



## Clinicoepidemiological Profile of Pemphigus

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### Abstract

**Introduction:** Pemphigus refers to a group of acquired, chronic autoimmune blistering disorders presenting clinically by flaccid intraepidermal blisters, erosions and ulcerations on skin and mucosa. The study was aimed at evaluating the epidemiological and clinical features of pemphigus.

**Methods:** After taking history of clinically diagnosed pemphigus cases, characteristics of the lesions were noted. Extent of involvement of both skin and mucosa was graded using appropriate methods. Data was analysed using statistical package for social science version 10.

**Results:** Among the 30 patients, onset of the disease was noted in the younger age group in 16.7%. In majority of the patients, duration of the disease was less than 6 months. Onset of the disease on mucosa alone was seen in 50% cases and only 3.3% had mucocutaneous onset. Pain or burning sensation was present in 60% cases and pruritus in 40%. Based on body surface area, 53.3% had moderate degree and 26.7% had severe degree of skin involvement. Nikolsky's sign was positive in only 75% patients. Two pemphigus foliaceus cases had oral lesions and 1 had nasal lesions. Grade 1 oral mucosal involvement was found in 40.2% cases and 16.7% had grade 3 involvements. Two pemphigus vulgaris patients had paronychia and oral candidiasis was present in 12 patients.

**Conclusion:** Severe mucocutaneous pemphigus and onset in the younger age group may have social and medical implications. Nikolsky's sign was found as less sensitive. Mucosal involvement was seen in pemphigus foliaceus patients.

**Keywords:** pemphigus vulgaris, foliaceus.

### Introduction

The term pemphigus refers to a group of autoimmune blistering diseases of skin and mucous membranes, characterized histologically by intraepidermal blisters due to acantholysis and immunopathologically by in vivo bound and circulating IgG directed against desmogleins, which are adhesion molecules functioning in

desmosomes of the keratinocytes. Primary lesion is a flaccid blister arising from normal – looking or erythematous skin which rupture easily.<sup>1</sup> Itching may precede the development of a fresh crop of lesions. Most common lesions are painful erosions having a tendency to extend peripherally. This finding can be elicited by the phenomenon, Nikolsky's sign.

Almost all pemphigus vulgaris patients have oral involvement. There may be candidal superinfection.<sup>2</sup> Erosions may involve pharynx and larynx causing hoarseness of voice. Other mucosae namely oesophageal, conjunctival, cervical, urethral and anal mucosa may be affected.

Pemphigus vulgaris and pemphigus foliaceus share the same chronic course. The factors which can be considered as predictive of the course of disease are severity at onset and response to treatment. Patients with mild to moderate disease enter complete and long remission twice as likely compared to those with severe disease.<sup>3</sup>

Even though the age group described for pemphigus is 40 - 60 years, many cases have an earlier age of onset with added medical and social implications. Various demographic and other epidemiological factors may be important in the progression of the disease. Some aim to study the clinical profile and various epidemiological factors in pemphigus.

## Materials and Methods

**Aim:** To study the clinical and epidemiological profile of pemphigus and its variants.

After getting consent, history of the patients were taken including the age of onset of the disease, duration and site of onset of the lesions. During dermatological examination, characteristics of lesions, sites and extent of involvement on both skin and oral lesions were recorded.

A simple scoring system was used to grade the severity of skin based on body surface area (BSA) involvement at the time of sampling.

Skin score -

1. Mild disease (<10% BSA involved)
2. Moderate disease (11-30% BSA)
3. Severe (>30% BSA).

The extent of oral lesions was rated into 3 grades as follows using Camacho-Alonso et al grading.<sup>4</sup>

- |         |   |                           |
|---------|---|---------------------------|
| Grade 1 | - | only 1 site involved      |
| Grade 2 | - | 2 sites involved          |
| Grade 3 | - | 3 or more sites involved. |

Investigations were done to confirm the diagnosis. All the data were entered in the proforma and were analysed using the computer software, statistical package for social science (SPSS) version 10. The data was given in frequency and percentage. The associations and comparisons between different parameters were elucidated using the nonparametric test- chi square (X<sup>2</sup>) test. A two-tailed distribution was assumed for all statistical evaluations and probability value <0.05 was considered significant.

