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# Prevalence of Pemphigus vulgaris and Pemphigus Foliaceus in tertiary care hospital in India: an update

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Pemphigus is a life threatening autoimmune chronic blistering disease that involves in the squamous epithelia and mucous membrane of skin. Pemphigus shows effect on quality of life of patients. The data was collected from in-patient record book in the Department of Dermatology in a tertiary care hospital in India for a period of 2 years (April 2011 to March 2013). In this study overall 274 inpatients were recorded in the in-patient record book in the Department of Dermatology in a tertiary care hospital for a period of 2 years (April 2011 to March 2013). Among the patients, 207(75.54%) had Pemphigus vulgaris, 67(24.46%) had Pemphigus foliaceus. In our study age group of 31-40 mainly having Pemphigus vulgaris and age group of 51-60 mainly having Pemphigus Foliaceus. In our study most of the females (129 patients out of 207 (62.32%)) having Pemphigus vulgaris and most of the males (44 patients out of 67 (65.67%)) having Pemphigus Foliaceus disease.

*Keyword:* Pemphigus Vulgaris (PV), Pemphigus Foliaceus (PF), Prevalence, male-female ratios.

## 1. Introduction

The word Pemphigus derived from pemphix, a Greek word that means blister. Pemphigus was first demonstrated in 1943, acantholysis is the characteristic feature of the bullae of Pemphigus vulgaris. It is a life threatening autoimmune chronic bullous disease that involves the squamous epithelia and mucous membranes, manifested as painful erosions, loose blisters, and ulceration of skin and mucous membrane [1-3].

Pathogenesis of Pemphigus is the formation of autoantibodies against desmoglein. Desmoglein forms the "glue" that attaches adjacent epidermal cells via attachment points called Desmosomes. When autoantibodies attack desmogleins, the cells become separated from each other and the epidermis becomes "unglued", a phenomenon called acantholysis [2]. (IgG autoantibodies cause intraepidermal blisters as a result of acantholysis. In immunopathology, we can find circulating IgG acting against desmoglein I and III) this causes blisters that slough off and turn into sores [4]. In some cases, these blisters can cover a significant area of the skin [1]. Recent studies have shown that acantholysis can also occur in presence of antibodies against 9-α nicotinic acetylcholine receptor. Apoptosis of keratinocyte may have a possible role in pathogensis [5].

The common types include Pemphigus vulgaris (PV); its variants Pemphigus vegetants (PVeg) and Pemphigus foliaceus (PF) with its variant Pemphigus Erythematosus (PE). Recent studies described the variants are Pemphigus herpetiformis, IgA Pemphigus, and paraneoplsatic Pemphigus [6]. The disease that mainly involved in different parts of the body. The onset of the disease, the major sites of involvement are as follows:

- Oral mucosa
- Skin surfaces
- Skin and oral mucosal involvement
- Other mucous membranes such as esophagus and vaginal mucosa without oral mucosa and skin involvement.

Pemphigus vulgaris (PV) is the most widespread form of the disorder and occurs when antibodies attack *Desmoglein* 3. PV may occur at any age and it is most common among people between the ages of 40 and 60 [7]. Pemphigus patient having large and flaccid bullae which break easily and are seen in oral mucosa, sternum, midface, scalp and groin. Sores frequently originate in the oral cavity. Identification of PV in a Patients with active disease have circulating and tissue-bound autoantibodies of both the immunoglobulin G1 (IgG1) and immunoglobulin G4 (IgG4) sub classes .The identification of PV by histopathologically and using immunofluorescence test. Histopathologically, there is characteristic formation of suprabasal clefts and appearance of the 'row of tombstone' [8, 9]. The immunofluorescent test is Positive direct immunofluorescence testing is the gold standard for diagnosis [8-10]. Immunofluorescence testing shows IgG antibodies in more than 75% of the cases and anti-nuclear antibodies in 30-80% of the patients [10, 11].

Pemphigus foliaceus (PF) generally affects middle-aged individuals like 40-60 old age people [7]. PF is the least severe of the three varieties. *Desmoglein 1*, the protein that is destroyed by the autoantibody, is only appeared in the top dry layer of the skin. PF is characterized by brittle sores that frequently begin on the scalp, and may progress to the chest, face, and back. The main difference may observe in PF compared with PV Mouth sores do not occur. Histopathology shows acantholysis in upper epidermis within or adjacent to granular layer leading to sub corneal bullae. In PF immunofluorescence test is used for the identification of disease, the test shows antibodies against desmoglein I [10].

