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# CLINICAL CORRELATION IN DIFFERENT HISTOPATHOLOGICAL TYPES OF PEMPHIGUS VULGARIS

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## Abstract

***Background*** *Intraepithelial bullae, skin and mucous membrane erosions are hallmarks of the autoimmune disease pemphigus vulgaris. Assessing the clinical and demographic characteristics of individuals with pemphigus vulgaris (PV) who came to our department was our objective.*

***Methods*** *A cross-sectional study was conducted in the Dermatology Department of the Pakistan Emirates Military Hospital from December 2023 to June 2024. It included patients aged 18 to 70 with pemphigus vulgaris (PV) confirmed by histopathology and immunofluorescence. Fifty patients were categorised into mild, moderate, and severe PV based on severity scores.*

***Results*** *The study involved 50 patients with various histopathological types of pemphigus vulgaris. The mean age was 52.3 years (SD = 14.6), ranging from 18 to 70 years. The cohort included 23 females (46.0%) and 27 males (54.0%). A family history of pemphigus vulgaris was present in 22 patients (44.0%). Clinically, 39 patients (78.0%) had mucocutaneous pemphigus vulgaris (MCPV), 10 (20.0%) had mucosal pemphigus vulgaris (MPV), and 1 (2.0%) had skin-only involvement. At presentation, 94.0% reported oral esions, and 34.0% experienced dysphagia. The disease duration averaged 44.5 onths. Severity ranged from mild in 42.0% to severe in 22.0%, with 42.0% xperiencing a relapse.*

***Conclusion*** *Our region's pemphigus vulgaris (PV) patients' clinical and demographic findings were consistent with those of earlier research.*

## INTRODUCTION

Pemphigus is an autoimmune condition characterised by the presence of erosions and intraepithelial bullae on the mucous membranes and skin [1]. A key feature of pemphigus is acantholysis, the loss of intercellular adhesion between epidermal keratinocytes, which is driven by autoantibodies present in pemphigus serum [2, 3]. Immunopathological studies have identified that these autoantibodies, predominantly of the IgG type, target desmoglein 1 and/or 3, a cell surface antigen of keratinocytes [1-3].

Pemphigus vulgaris (PV), IgA pemphigus, pemphigus foliaceus (PF), and paraneoplastic pemphigus (sometimes called paraneoplastic autoimmune multiorgan syndrome, or PAMS) are the four types of pemphigus that can be distinguished based on their clinical and immunopathological characteristics [4, 5].

The most common clinical manifestation of PV is defined by loose, readily ruptured bullae on mucous membranes and skin that would otherwise be normal or erythematous [1].

Pemphigus is a globally distributed disease, most commonly affecting individuals between the ages of 40 and 60, with an equal prevalence in both females and males [6]. In addition, the incidence of pemphigus vulgaris (PV) ranged from 0.098 to 5 cases per 100,000 individuals, while bullous pemphigoid (BP) ranged from 0.21 to 7.63 per 100,000. PV and BP prevalence ranged from 0.38 to 30 and 1.46 to 47.99 per 100,000, respectively. For PV and BP, the mean age of onset ranged from 36.5 to 71 years and 64 to 82.6 years, respectively. In PV and BP, the female-to-male ratios varied from 0.46 to 4.4 and 1.01 to 5.1, respectively [6]. Additionally, according to a different study, the incidence varies between 0.7 and 5 new cases per million year [7].

Pemphigus vulgaris disease has a complex etiopathogenesis that includes polygenic genetic risk factors and mainly unknown environmental variables. HLA class II molecules are a crucial hereditary component that is required but not entirely accountable for the development of disease. Pemphigus vulgaris (PV) has been linked to the alleles DRB10402 and DQB10503 in Ashkenazi Jewish and Caucasian individuals [8]. Because pemphigus and pemphigoid diseases are rare, their risk and development are complex, and disease expression varies from person to person, it is difficult to accurately assess global epidemiological and genetic risk factors. Additionally, the lack of centralised and standardised epidemiological, clinical, and genetic data further complicates assessment. A critical analysis of the current knowledge and its gaps is essential to develop informed strategies for advancing our understanding of the etiopathogenic factors relevant to these diseases. The primary aim of pemphigus treatment is to achieve rapid disease remission and reduce the synthesis of autoantibodies. Early and systematic treatment is essential for controlling the disease and maintaining long-term remission [9, 10]. Therefore, in this study we aimed to evaluate of clinical correlation of severity of different histopathological types of pemphigus vulgaris.

