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PEMPHIGUS VULGARIS: PATHOGENESIS, CLINICAL MANIFESTATIONS, AND EMERGING THERAPEUTIC STRATEGIES

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Abstract:

Intraepithelial blister production affecting the skin and mucous membranes is the hallmark of Pemphigus vulgaris (PV), an uncommon and possibly fatal chronic autoimmune blistering illness. Autoantibodies against desmoglein-3 and desmoglein-1 are the main cause of the disease, which results in keratinocyte adhesion loss (acantholysis). Clinically, PV presents as painful oral erosions and easily ruptured flaccid skin blisters. The diagnosis is based on clinical presentation, histology showing suprabasal acantholysis, and direct immunofluorescence showing intercellular IgG deposition. Systemic corticosteroids remain the mainstay of treatment, but immunosuppressive medications and biologics like rituximab have significantly improved disease outcomes. Potential alternatives with less adverse effects are presented by targeted therapies and fresh discoveries about biological processes. The current understanding of pemphigus vulgaris epidemiology, immunopathogenesis, clinical characteristics, diagnostic techniques, and developing treatment approaches is compiled in this study.

Index Term:

Pemphigus vulgaris, Autoimmune blistering disorder, Desmoglein, Acantholysis, Rituximab, Immunotherapy

1. Introduction

The characteristic of Pemphigus vulgaris (PV), an uncommon, chronic, and potentially fatal autoimmune blistering disease, is intraepidermal blister development affecting the skin and mucous membranes. The primary targets of pathogenic immunoglobulin G (IgG) autoantibodies that induce the disease are desmoglein-3 (Dsg3) and desmoglein-1 (Dsg1), cadherin-type adhesion molecules required for keratinocyte cell–cell interaction within desmosomes [1,2].
PV is the most severe kind of pemphigus, accounting for most occurrences worldwide [3]. With an annual incidence ranging from 0.1 to 0.5 cases per 100,000 individuals, the prevalence has been reported to be higher in Mediterranean, Middle Eastern, and South Asian communities, particularly those in India. The condition often appears between the ages of 40 and 60, with a little female predominance.[4]

Clinically, PV typically begins with excruciating oral erosions before spreading to the skin, where flaccid bullae easily rupture and result in widespread erosions. A positive Nikolsky sign is common [5]. Sepsis, electrolyte imbalance, dehydration, and secondary infections are among the major side consequences of untreated PV that have previously been associated with high death rates [6]. The introduction of systemic corticosteroids in the middle of the 20th century significantly reduced mortality. Long-term steroid use, however, is associated with major adverse consequences, including osteoporosis, diabetes mellitus, hypertension, and an elevated risk of infection [7]. Over the past ten years, targeted biologic treatments—particularly anti-CD20 monoclonal antibodies like rituximab—have revolutionized treatment strategies and raised long-term remission rates [8,9].

Recent discoveries (2020–2025) that have expanded our knowledge of PV immunopathogenesis include B-cell dysregulation, cytokine imbalance, intracellular signaling cascades, and the role of neonatal Fc receptor (FcRn) in IgG recycling [2,10]. These discoveries have led to the development of novel therapeutic treatments such cellular immunotherapies, FcRn inhibitors. Given the rapid development of therapeutic approaches and expanding molecular discoveries, a thorough review of recent literature is required to gather the most recent information on the pathogenesis, clinical outcomes, and novel treatment modalities of pemphigus vulgaris.

2. Epidemiology

There are significant regional and ethnic differences in the frequency and prevalence of pemphigus vulgaris, a rare autoimmune blistering condition. Despite being classified as an uncommon disease globally, PV is the most prevalent subtype within the pemphigus group [11].

