DERMATOLOGY

Blistering Diseases

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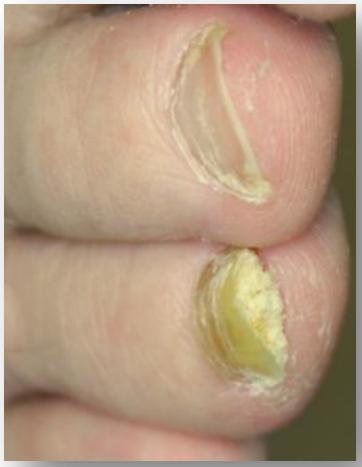
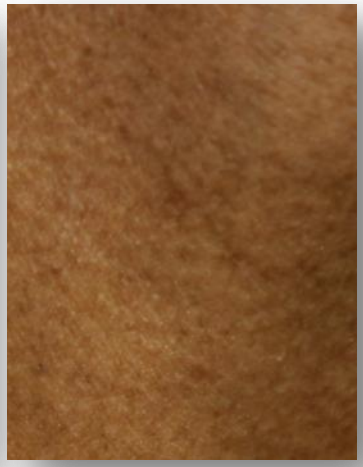
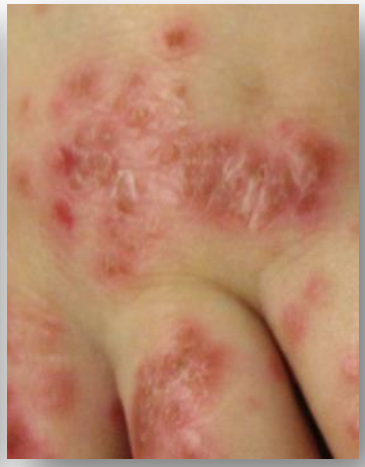
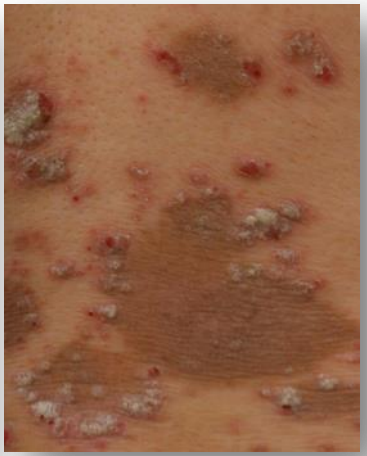
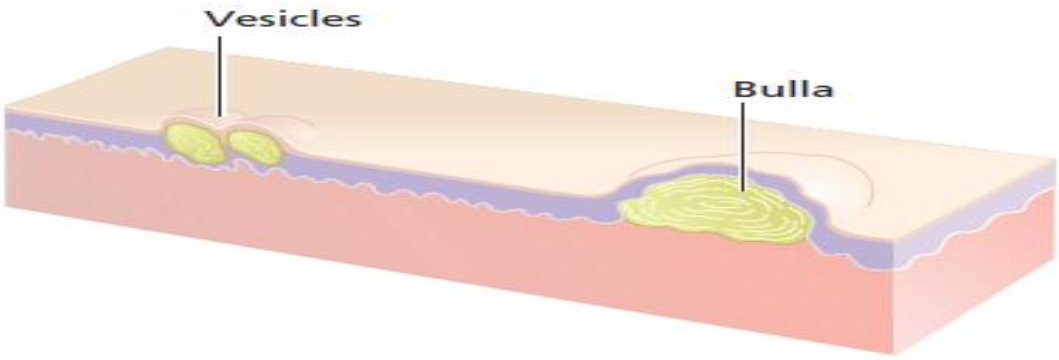





FROM SLIDES DOCTOR'S NOTES TEAM'S NOTES FROM BOOK IMPORTANT

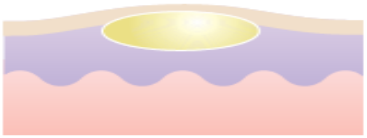
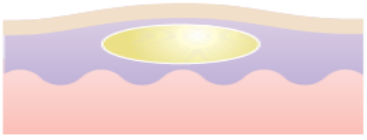
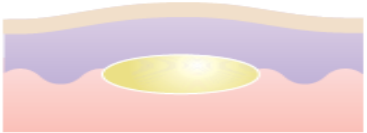
Blistering Diseases "Vesico-Bollous Diseases"

- 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 management lines of these diseases.

