

**Original Article**

**Pemphigus diseases in a tertiary care hospital: epidemiology and clinical profile**

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**Abstract**

*This cross-sectional study was done to evaluate the pattern of patients suffering from pemphigus disease attending in the department of Dermatology and Venereology, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh from 2006 to 2010. Thirty patients of pemphigus diagnosed clinically and confirmed by histopathology and immunology, were included in the study. Among the thirty patients, 18(60%) had pemphigus vulgaris. Among the patients of pemphigus foliaceus, 13.33% patients were in the age group of 30-45 years, among the patients of pemphigus erythematosus, 6.67% patients were in the age group of 30-45 years and among the patients of paraneoplastic pemphigus, 3.33% patients were in the age group of 46-60 years. Among the different varieties of pemphigus, highest 66.67% had the disease duration of 1 to 6 months. Male were more than female. Highest 76.50 % showed both mucosal and skin involvement as initial clinical presentation. In future, this study should be carried out on a large scale with prolong period, large sample size and in different hospitals in Bangladesh.*

**Key words :** Pemphigus, blistering disorder, pemphoid

**Introduction**

Pemphigus is a group of potentially life threatening rare autoimmune blistering disorder of the skin and mucous membrane, clinically characterized by flaccid, easily ruptured bullae on normal appearing skin or

erythematous base.<sup>1,2,3</sup> Pemphigus vulgaris (PV) frequently begins with oral lesions and progresses to skin lesions.<sup>4</sup> Autoimmune bullous skin disorders are associated with IgG or IgA auto- antibodies against distinct adhesion molecules of the epidermis and dermal epidermal basement membrane zone, respectively. These auto- antibodies lead to a loss of skin adhesion which shows up clinically as the formation of blisters or erosions. In pemphigus, loss of adhesion occurs within the epidermis while in the pemphigoids, linear IgA dermatosis, epidermolysis bullosa acquisita and dermatitis herpetiformis, loss of adhesion takes place within or underneath the basement membrane zone. The auto-antigens of these disorders are largely identified and characterized.<sup>1-4</sup> The diagnosis of autoimmune bullous skin diseases is based on histology and direct immunofluorescence of perilesional skin and the serological detection of auto antibodies by indirect immunofluorescence and recombinant autoantigens.<sup>5</sup> Two major variants of pemphigus is identified by the level of cleavage within the epidermis, by different clinical pattern and by the auto antibodies. Suprabasal cleaving is seen in pemphigus vulgaris (PV) and its variants pemphigus vegetans. Most superficial subcorneal bullae are formed in pemphigus foliaceus (PF) and pemphigus erythematosus (PE) and presented as superficial blisters on the seborrheic areas of the body. In addition there may be paraneoplastic pemphigus, drug induced pemphigus, IgA pemphigus.<sup>4-6</sup> The aim of this study was to evaluate the clinical presentation of pemphigus group of patients attending in the tertiary care hospital.

**Methods**

This cross-sectional study was done in the department of Dermatology and Venereology, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh from 2006 to 2010. Thirty patients of more than 30 years of age, both male and female, suffering from pemphigus diagnosed clinically and confirmed by histopathology and immunology, were included in the study. Patients with systemic disorder such as diabetes mellitus, hematologic disturbances and severe physically debilitated subjects were excluded from the study. The patients were selected purposively. Data were collected through face to face interview by using a semi-structured questionnaire.

**Results**

Among the patients, 18(60%) had pemphigus vulgaris,

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8(26.67%) had pemphigus foliaceus, 2 (6.67%) had pemphigus erythematosus and 2(6.67%) had paraneoplastic pemphigus (Figure -1). Among the patients of pemphigus vulgaris, 10% patients were in the age group of 30-45 years, 20% patients were in the age group of 46-60 years, 30% patients were in the age group of 61-75 years. Among the patients of pemphigus foliaceus, 13.33% patients were in the age group of 30-45 years, 10% patients were in the age group of 46-60 years, 3.33% patients were in the age group of 61-75 years. Among the patients of pemphigus erythematosus, 6.67% patients were in the age group of 30-45 years. Among the patients of paraneoplastic pemphigus, 3.33% patients were in the age group of 46-60 years. (Table-I)

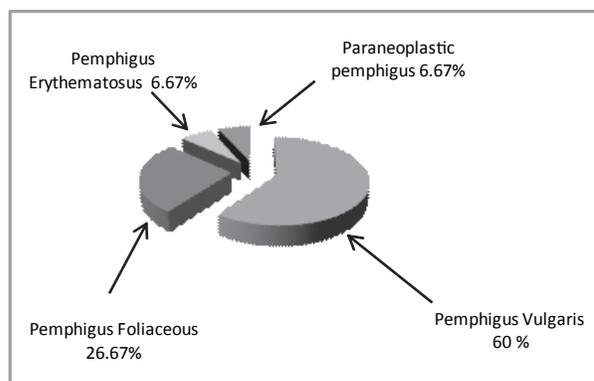


Figure -1: Distribution of the patients by different types of pemphigus

Table - I: Distribution of patients by age

Age in years	Pemphigus vulgaris	Pemphigus foliaceus	Pemphigus erythematosus	Paraneoplastic pemphigus
30-45	3(10%)	4(13.33%)	2(6.67%)	0
46-60	6(20%)	3(10%)	0	1(3.33%)
61-75	9(30%)	1(3.33%)	0	1(3.33%)

