433 Teams

DERMATOLOGY

Lecture (11+12)

Blistering Diseases

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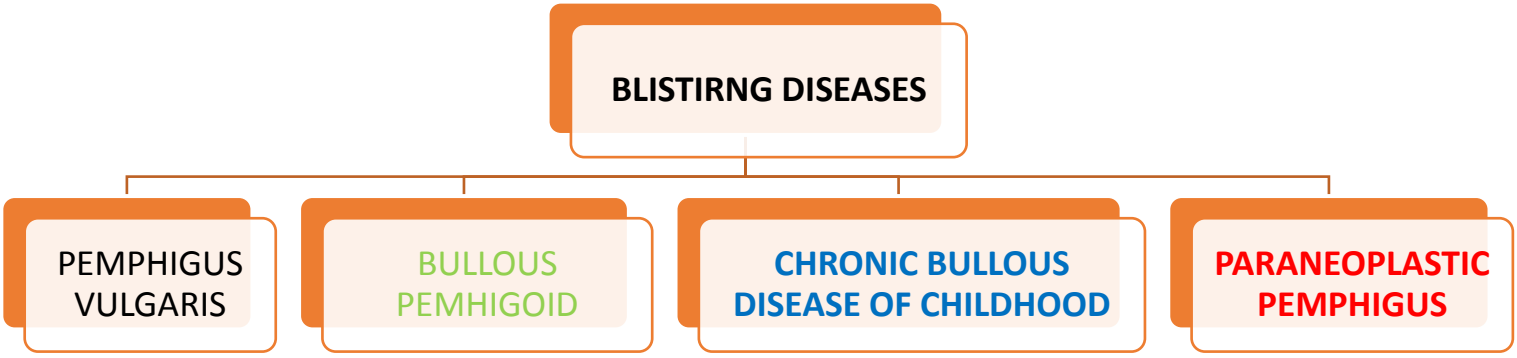
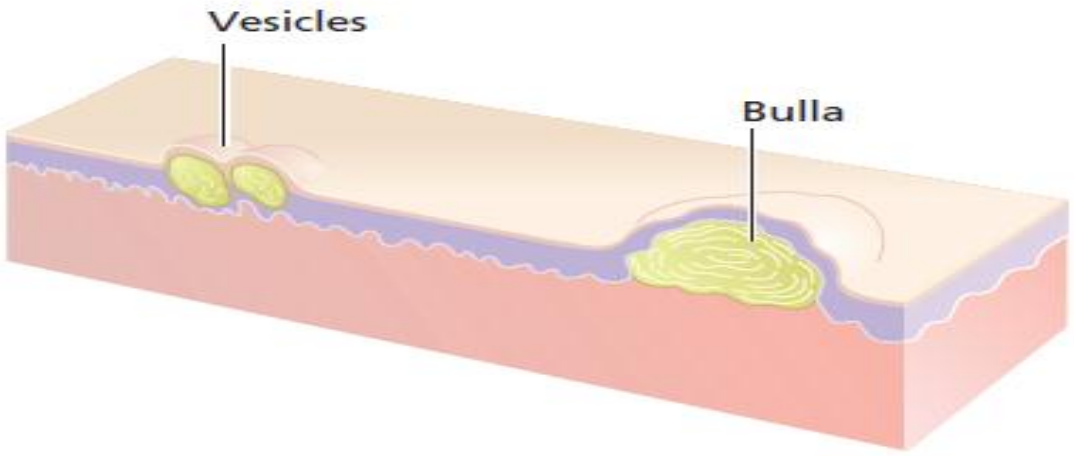
Objectives:

- To know the definition& classification of Blistering diseases
- To recognize the primary presentation of different types of main blistering diseases
- To understand the possible pathogenesis of the main types of blistering diseases
- To have an overview about managements lines of these diseases