2.1 Incidence and Prevalence

The reported annual incidence of PV ranges from 0.1 to 0.5 cases per 100,000 individuals, depending on the region [11,12]. Population-based studies from Europe and North America report incidence rates between 0.1 and 0.3 per 100,000 people yearly, however higher rates have been observed in Middle Eastern, Mediterranean, and South Asian communities [12,13]. Prevalence estimates range from 1 to 5 instances per 100,000 people, indicating that modern immunosuppressive treatment has improved survival rates [13]. Due to increased accessibility and understanding of diagnostic immunofluorescence testing, identification rates have also increased in recent years.

2.2 Geographic and Ethnic Distribution

There has been noticeable spatial clustering. There are reports of higher incidence rates among:

- Ashkenazi Jewish populations
- Mediterranean countries
- Middle Eastern populations
- South Asian populations, including India

Genetic susceptibility linked to specific human leukocyte antigen (HLA) alleles partially explains this variation [12,14].
Strong correlations have been confirmed by recent epidemiological research (2020–2023) with:

- HLA-DRB1*0402
- HLA-DQB1*0503

These genotypes are thought to promote autoreactive T-cell activation against desmoglein peptides and are especially common in high-incidence groups [14].

2.3 Age and Gender Distribution

PV often appears between the ages of 40 and 60, while cases have been reported in both younger and older individuals [11,13]. Pediatric and juvenile cases are still rare.

A slight female predominance has been consistently reported, with female-to-male ratios in several cohorts ranging from 1.1:1 to 2:1 [12]. Hormonal and immunological elements may be involved, even if the precise mechanisms causing this gender gap are still unknown.

2.4 Mortality and Disease Burden

Prior to the introduction of systemic corticosteroids, mortality rates exceeded 70%. Thanks to contemporary immunosuppressive and biologic medications, mortality has sharply decreased to less than 10% in wealthy countries [13].

But morbidity is still high because of:

- Chronic relapsing disease course
- Treatment-related adverse effects
- Secondary infections
- Reduced quality of life

According to recent longitudinal research, infection continues to be the primary cause of death for PV patients, especially for those on high doses of corticosteroids or other immunosuppressive medications [15].

2.5 Environmental and Triggering Factors

Environmental factors are becoming more widely acknowledged, despite the fact that PV is predominantly autoimmune and genetically determined. Among them are:

- Drugs (penicillamine, captopril, ACE inhibitors)
- Viral infections
- Ultraviolet radiation
- Physical trauma
- Emotional stress

Drug-induced pemphigus represents a small but clinically significant subset, often reversible upon withdrawal of the offending agent [14].

3. Immunopathogenesis

Pathogenic autoantibodies that target desmosomal cadherins cause keratinocyte adhesion to be lost (acantholysis) in pemphigus vulgaris, a classic antibody-mediated autoimmune illness. Clarifying the cellular and molecular mechanisms underlying the onset and course of disease has advanced significantly over the last ten years [16,17].

3.1 Role of Desmoglein Autoantibodies

The hallmark of PV is the production of IgG autoantibodies directed against:

- Desmoglein-3 (Dsg3) – predominantly expressed in mucosal epithelium
- Desmoglein-1 (Dsg1) – expressed in the superficial layers of epidermis

The "desmoglein compensation theory" explains clinical presentation:

- Anti-Dsg3 antibodies → mucosal-dominant PV
- Anti-Dsg1 + Anti-Dsg3 antibodies → mucocutaneous PV [16,18]

Intraepidermal blister formation results from the disruption of desmosomal adhesion between keratinocytes caused by these pathogenic IgG (mostly IgG4 subclass) binding to extracellular domains of desmoglein's [17].

Antibody titers are correlated with disease activity and may be used as biomarkers to track treatment response, according to recent research [19].

3.2 Mechanisms of Acantholysis

In the past, adhesion molecules' steric hindrance was the only explanation for blister formation. Acanthosis, however, is a complicated signaling-mediated process, according to recent study.

Autoantibody binding triggers:

- p38 mitogen-activated protein kinase (MAPK) activation
- Src kinase signaling
- Epidermal growth factor receptor (EGFR) activation
- RhoA pathway inhibition

These intracellular cascades lead to:

- Cytoskeletal reorganization
- Desmosomal disassembly
- Keratin filament collapse
- Loss of intercellular cohesion [17,20]

According to recent molecular analysis (2022–2024), p38 MAPK activation is crucial and could be a therapeutic target.