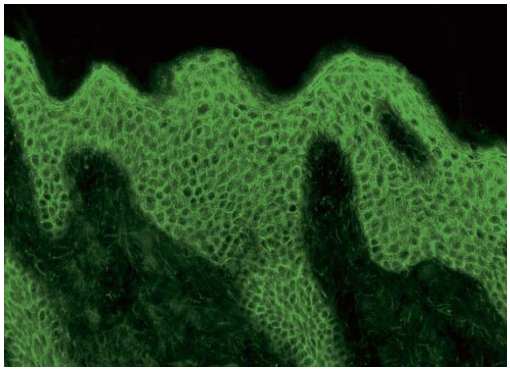
- Definition:
  - Vesicles and bullae are raised lesions that contain fluid.
  - A vesicle is less than 0.5 cm in diameter.
  - A bulla is more than 0.5 cm in diameter.



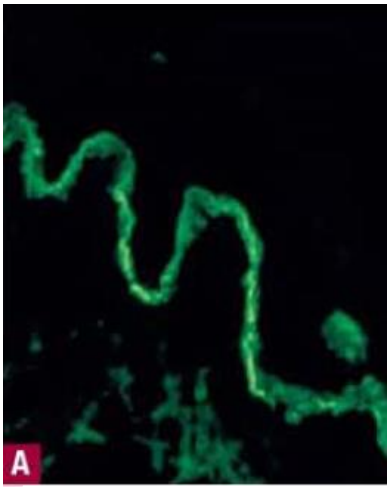
- Classification:
  - Intra-epidermal blisters: the lesion is within the epidermis.
  - Sub-epidermal blisters: the lesion is between the epidermis & dermis.

Location of bullae	Diseases
 Subcorneal bulla	Bullous impetigo Miliaria crystallina Staphylococcal scalded skin syndrome
 Intra-epidermal bulla	Acute eczema Viral vesicles Pemphigus Miliaria rubra Incontinentia pigmenti
 Subepidermal bulla	Bullous pemphigoid Cicatricial pemphigoid Pemphigoid gestationis Dermatitis herpetiformis Linear IgA disease Bullous erythema multiforme Bullous lichen planus Bullous lupus erythematosus Porphyria cutanea tarda Toxic epidermal necrolysis Cold or thermal injury Epidermolysis bullosa

- Diagnosis:
  - Routine histology: lesion sample (small bulla or edge of large one).
  - Direct immunofluorescence: perilesional sample.
  - Indirect immunofluorescence: Patients serum is added to specific substrates that express antigen of interest.
  - Electron microscopy.



intra-epidermal



Sub-epidermal

Pemphigus Vulgaris:

- "pemphigus" is a group of diseases characterized by blistering of the skin & mucus membranes. (Vulgaris, Folacious, Vegetens, Erythematosus)
- Pemphigus vulgaris is the most common pemphigus variant and is the one usually responsible for oral ulcers.
- Middle age, 40-60 years.
- Auto-antibodies directed against epidermal & mucosal desmosomes (IgG & C3)
- Blisters are flaccid & thin, and usually easily blanch, and that's why the usual presentation is EROSIONS.
- Nicholsky sign: twisting pressure on normal skin shears skin.
- Very painful.
- Secondary infection & disturbance of fluid and electrolyte balance are common complications.
- Acantholysis "Floating cells" is a characteristic feature in routine histology. (Acantholysis is the loss of intercellular connections, such as desmosomes, resulting in loss of cohesion between keratinocytes)
- FATAL in all cases if not treated.

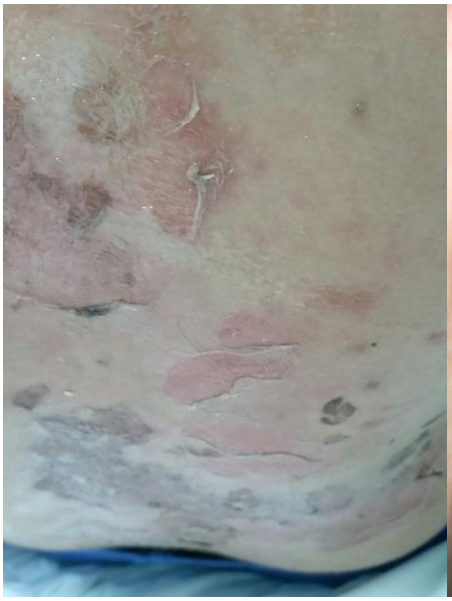
Drug-induced pemphigus vulgaris: penicillamine & captopril.

