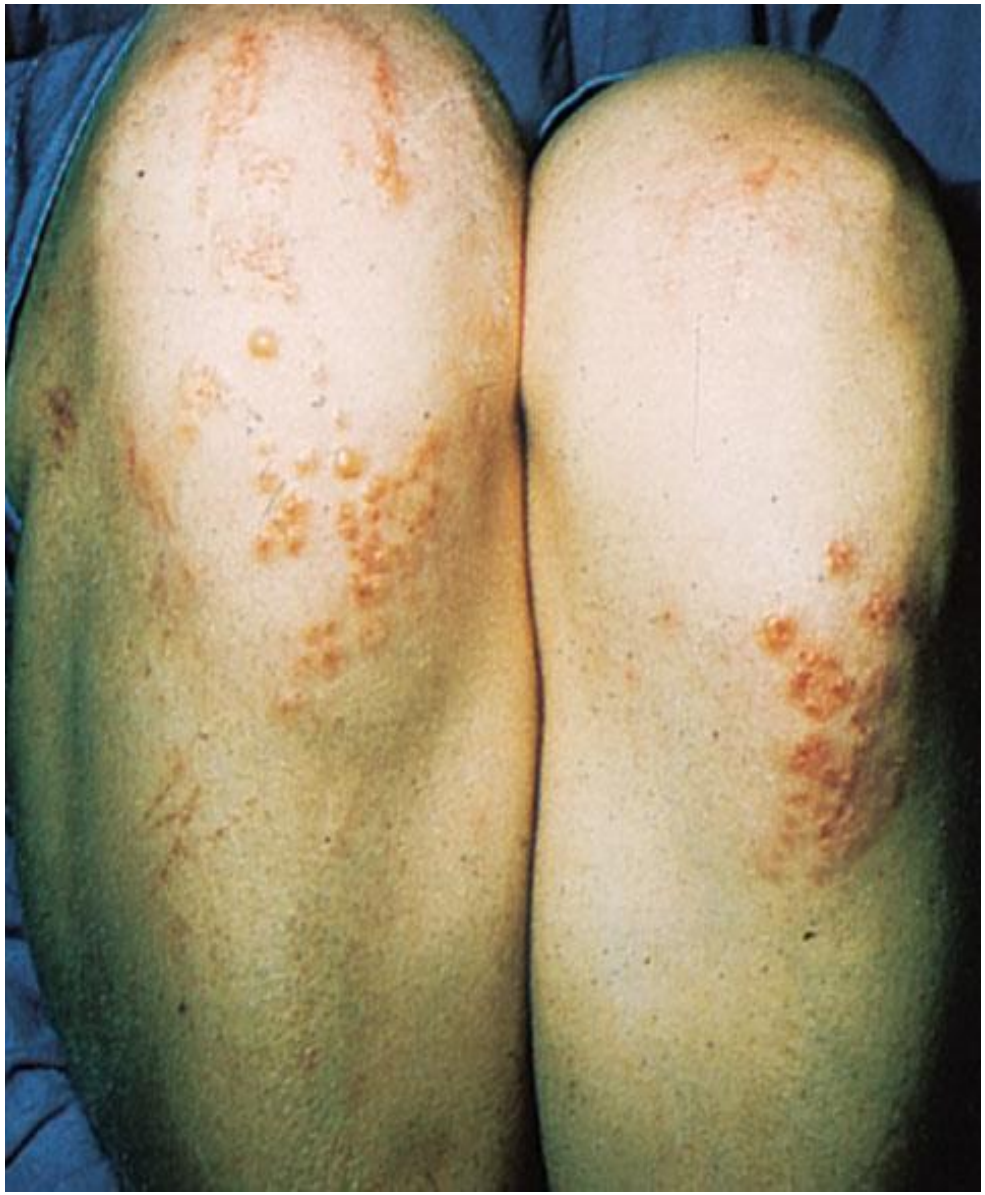
DH, affects all ages most commonly (20-50 years).
Both sexes are equally affected.
Commonly associated with asymptomatic gluten entheropathy.

Clinical Features

The primary lesions in DH is intensely itchy, symmetrically distributed, polymorphic, grouped (hence the name herpetiformis) papulovesicular erpution. Characteristically involving the extensor aspects.

