Derm Path IDM

TOPIC - Pemphigus vulgaris

PRESENTER - Dr. Afrin

summary of Pemphigus Vulgaris:

Pemphigus vulgaris is a chronic, potentially life-threatening autoimmune blistering disorder that primarily affects middle-aged and older individuals. It is characterized by the formation of flaccid bullae and painful erosions on the skin and mucous membranes, particularly the oral mucosa, which is involved in nearly all cases and often precedes skin lesions. These bullae rupture easily, resulting in widespread denuded areas, and the Nikolsky sign (shearing of the epidermis with lateral pressure) is typically positive. The pathogenesis involves IgG autoantibodies directed against desmoglein 3 and sometimes desmoglein 1, which are cadherin-type adhesion molecules in desmosomes. This antibody-mediated disruption leads to suprabasal acantholysis, where keratinocytes lose cohesion, forming intraepidermal clefts. Histologically, this appears as separation just above the basal layer with “tombstone” basal cells. Direct immunofluorescence (DIF) reveals a characteristic intercellular (fishnet or lace-like) deposition of IgG and C3 throughout the epidermis. Indirect immunofluorescence (IIF) demonstrates circulating autoantibodies against epithelial cells. The disease must be differentiated from other blistering conditions such as bullous pemphigoid, Hailey-Hailey disease, and IgA pemphigus. Management typically involves systemic corticosteroids and immunosuppressive agents such as azathioprine or mycophenolate mofetil, with rituximab (an anti-CD20 monoclonal antibody) being increasingly used for resistant cases. Early diagnosis and aggressive immunosuppressive therapy are essential for reducing mortality and preventing complications such as sepsis or fluid imbalance.