## Results

There were 27 pemphigus vulgaris (PV) and 3 pemphigus foliaceus (PF) cases. Youngest age of onset noted was 16 years and oldest being 87 years with a mean age of onset 45.77 years. In 5 cases (16.7 %), onset of disease was noted in younger age group (15-29). Out of these 5 patients, 4 were females.

Majority (50%) of the patients presented within 6 months of onset of the disease with a range of 1 month to 2 and half years (Table 1). Pain or burning sensation was present in 60% cases and pruritus in 40%. Eleven (40.7%) pemphigus vulgaris patients presented with pain and 66.7% of pemphigus foliaceus presented with pruritus.

Onset of the disease was noted in mucosa alone in 15 cases (50%) and on skin alone in 14 patients (46.7%). In one patient, onset was on skin and mucosa simultaneously (3.3%). Eighteen patients (60%) had a wide range of interval between onset of lesions on mucosa and skin, of 1 week to 1 year (Figure 1). Two patients had only oral lesions and 3 had no oral lesions.

Either vesicles or bullae as primary lesion were present in 80% cases. One patient with pemphigus vulgaris had only tense vesicles. Exfoliative dermatitis like picture was seen in a patient with pemphigus foliaceus. In 3 pemphigus vulgaris cases, vegetative lesions were present. Nikolsky's sign could be elicited in 28 patients and positive result was seen in 75 % cases. Based on body surface area, 53.3% had moderate degree and

26.7% had severe degree of skin involvement (Table 2).

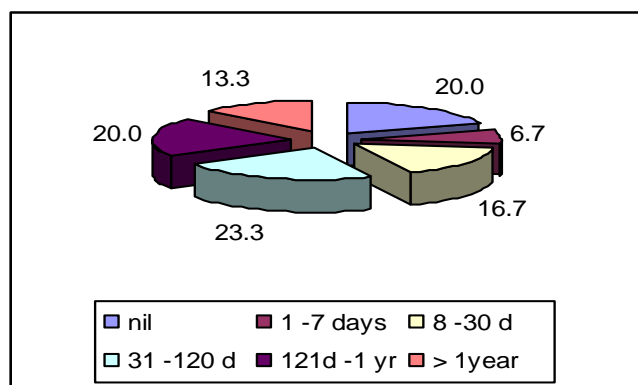
Twenty seven cases had mucosal involvement. Among them, oral mucosa was involved in majority (86.7%). All the 3 pemphigus foliaceus cases had mucosal involvement. Among them, 2 patients had oral lesions and 1 had nasal involvement. Grade 1 oral mucosal involvement was found in 40.2% patients and 16.7% had severe grade 3 involvements according to number of sites involved. Two pemphigus vulgaris cases had paronychia and oral candidiasis was present in 12 patients.

**Table 1** Duration of disease

Duration	Count	Percent
1- 6months	15	50.0
7-12m	7	23.3
13 -18 m	1	3.3
19- 24 m	4	13.3
.>25 m	3	10.0

Duration of the disease in months.

**Figure 1** Distribution of interval



Interval between the onset of skin and mucosal lesions /mucosa and skin lesions.

**Table 2** Extent of skin lesions

Extent	Count	Percent
Nil	2	6.7
< 10 %	4	13.3
11- 30 %	16	53.3
> 30 %	8	26.7

Extent of skin involvement based on body surface area (BSA).

### Discussion

Out of the 30 patients with pemphigus, 27(90%) were pemphigus vulgaris and 3(10%) were

pemphigus foliaceus cases. In the study by Ramassamy S et al<sup>5</sup> consisting of 132 patients with pemphigus, 89.4% had pemphigus vulgaris and 10.6% had pemphigus foliaceus which was similar to the present study. Youngest patient was 16 years and oldest was 87 years with a mean of 45.77 years. The minimum age was found to be 11 and maximum was 80 years with an average age of 45.9 years, in the study done by Asia AJ et al<sup>6</sup> which was almost conforming to this study. There were a sizeable number of patients (16.7%) with onset in the younger age group of 15 -29 years with female predominance. The number of patients in the age group less than 30 years was at a lower rate of 11.76% (26/ 221) in a study done by Xiaoling Z et al, but here also females predominated.<sup>7</sup> Also in the study by Mahajan A et al, it was only 11.4%( 4/35 ).<sup>8</sup>

Majority (50%) of the patients presented within 6 months of onset of the disease. The duration of the illness was longer in the study by Chowdhury J et al which ranged upto 108 months.<sup>9</sup> In a study done by Mahajan A et al also the duration of the disease was more, where the mean duration of pemphigus was 9.24 months.<sup>8</sup> Pain or burning sensation was present in 60% cases and pruritus in 40% in the present study. In the 11 year review by Chmurova N et al<sup>2</sup>, pain was present in 68.3%, burning sensation in 30.7% and itching in 30.6% patients, which was almost similar to this study.