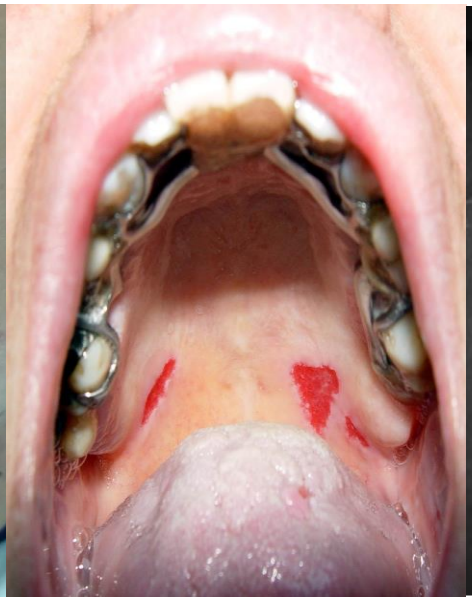
Investigations:

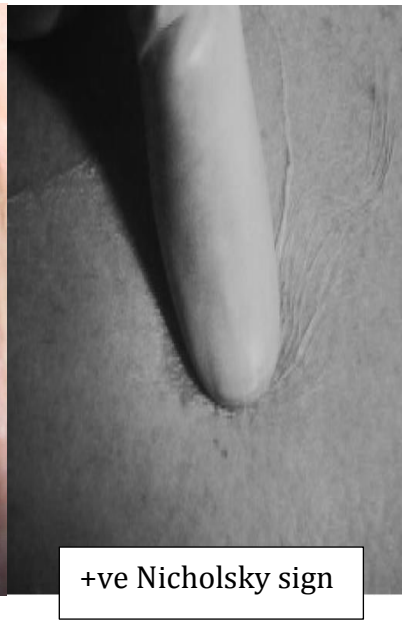
o Biopsy shows that vesicles are intra-epidermal with rounded keratinocytes floating freely within the blister cavity (acantholysis)
o Direct IF of adjacent normal skin shows intercellular epidermal deposits of IgG and C3

Treatment:

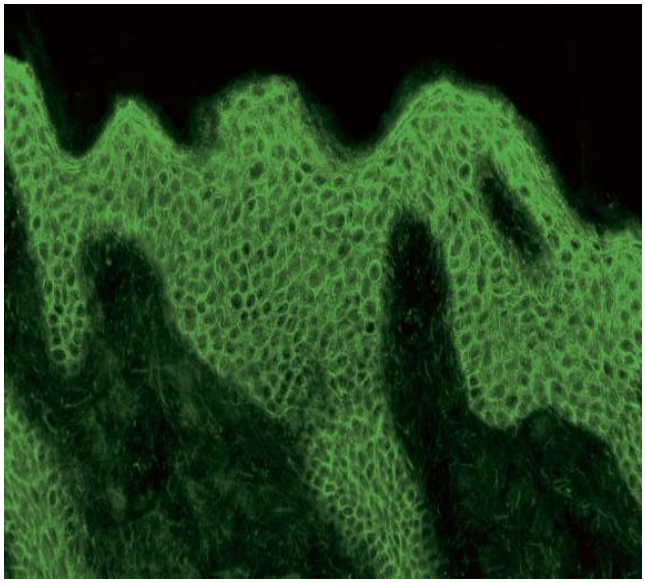
- High dose systemic steroids (prednisolone 60-100 mg)
- Immunosuppressive therapy (azathioprine, cyclophosphamide, methotrexate, or mycophenolate).
- Topical therapy is mainly symptomatic.
- Antibiotics
- Biologic Rx: Rituximab or IVIG.
- IF MILD: class III/IV corticosteroid creme / intralesional injection.
- IF SEVER: prednisolone 80 mg & taper in 5 months, immunosuppressive, biological.