3.3 B-Cell Dysregulation

The pathogenesis of PV heavily relies on B lymphocytes. After being triggered and differentiating into plasma cells, autoreactive B cells create pathogenic IgG autoantibodies.

Current studies reveal:

•Decreased regulatory B-cell (Breg) function.
• Memory B-cell populations that continue to exist after remission.
• Dsg-specific B lymphocyte clonal growth.

This explains high relapse rates following anti-CD20 treatment and B-cell reconstitution [17, 21]. Moreover, long-lived plasma cells in bone marrow may promote extended autoantibody generation, which poses a therapeutic challenge.

3.4 T-Cell Involvement

T helper cells have a crucial role in the initiation and maintenance of autoimmunity, despite the fact that PV is mainly antibody-mediated.

Evidence shows:
- Th2 polarization (increased IL-4, IL-10)
- Elevated IL-21 promoting B-cell activation
- Reduced regulatory T-cell (Treg) function

HLA-DRB1*0402 and HLA-DQB1*0503* alleles present Dsg peptides to autoreactive CD4+ T cells, facilitating autoantibody production [18,22].

3.5 Cytokine and Inflammatory Network

Recent immunological profiling has revealed elevated serum levels of the following:

• TNF-α, IL-17, IL-6, IL-10, and IL-4
These cytokines create a pro-inflammatory environment that promotes B-cell survival and antibody production [20].
Research into targeted therapies, such as JAK inhibitors and IL-4/IL-13 inhibitors, has been spurred by the expanding knowledge of cytokine involvement.

3.6 Neonatal Fc Receptor (FcRn) and IgG Recycling

The neonatal Fc receptor (FcRn) is crucial for prolonging the half-life of IgG by blocking lysosomal degradation.

In PV:

- Pathogenic IgG autoantibodies are recycled via FcRn
- Increased IgG persistence sustains disease activity

This information led to the development of FcRn inhibitors such as efgartigimod.
By blocking FcRn, these medications rapidly reduce pathogenic antibody titers and circulating IgG levels [23].

3.7 Emerging Molecular Targets

Additional possible targets are highlighted by recent translational research:
• B cell signaling via Bruton's tyrosine kinase (BTK)
• Pathways for complement activation
• Patterns of autoantibody glycosylation
• Epigenetic regulation of immune cells

By focusing on specific routes, immunomodulation may be more selective and have less negative systemic consequences.

4. Clinical Manifestations

Mucocutaneous blistering that is persistent, recurrent, and has a high morbidity rate is a hallmark of pemphigus vulgaris. The distribution and titers of anti-desmoglein autoantibodies determine the clinical symptoms, however in the majority of cases, mucosal involvement is the defining characteristic [24].

4.1 Mucosal Involvement

About 50–70% of cases begin with mucosal lesions, which frequently appear weeks or months before cutaneous involvement [24,25].

