	Notes	Clinical Features	Diagnosis	Management
Pemphigus Vulgaris	- Rare, lethal, autoimmune blistering disease. - Age - sixth decade	**Presentation:** Flaccid blisters that break and expand to form large erosions with weeping painful erosions Starts with **mucosal membrane involvement** [Painful mouth erosions]	**Clinically:** Flaccid blisters Nikolosky's sign +ve Asboe - Hansen sign or (bulla spread sign) +ve **Biopsy** [light microscope]: Vesicles are intra-epidermal with rounded keratinocytes floating freely within the blister cavity [acantholysis] **DIF**: IgG, C3 in intercellular space of epidermis. **IF:** S.Abs to epidermal cell surface antigens - Titer reflects disease Activity.	Supportive care Corticosteroids: High dose > 1st choice Adjuvant Immunosuppressive therapy: (Azathioprine, mycophenolate, mofetil) Plasmapheresis / IVIG Biological: Anti CD-20, Rituximab Extracorporeal photochemotherapy
Pemphigoid Group of Diseases: Autoimmune sub-epidermal blistering diseases with circulating IgG and BMZ-bound IgG ABS α C3.	**Bullous Pemphigoid:** Sub-epidermal blister Seen in elderly Relatively benign untreated pemphigoid may remain localized and undergo spontaneous remission	**Presentation:** Large tense bullae on erythematous plaques or normal appearing skin. Pruritic **Location:** Lower part of abdomen, groin, flexors of arms & legs. No or only mild involvement of mucosa	**Clinically:** Tens blisters Local Erythema, urticaria & pruritus -ve Nikolsky's sign **Labs:** CBC [Esinophilia 50% | IgE 70%] **Biopsy:** Deeper blister owing to a sub-epidermal split through the BM **DIF:** linear band of IgG and C3 along dermal-epidermal junction. **IF:** IgG antibodies against BMZ [doesn't correlate with the disease activity]	Topical steroids [localized] Systemic steroid [sever] Antibiotics: Tetracycline, Minocin Dapsone Immuno Suppressive (Azathioprine, Mycophenolate, Mofetil)
	Pemphigoid Gestationis: Sub-epidermal blister Starts in the 2nd & 3rd trimester Flares at post partum , OCP	**Presentation:** Erythematous urticarial Plaques, alone or with papules, vesicles, blisters & erosions. **Location:** Most on abdomen, proximal extremities. Intense pruritus	**DIF:** C3, IgG at BMZ H.G. Factor in Patient's Sera	Steroids [Topical or Systemic]
	Dermatitis Herpetiformis: Associated with gluten sensitive enteropathy	**Presentation:** Small grouped papules, Vesicles, Blisters, Erosions on erythematous plaques. **Location:** Extensors of large joints	**Clinically**: Pruritic tense blisters on extensors **Biopsy [Microscope]:** Sub epidermal blister, prominent neutrophil infiltration **DIF:** granular IgA in dermal papillae	Gluten free diet Dapsone - 1st line of treatment Sulfa pyridine Tetracycline and Nicotinamide

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Notes From Clinical Sessions:

Pemphigus Vulgaris:

Affects mouth mucosa
Nail changes: Chronic infection [candida] + periungual edema
Lymph node enlargement [submandibular, submental, cerviacal, supraclavicular & inguinal]
Mouth erosions seen in: [TEN, SJS, pemphigus vulgaris & para-neoplastic pemphigoid]

Investigations:

Biopsy:

H&E:

Pemphigoid Vulgaris → epidermal [suprabasal]
Pemphigoid Group →Sub-epidermal [sub-basal]
Drug induced → ↑ Esinophills

DIF:

Pemphigus Vulgaris → IgG between keratocytes
Pemphigoid Group → IgG & C3 on the basal membrane.

Tzank Smear [Only if needed]

Others:

Nail swab → Fungal bacterial
Mouth swab → R/o infection
If associated with fever → blood work up
FBS + CBC + U&E

NB: if mild → topical steroids | if mucous membrane → mouth wash + lidocaine