**Methods**

A cross-sectional study was conducted in the Dermatology Department of the Pakistan Emirates Military Hospital. The study spanned six months, from December 2023 to June 2024. Participants were aged between 18 and 70 years, and both sexes were included. Patients with a diagnosis of pemphigus vulgaris (PV) confirmed through histopathological and immunofluorescent examinations were selected for inclusion.

A total of 50 confirmed PV patients meeting the inclusion criteria were enrolled in the study between August 2023 and August 2024. Data collection was carried out by a principal investigator who gathered information on medical history, familial diseases, sociodemographic characteristics (such as age), clinical data (including age at disease onset, PV phenotype, treatments received, and number of relapses).

The Autoimmune Bullous Skin Disorder Intensity Score (ABSIS), which yielded a total score ranging from 0 to 206, was used to evaluate the disease's severity. This score took into consideration subjective discomfort during eating and drinking (0–45), oral involvement (0–11), and cutaneous involvement (0–150). PV cases were categorised into three severity levels based on the overall score: mild (scores between 1 and 10), moderate (scores between 11 and 40), and severe (scores between 41 and 206).

IBM-SPSS 21.0 was used to validate, code, and analyse the gathered data (IBM-SPSS Inc., Chicago, IL, USA). For the analysis, descriptive statistics such as standard deviations, means, ranges, medians, and percentages were calculated.

**Results**

Table 1 presents the demographic characteristics of 50 patients diagnosed with various histopathological types of pemphigus vulgaris. The mean age of the patients was 52.3 years, with a standard deviation of 14.6 years, ranging from 18 to 70 years. The cohort comprised 23 females (46.0%) and 27 males (54.0%). A family history of pemphigus vulgaris was absent in 28 patients (56.0%) and present in 22 patients (44.0%). Regarding clinical phenotypes, 39 patients (78.0%) presented with mucocutaneous pemphigus vulgaris (MCPV), 10 patients (20.0%) with mucosal pemphigus vulgaris (MPV), and 1 patient (2.0%) had skin involvement only as shown in Table 1.

**Table 1 Demographical characteristics of all the patients presented with different histopathological types of pemphigus vulgaris.**

| Variables | Categories | Total (N = 50 (100.0%)) |
|---|---|---|
| **Age (years)** | | |
| | Mean ± SD* | 52.3 ± 14.6 |
| | Minimum-Maximum | 53 (18-70) |
| **Sex** | | |
| | Female | 23 (46.0) |
| | Male | 27 (54.0) |
| **Family history** | | |
| | Absent | 28 (56.0) |
| | Present | 22 (44.0) |
| **Clinical phenotype** | | |
| | Mucocutaneous | 39 (78.0) |
| | Mucosal | 10 (20.0) |
| | Skin | 1 (2.0) |

***SD*** (standard deviation).

Table 2 outlines the clinical parameters of 50 patients diagnosed with various histopathological types of pemphigus vulgaris. At presentation, 47/50 patients (94.0%) reported lesions in the mouth, 17/50 (34.0%) experienced dysphagia and pain, 9/50 (18.0%) had nosebleeds, 12/50 (24.0%) presented with hoarseness, 39/50 (78.0%) had lesions or blisters on the skin, and 7/50 (14.0%) reported red eyes. The onset of the disease was most commonly located in the oropharynx (64.0%), followed by a combination of skin and oropharynx (16.0%), skin alone (18.0%), and the anal region (2.0%). The mean duration of the disease was 44.5 months, with a standard deviation of 39.4 months. The severity of the disease, assessed using the Autoimmune Bullous Skin Disorder Intensity Score (ABSIS), revealed that 21 patients (42.0%) had mild disease, 18 (36.0%) had moderate disease, and 11 (22.0%) had severe disease. Additionally, 21 patients (42.0%) experienced a relapse, while 29 patients (58.0%) did not.