Oral Lesions

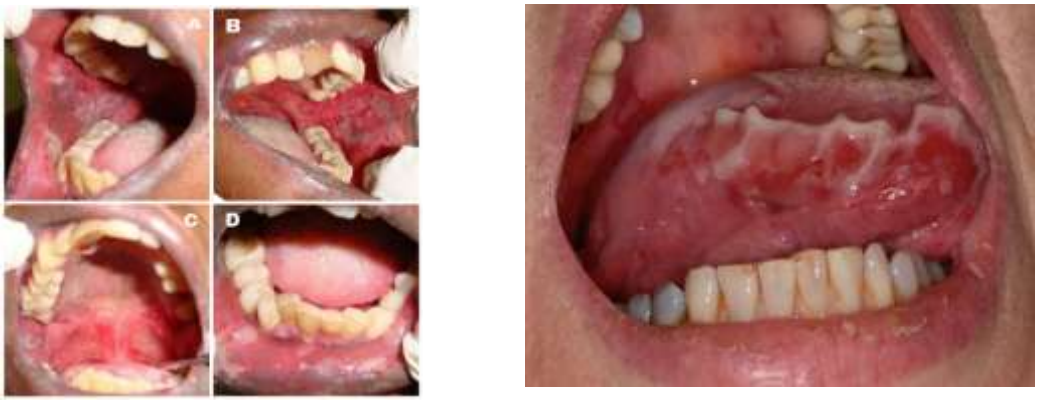

Fig: 1 Oral Lesions

The oral cavity is the most commonly affected site. Lesions typically present as:

- Painful erosions or shallow ulcers
- Fragile vesicles that rupture rapidly
- Desquamative gingivitis
- Persistent non-healing ulcers

Eating, swallowing, and speaking are severely hampered by these lesions, which leads to weight loss and a lower quality of life [25].

Other mucosal sites that may be involved include:

- Oropharynx, Nasal mucosa, Conjunctiva, Genital mucosa, Esophagus

Esophageal involvement may present with dysphagia and odynophagia, though less commonly reported.

4.2 Cutaneous Manifestations

Fig: 2 Cutaneous Manifestations

Cutaneous involvement develops in the majority of patients during disease progression.

Characteristic features include:

- Flaccid bullae on normal or erythematous skin
- Easily ruptured blisters
- Extensive erosions with crusting
- Positive Nikolsky sign (epidermal shearing with lateral pressure)

Because of intraepidermal cleavage at the suprabasal level, blisters are usually superficial, which accounts for their fragility [24].

Common sites include: Scalp, Face, Chest, Back, Intertriginous areas

4.3 Disease Variants

Clinical presentation may vary:

4.3.1 Mucosal-Dominant PV

- Primarily anti-Dsg3 antibodies

- Limited or absent skin involvement

4.3.2 Mucocutaneous PV

- Both anti-Dsg1 and anti-Dsg3 antibodies
- Widespread skin and mucosal disease

These clinical phenotypes correlate with immunological findings and antibody profiles [26].

4.4 Disease Severity and Course

PV typically follows a chronic relapsing course.

Disease severity is assessed using validated scoring systems such as:

- Pemphigus Disease Area Index (PDAI)
- Autoimmune Bullous Skin Disorder Intensity Score (ABSIS)

Untreated disease can lead to:

- Secondary bacterial infections
- Fluid and electrolyte imbalance
- Sepsis
- Malnutrition

Although mortality has decreased because to modern biologic medicines, morbidity is still high because of the burden of chronic diseases and side effects from therapy [27].

4.6 Clinical Red Flags

The following characteristics should make one suspicious of PV:
- Persistent oral ulcers resistant to standard therapy
- Flaccid blisters that rupture easily
- Positive Nikolsky sign
- Coexisting mucosal and cutaneous erosions

In order to avoid problems and start immunosuppressive therapy on time, early detection is essential.

5. Diagnosis of Pemphigus Vulgaris

The diagnosis of Pemphigus vulgaris (PV) requires histopathology, immunopathology, serological testing, and clinical evaluation. Early and accurate diagnosis is crucial since delaying treatment increases morbidity and mortality. Modern diagnostic techniques combine conventional microscopy with advanced immunological and molecular techniques.

5.1. Clinical Evaluation

PV usually manifests as:
- Flaccid blisters and erosions on skin
- Painful oral erosions (often the first manifestation)
- Positive Nikolsky's sign (shearing of epidermis with lateral pressure)
- Chronic, relapsing course

Months may pass between oral lesions and skin involvement. Lesions can break easily due to their brittleness, causing agonizing erosions.

