

#MADEEASY



**PEMPHIGUS VULGARIS AND
PEMPHIGUS FOLIACEUS**





PEMPHIGUS VULGARIS AND PEMPHIGUS FOLIACEUS

- *Pemphigus vulgaris and pemphigus foliaceus are autoimmune blistering disorders characterized by the presence of autoantibodies against desmosomes.*
- *Despite their similarities, these two conditions have distinct clinical, histological, and immunopathological features.*

Can you explain with an easy example?

- *Think of a brick wall where the bricks (keratinocytes) are held together by cement (desmosomes).*
- *Pemphigus Vulgaris: Here, the cement between the bricks at the foundation level (lower epidermis) separates, causing the wall to collapse (blisters and erosions)*
- *Pemphigus Foliaceus: This time the cement separates at the surface level (superficial epidermis), causing only the outer layer of bricks to peel away (crusted erosions).*

Pemphigus Vulgaris

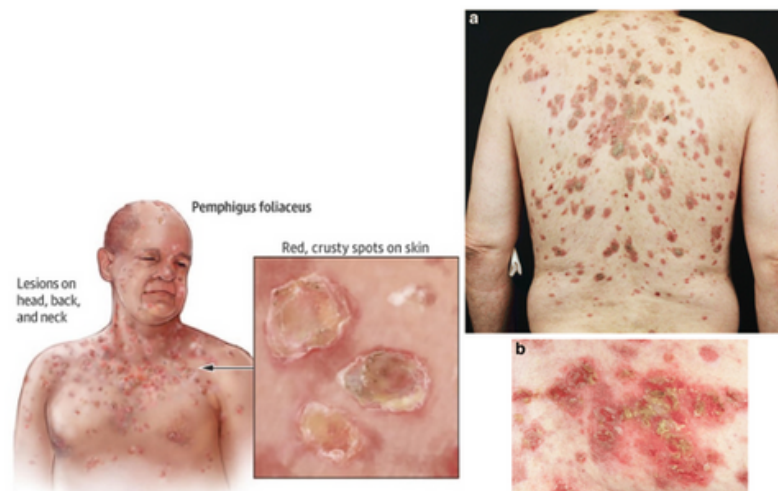




PEMPHIGUS VULGARIS

- **Clinical Features**
- **Intraepidermal blister, often in the lower epidermis.**
- **Lesion Appearance: Flaccid blisters that easily rupture, leading to painful erosions.**
- **Distribution: Commonly affects mucous membranes (oral cavity, esophagus) and skin.**
- **Nikolsky Sign: Positive** (slight rubbing of the skin results in exfoliation of the outermost layer).
- **Histopathology**
- **Acantholysis: Loss of keratinocyte adhesion, resulting in separation of the epidermal cells, particularly above the basal layer.**
- **Row of Tombstones: Appearance of the basal cells that remain attached to the basement membrane while the upper layers are detached.**
- **Immunopathology**
- **Direct Immunofluorescence (DIF): IgG and C3 deposition in the intercellular spaces of the epidermis.**
- **Autoantibodies: Target desmoglein 3 (and sometimes desmoglein 1).**
- **Management**
- **High-dose systemic corticosteroid**
- **immunosuppressants (azathioprine, mycophenolate mofetil)**

Pemphigus Foliaceus

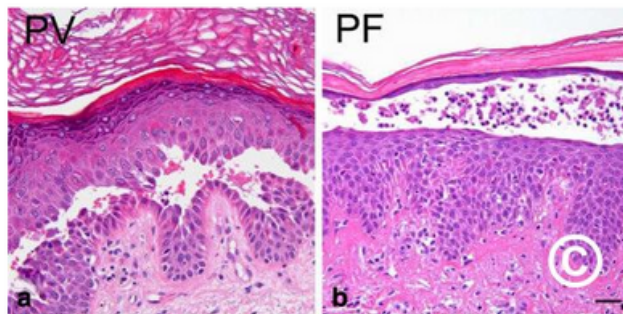




PEMPHIGUS FOLIACEUS

- **Clinical Features**
- **Intraepidermal, in the superficial layers of the epidermis.**
- **Lesion Appearance: Superficial, crusted erosions rather than intact blisters.**
- **Distribution: Typically affects the skin, sparing the mucous membranes.**
- **Nikolsky Sign: Positive**
- **Histopathology**
- **Acantholysis: Separation of keratinocytes occurs in the upper layers of the epidermis (subcorneal).**
- **Less Prominent basal layer changes**
- **Immunopathology**
- **Direct Immunofluorescence (DIF): IgG and C3 deposition in the intercellular spaces of the superficial epidermis.**
- **Autoantibodies: Predominantly target desmoglein 1.**
- **Management**
- **Systemic corticosteroids**
- **Topical steroids for localized lesions**
- **Immunosuppressants (similar to pemphigus vulgaris)**

Difference between pemphigus vulgaris and pemphigus foliaceus



Feature	Pemphigus Vulgaris	Pemphigus Foliaceus
Blister Location	Lower epidermis	Superficial epidermis
Lesion Appearance	Flaccid blisters, painful erosions	Superficial, crusted erosions
Distribution	Mucous membranes and skin	Skin, sparing mucous membranes
Nikolsky Sign	Positive	Positive
Histopathology	Acantholysis above the basal layer, row of tombstones	Acantholysis in the superficial epidermis
Direct Immunofluorescence	IgG and C3 in intercellular spaces of epidermis	IgG and C3 in intercellular spaces of superficial epidermis
Autoantibodies	Desmoglein 3 (and sometimes desmoglein 1)	Desmoglein 1
Simple Example	Like the foundation of a wall breaking apart	Like the surface of a wall peeling off

