Pemphigus vulgaris

Abdulkasymova Dinara Abdulazizovna¹, Javed Shekh²

¹ Osh State University, International medical faculty of Kyrgyzstan; Republic of Kyrgyzstan
² Osh State University, International medical faculty of Kyrgyzstan; Republic of Kyrgyzstan

Abstract.
Pemphigus vulgaris is a chronic autoimmune bullous dermatosis that results from the production of autoantibodies against desmogleins 1 and 3. It is the most frequent and most severe form of pemphigus, occurring universally, usually between 40 and 60 years of age. It usually begins with blisters and erosions on the oral mucosa, followed by lesions on other mucous membranes and flaccid blisters on the skin, which can be disseminated. There is a clinical variant, pemphigus vegetans, which is characterized by the presence of vegetating lesions in the large folds of the skin.

Keywords:
Autoantibodies Vesiculobullous skin diseases
INTRODUCTION

Phemphigus diseases are a group of rare autoimmune bullous diseases that affects the skin and mucous membranes. Their estimated incidence is two new cases/million inhabitants/year in central Europe. They present chronic evolution, with significant morbidity and mortality, as well as an important impairment in quality of life.1., 2. They originate from the production of pathogenic autoantibodies (usually of the IgG class) directed against different proteins of desmosomes (desmogleins). The union of these autoantibodies to the components of the desmosomes compromises intraepidermal adhesion, leading to acantholysis and formation of vesicles, blisters, and erosions on the skin and/or mucous membranes.3., 4., 5.

The formation of autoantibodies against components of desmosomes has long been considered the main process in pemphigus pathogenesis. In addition to the important role of humoral immunity, cellular immunity has also been highlighted in the literature.7 PV is the main clinical form of pemphigus, accounting for approximately 70% of cases; it is also considered the most severe form of the disease.

PEMPHIGUS VULGARIS

Pemphigus vulgaris (PV) is a rare, chronic autoimmune disease that causes painful blisters on the skin and mucous membranes. It is the most common type of pemphigus, affecting about one in 40,000 people worldwide. PV is caused by the production of autoantibodies that attack desmogleins [DGS 3>>DSG1], proteins that hold skin cells together. This attack causes the skin cells to separate, leading to the formation of blisters.

**Symptoms**

The main symptom of PV is the formation of large, painful blisters on the skin and mucous membranes. The blisters can be clear or filled with fluid, and they can rupture easily, leaving raw, erosive lesions.

The blisters most commonly appear on the mouth, lips, groin, armpits, and around the umbilicus. They can also affect the eyes, nose, throat, and genitals.

In addition to blisters, other symptoms of PV may include:

- Fever
Fatigue
Weight loss
Pain
Causes

The exact cause of PV is unknown, but it is thought to be an autoimmune disease. This means that the body's immune system mistakenly attacks healthy tissues.

In PV, the immune system attacks desmogleins, which are proteins that help to hold skin cells together. This attack causes the cells to separate, leading to the formation of blisters.

**DESIGN**

Pemphigus vulgaris had a mean/median age of 10.9/12 years, linear IgA disease of 5.5/3 years and bullous pemphigoid of 5.5/3.5 years. Two of the three patients with mucous membrane pemphigoid were in the first year of life, and one girl was 5 years old. The two patients with epidermolysis bullosa acquisita were 8 and 15 years of age. The age distributions for the most prevalent AIBD, i.e. pemphigus vulgaris, linear IgA disease and bullous pemphigoid, are detailed in Fig.

Frequency of signs and symptoms when continuously monitored in a group of infected patients appear as
SIGNS AND SYMPTOMS

Who gets Pemphigus vulgaris?
It is more common in adults between the ages of 50 and 60. PV is also slightly more common in women than in men.

Here are some of the risk factors for PV:
Age: PV is most common in adults between the ages of 50 and 60.
Sex: PV is slightly more common in women than in men.
Ethnicity: PV is more common in people of Jewish descent, especially Ashkenazi Jews.
Genetics: People with a family history of PV are at increased risk of developing the disease.
Certain medications: Certain medications, such as penicillins and nonsteroidal anti-inflammatory drugs (NSAIDs), can trigger PV in people who are already at risk.
Environmental factors: Exposure to certain environmental factors, such as ultraviolet radiation, may also trigger PV in people who are already at risk.

INVESTIGATION

1. Medical History and Physical Examination
Your doctor will start by taking a thorough medical history to learn about your symptoms, including when they began, their location, and any factors that seem to aggravate them. They will also perform a physical examination to look for blisters and other signs of PV.

2. Skin Biopsy
A skin biopsy is the most common test used to diagnose PV. During a skin biopsy, a small sample of skin is removed and examined under a microscope. The pathologist will look for the characteristic features of PV, such as acantholysis, which is the separation of skin cells due to the loss of
desmoglein adhesion.

3. Immunofluorescence Studies

Immunofluorescence studies can detect the presence of autoantibodies in the skin and blood. Autoantibodies are antibodies that attack the body's own tissues. In PV, the autoantibodies target desmogleins, which are proteins that hold skin cells together.