Color index: **slides,** **important,** **432 notes**


BLISTIRNG DISEASES
PEMPHIGUS VULGARIS
BULLOUS PEMHIGOID
CHRONIC BULLOUS DISEASE OF CHILDHOOD
PARANEOPLASTIC PEMPHIGUS

Blistering Diseases



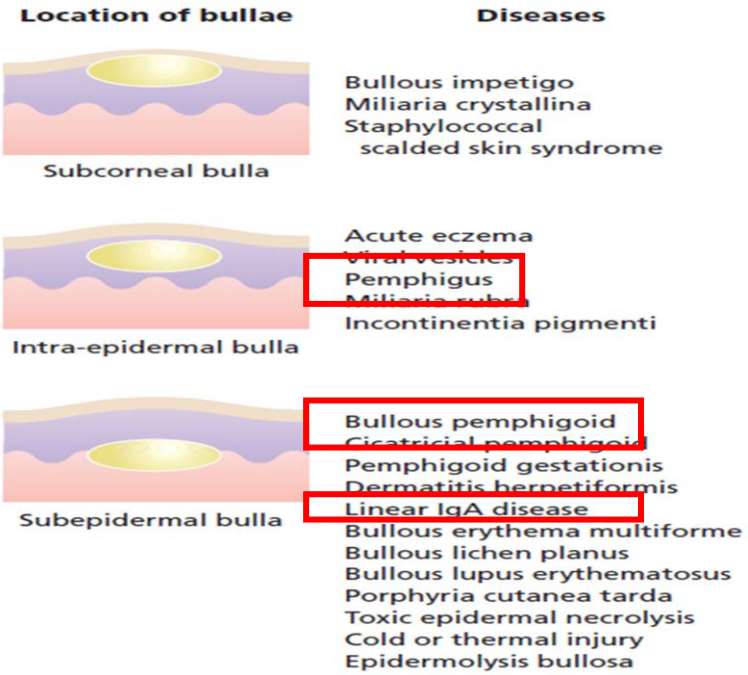
• Definition:

- Vesicles and bullae are raised lesions that contain fluid.
- A vesicle is less than 0.5 cm in diameter.
- A bulla is larger than 0.5 cm in diameter.

• Classification Of Vesiculobullous Diseases:

Intra Epidermal Blisters:
The lesion is formed within the Epidermis

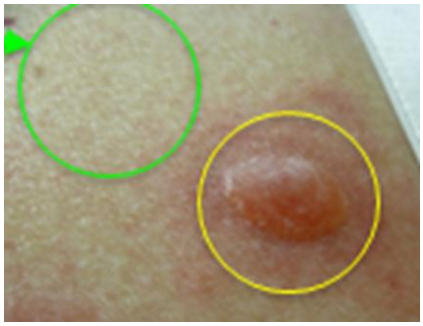
Sub Epidermal Blisters :
Lesions formed between the epidermis and the dermis



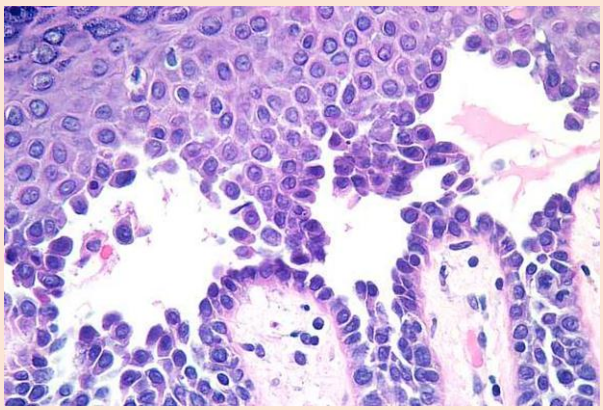
- Accurate pathological diagnosis requires 2 biopsies of a small newly formed lesion and perilesional skin for immunopathological studies.

