VESICULOBULLOUS DISEASES

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CLASSIFICATION OF VESICULOBULLOUS DISEASES

- **VESICLE&BULLA**

  A clear fluid lesion just below the epithelium which ruptures to form an ulcer, if this is smaller than 5mm then it is a vesicle ,if larger than 5mm than it is a bulla

Blistering (Bullous) Diseases

- Inflammatory Types
  - Pemphigus
  - Bullous Pemphigoid
  - Dermatitis Herpetiformis
- Noninflammatory Types
  - Epidermolysis Bullosa
  - Porphyria

CLASSIFICATION OF VESICULOBULLOUS DISEASES

CLASSIFICATION

INTRA EPITHELIAL VESICLES: The lesion is formed within the epithelium

- **Acantholytic vesicles :** This is because of the break down of specialized attachments called the desmosomes

- **Nonacantholytic vesicles**: It is usually in the viral infections because of the death or the rupture of the group of cells.

PEMPHIGUS VULGARIS

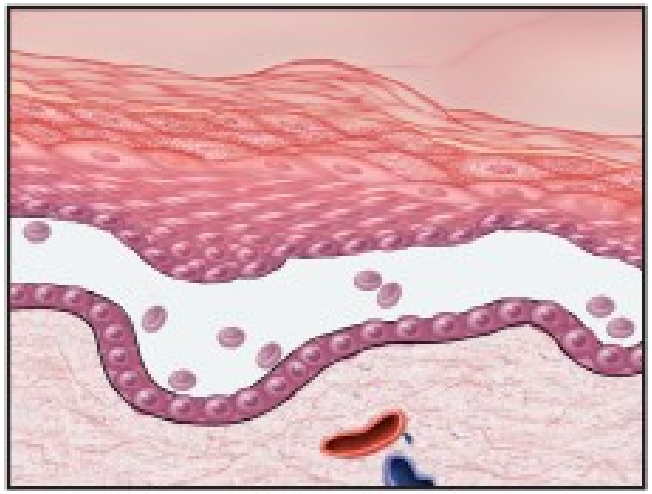
- Autoimmune disease.
- Common in Ashkenazi and Mediterranean jews .
- Middle aged females.
- Other variants are:

  Pemphius Vegitans

  Pemphigus Foliaceus & Erthematosus

  Paraneoplastic pemphigus.

Suprabasal Blister
Pemphigus Vulgaris



B Suprabasal

PEMPHIGUS VULGARIS

CLINICAL FEATURES:

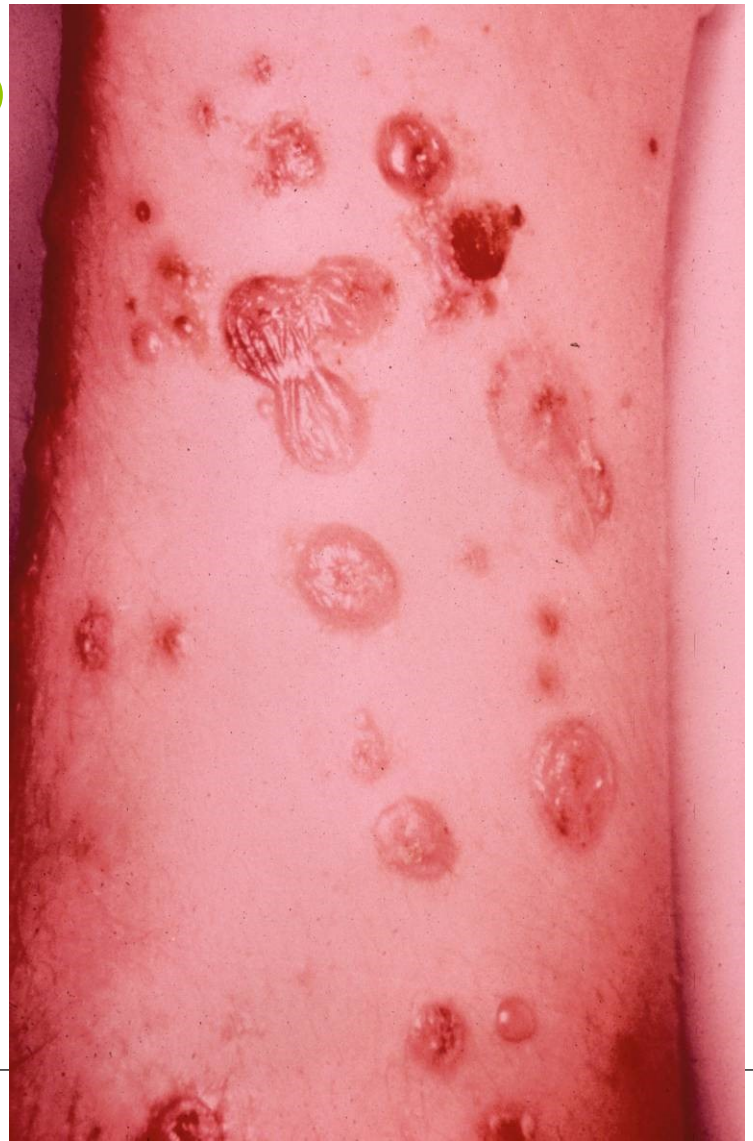
- Painful ulcers or bulla are formed which are fluid filled.
- They can be formed any where in the oral cavity .
- The bulla is rapidly ruptured leaving a collapsed roof of grayish membrane with a red ulcerated base.The ulcer may look like an apthous ulcer or may be large map shaped.
- Nikolsky sign is positive.

C/F con…d

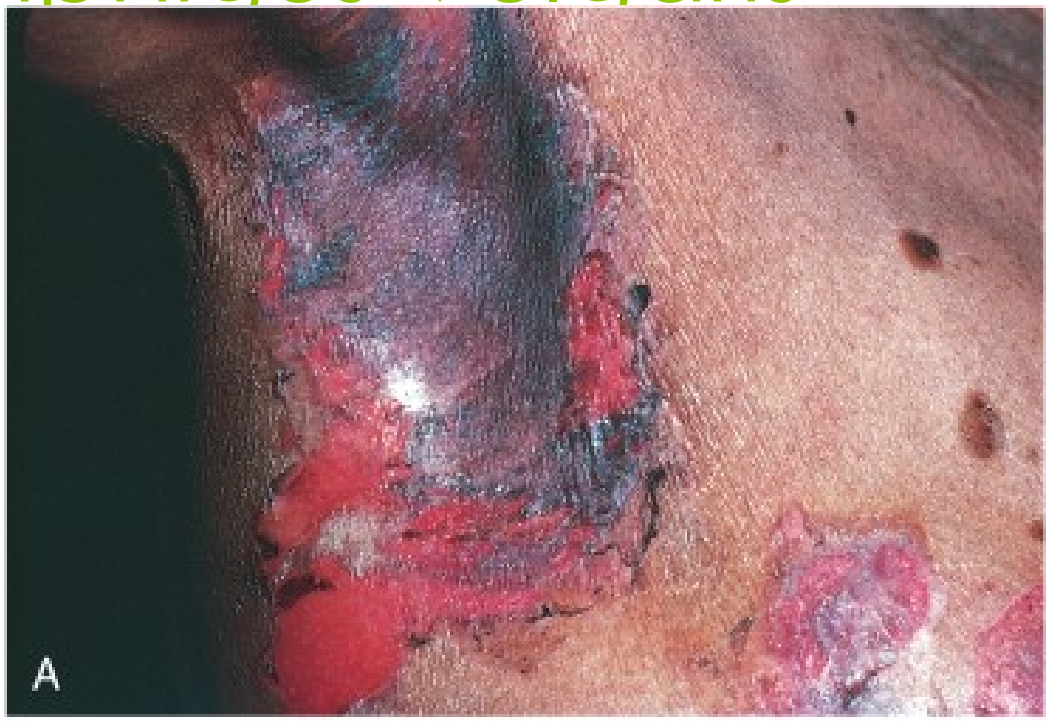
PEMPHIGUS VULGARIS

- Some time the ulcers are joined together to make a confluence this condition is very painful.
- It has a variable course might involve skin, oesophagus, cervix.
- Protein/fluid,electrolyte and weight loss /secondary infections.
- Fatal if untreated.