## 2. Epidemiology

The disease has worldwide distribution affecting 0.1–0.5 patients per 100 000 population per year. The majority of patients have been diagnosis to have Pemphigus Vulgaris (PV) and some of patients have Pemphigus Foliceus (PV) [12, 13]. In an epidemiological study of patients treated in a Finnish hospital between 1969 and 1978, the mean age of onset was found to be 57.5 years and the male to female ratio was 0.9:1.0 [14]. In a survey of 110 cases in Italy, the median age at diagnosis was 54 years and the male to female ratio was 0.7:1.0 [13].

The Observational non analytical retrospective study was conducted by observation of the medical records of all the newly registered patients with Pemphigus at Sanglah General Hospital in Bali-Indonesia during the period of January 1995 and December 2002 Analyzed with regard to personal statistic: During the 8-years period of studies, 33 Pemphigus patients were admitted. Among the patients 20 (60.6 %) female patients and 13 (39.4 %) male patients. [14,15] A number of studies have suggested a higher incidence in women [16, 17].

A cross-sectional study was done in the department of Dermatology and Venereology in three different hospitals in Bangladesh for a duration of 2006 to 2010.Among the patients, 18 (60%) patients had Pemphigus vulgaris, 8(26.67%) patients had Pemphigus foliaceus and reaming other variants [18]. In a comparative study of Pemphigus in Tunisia and France, it was observed that in France Pemphigus vulgaris accounted for 73% of all cases, incidence increased with age and the male to female ratio was 1:1.2 [13, 14].

Literature on Pemphigus has flourished subsequently in India. Due to lack of facilities to elucidate the molecular aspects of the disease in most of the centers, Indian literature on Pemphigus has largely remained restricted to epidemiology and treatment with some recent research on immunology [19, 20].

## 3. Patients and methods

The data was collected from in-patient record book in the Department of Dermatology in a tertiary care hospital in India (Gauhati Medical College and Hospital) for a period of 2 years (April 2011to March 2013).

## 4. Discussion

A total of 274 patients were recorded in the in-patient record book of the Department of Dermatology in a tertiary care hospital for a period of 2 years (April 2011to March 2013). Among the patients, 207(75.54%) had Pemphigus vulgaris, 67(24.46%) had Pemphigus foliaceus (Table 1).

Among the patients of Pemphigus vulgaris (207), 13 (6.28%) patients were in the age group of 11-20 years, 35 (16.90%) patients were in the age group of 21-30 years, 82 (39.61%) patients were in the age group of 31-40 years. 24(11.59%) patients were in the age group of 41-50 years. 35 (16.90%) patients were in the age group of 51-60 years (Table 2)

**Table 1:** The no of Pemphigus Vulgaris (PV) and Pemphigus Foliaceus (PF) reported in 2011-2013

| | PEMPHIGUS VULGARIS (PV) | | PEMPHIGUS FOLIACEUS(PF) | |
|---|---|---|---|---|
| MONTH | 2011-12 | 2012-13 | 2011-12 | 2012-13 |
| APR | 6 | 12 | 2 | 4 |
| MAY | 7 | 17 | 1 | 3 |
| JUN | 4 | 14 | 1 | 8 |
| JUL | 4 | 10 | 2 | 4 |
| AUG | 3 | 10 | 1 | 2 |
| SEP | 9 | 9 | 3 | 3 |
| OCT | 6 | 8 | 1 | 4 |
| NOV | 8 | 9 | 2 | 4 |
| DEC | 9 | 5 | 3 | 2 |
| JAN | 10 | 8 | 4 | 3 |
| FEB | 12 | 9 | 1 | 2 |
| MAR | 10 | 8 | 1 | 2 |
| TOTAL | 88 | 119 | 23 | 44 |
| OVERAL TOTAL | | 88+119=207 | | 23+44=67 |
| | | | | 207+67=274 |

**Table 2:** Age and sex distribution of Pemphigus Vulgaris (PV) patients from April 2011 to March 2013. M/F= Male Female ratio.