5.2) Histopathology (Gold Standard Initial Test

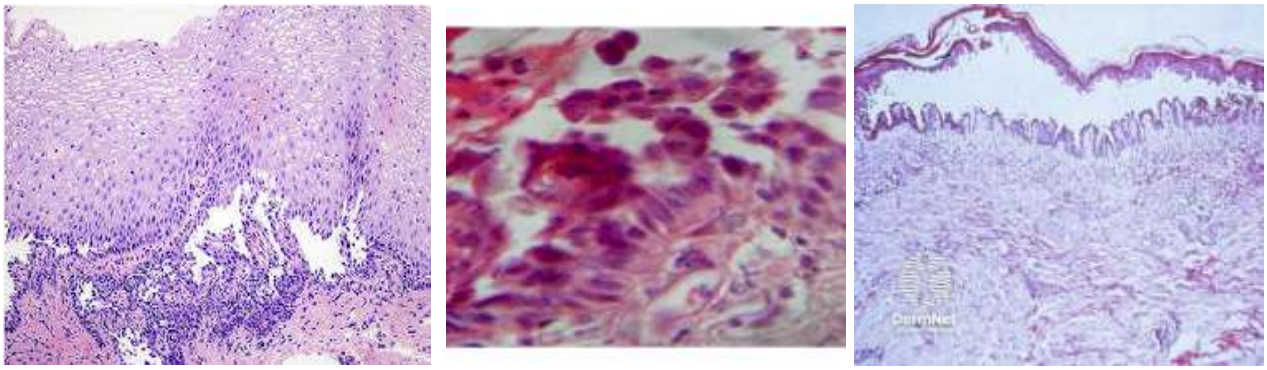

Fig: 3 Histopathology (Gold Standard Initial Test)

Results of a skin biopsy (H&E staining):

• The suprabasal intraepidermal cleft
• Loss of keratinocyte adhesion, or acantholysis
• Basement membrane-attached basal keratinocytes

5.3. Direct Immunofluorescence (DIF) – Diagnostic Gold Standard

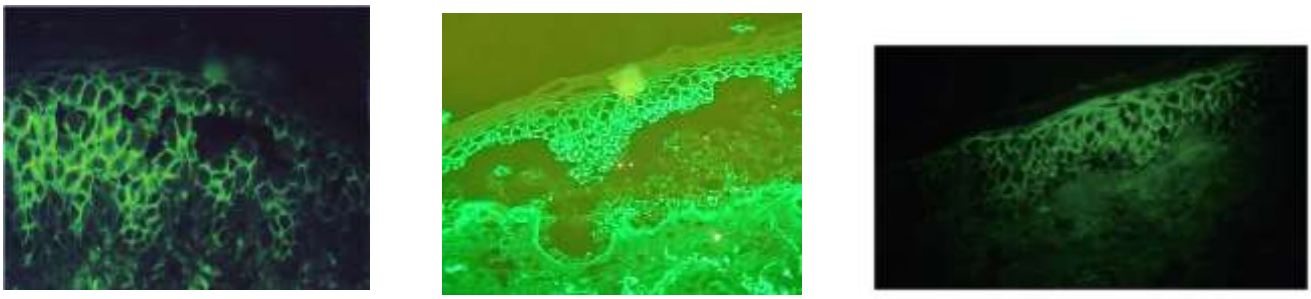

Fig: 4 Direct Immunofluorescence (DIF) – Diagnostic Gold Standard

Perilesional skin's direct immunofluorescence reveals:

- Intercellular deposition of IgG
- Often C3 complement deposition
- Classic “fish-net” or “chicken wire” pattern

DIF confirms autoimmune etiology and is still the most sensitive diagnostic test.

5.**4. Indirect Immunofluorescence (IIF)**

- Detects circulating anti-desmoglein antibodies
- Substrate: monkey esophagus or human skin
- Titers often correlate with disease activity

Sensitivity: ~80–90%.

5.5. Enzyme-Linked Immunosorbent Assay (ELISA)

ELISA detects:
- Anti-desmoglein 3 (Dsg3) antibodies
- Anti-desmoglein 1 (Dsg1) antibodies

Clinical correlation:
- Dsg3 positive → mucosal PV
- Dsg1 + Dsg3 positive → mucocutaneous PV

ELISA titers are helpful for tracking relapses and correspond with the severity of the illness.