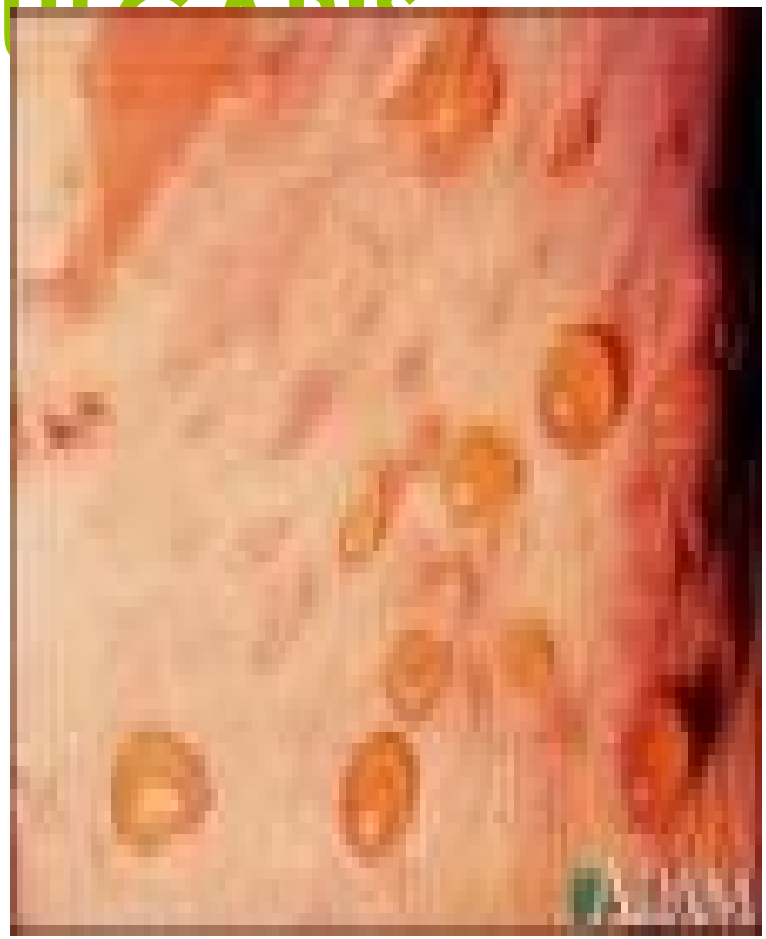
Pemp

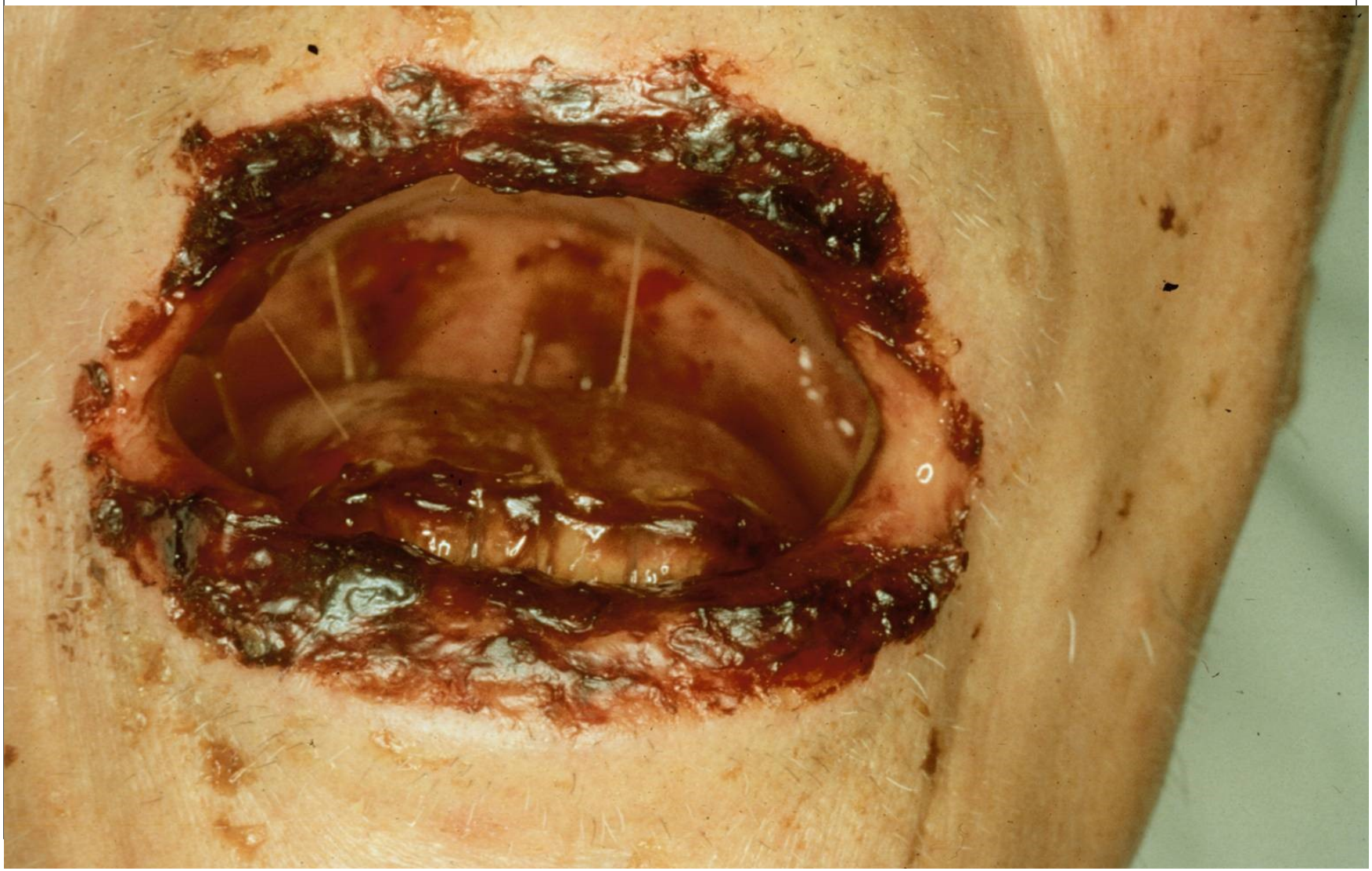


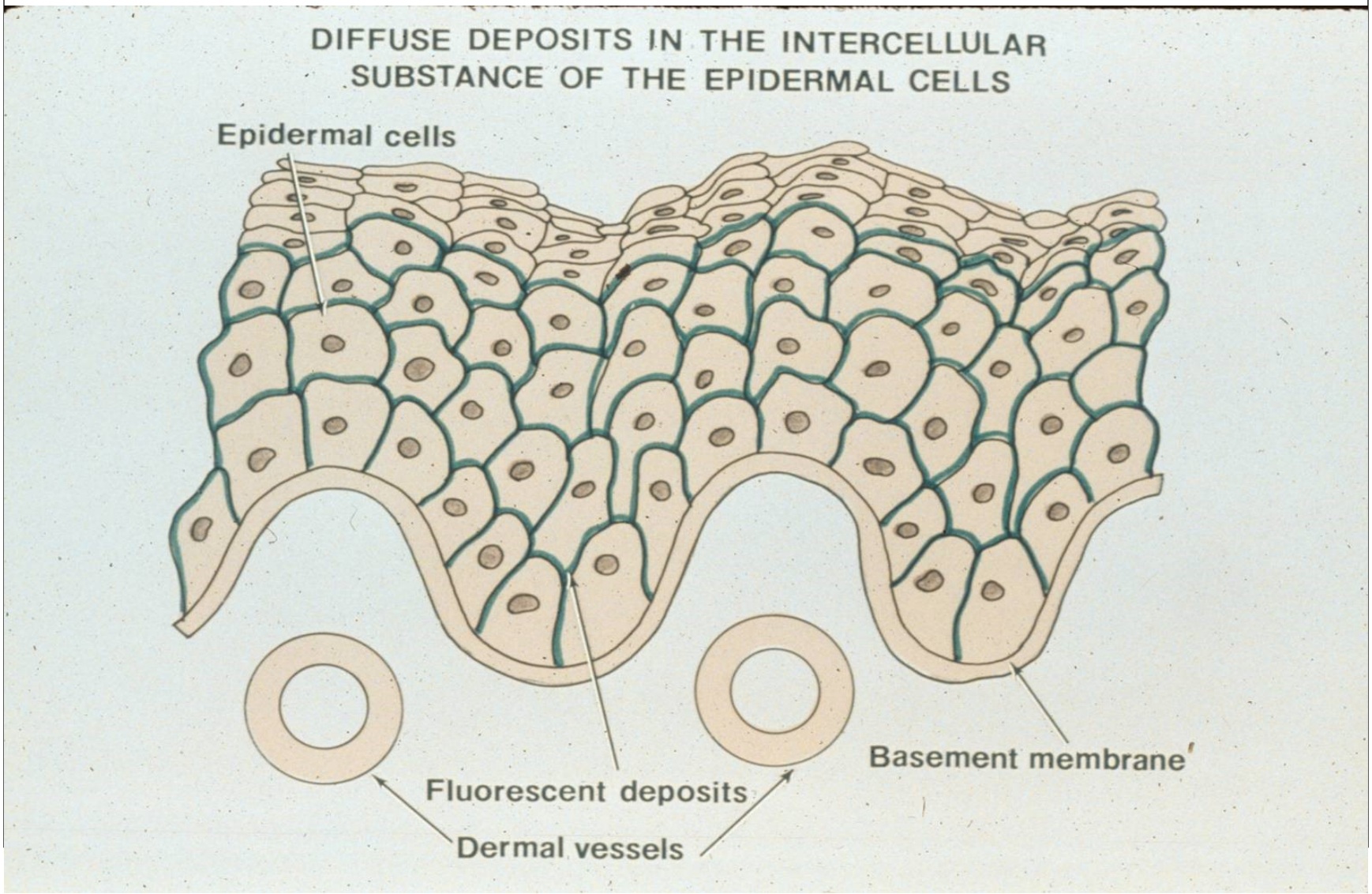
Pemphigus Vulgaris



PEMPHIGUS VULGARIS






DIFFUSE DEPOSITS IN THE INTERCELLULAR SUBSTANCE OF THE EPIDERMAL CELLS
Epidermal cells
Basement membrane
Fluorescent deposits
Dermal vessels

PEMPHIGUS VULGARIS

PATHOGENESIS:

- It is an autoimmune disease
- There are circulating antibodies of type IgG.
- These antibodies are reactive against the desmosomes or the tonofilament complex.
- path.cont…d

PEMPHIGUS VULGARIS