4. Blood Tests

Blood tests can be used to look for certain antibodies that are associated with PV. These antibodies can also be found in other autoimmune diseases, so the results of blood tests alone are not enough to confirm the diagnosis of PV.

5. Other Tests

In some cases, other tests may be used to diagnose PV or rule out other conditions. These tests may include:
- Direct immunofluorescence (DIF) of the skin
- ELISA (enzyme-linked immunosorbent assay) to detect antibodies in the blood
- Western blot to confirm the identity of the antibodies

PV can be difficult to distinguish from other blistering skin diseases, such as:
- Pemphigus foliaceus
- Bullous pemphigoid
- Epidermolysis bullosa
- Stevens-Johnson syndrome

TREATMENT

NO CURE FOR PEMPHIGUS VULGARIS

SUPPORTIVE THERAPY

Initial Therapy

Initial therapy for PV typically involves corticosteroids, such as prednisone. Corticosteroids are powerful drugs that can suppress the immune system and reduce inflammation. However, they can also cause side effects, such as weight gain, bone loss, and high blood pressure.

Maintenance Therapy

Once the blisters have healed, the dosage of
Corticosteroids can be gradually reduced and tapered off. However, many people with PV require long-term maintenance therapy to prevent new blisters from forming. Maintenance therapy may involve corticosteroids at a lower dose or other immunosuppressant medications, such as:

- Azathioprine
- Mycophenolate mofetil
- Rituximab

**Other Treatments**

In addition to corticosteroids and immunosuppressant medications, other treatments that may be used to treat PV include:

- Plasmapheresis: This procedure removes antibodies from the blood.
- Intravenous immunoglobulin (IVIG): This blood product contains antibodies from healthy donors that can help to suppress the immune system.
- Local therapy: This may include topical corticosteroids, antibiotics, or wound care.

**Latest developments in pemphigus vulgaris research**

**Nanotech-based strategy for long-term treatment:** Researchers at the University of California, San Francisco, have developed a nanotech-based strategy that could enable safe, long-term treatment for autoimmune diseases like pemphigus vulgaris. The approach involves using nanoparticles to deliver small interfering RNA (siRNA) molecules directly to the immune cells responsible for causing the disease.

**NIH begins clinical trial for extra COVID-19 vaccine dose in people with autoimmune disease:** The National Institutes of Health (NIH) has begun a clinical trial to assess whether an extra dose of COVID-19 vaccine can improve immune responses in people with autoimmune diseases like pemphigus vulgaris. The trial will enroll up to 240 adults with autoimmune diseases who have not responded adequately to the standard COVID-19 vaccine regimen.

**OUTCOMES**

The outcome of pemphigus vulgaris (PV) varies depending on the severity of the disease and the individual's response to treatment. With proper treatment, most people with PV can achieve remission and control their symptoms. However, PV can
be a lifelong condition and there is a risk of recurrence.

Factors Affecting Prognosis

Several factors can affect the prognosis of PV, including:
- Age of onset: Younger patients tend to have a better prognosis than older patients.
- Severity of the disease: Patients with more severe PV may have a longer and more difficult course of treatment.
- Response to treatment: Patients who respond well to treatment are more likely to have a good prognosis.
- Presence of comorbidities: Patients with other medical conditions may have a more difficult time controlling their PV.

With treatment, most people with PV can achieve remission and control their symptoms. Remission is defined as the absence of blisters for at least six months. Some people may experience long-term remission, while others may have periods of remission and relapse.

In some cases, PV can be a serious and life-threatening condition. Complications of PV can include infection, dehydration, and malnutrition. In rare cases, PV can be fatal.

CONCLUSION

Pemphigus vulgaris is a challenging autoimmune condition that requires a comprehensive approach involving dermatologists, immunologists, and other healthcare professionals. While treatments aim to manage symptoms and suppress the immune response, ongoing research is exploring novel therapies to improve outcomes and enhance the quality of life for individuals affected by this condition. Early diagnosis, prompt intervention, and a multidisciplinary approach are pivotal in addressing pemphigus vulgaris and mitigating its impact on patients' lives.

References:
[2] Google Scholar: Use [Google Scholar](https://scholar.google.com/) to search for scholarly articles, theses, books, and more on pemphigus vulgaris. It can often provide direct links to articles or publications.
MEDICINE AND PHARMACY

[3] Dermatology Journals: Journals like the Journal of the American Academy of Dermatology (JAAD), Journal of Investigative Dermatology (JID), Dermatology, etc., often publish articles and studies on pemphigus vulgaris. Visiting their respective websites or using databases through university access can provide access to these articles.

[4] Library Databases: University or medical school libraries often grant access to various medical journals and databases. If you have access, check library databases for articles on pemphigus vulgaris.

[5] ResearchGate: ResearchGate is a platform where researchers share their publications. Searching for pemphigus vulgaris on [ResearchGate](https://www.researchgate.net/) might lead you to articles or studies shared by experts in the field.