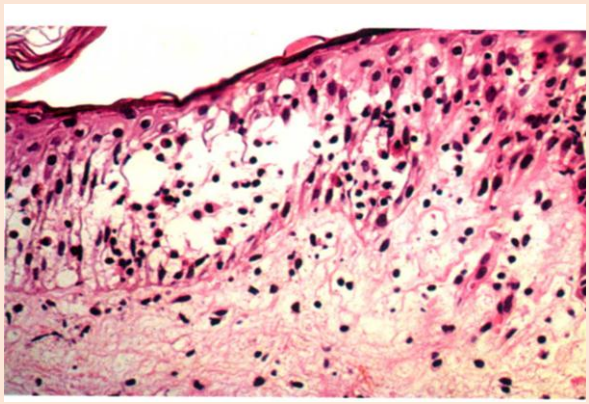
- Diagnostic tests

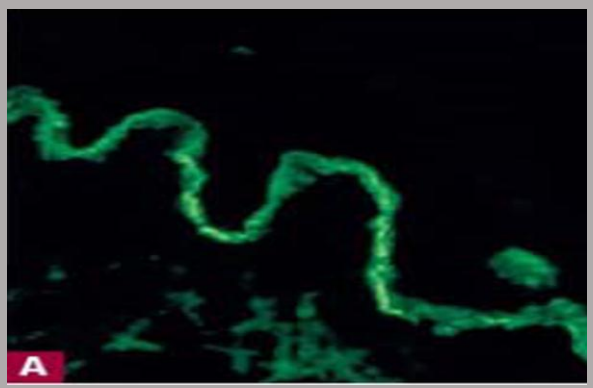
1. **Routine histology (Yellow circle)**
   Lesional sample –small bulla or edge of large one.
2. **Direct immunofluorescence (Green Circle)**
   Perilesional sample
3. **Indirect immunofluorescence (Blood)**
   Patient's serum is added to specific substrates that express antigen of interest.
4. **Electron microscopy.**



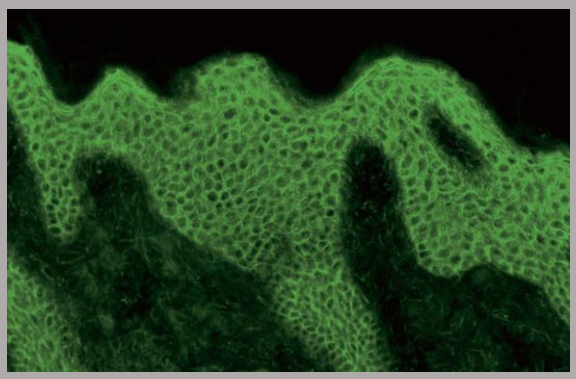
Routine Histology







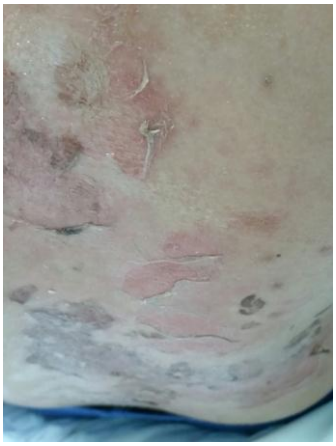
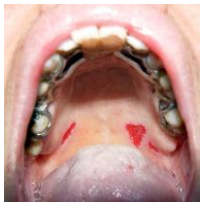
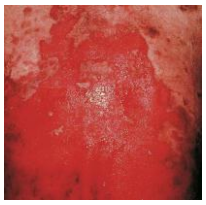
(Sub-Epidermal)



(Intra-Epidermal)

Pemphigus Vulgaris

- Pemphigus is a group characterized by blistering of the skin and mucous membranes.
- Age of Onset. 40-60 years also in children and young adults.
- Secondary infection and disturbance of fluid and electrolyte balance are common complications

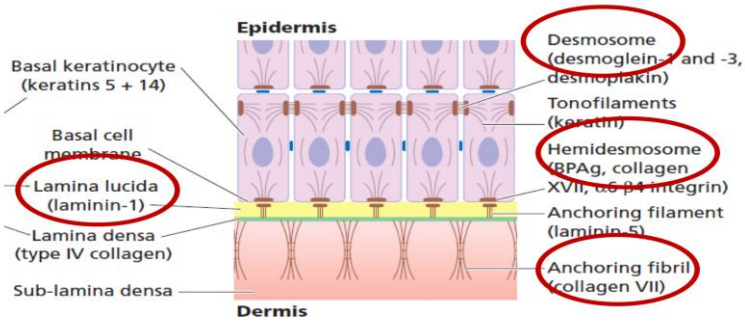




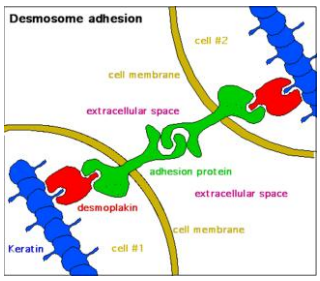
• Four sub-clinical varients:

- **Pemphigus Vulgaris**: is the most common Pemphigus variant, and the form usually responsible for oral lesions
- **Folacious, Vegetens, Erythematosus**

• Etiology and Pathogenesis:

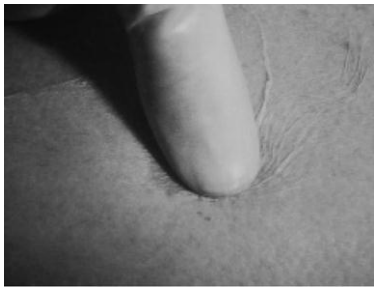
- An autoimmune against desmosomes in epidermis and mucosal surface.
- Loss of cell-to-cell adhesion in the epidermis (acantholysis).
- Occurs as a result of circulating antibodies of the IgG class, which bind to desmogleins, transmembrane glycoproteins in the desmosomes, members of the cadherin superfamily.
- In Pemphigus vulgaris , desmoglein 3 (in some, also desmoglein 1).
- In Pemphigus Foliaceus , desmoglein 1.
- Autoantibodies interfere with calcium-sensitive adhesion function and thus induce acantholysis.

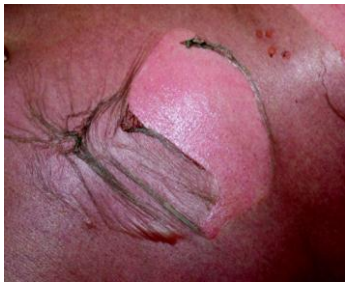




• Clinical Manifestation

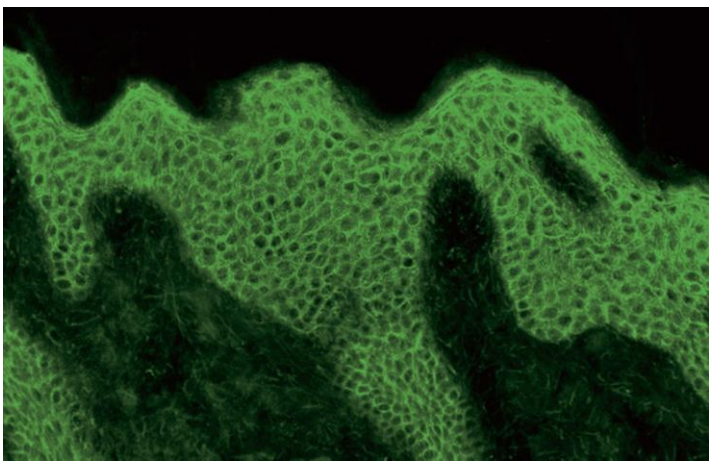
- Begins with erosions on mucous membrane then other skin areas.
- Very painful.
- +ve Nikolsky's sign (Twisting pressure on normal skin shears skin).



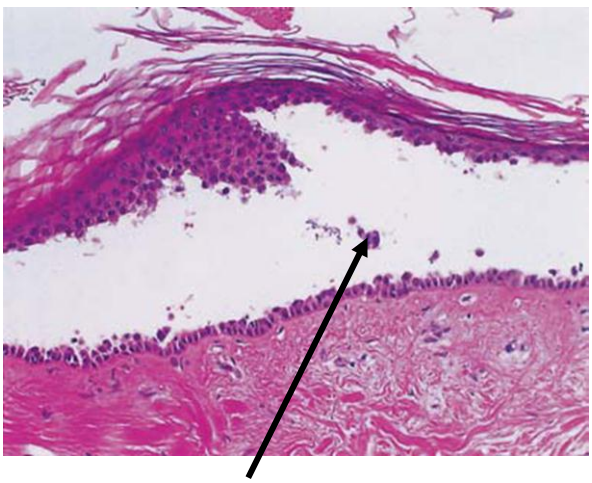


- **Pemphigus Foliaceus:** has no mucosal lesions and starts with scaly, crusted lesions on an erythematous base, initially in seborrheic areas(oily areas of their body, such as the face, upper chest and back).

Nikolsky's sign is almost always present in toxic epidermal necrolysis and is associated with pemphigus vulgaris.