- The epithelial damage is directly proportion to the number of the circulating antibobies.

The cell to cell break down also takes place through a complement system but this process is not clearly understood .

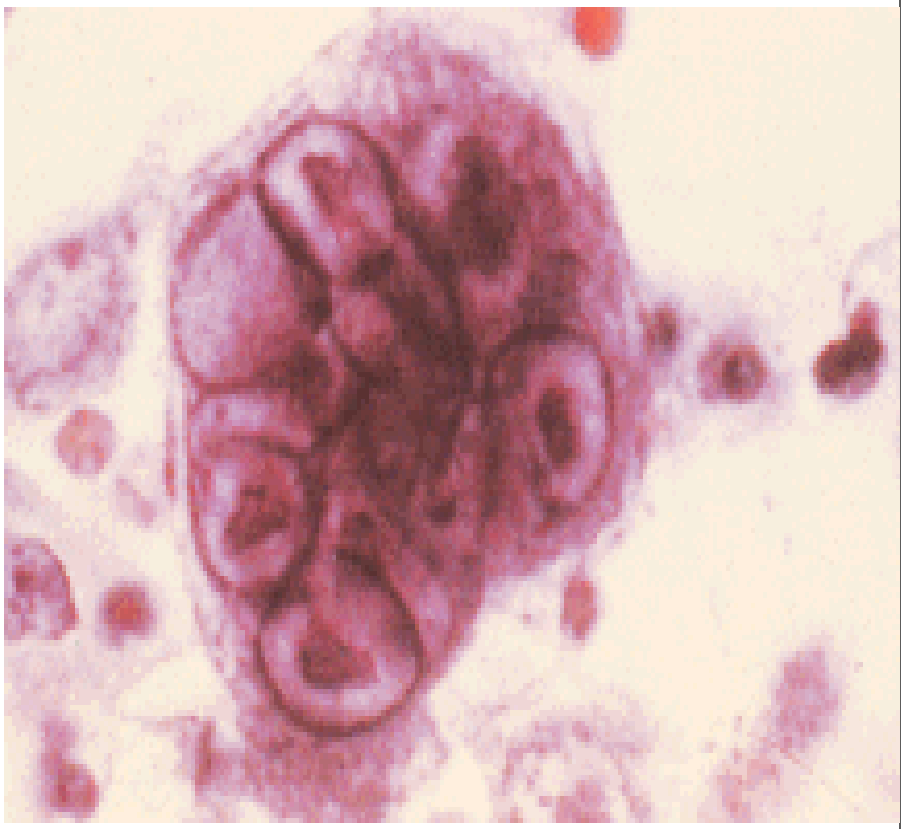
PEMPHIGUS VULGARIS

HISTOPATHOLOGY:

- Intra epithelial vesicles or bulla and cleft like spaces are produced by acantolysis
- These changes are in the stratum spinosum or the prickle cell layer
- The basal cell remain attach to the lamina propria and project into the bulla like tombstones.
- Inflammatory cells are very scanty however eosinophils may be seen.

