

Clinical Study of Pemphigus in Lumbini Medical College

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ABSTRACT

Background: Pemphigus is a worldwide disease and varies in its clinical profile and epidemiology in different regions of the world. The disease is rare and few epidemiological data are available. **Objective:** The purpose of this study was to evaluate the epidemiologic, clinical, and therapeutic features of pemphigus in Lumbini Medical College. **Methods:** We retrospectively investigated a total of 70 patients with pemphigus. The parameters including age at diagnosis, sex, diagnostic methods and treatment outcome of the disease. **Results:** The mean age at the onset was 41.3 years. The male-to-female ratio was 1.18:1. Most of the patients were cured at the end of the treatment. Rest were clinically improved. **Conclusion:** Pemphigus vulgaris (PV) is the most common clinical subtype in our centre. The disease is more frequent in the fifth decade of life and has a male predominance in our region.

Keywords: Pemphigus, dermatological disease, epidemiology, clinical, therapeutic

INTRODUCTION

The term pemphigus refers to a group of autoimmune blistering diseases of the skin and mucous membranes characterized histologically by intraepidermal blister and acantholytic cells and immunopathologically by the finding of IgG, IgA and C₃ deposited on the surface of the keratinocytes.^{1,2,3,4} In the patient's serum, indirect immunofluorescence demonstrates the presence of circulating pemphigus autoantibodies that bind to epidermis. The titer of circulating antibody correlates with disease course.^{5,6,7}

Pathogenesis of pemphigus is the binding of autoantibodies to keratinocyte cell surface molecules. These intercellular or pemphigus antibodies bind to keratinocyte desmosomes and to desmosome-free areas of the keratinocyte cell membrane. The binding of autoantibodies results in blisters and a loss of cell-cell adhesion, a process termed acantholysis.^{8,9}

Pemphigus has the tendency to recurrent and its clinical symptoms is varied. Prognosis is worse in patients with extensive lesion, especially when associated with infection.¹⁰ Therefore, the diagnosis and treatment has been one of the issues to focus on to the dermatologists. That is why we choose pemphigus to study its features and treatment effect.

We analyzed and summarized the clinical manifestations and the treatment of pemphigus to supply the reference of clinical diagnosis and treatment.

MATERIAL AND METHODS

All patients diagnosed as pemphigus from the out and

in-patient Department of Dermatology and Venereology of Lumbini Medical College were selected for the study. Their records were reviewed retrospectively. Patients visiting from 1st of June 2009 to 31st of May 2013 were included in the study.

Socio-Demographic data and subtype of disease were studied. All the data was entered into Microsoft Excel and statistical analyses were performed by the SPSS17.0 software package and expressed with mean and standard deviation.

RESULTS

There were a total of 70 patients enrolled in the study. Out of those, 38(54%) were male and 32(46%) were female. The extent of patient's age was from 16 to 82 years old (mean 41.3 ± 28.5).

Disease was classified into following types:

1. Pemphigus vulgaris
2. Pemphigus vegetans
3. Pemphigus foliaceus
4. Pemphigus erythematosus
5. IgA pemphigus
6. Herpetiform pemphigus
7. Paraneoplastic pemphigus

There were 42 cases of Pemphigus vulgaris accounting for 60% of all those cases. Sixteen cases (22.8%) were diagnosed as pemphigus foliaceus. Eight cases (11.4%) were of Pemphigus erythematosus variety. There were 4 cases (5.8%) diagnoses as Pemphigus vegetans. There

were no cases of IgA, Paraneoplastic and Herpetiform pemphigus.

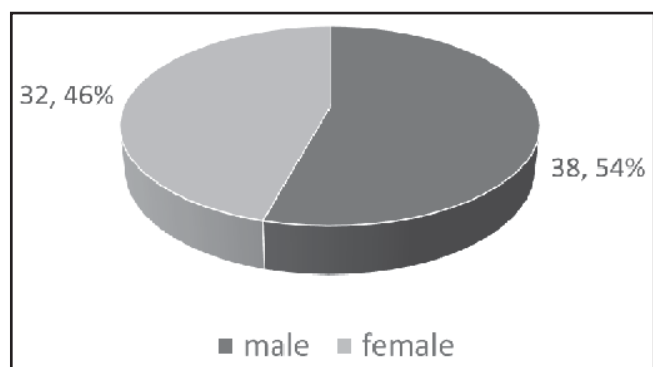


Fig 1: Sex distribution

THERAPEUTIC EFFECT:

Pemphigus vulgaris: Out of 42 cases, 32 were clinically cured and 8 cases were improved.

Pemphigus foliaceus: All the 16 cases were cured.

Pemphigus erythematosus: All 8 cases were cured.

Pemphigus vegetans: All 4 cases were cured as shown in table 2.

Table-1: Types of Pemphigus

Disease	Number	Percentage
Pemphigus vulgaris	42	60
Pemphigus vegetans	4	5.8
Pemphigus foliaceus	16	22.8
Pemphigus erythematosus	8	11.4
IgA pemphigus	0	0
Herpetiform pemphigus	0	0
Paraneoplastic pemphigus	0	0

Table 2: Results after treatment

Disease	Total	Cured	Improved
Pemphigus vulgaris	42	34	8
Pemphigus vegetans	4	4	NA
Pemphigus foliaceus	16	16	NA
Pemphigus erythematosus	8	8	NA
IgA pemphigus	NA	NA	NA
Herpetiform pemphigus	NA	NA	NA
Paraneoplastic pemphigus	NA	NA	NA

NA= Not available or Non applicable

DISCUSSION

Pemphigus is a worldwide disease and varies its presentation, clinical profile, and incidence in different regions of the world. In general, the incidence ranges from 0.76 to 5 new cases per million per year.¹¹

There was a male dominance in our study with M:F ratio of 1.18. Overall, the sex ratio for pemphigus appears to be equivalent or close to equivalent. However, a few studies have found large imbalances in the sex

distribution, such as a study that found a 4:1 ratio of females to males with pemphigus foliaceus in Tunisia and a study that found a 19:1 ratio of males to females in an endemic location in Columbia.^{12,13}

Pemphigus vulgaris was the most common variety in our study. This result was comparable to those reported in the majority of the studies.^{12,13}

Treatment was very effective in early and limited diseases with 100% cure rate in many cases. Some advanced cases were partially improved. Similar results were shown by studies.^{14,15,16}

CONCLUSIONS

Pemphigus is a common dermatological disease. It affects males more than females. Pemphigus vulgaris is the most common clinical type of Pemphigus, accounting for 60% of the patients, followed by pemphigus foliaceus, accounting for 22.8%. IgA pemphigus, Paraneoplastic and herpetiform pemphigus are rare.

The total cure rate is 88.6%. Treatment is very effective for vegetans, foliaceus and erythematosus types. Whereas it is effective for Pemphigus vulgaris with cure rate of 81%. The effect of treatment in patients with mild to moderate condition of pemphigus vulgaris, pemphigus foliaceus, pemphigus erythematosus and pemphigus vegetans was best, while in serious condition of pemphigus vulgaris it was partial.

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