Among the different varieties of pemphigus, 66.67% had the duration 1 to 6 months, 16.67% had the duration 7

months to 1 year, 10% had the duration 1 to 2 years and 6.67% had the duration more than 2 years (Table-II)

Table-II: Distribution of patients by duration of the diseases

Duration of disease	Pemphigus vulgaris	Pemphigus foliaceus	Pemphigus erythematosus	Paraneoplastic pemphigus
1 to 6 months	14(46.67%)	4(13.33%)	2(6.67%)	0
7 to 12 months	2(6.67%)	2(6.67%)	0	1(3.33%)
13 to 18 months	1(3.33%)	1(3.33%)	0	1(3.33%)
More than 18 months	1(3.33%)	1(3.33%)	0	0

Among the patients of pemphigus vulgaris, male- female ratio was 1.3:1, among the patients of pemphigus foliaceus, male- female ratio was 1.6:1, and only 2 male

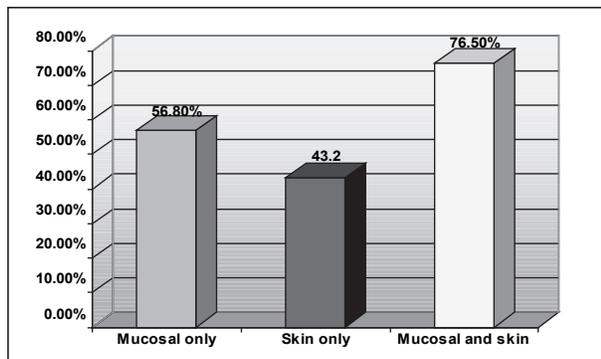
patients were in each pemphigus erythematosus and paraneoplastic pemphigus group. (Table-III)

Table-III: Distribution of the patients of pemphigus by sex.

Type of pemphigus	Sex		Ratio
	Male	Female	
Pemphigus vulgaris	10(33.33%)	8(26.67%)	1.3:1
Pemphigus foliaceus	5 (16.67%)	3(10.0%)	1.6:1
Pemphigus erythematosus	2(6.67%)	0	2:0
Paraneoplastic pemphigus	2(6.67%)	0	2:0
Total	19(63.33%)	11(36.67%)	

In 56.8% of patients, the initial presentation of the disease was only mucosal involvement, in 43.2% of the

disease initially presented by the skin only and 76.50 % showed both mucosal and skin involvement. (Figure-2)



**Figure-II:** Distribution of patients by initial clinical presentation of the disease

### Discussion

Among the patients, 18(60%) had pemphigus vulgaris, 8(26.67%) had pemphigus foliaceus, 2(6.67%) had pemphigus erythematosus and 2(6.67%) had paraneoplastic pemphigus. Yazdanfar conducted a study with two hundred new patients of pemphigus during the 10 year period in Iran, also observed pemphigus as most common form in his study.<sup>7</sup> Among the patients of pemphigus the mean age of onset was  $56.1 \pm 9.7$  years, ranging from 36 to 68 years. This finding is not similar to the study finding of Asilian et al. they conducted a study on 188 patients of pemphigus vulgaris in Alzahra University Hospital, Iran for 10 year period. Their mean age of onset was  $41.1 \pm 13.7$  years.<sup>8</sup>

Among the patients of pemphigus vulgaris, male - female ratio was 1.3:1; among the patients of pemphigus foliaceus, male- female ratio was 1.6:1; and only 2 male patients were in each pemphigus erythematosus and paraneoplastic pemphigus group. This finding is not similar to the study finding of Asilian et al and Esmaili et al. Asilian et al found a male : female ratio of 1.6: 1 in his study n pemphigus vulgaris patients in Iran.<sup>8</sup> Esmaili et al conducted a study of on 140 patients with a 1.59: 1 ratio of women compared with men among pemphigus vulgaris patients in Iran.<sup>9</sup>

In 56.8% of patients, the initial presentation of the disease was only mucosal involvement. On the other hand, 43.2% of the disease initially presented in the skin only and 76.50 % showed both mucosal and skin involvement. Asilian et al found mucosal involvement in 74% of patients as initial presentation of the disease and in the remaining 26%, the disease initially presented on the skin.<sup>8</sup> Esmaili et al observed oral cavity as initial localization in 77.5% cases.<sup>9</sup> Yazdanfar was also seen mucosal involvement as first manifestation in 60% cases among the total mucosal involvement of 84.5% cases.<sup>7</sup> The study showed that among the thirty cases of the

pemphigus patients, the most common clinical variant was pemphigus vulgaris. Pemphigus vulgaris was common in the age group of 61-75 years and pemphigus foliaceus, pemphigus erythematosus and paraneoplastic pemphigus were common in the age group of 30-45 years. Male was more than female and both mucosal and skin involvement as initial clinical presentation was highest among the pemphigus group of diseases.

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