PEMPHIGUS VULGARIS

tazank cells



PEMPHIGUS VULGARIS

DIFFRENTIAL DIAGNOSIS:

- Pempegiod
- Erthema multiforme
- Bullous lichen plannus

PEMPHIGUS VULGARIS

TREATMENT:

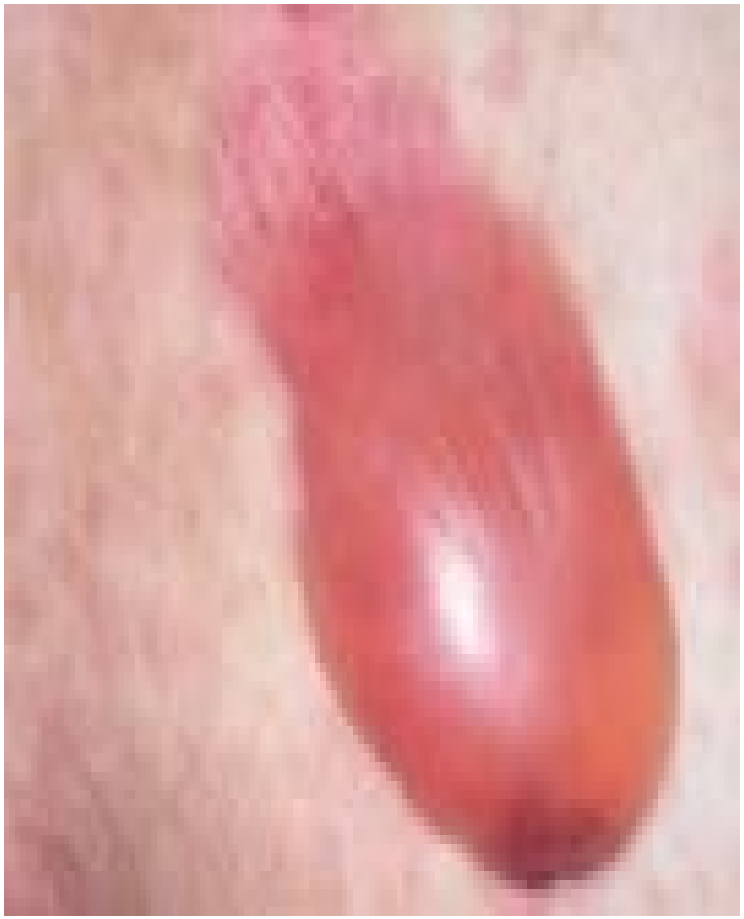
- High mortality rates previously
- Introduction of systemic corticosteroids like prednisolone in stable cases.
- Prednisolone plus azathioprine methotrexate and cyclophospamide in progressed or advance cases.