| AGE AND SEX DISTRIBUTION OF THE PV PATIENTS FROM APRIL 2011 TO MARCH 2013 | | | | |
|---|---|---|---|---|
| | PEMHIGUS VULGARIS(PV) | | | |
| AGE GROUP (YEARS) | MALE | FEMALE | MALE+FEMALE | M/F SEX RATIO |
| UPTO 10 yr | 0 (0%) | 0(0%) | 0(0%) | 0 |
| 11yr-20yr | 1(0.48%) | 12(5.80%) | 13(6.28%) | 1.0:12.0 |
| 21yr-30yr | 11(5.31%) | 24(11.59%) | 35(16.90%) | 1.0:2.18 |
| 31yr-40yr | 26(12.56%) | 56(27.05%) | 82(39.61%) | 1.0:2.15 |
| 41yr-50yr | 16(7.73%) | 8(3.86%) | 24(11.59%) | 1.0:0.5 |
| 51yr-60yr | 12(5.80%) | 23(11.10%) | 35(16.90%) | 1.0:1.9 |
| 61yr-70yr | 11(5.31%) | 5(2.41%) | 16(7.72%) | 1.0:0.45 |
| 71yr-80yr | 1(0.48%) | 1(0.48%) | 2(0.96%) | 1.0:1.0 |
| TOTAL | 78(37.68%) | 129(62.32%) | 207(100%) | 1.0:1.65 |
| | | | | |
| | | | 78+129=207(75.54%) | |

Among the patients of Pemphigus foliaceus (67%), 1 (1.50%) patients was in the age group of 11-20 years, 16(23.95%) patients were in the age group of 21-30 years, 11(16.43%) patients were in the age group of 31-40 years. 8 (11.95%) patients were in the age group of 41-50 years. 20(29.85%) patients were in the age group of 51-60 years (Table 2).

Among the patients of Pemphigus vulgaris, male female ratio was 1:2.18 in the age group of 21-30 years; male-female ratio was 1:2.15 in the age group of 31-40 years and 1:1.9 in the age group of 51- 60 years. Among the patients of Pemphigus foliaceus, male-female ratio was 1:0.6 in the age group of 21-30 years, male- female ratio was 1:0.22 in the age group of 31-40 years and 1:0.33 in the age group of 51- 60 years (Table 3).

**Table 3:** Age and sex distribution of Pemphigus Foliaceus (PF) patients from April 2011 to March 2013. M/F= Male Female ratio.

| AGE AND SEX DISTRIBUTION OF THE PF PATIENTS FROM APRIL 2011 TO MARCH 2013 | | | | |
|---|---|---|---|---|
| | PEMPHIGUS FOLIACEUS(PF) | | | |
| AGE GROUP (YEARS) | MALE | FEMALE | MALE+FEMALE | M/F SEX RATIO |
| UPTO 10 | 0(0%) | 0(0%) | 0(0%) | 0 |
| 11yr-20yr | 0(0%) | 1(1.50%) | 1(1.50%) | 0 |
| 21yr-30yr | 6(8.95%) | 10(15.0%) | 16(23.95%) | 1.0:0.6 |
| 31yr-40yr | 9(13.43%) | 2(3.0%) | 11(16.43%) | 1.0:0.22 |
| 41yr-50yr | 6(8.95%) | 2(3.0%) | 8(11.95%) | 1.0:0.33 |
| 51yr-60yr | 15(22.39%) | 5(7.46%) | 20(29.85%) | 1.0:0.33 |
| 61yr-70yr | 7(10.45%) | 3(4.47%) | 10(14.92) | 1.0:0.42 |
| 71yr-80yr | 1(1.50%) | 0(0%) | 1(1.50%) | 0 |
| TOTAL | 44(65.67%) | 23(34.33%) | 67(100%) | 1.0:0.52 |
| | | | | |
| | | 44+23=67(24.46%) | | |

Among the patients of Pemphigus Vulgaris (PV) the dominating age group was 31 to 40 years whereas in the case of Pemphigus Foliaceus (PF) the patients are mainly in the age group of 51 to 60 years.

## 5. Conclusion

The study showed that among the 274 patients 207 patients were having Pemphigus Vulgaris (PV) and 67 patients were having Pemphigus Foliaceus (PF). In our study, the patients in the age group of 31- 40 are mainly having Pemphigus Vulgaris and the patients in the age group of 51-60 are mainly having Pemphigus Foliaceus. In the study most of the females (129 patients out of 207 (62.32%) are having Pemphigus Vulgaris and most of the males (44 patients out of 67 (65.67%) are having Pemphigus Foliaceus disease.

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## 7. Conflicts of Interest

Nil

## 8. References

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