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Commentary

# Symptoms and Autoimmune Mechanisms of Pemphigus Vulgaris

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## DESCRIPTION

Pemphigus is a blistering autoimmune disorder which affects the skin and mucous membranes. Pemphigus vulgaris is the most common type of disorder. It is the chronic disorder of the skin and oral mucous membrane which is characterized by blister or bullae formation. It occurs when antibodies attack desmoglein 3 (a protein that is present in the epidermal layer of the skin).

### Signs and symptoms of pemphigus vulgaris

**Blisters:** Sudden development of blisters and spreads widely which would be life threatening. These blisters make the healthy person to be extremely sick, incredibly tired, and cause pain. The blisters usually begin in one area and spread to other organs of the body. The fluid oozes out of ruptured blisters that leave painful sores. The sores are non-itchy but painful. The sores heal slowly by leaving dark spots which fade by their own. Sometimes, the lesions might not heal in persistent.

**Mouth and throat:** In almost 50-70% of the cases, the development of mouth sores begins prior to the appearance of blisters on the skin. The mouth sores also begin as blisters and burst which leads to formation of painful sores thus cause difficulty in intake of food. These sores even develop in throat and induce dysphagia. Often, these blisters spread to lips and to skin. In case of pemphigus vulgaris the sores develop repeatedly at the same point.

**Nails:** The skin surrounding the nails gets infected and it leads to slowly disappearing of the nails and causes nail related problems.

**Moist tissues:** Painful sores develop in tissue lining of eyes, nose, genitals, anus and other areas. Sometimes it can also include the sores or blister formation in the oesophagus of the pemphigus patients.

### Autoimmune mechanisms of pemphigus vulgaris

**Outside-in signalling:** This mechanism explains the phosphorylation of DSG3 by binding of autoantibodies leads to dissociation of plakoglobin and endocytosis mediated degradation. The protein kinase c isomer triggers the pemphigus vulgaris IgG antibodies to bind to DSG3 that leads to intracellular signalling. The p38 MAPK (mitogen activated protein kinase) provided the guidance of interaction of DSG and EGFR (Epidermal Growth Factor Receptor). The dysregulation of signalling pathways like Src (proto-oncogene tyrosine protein kinase), Rho (Ras homologous) GTPases (Guanosine Tri Phosphatase) and actin disorganization leads to pemphigus vulgaris. The signalling pathways like Src, Rho GTPases are involved in desmosome remodelling.

**Non-desmoglein antibodies:** The non DSG antibodies have implicated the blister formation in pemphigus vulgaris patients. Autoantibodies against muscarinic acetylcholine receptors (mAChRs) are involved in regulation of cell adhesion and motility whose inactivation can lead to keratinocyte detachment and acantholysis. Anti-Mitochondrial Antibodies (AMA) trigger the intrinsic apoptotic pathway to release cytochrome C which in turn activates caspase-9 aiding acantholysis.

Dysfunction of mitochondrial pathway increase ROS which cause cellular oxidative stress. Antibodies against Plakophilin-3 (PKP3) which involves in desmosome assembly and stabilization shows altered desmosome and adherent junctions.

**T-cell dysregulation:** The mechanism is associated with specific genotypes of HLA (Human Leukocyte Antigen) and non-HLA genes. The pemphigus vulgaris HLA class II genes are widely associated where there is no appropriate mechanism to explain. The T cell plays a role in immune response, where its defects have identified to contribute pemphigus vulgaris immunopathogenesis. Th1 and Th2 cytokine concentrations are altered and T helper 17 is elevated in pemphigus vulgaris patients when compared to healthy controls. Regulatory T cells that involve in preventing autoimmune response and controls the antigen mediated inflammation and immune response which was identified to be dysregulated in pemphigus vulgaris patients.

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