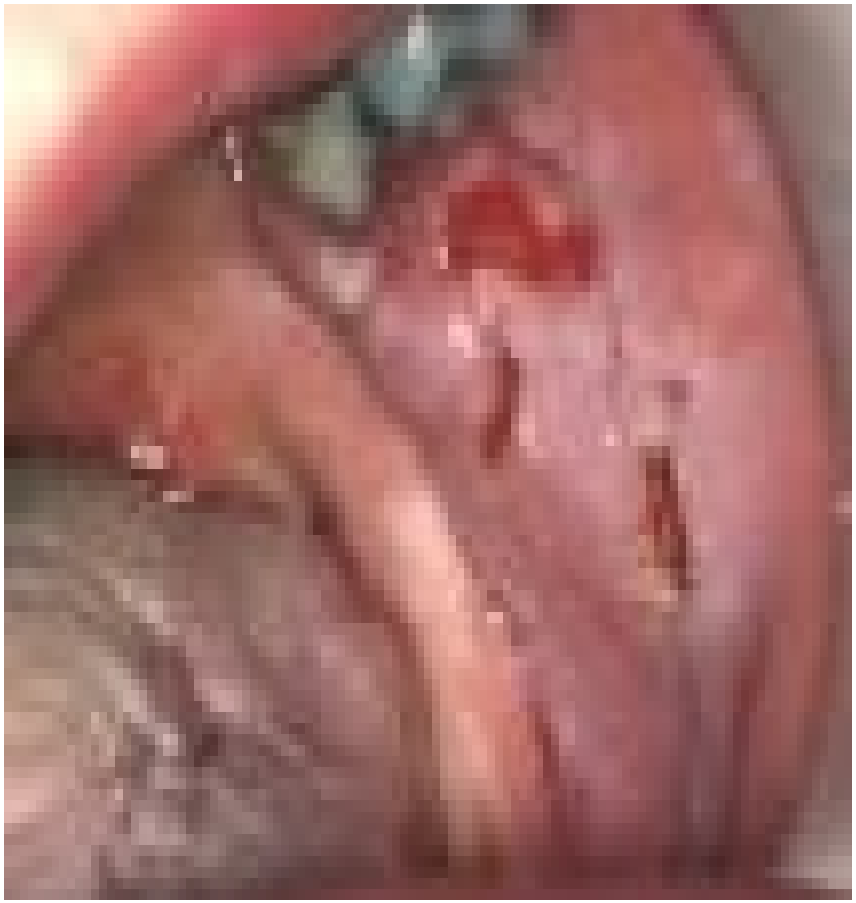
PEMPHGOID

PEMPHGOID

- Mucous membrane pemphigoid (cicatricial) CIKA-TRI-CIAL
- Bullous pemphigoid

PEMPHGOID



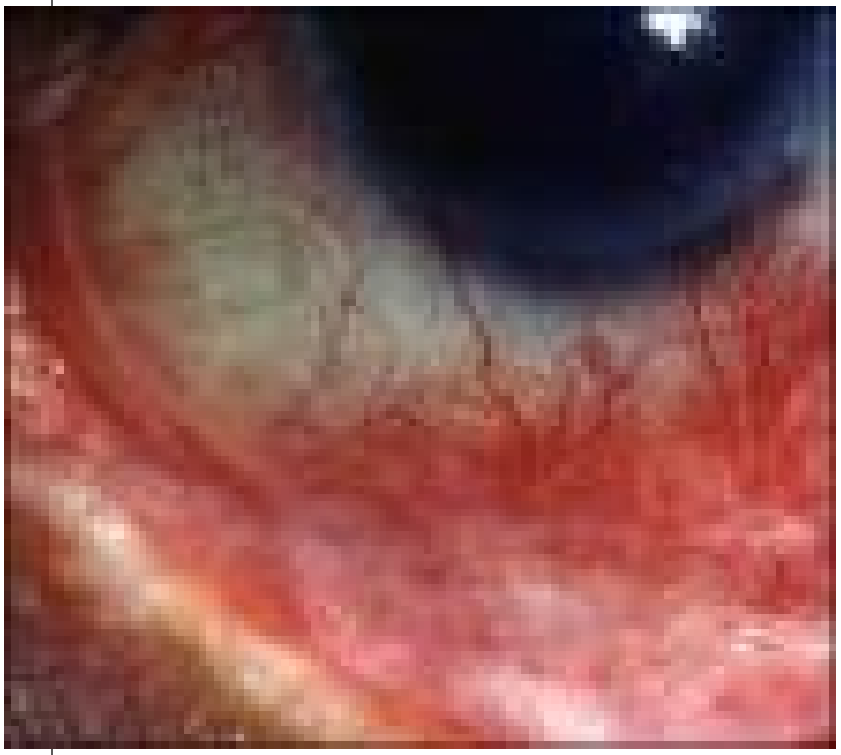


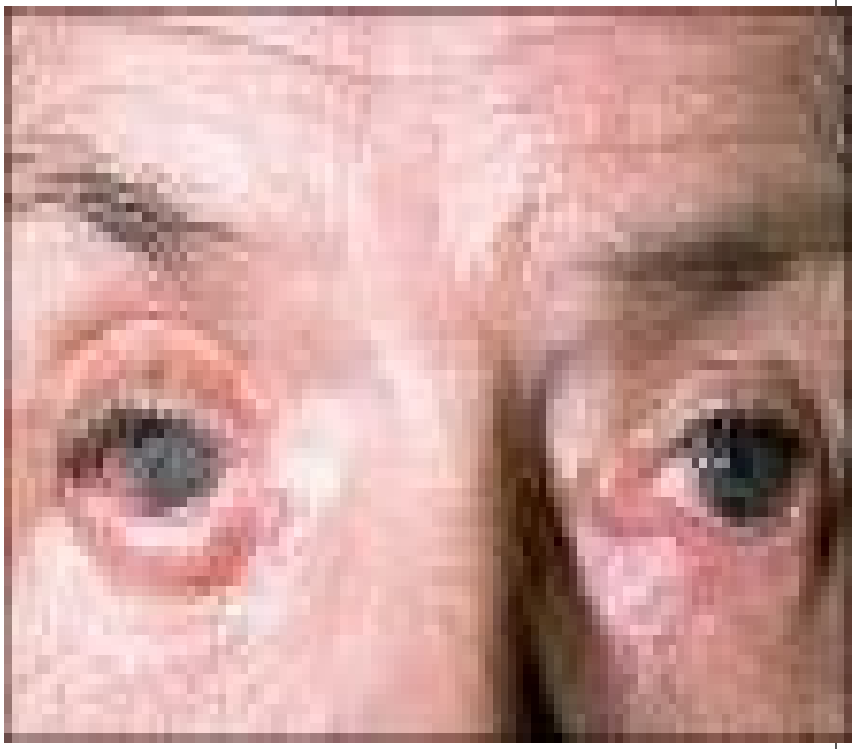
PEMPHGOID

CLINICAL FEATURES(MMP)

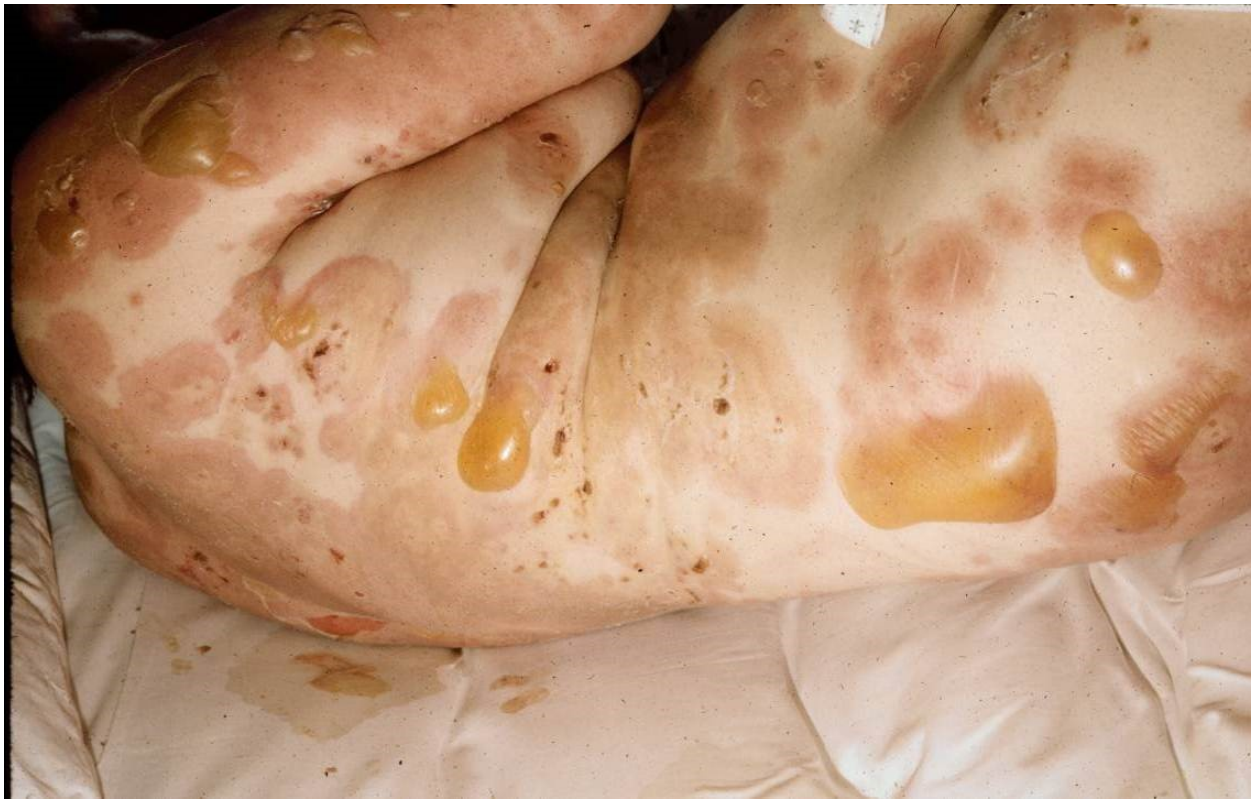
- Oral mucosa is the first site- lesions are rarely wide spread
- Subepithelial bullae, ruptured in the later stages.
- Bleeding in the bullae – bleeding blisters
- Slow progress, skin involvement absent or rare
- Involvement of eyes, nose larynx, pharynx and osephaghus
- Nikolsky sign is positive