**Table 2 Clinical parameters of all the patients presented with different histopathological types of pemphigus vulgaris.**

| Variables | Categories | Total (N = 50 (100.0%)) |
|---|---|---|
| **Complaints at presentation** | | |
| | Lesion in the mouth | 47/50 (94.0) |
| | Dysphagia, pain | 17/50 (34.0) |
| | Nosebleed | 9/50 (18.0) |
| | Hoarseness | 12/50 (24.0) |
| | Lesion/blister on the skin | 39/50 (78.0) |
| | Red eye | 7/50 (14.0) |
| **Onset of location** | | |
| | Oropharynx | 32 (64.0) |
| | Skin + Oropharynx | 8 (16.0) |
| | Skin | 9 (18.0) |
| | Anal Region | 1 (2.0) |
| **Duration of disease (months)** | | |
| | Mean ± SD* | 44.5 ± 39.4 |
| **Severity of disease** | | |
| | Mild | 21 (42.0) |
| | Moderate | 18 (36.0) |
| | Severe | 11 (22.0) |
| **Relapse cases** | | |
| | No | 29 (58.0) |
| | Yes | 21 (42.0) |

***SD*** (standard deviation), severity of the disease was assessed using the Autoimmune Bullous Skin Disorder Intensity Score (ABSIS). PV cases were classified into three severity levels: ***mild*** (scores from 1 to 10), ***moderate*** (scores from 11 to 40), and ***severe*** (scores from 41 to 206).

## Discussion

Pemphigus is a globally prevalent disease, most commonly affecting individuals aged 40 to 60 years, although it can occasionally be seen in children and older adults. Research indicates that the mean age of onset varies by region, being 46 years in South Africa, 36.7 years in Tunisia, 52 years in Macedonia, and 57 years in Finland [11-15]. Consistent with these findings, the mean age of our patients at diagnosis was 52.3 years, with ages ranging from 18 to 70 years.

While it is generally accepted that pemphigus affects males and females equally, some studies suggest a higher prevalence in females, whereas others report a greater prevalence in males [7, 16-18]. In our study, female versus male proportion was 23 (46.0%) and 27 (54.0%), respectively. Despite the slightly higher prevalence in males, this finding aligns with literature indicating a roughly equal prevalence of pemphigus vulgaris across both sexes.

Although pemphigus can affect other mucosal tissues, it most commonly affects the oral mucosa [16, 19, 20]. According to certain research, up to 100% of patients had mucosal involvement [13, 15]. The study found

that 98% of patients had mucosal involvement, with 94% presenting with oral lesions, 34% with pain or

dysphagia, 18% with nosebleeds, 24% with hoarseness, 78% with skin lesions or blisters, and 14% with red eyes. The oral mucosa was the most frequently impacted region, and mucosal involvement was highly prevalent, in line with the literature [18].

In pemphigus vulgaris (PV), skin symptoms typically manifest after oral mucosa involvement [21]. In our study, 43% of patients first exhibited symptoms in the oral mucosa before they appeared on the skin, while 18% experienced initial skin symptoms followed by oral mucosa involvement. Sixteen percent of individuals experienced simultaneous involvement of the oral mucosa and skin. While 8% of patients had both oropharyngeal and other mucosal involvement, 2% of cases had oral involvement followed by involvement of other mucous membranes (such as the conjunctiva, nasal, and anogenital). Exclusively the oral mucosa was impacted in 10% of individuals, while 2% exclusively experienced dermatological symptoms. These results align with those reported in the literature [18]. In our study, 78% of patients with pemphigus vulgaris (PV) had lesions on both their skin and mucosae. While a Bulgarian study found a lower incidence (64.8%) and another study from our nation claimed a greater prevalence (82.5%), a Macedonian study showed similar results (76%). Additionally, 20% of PV patients had lesions that were limited to the mucosae, according to our data. Although the Bulgarian study showed a lower incidence (10.8%) and a Spanish study a considerably greater prevalence (71%) [15, 22, 23], this is equivalent to the Macedonian study (24%). According to a Bulgarian study, 24.3% of patients had lesions that were only on their skin, while studies from Turkey (6.5%) and Iran (6.4%) showed significantly lower rates of skin-only lesions [22–24]. In our study, only 2% of patients had skin-only lesions, a rate lower than those reported in the literature. The notion that mucosal involvement is more prevalent in PV is supported by this study.

**Conclusion**

The clinical and demographic characteristics of pemphigus vulgaris (PV) patients in our region align closely with findings from other studies conducted in different geographic locations. This consistency across various studies suggests that the clinical presentation and demographic profiles of PV patients may exhibit common patterns irrespective of regional differences. This agreement underscores the reliability of our findings and supports the notion that PV exhibits relatively uniform clinical and demographic features across diverse populations.

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