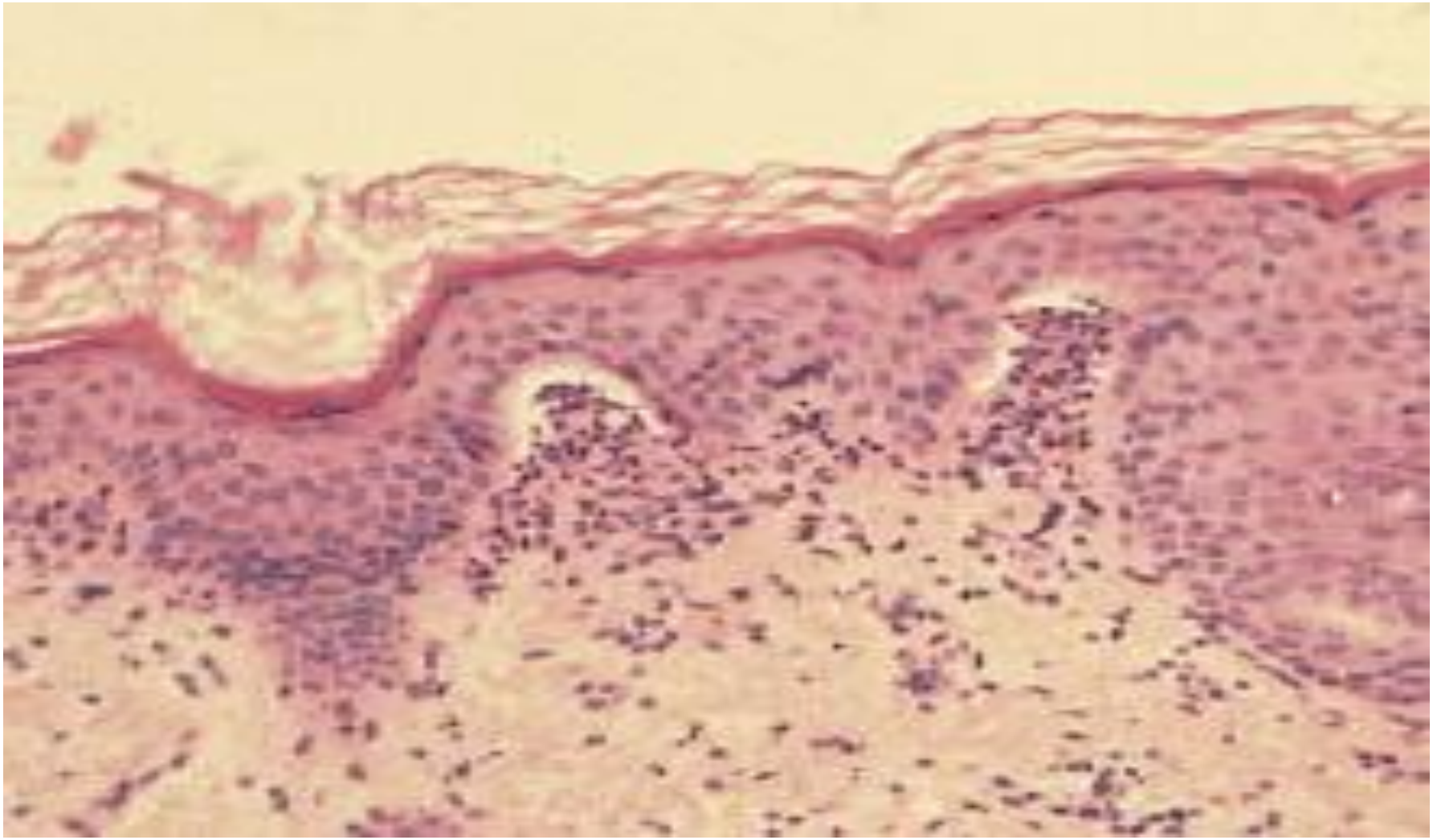






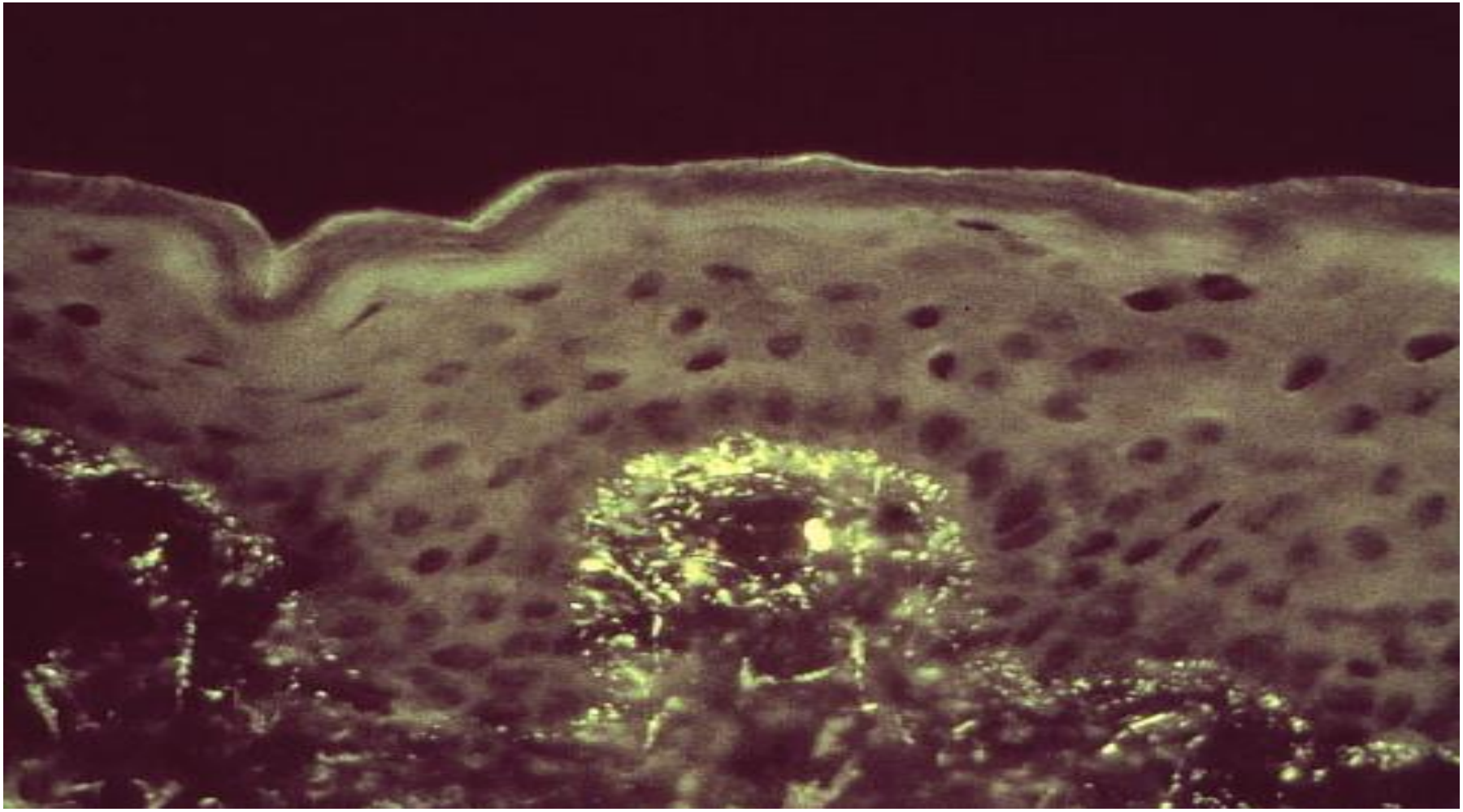
Grouped papulo-vesicular eruption involving the extensors aspects of the body

Histopathology



Subepidermal bullae with papillary neutrophilic microabscesses.

Immunopathology



DIF: S/E granular deposition of IgA

IIF : Negative in DH.

Treatment of DH

- The drug of 1st. Choice in DH is DAPSON.
- Gluten free diet may help some patients