PEMPHGOID

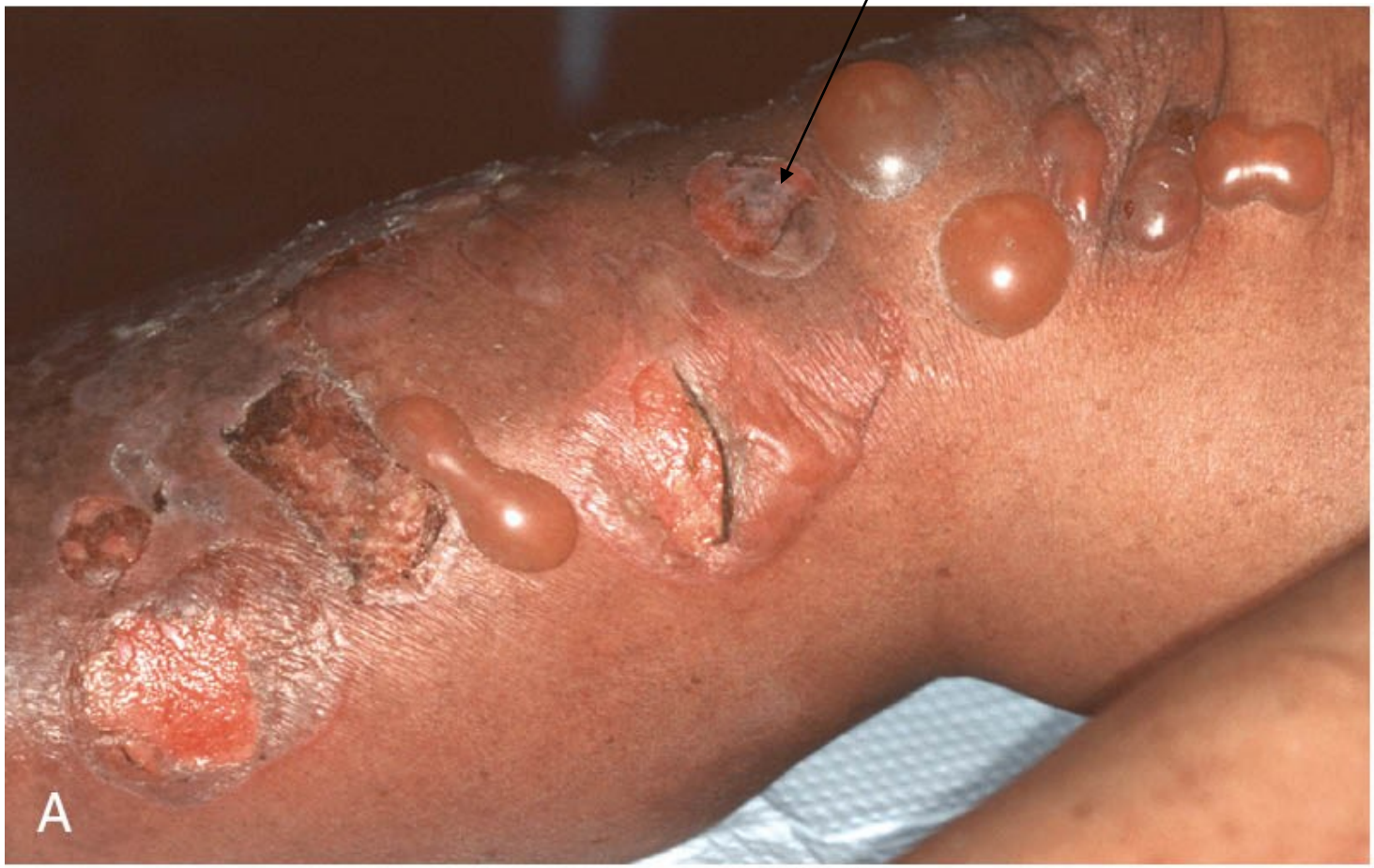




Bullous Pemphigoid
Tense Bullae



Bullous Pemphigoid-Clinical



PEMPHGOID

- CASCADEOF EVENTS

Antibody antigen complex

↓

Complement activation

↓

Neutrophils & Eosinophils recruited

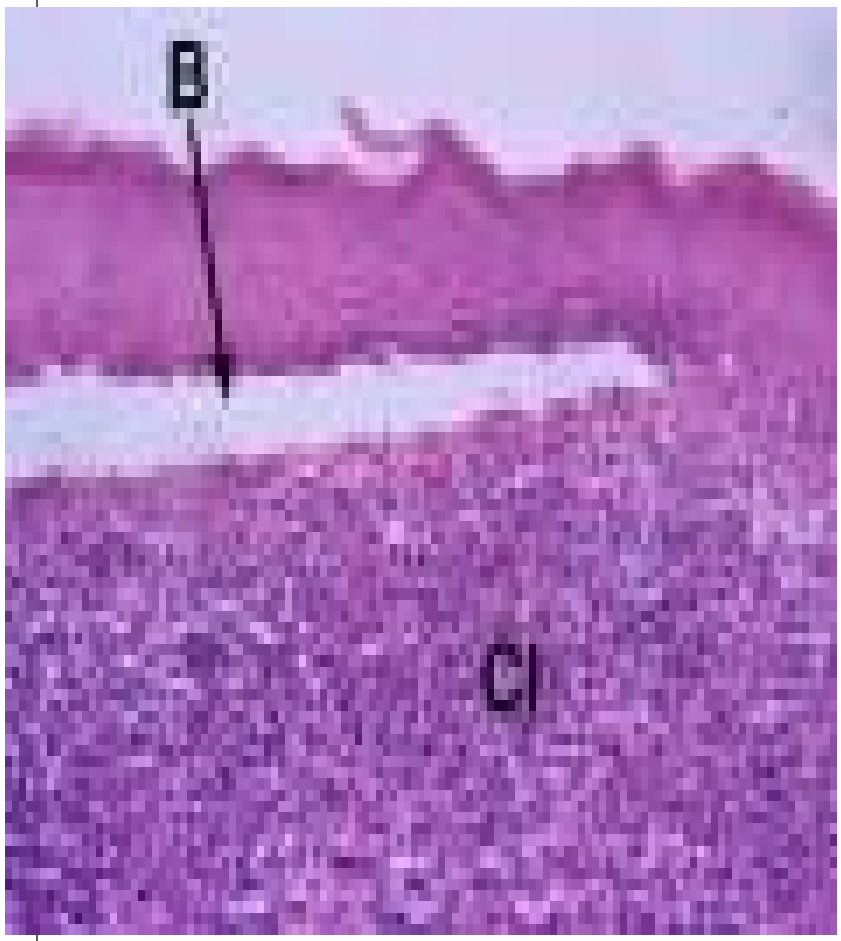
↓

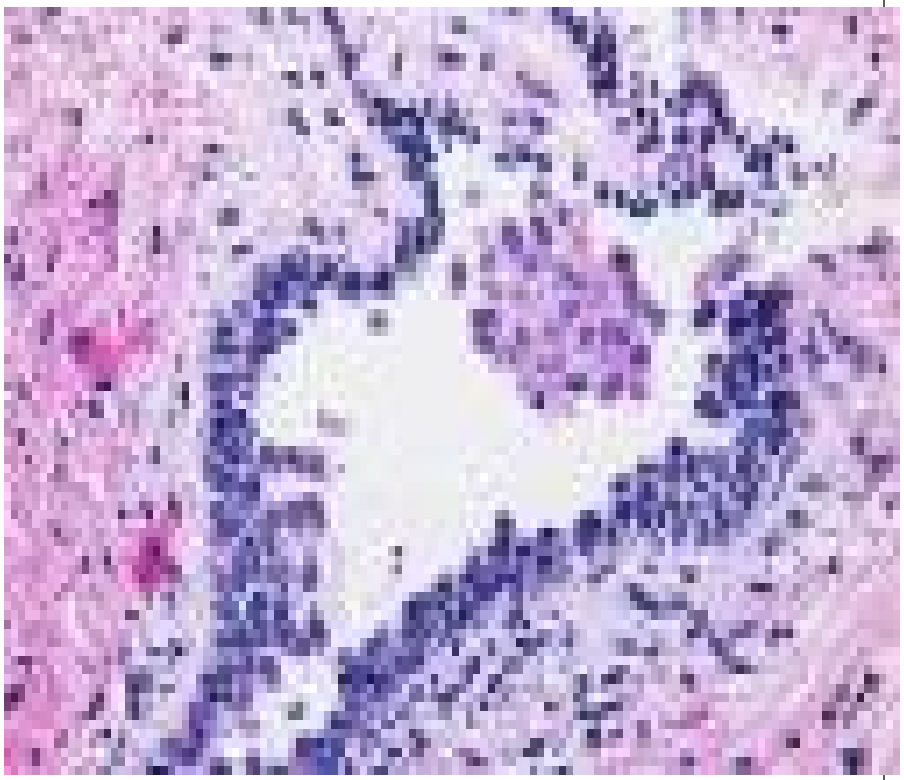
Release of proteases by the recruited cells