Immunofluorescence (IgG and C3)



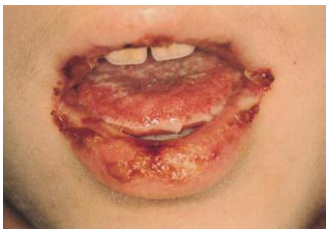
Acantholysis(floating cells)

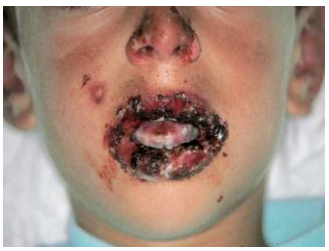
Management:

- High dose systemic steroids 60-100 mg of **prednisolone**.
- Immunosuppressive agent such as **azathioprine cyclophosphamide , Methotrexate or mycophenolate**
- Patient will probably have to remain on systemic steroids for long time.
- **Antibiotics**; to treat superinfection
- Biological: **Rituximab** (IV 86% free of disease after 3 y) and **IVIG (**intravenous immunoglobulin).
- If Mild: class III/IV corticosteroid creme / intralesional injection.
- If Sever: prednisolone 80 mg & taper in 5 months, immunosuppressive, biological.

- ## Drug-induced PV
  - **Drugs can induce PV**
  - **Drugs reported most significantly in association with PV are;**
  - **Penicillamine**
  - **Captopril**

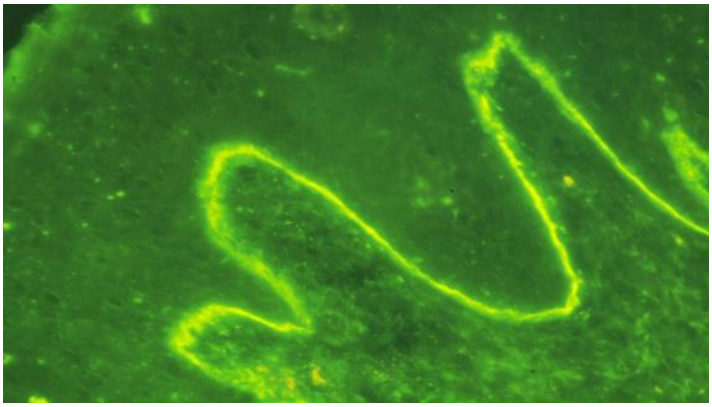
Paraneoplastic Pemphigus

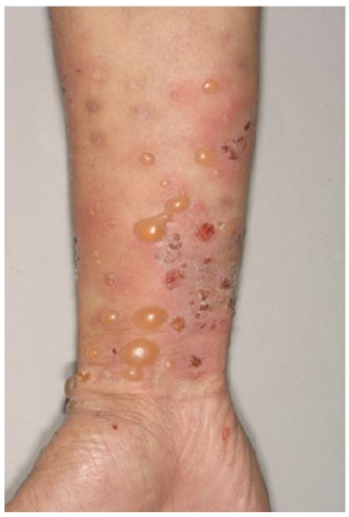




- The least common and most severe type of pemphigus is **Paraneoplastic Pemphigus** (PNP).
- This disorder is a complication of cancer,
- Usually lymphoma and Castleman's disease. It may precede the diagnosis of the tumor. Painful sores appear on the mouth, lips, and the esophagus.
- Complete removal and/or cure of the tumor may improve the skin disease,
- Both Intra epidermal and sub epidermal blister.
- Autoantibody (IgG , IgA , C3)