5.6. Emerging Diagnostic Techniques (Post-2020 Advances)

Recent studies highlight:
- Multiplex immunoassays
- Biochip mosaic indirect immunofluorescence
- Desmocollin antibody detection
- Autoantibody epitope mapping
- Serum cytokine profiling

These instruments enhance prognosis and early diagnosis.

5.7. Diagnostic Algorithm (Stepwise Approach)

1. Clinical suspicion (oral erosions ± skin lesions)
2. Skin biopsy for histopathology
3. Perilesional biopsy for DIF
4. ELISA for anti-Dsg1/Dsg3 antibodies
5. IIF for antibody titers
6. Monitoring with serial ELISA titers

5.8. Conclusion

Pemphigus vulgaris diagnosis depends on:
- Clinical suspicion
- Histopathological confirmation
- Direct immunofluorescence (gold standard)
- Serological antibody detection

Modern immunological testing has improved early detection and illness surveillance, allowing for customized therapy options.

6. Management and Treatment of Pemphigus Vulgaris

Targeted biologic therapies have replaced long-term high-dose corticosteroid therapy as the primary treatment for Pemphigus vulgaris (PV) in the last decade. Current approaches seek to minimize recurrence, produce prolonged remission, establish quick disease management, and lessen the toxicity of long-term treatment.

6.1. Treatment Goals

- Rapid cessation of new blister formation.
- Healing of existing lesions.
- Reduction of circulating anti-desmoglein antibodies.
- Prevention of relapse.
- Minimization of corticosteroid-related adverse effects.

The absence of new lesions and the start of established lesions' healing are considered to be indicators of disease control.

6.2. First-Line Therapy

6.2.1. Systemic Corticosteroids

Systemic corticosteroids remain a fundamental component of therapy.

Typical dosage:

Depending on the severity of the disease, prednisolone dosages range from 0.5 to 1.5 mg/kg/day.

Autoantibody-mediated acantholysis and inflammation are quickly suppressed by corticosteroids. However, prolonged use is linked to:

- Diabetes mellitus
- Hypertension
- Osteoporosis
- Increased infection risk
- Cushingoid changes

Steroid-sparing medications are highly advised in cases of moderate-to-severe illness due to these hazards (28,29).

6.2.2. Rituximab (Current First-Line Biologic Therapy)

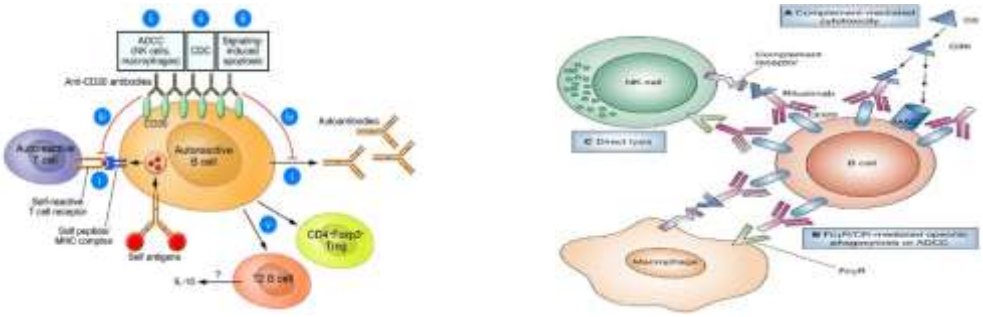

Fig: 5 Rituximab (Current First-Line Biologic Therapy)

For moderate-to-severe PV, rituximab, a chimeric monoclonal antibody that targets CD20-positive B cells, is now advised as the initial treatment.