↓

Sub epithelial blister formation

PEMPHGOID





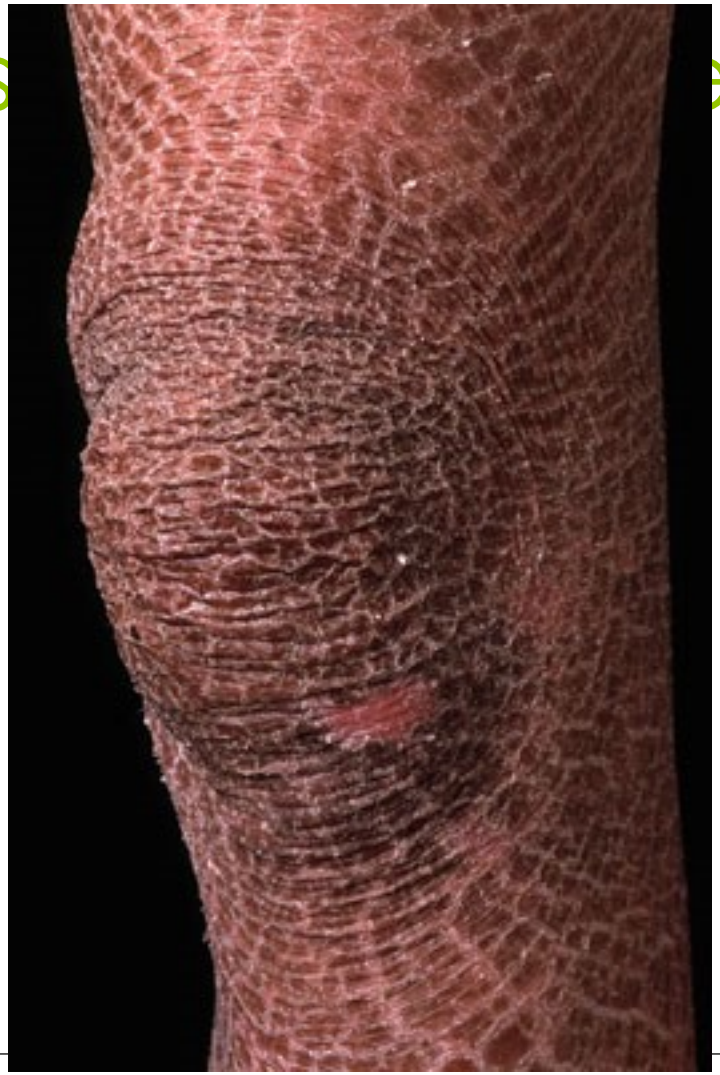
PEMPHGOID

MANAGEMENT

- Confirm diagnosis
- Topical corticosteroids
- Ocular involvement –systemic steroids.

Ichthyosis Vulgaris

Ichthyosis e Knee



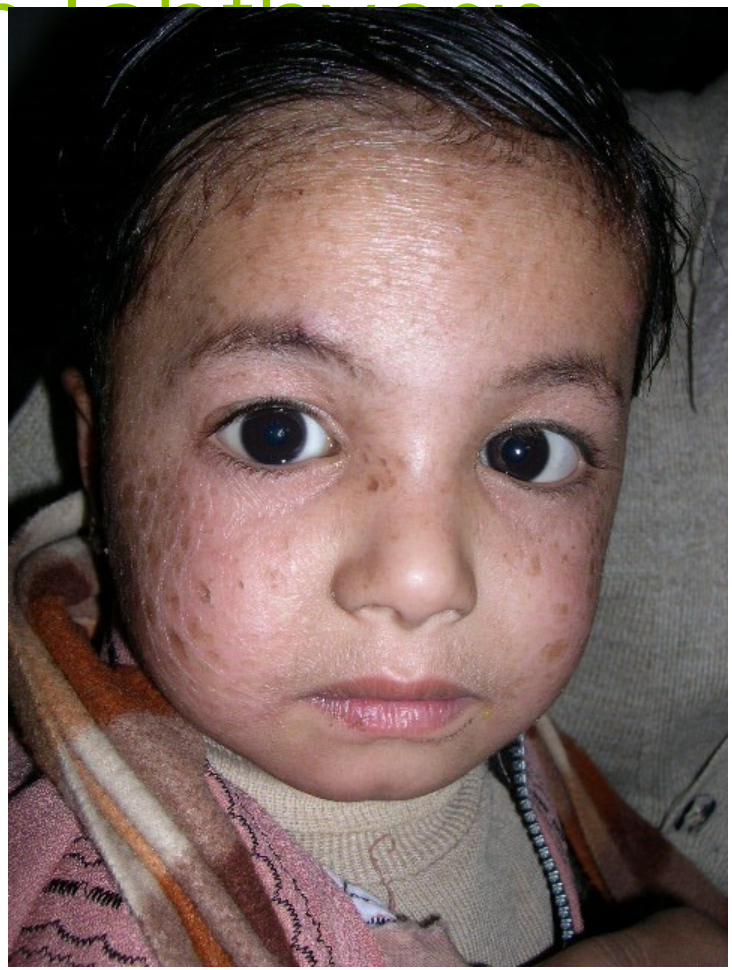
How does Ichthyosis Vulgaris occur?

- Icthyosis Vulgaris is the most common genetic skin disorder because only one of your parents needs to have the abnormality in the gene. The abnormality in the gene causes skin to regenerate (go through mitosis) faster than it sheds.

History of Ichthyosis Vulgaris

- Ichthyosis Vulgaris has been around for many years, and with so many people affected by it (1 out of every 250), it's hard to find out when and where the disorder originated.

Girl with



Passing to Other Generations…

- Icthyosis Vulgaris can be passed down to other generations quite easily. The gene is inherited in an autosomal dominate pattern, so the child only needs one mutated gene. This is a contributing factor to the great number of people that are living with the disorder.

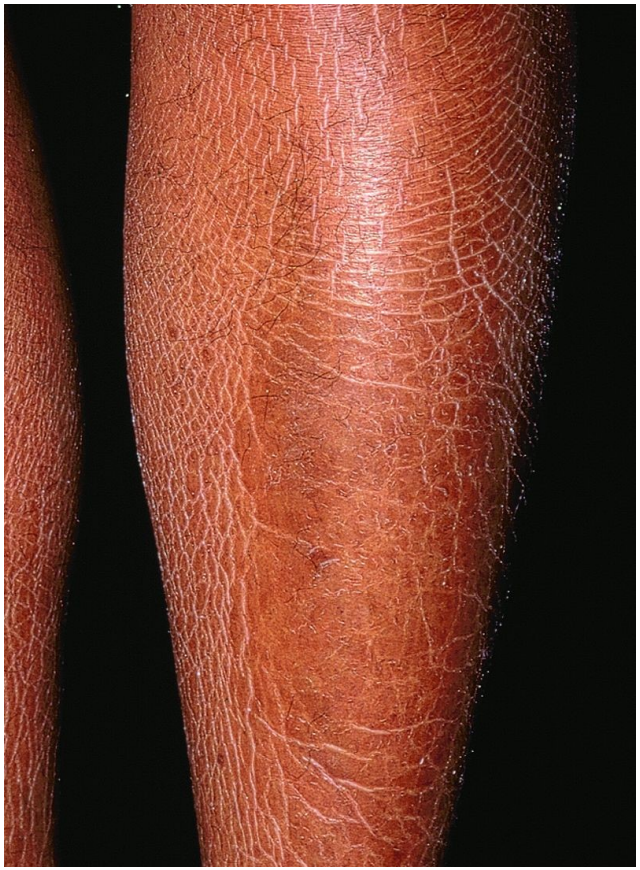
What kind of mutation is it?

- Ichthyosis Vulgaris is caused by a genetic mutation. However, ichthyosis that is not due to a genetic mutation is called acquired ichthyosis, and is extremely rare.

Phenotypic effects

- Effects of Ichthyosis Vulgaris include:
  - Dry Skin
  - Scaly Skin
  - Discoloration of Skin
  - Flaky Scalp
  - Painful deep cracks in palms and soles

Ichthyosis



How is it Diagnosed?

- Although many cases of Ichthyosis go unreported, it can be diagnosed through:
  - Skin examinations
  - Skin biospy
  - Researching famliy history (has anyone in your family had Ichthyosis before?)