Bullous pemphigoid



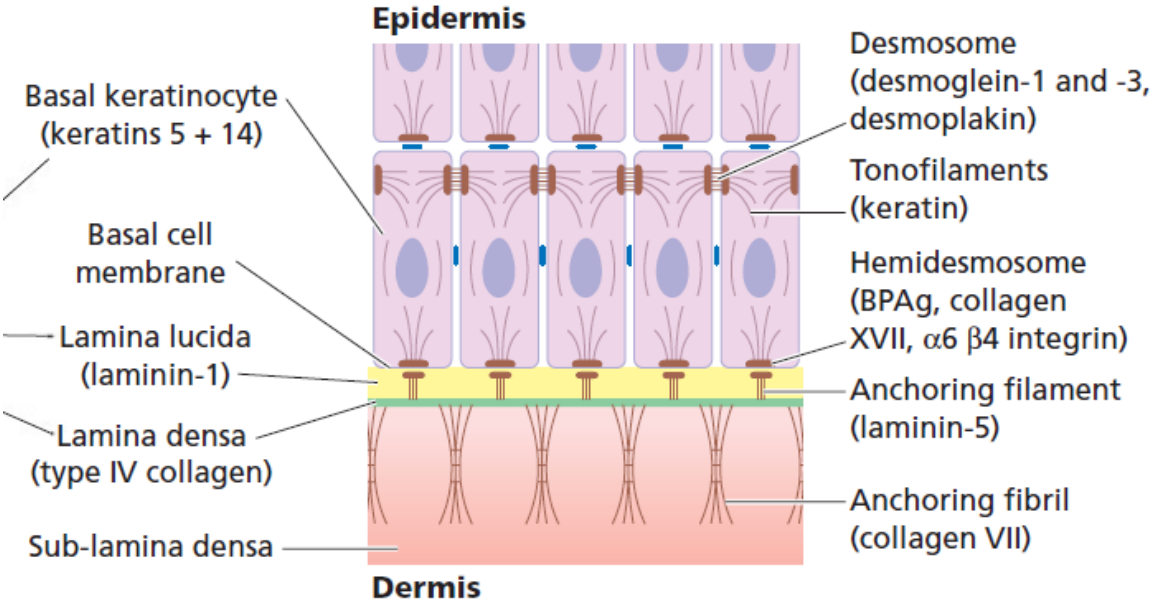


Characterized by **large, tense, intact blisters** on an erythematous base. it can errupt to form erosions

- Mainly in older age group.
- more than 60 y.
- The prognosis is usually good.
- **Linear band on immunofluorescence.**
- **Antigens identified are in hemidesmosomes.**

Clinical features :

- Elderly patents.
- Large tense blisters on upper arms and thighs.
- Eczematous base .
- Itch rather than pain.
- Oral lesions are less frequent than pemphigus.



Pathology :

• Sub epidermal between epidermis and dermis the epidermis forms the roof of the blister.

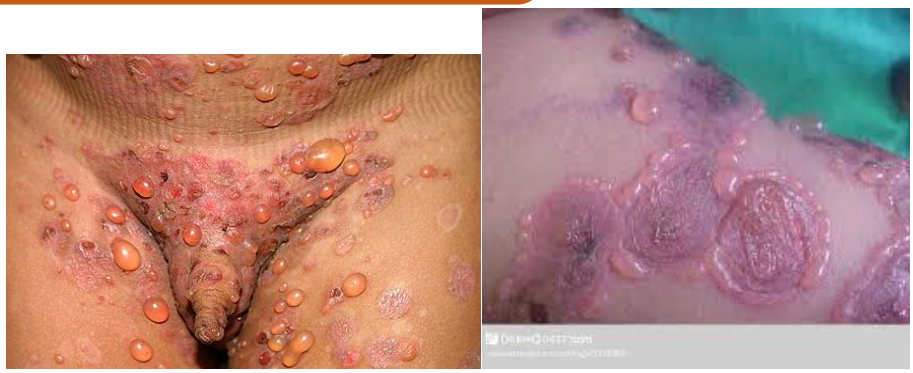
• Antigens identified are in **hemidesmosomes**.

• Immunoglobulin and complement are deposited in the lamina lucida of the basement membrane in a linear band.

Treatment :

- Topical Steroids
- Severe pemphigoid :Systemic steroids , but unlike pemphigus, usually possible to discontinue Rx.
- The addition of either azathioprine enable the oral steroid dose to be reduced more rapidly.
- Milder may also respond very well to potent or moderately potent topical steroids alone.
- Other treatment: Antibiotics like tetracyclines group

CHRONIC BULLOUS Disease OF CHILDHOOD



• Chronic blistering dis. which occur in children, usually starts before the age of 5yrs

• Small and large blisters appears predominantly on the lower trunk, genital area, and thighs

• May also affects the scalp and around the mouth

• New blisters form around healing old blisters forming **a CLUSTER OF JEWELS**

Course: is chronic and spontaneous remission usually occurs after an average of 3-4 yrs

• **IgA autoantibodies** binds to proteins at dermo epidermal junction as linear pattern like the pattern

of bullous pemphigoid

CLINICAL FEATURES :

• Circular clusters of large intact blisters and can erupt to form erosions
• It involves the perioral area, lower trunk, inner thighs and genitalia
• Blistering may spread all over the body

INVESTIGATION :

• Skin Biopsy will show **subepidermal splits**
• Direct IF reveals **IgA** along the BM of the epidermis in a **linear pattern**

TREATMENTS :

• Oral dapsone

(may cause hemolaysis , check G6PD or methemoglobinemia)

• Sulphonamides and immunosupressants
• Erythromycine
• Flucloxacillin

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