Onset of the disease was noted in mucosa alone in 50%, on skin alone in 46.7% and on both skin and mucosa in 3.3%. Compared to the present study, number of patients with onset of the disease on skin alone was less (33.3 %) in the study by Suliman NM et al among 16 patients.<sup>10</sup> They reported 50.0% with oralmucosa as the initial site of involvement and 5.5% had simultaneous involvement of both skin and oral mucosa which was comparable to this study. Shamim et al got primary site of involvement as oral cavity in 53.52 % cases which was similar to the present study but skin (22.53%) as initial site and simultaneous mucocutaneous onset (23.94 %) was not in conformity.<sup>11</sup> Kavusi S has shown

lower rate of remission (31.9%) in mucocutaneous pemphigus patients compared to those with only mucosal or cutaneous (48.6%) involvement. Relapses were also more frequent in mucocutaneous subtype.<sup>12</sup>

The interval between onset of lesions on mucosa and skin was between 1 month and 3 months in 23.3 % patients. Skin lesions may appear several weeks to a year later, causing a delay in diagnosis. The study by Huda MM et al has shown development of cutaneous lesions on an average of 4.5 months after the onset of oral lesions.<sup>13</sup>

Either vesicles or bullae as primary lesion were present in 80 % patients. Three pemphigus vulgaris patients had vegetative lesions. Flaccid bullae were found in all 68 cases by Arya S.R et al in contrast to this study. They have also found blisters on non erythematous skin in 86.76% and erosions in 55.88%. Crusted lesions, erythematous plaques, vegetations and pustules were present less frequently in their study.<sup>14</sup> More number of pemphigus vulgaris presented with only vesicles (19.5%) and majority of the patients (65.9%) presented with flaccid vesicles and bullae followed by concomitant pustules (4.9%) in the study done in Eastern India (Chowdhury J et al).<sup>9</sup>

Nikolsky's sign was positive in only 75 % cases. In the study by Arya SR et al<sup>14</sup> it was found positive in 97.2%, not conforming to the present study. Uzun S et al found Nikolsky's sign as moderately sensitive (69%) and highly specific (100%) for pemphigus.<sup>15</sup> Based on body surface area, 53.3% had moderate degree and 26.7% had severe degree of skin involvement. Asia AJ et al got almost similar values with severe form of skin disease (28.9%) but 27.7 % of patients had mild and only 43.3% had moderate skin involvement.<sup>6</sup>

Three pemphigus foliaceus cases had mucosal involvement. Similarly out of 8 PF cases, two (25%) had lesions in oral mucosa (Chowdhury J et al).<sup>9</sup> There was no mucosal involvement in patients with pemphigus foliaceus in the study done in Iran by Javidi Z et al.<sup>16</sup> Grade 1 oral mucosal involvement was present in 40.2% and 16.7% had severe grade 3 involvement. Contrary

to this, Shamim et al had 53.53% with grade III oral mucosal involvement followed by grade II in 28.17% and grade I in 18.30%.<sup>11</sup>

The number of patients with nail apparatus involvement (paronychia) was not consistent to the study by Pietkiewicz P et al <sup>17</sup>(13.4%) which was only 6.6 % in the present study. Oral candidiasis was present in 40 % patients which was not in conformity to the study by Esmaili N et al (23.87 %).<sup>18</sup>

### Conclusion

Pemphigus vulgaris was the common form in our study. Disease onset in the younger age group was found and pemphigus foliaceus had mucosal lesions. Nikolsky's sign was not found as sensitive in the study. Apart from the known pathogenic mechanisms, other factors could be implicated for these varying presentations. Course of the disease may be prolonged with severe and extensive cutaneous and mucosal lesions. These can have both medical and social consequences. Further studies are required aimed at these factors.

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