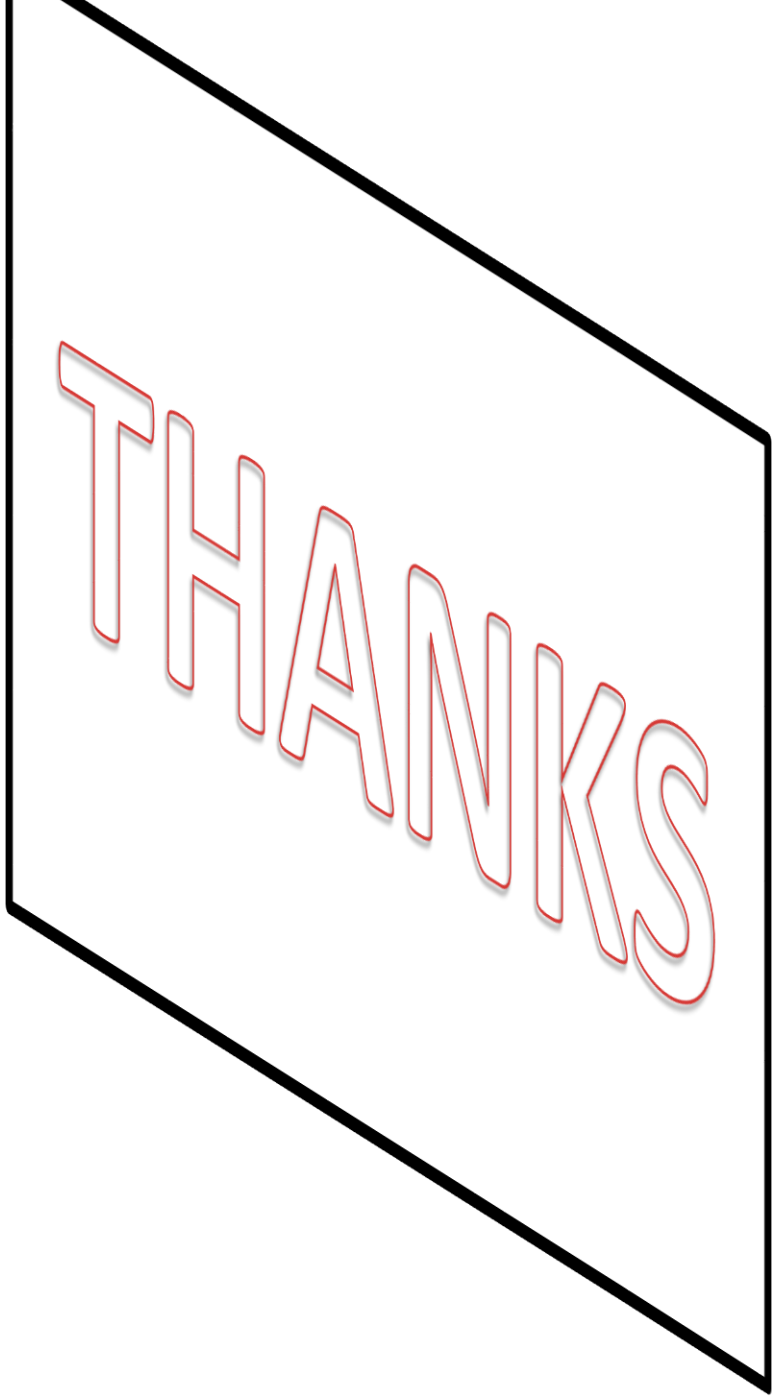
Side effects of Dapson.

Before you start dapson you must do G -6- P Dehdrogenase enzyme.

1- Hemolytic anemia
2 -Methhemoglobinemia.
3- Agranulocytosis.
4- Perpherral neuorpathy
5- Drug rashes

Feature	Pemphigus	Pemphigoid	D.Herpitiformis
Age	Middle age	Elderly	Young age
Race & HLA	HLA	Non	HLA
Bullae	Flaccid	Tense	Papulo - vesicles
Symptoms	Non	Non	Sever itching
Distributions	Trunk	Extremities	Extensors
M.M	Involved	Spared	Spared
Histopathology	Intraepidemal	Subepidermal	Subepidermal
DIF	Ig G I/E	Ig G DEJ	Ig A dermal tips
IIF	Positive	Positive	Negative
Treatment	Steroid High	Steroid low	Dapson

Feature	Pemphigus	Pemphigoid	D.Herpitiformis
Age	Middle age	Elderly	Young age
Race & HLA	HLA	Non	HLA
Bullae	Flaccid	Tense	Papulo - vesicles
Symptoms	Non	Non	Sever itching
Distributions	Trunk	Extremities	Extensors
M.M	Involved	Spared	Spared
Histopathology	Intraepidemal	Subepidermal	Subepidermal
DIF	Ig G I/E	Ig G DEJ	Ig A dermal tips
IIF	Positive	Positive	Negative
Treatment	Steroid High	Steroid low	Dapson


THANKS