Pemphigus Vulgaris

Pemphigus Vulgaris is an autoimmune disease where patients form autoantibodies against desmosomes, a protein complex involved in keratinocyte adhesion. Destruction of the desmosomes, a type II hypersensitivity reaction, leads to acantholysis of the stratum spinosum layer of the skin and the formation of thin bullae on the skin which rupture with gentle traction over the top of the blister (positive Nikolsky sign). The bullae most commonly form on the oral mucosa, but also may be seen on the eyes, nose, esophagus, and genital mucosa. Common complications include fluid loss and infection, secondary to bullae rupture. Diagnosis is confirmed on biopsy which demonstrates immunofluorescence of the autoantibodies in a reticular pattern. Treatment of Pemphigus Vulgaris involves a combination of immunosuppression and immunoglobulin replacement to prevent inflammation and wound care.



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Pathophysiology

Autoantibodies against desmosomes

Anti-tie-body against Diamonds

Desmosomes, also known as macula adherens, are protein complexes that aid in cell-to-cell adhesions by anchoring keratin cytoskeleton components in adjacent keratinocytes (squamous epithelial cells). The IgG autoantibodies in Pemphigus Vulgaris specifically target desmoglein, a type of cadherin protein, which has a transmembrane domain that aids in homophilic interactions to facilitate adjacent cell adhesion. These antibodies cause destruction, or acantholysis of the stratum spinosum, via a Type II Hypersensitivity reaction.

Symptoms

Flaccid Bullae on Skin

Floppy Blisters

These are suprabasal fluid-filled blisters greater than 1 centimeter in size. In Pemphigus Vulgaris, they rupture easily because desmosome destruction occurs above the basement membrane, creating a thin-walled blister. After rupturing, the bullae turn to erosions with dried crust.

Oral Mucosa Involvement

Mouth

Because this mucosa is lined with squamous epithelium, bullae commonly appear in the oral cavity, causing difficulty while eating or speaking. The bullae may spread to the larynx, leading to voice hoarseness.

Intraepidermal Acantholysis

In-skin A-candle-lysing

Acantholysis, the separation of keratinocytes due to desmosome destruction, occurs specifically in the stratum spinosum layer of skin.

Fluid Loss

Fluid Loss

If blood vessels underneath the skin are damaged, significant fluid loss can occur in these patients.

Infection

Infectious-bacteria

When bullae rupture, bacteria from the skin and environment may enter at these vulnerable sites, causing infection.

Nikolsky Positive

Nickel-ski Positive

A positive Nikolsky sign indicates that when bullae are lightly touched, the superficial layers of skin separate from basolateral membrane, facilitated easy rupture.

Diagnosis

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Biopsy showing Autoantibodies

Biopsy-needle showing Ant-tie-body

Skin biopsy demonstrates autoantibody immunofluorescence surrounding the keratinocytes with a “net-like” pattern.

Treatment

IVIG and Plasmapheresis

Ivy-gold-goblin and Plasma-fairy

Administration of IVIG helps to suppress inflammation, with its effect lasting 2 to 3 months. The mechanism of action is unknown, but may involve binding the patient’s autoantibodies to prevent them from binding desmosomes. Plasmapheresis involves removing and replacing the patient’s plasma with donor plasma that does not contain the autoantibodies. Both treatment modalities help to prevent inflammation, infection, and fluid loss.

Oral Steroids

Steroid-stairs

Because Pemphigus Vulgaris is an autoimmune condition, treatment is directed as suppressing the patient’s immune system to prevent further exacerbation of the condition. Moreover, inflammation is left behind after bullae rupture. Therefore patients may be treated with oral steroids, or other immunosuppressants, such as Rituximab, Mycophenolate, Azathioprine to suppress the immune system and alleviate inflammation.

Immunosuppressants

Moon-suppressed

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