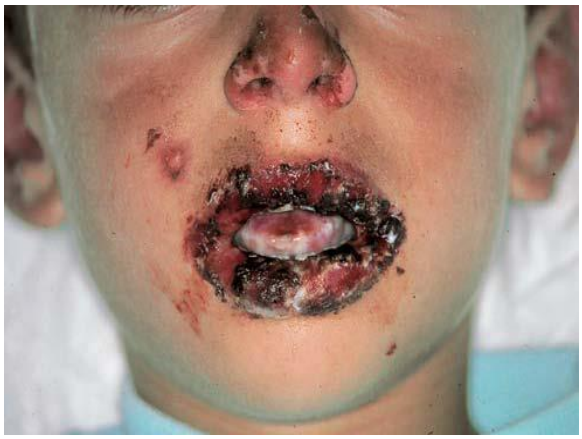
+ve Nicholsky sign



intra-epidermal

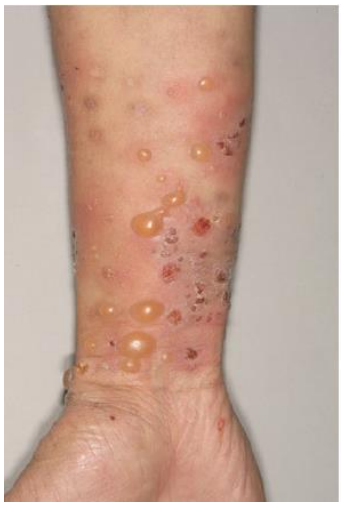
Paraneoplastic pemphigus:

- The least common type of pemphigus and is usually a complication of cancer (lymphoma & castleman's disease).
- Painful sores appear on the mouth, lips, and the esophagus.
- Complete removal or cure of the tumor may improve the skin disease.
- Blisters are both intra & sub-epidermal.
- Autoantibodies (IgG, IgA, & C3).



Bullous pemphigoid

- Mainly in old age group, 60 years.
- Prognosis is usually good.
- Characterized by LARGE, TENSE, INTACT blisters on an erythematous base, it can erupt to form erosions.
- On upper arms and thighs
- Itch rather than pain.
- Sub-epidermal blisters, autoantibodies (IgG) affecting hemidesmosomes.
- Linear band on immunofluorescence.
- Antigens identified are in hemidesmosomes.



- Negative Nikolsky's sign
- Mucosal lesions unusual, patients well
- Skin biopsy shows a deeper blister (than in pemphigus) owing to a sub-epidermal split through the BM

- PROGNOSIS: Good; relatively benign

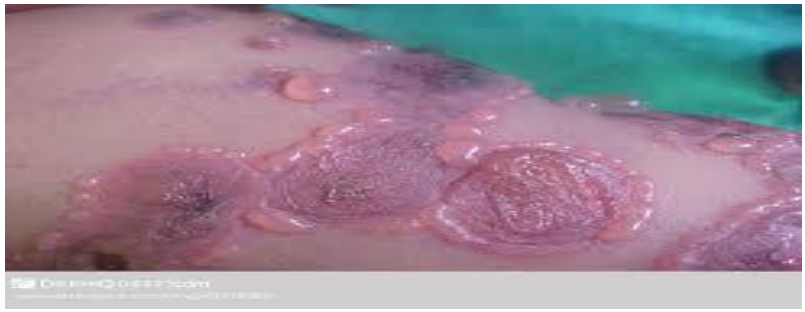
Treatment:

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

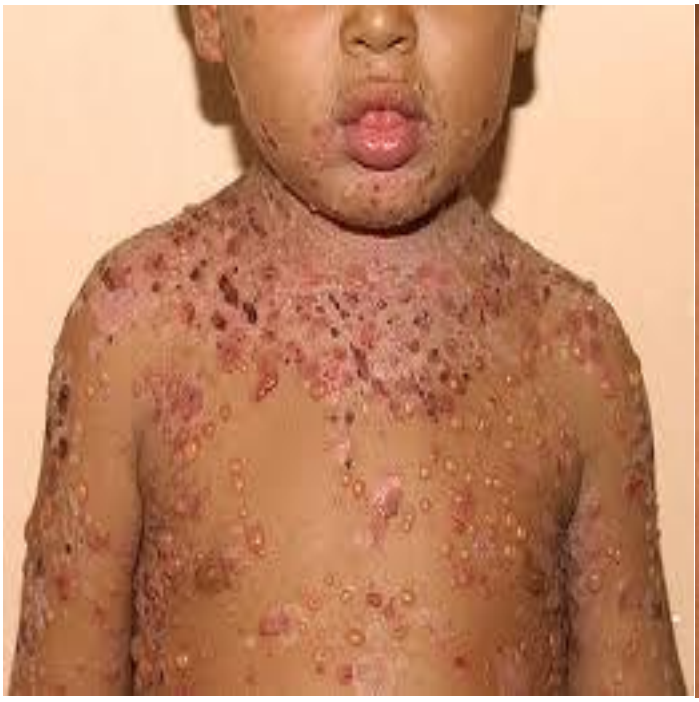
Chronic bullous disease of childhood

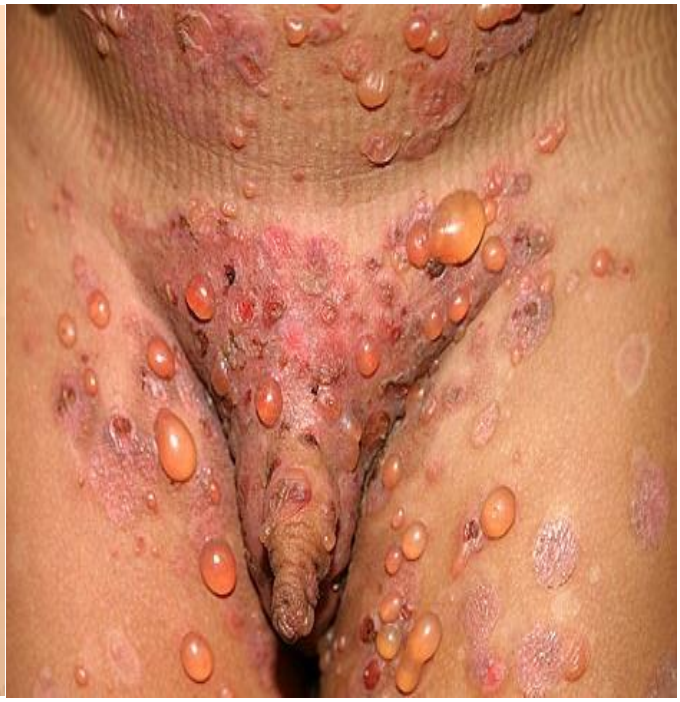
- Chronic blistering disease in children younger than 5 years, there is spontaneous remission occurs after an average of 3-4 years.
- Small and large TENSE blisters appears predominantly on the lower trunk, genital area, and thighs
- May also affects the scalp and around the mouth

- New blisters form around healing old blisters forming a CLUSTER OF JEWELS.



- IgA autoantibodies bind to proteins at dermo-epidermal junction as linear pattern like the pattern of bullous pemphigoid.
- Blistering may spread all over the body.
- Skin Biopsy will show sub-epidermal splits.
- Direct IF reveals IgA along the BM of the epidermis in a linear pattern.
- <u>Treatment:</u>
  - Oral dapsone, may cause hemolysis (check for G6PD) or methemoglobinemia.
  - Sulfonamides & immunosuppressive.
  - Erythromycin, Flucloxacillin.





Questions

1 . Fifty five year old teacher presented with sever oral mucusal ulceration. Moreover, he developed numerous flacid blisters which ruptured easily leaving only erosions. The most likely diagnosis is:

a. Bullous Pemphigoid
b. Pemphigus Vulgaris
c. Pyoderma gangrenosum
d. Stevens-Johnson syndrome
e. Toxic Epidermal Necrolysis

2. The mortality in Pemphigus Vulgaris is due to:

a. Inadequate food intake
b. Renal failure
c. Secondary Infection
d. Squamous cell carcinoma
e. None of the above

3. keratinocytes detach from their neighbors & float free in the blister a process called:

a. Acantholysis.
b. CLUSTER OF JEWELS.
c. Sub-epidermal splits
d. Acanthosis

4. Patient diagnosed with blister disease, the IF showed aggregation of IgG along with the basement membrane zone, what's the Most likely DX?

a. Pumphigus vulgaris.
b. Bollus Pumphigoid
c. Dermatitis herpetiformis
d. Chronic bulls disease of childhood

ANS :

1 – B
2 – C
3 – A
4 – B