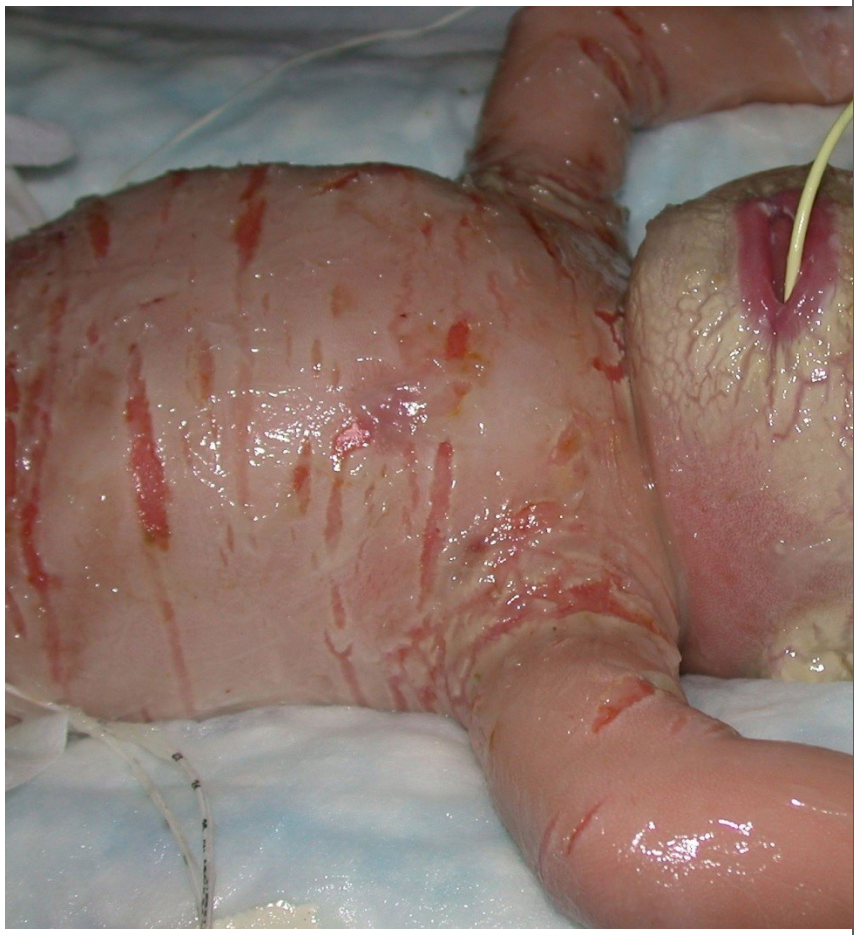
Treatment of Ichthyosis Vulgaris

- There is no known cure, but the following can aid in easing the pain of ichthyosis vulgaris:
  - Retinoids (from Vitamin-A)
  - Constant moisture to affected area

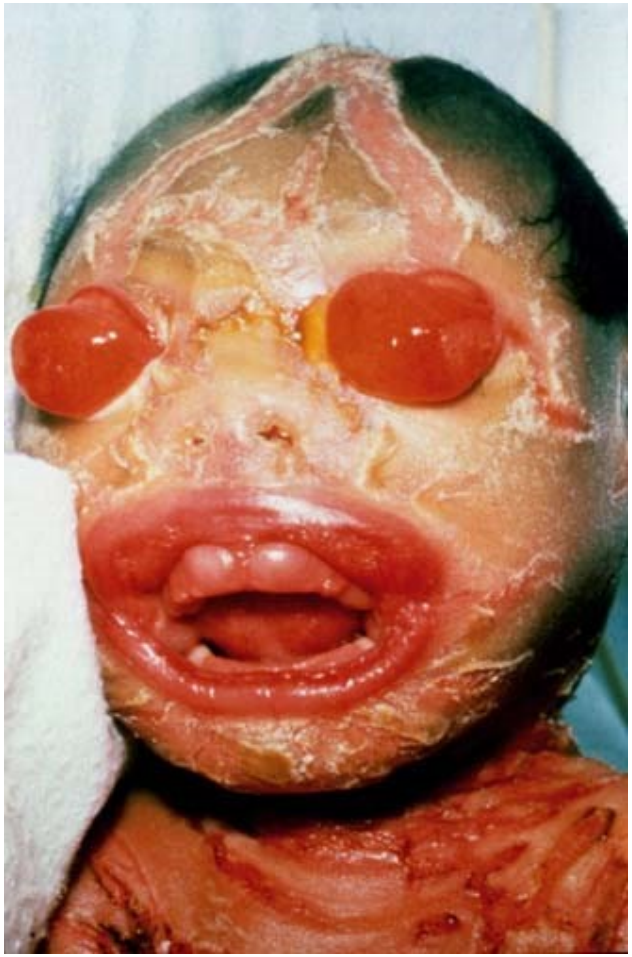
Harlequin Ichthyosis

Biological Basis

- Intro:
  - Harlequin Ichthyosis, most commonly known as harlequin's disease
  - The condition is characterized by a profound thickening of the external layer of the skin.

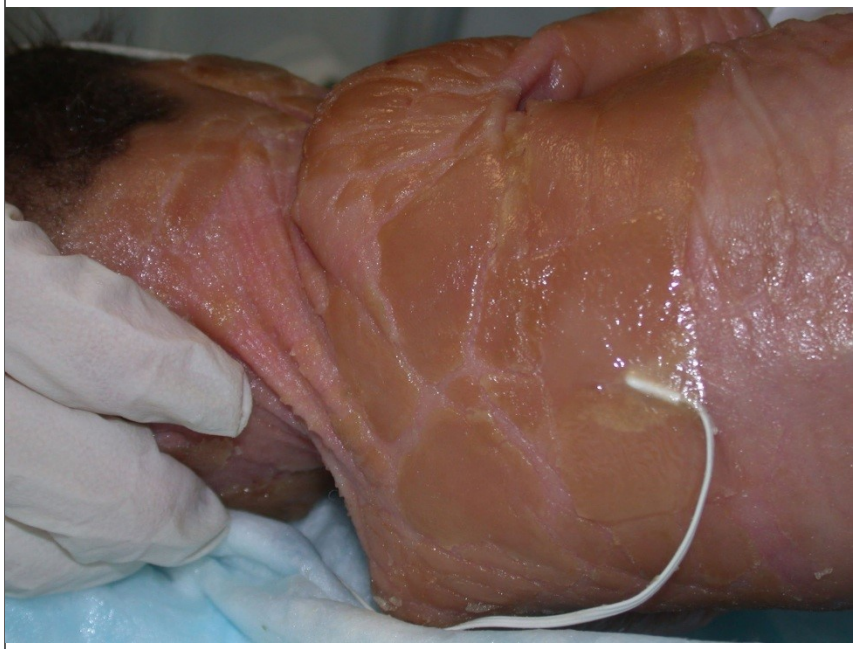


The Condition



- The babies are born with a massive hard shell like covering containing many deep fissures.
- The babies also have problems contracting their eyes, mouths, and appendages.
- They have a hard time moving because where there are folds on a normal baby there are cracks on an HI child.

Surviving HI



- With improved neonatal care more HI children have the chance of survival.
- The few survivors have skin that turns dark red and very scaly.
- The skin will eventually from a scaly layer and dry out so water retention and sun protection are concerns for the survivors.

Treatment

- The treatment of HI babies is quite extensive.
- The vitals of an HI baby must be constantly monitored.
- Their eyes must be constantly kept moist and shielded from the sun.
- The environment they live in must be constantly kept sterile to protect from infection.
- They must have a salt solution applied followed by a lubricant to facilitate the skin cells to fall off and to keep the cells moist.
- Antibacterial pills and creams are also used to fight infection.

Life of a Harlequin

- He competes in triathlons.
- To keep his skin from hardening, he covers himself in lotion 7 times per day.
- He has to consume 7,500 calories per day.
- He tube feeds himself pure protein every night while he sleeps to help him produce extra skin cells.