Mechanism of action:

- Depletes autoreactive B cells
- Reduces anti-desmoglein antibody production
- Induces long-term remission

Two commonly used regimens:

- Rheumatoid arthritis protocol (1 g on days 1 and 15)
- Lymphoma protocol (375 mg/m² weekly for 4 weeks)

The recommended initial treatment is rituximab plus short-term corticosteroids, according to recent international guidelines (1-4). Research conducted after 2020 confirms:

- Higher complete remission rates
- Reduced cumulative steroid exposure
- Longer relapse-free survival

6.3. Conventional Steroid-Sparing Immunosuppressants

When biologics are unavailable or contraindicated, conventional immunosuppressants are used.

Azathioprine

- Dose: 1–3 mg/kg/day
- Inhibits purine synthesis
- Requires TPMT testing before initiation
- Effective as adjunct therapy (29)

Mycophenolate Mofetil (MMF)

- Dose: 2–3 g/day
- Inhibits lymphocyte proliferation
- Favorable safety profile
- Comparable efficacy to azathioprine (30)

Cyclophosphamide

- Reserved for severe or refractory disease
- Associated with gonadal toxicity and malignancy risk

Adjunctive and Supportive Therapy

- Topical corticosteroids for localized lesions
- Antiseptic mouthwashes for oral lesions
- Nutritional support
- Pain management
- Infection prophylaxis

Hospitalization may be necessary for patients with severe erosions in order to control their fluid and electrolyte levels.

7. Emerging Targeted Therapies

Recent therapeutic developments include:

7.1 FcRn Inhibitors

Neonatal Fc receptor (FcRn) inhibitors shorten the half-life of IgG, which causes the amount of autoantibodies in circulation to drop quickly (31).

7.2 Bruton Tyrosine Kinase (BTK) Inhibitors

They are being evaluated clinically and target B-cell receptor signaling pathways (32).

7.3 Anti-BAFF Therapy

Inhibits B-cell activating factor to reduce autoantibody production.

7.4 Next-Generation Anti-CD20 Agents

- Ofatumumab
- Obinutuzumab

The goal of these treatments is to increase remission persistence and safety.

7.5. Monitoring During Treatment

Monitoring includes:

- Clinical lesion assessment
- Anti-Dsg1 and Dsg3 ELISA titers
- Complete blood count
- Liver and renal function tests
- Screening for hepatitis B prior to rituximab

Serological titers often correlate with disease activity and relapse risk (33).

7.6. Prognosis

Before corticosteroid therapy, mortality exceeded 70%. With modern treatment:

- Mortality <10% in specialized centers
- High remission rates with rituximab
- Relapses remain common but manageable

Early rituximab initiation is associated with improved long-term outcomes (33).

8. Conclusion:

Autoantibodies that target desmosomal cadherins cause pemphigus vulgaris, a potentially fatal autoimmune blistering illness that causes mucocutaneous blisters and a lack of cell adhesion. Both diagnosis and therapy approaches have been improved by a better understanding of its immunological underpinnings, such as genetic predisposition and B-cell-mediated responses. Systemic corticosteroids are still the mainstay of treatment, although more recent targeted therapies like anti-CD20 monoclonal antibodies have produced far better results with fewer adverse effects. It is anticipated that further studies on immune modulation and molecular pathways will aid in the creation of more accurate, successful, and long-lasting remission-based treatment strategies for pemphigus vulgaris patients.

9. Abbreviations:

PV - Pemphigus vulgaris

Dsg3- Desmoglein-3

Dsg1- Desmoglein-1

FcRn - Neonatal Fc receptor

BTK- Bruton's tyrosine kinase

HLA-Human leukocyte antigen

EGFR- Epidermal growth factor receptor

MAPK- mitogen-activated protein kinase

Breg- Regulatory B-cells

Treg- Regulatory T-cell

PDAI - Pemphigus Disease Area Index

ABSIS - Autoimmune Bullous Skin Disorder Intensity Score

TPMT- Thiopurine Methyltransferase

ELISA- Enzyme-linked immunosorbent assay

IIF- Indirect immunofluorescence

MMF- Mycophenolate Mofetil

BAFF- Anti B-cell Activating Factor

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