

Original Research Article

Prevalence of oral lesions in pemphigus vulgaris: a retrospective study

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ABSTRACT

Background: Pemphigus vulgaris is an autoimmune vesiculobullous disease characterized by vesicles and or bullae affecting skin and mucosa. Oral lesions are the first manifestation of the disease in majority of cases according to literature. This study is conducted to know the prevalence of oral lesions in pemphigus vulgaris patients at the time of presentation.

Methods: This was a retrospective study. Departmental records were evaluated from January 2013 to January 2018. All the demographic data of pemphigus vulgaris patients at the time of first presentation, duration of disease, were assessed and tabulated. The clinical data of these patients during their subsequent visits to clinic were assessed for development of new oral lesions during course of disease.

Results: Total number of patients were 55. The mean age was 42.5 years. 12 (21.8%) were males and 4 (78.1%) were females. Out of 55 patients 37 (67.27%) cases had oral involvement and 18 (32.72%) cases did not have oral lesions at the time of presentation. Out of 37 patients who had oral lesions, 34 (91.89%) patients had both oral and skin lesions and 3 (8.10%) patients had only oral lesions at the time of presentation. Out of the 18 patients with no oral lesions, 3 (16.66%) patients developed oral lesions within 3 months. Total number of patients with oral lesions out of 55 were 40 (72.7%).

Conclusions: Our study shows that oral lesions as initial manifestation of pemphigus vulgaris is 67.2%.

Keywords: Pemphigus, Oral lesions

INTRODUCTION

The term pemphigus is derived from the Greek word pemphix, meaning vesicle or blister.¹ Pemphigus is a chronic, acquired, autoimmune vesiculobullous disease clinically characterized by vesicles and or bullae affecting skin and mucosa.²

It is caused due to auto-antibodies against bridging substance between the keratinocytes i.e., mainly desmosomal proteins like desmoglein.^{1,3} This results in destruction of desmogleins thereby leading to the formation of intra-epidermal blister.

They are mainly two types of pemphigus, pemphigus vulgaris and its variant pemphigus vegetans, pemphigus foliaceus and its variant pemphigus erythematosus. Pemphigus vulgaris is the commonest type of pemphigus and comprises approximately 80% of all pemphigus patients, affecting women more than men with the onset of the disease between the fourth and sixth decade of life.⁴

Pemphigus vulgaris is characterized by flaccid vesicles/bullae on normal or on an erythematous base, which rupture within 3 to 5 days forming painful erosions which do not show a tendency to heal unless treated.

The diagnosis is usually made on clinical grounds based on the morphology, sites affected, Nikolsky sign, histopathological examination to know the level of split and fish net pattern of IgG deposition on direct immunofluorescence microscopy.^{1,3}

Oral lesions are the first manifestation of the disease in majority of cases according to literature.^{5,6} In patients with early onset of oral lesions, these remain the sole symptoms of disease for a period of 2 to 6 months, until the appearance of cutaneous lesions.

Some studies have found major differences in the prevalence of oral lesions as first manifestations of the disease among different geographical areas.

Presence of this disease should be suspected in cases of persistent gingivostomatitis, multiple oral erosions or erosive gingivitis. Timely recognition and therapy of oral lesions is critical as it may prevent skin involvement. If treatment is instituted during this time, the disease is easier to control.

This study is conducted to know the prevalence of oral lesions in pemphigus vulgaris patients at the time of presentation.

METHODS

This was a retrospective study conducted in the Department of Dermatology venereology and leprosy at Hassan institute of medical sciences, Hassan, Karnataka based on departmental records of patients attending autoimmune vesiculobullous disease clinic. Institutional ethics committee clearance was obtained before study was started. Departmental records were evaluated from January 2013 to January 2018.

Clinical data of patients diagnosed as pemphigus vulgaris were screened, patients diagnosed with pemphigus vulgaris were included and patients with other vesiculo bullous disease were excluded from the study.

All the demographic data like age, sex, sites affected at the time of first presentation, duration of disease, were assessed and tabulated. The clinical data of these patients during their subsequent visits to clinic were assessed for development of new oral lesions during course of disease. Descriptive statistical analysis was used to calculate prevalence of oral lesions in pemphigus vulgaris.

RESULTS

Total number of patients who satisfied inclusion criteria was 55. The mean age of the patients in the study population was found to be 42.5 years. Out of 55, 12 (21.8%) patients were males and 43 (78.1%) were females (Figure 1). Female to male ratio was 3.5:1. Youngest patient was 21 years old and oldest was 79 years old. Nikolsky sign was positive in all cases. Out

of 55 patients 37 (67.27%) cases had oral involvement at the time of presentation. 18 (32.72%) patients did not have oral lesions at the time of presentation.

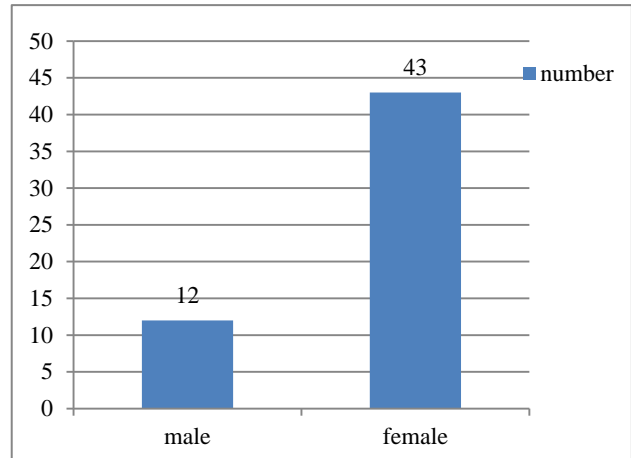


Figure 1: Gender distribution.

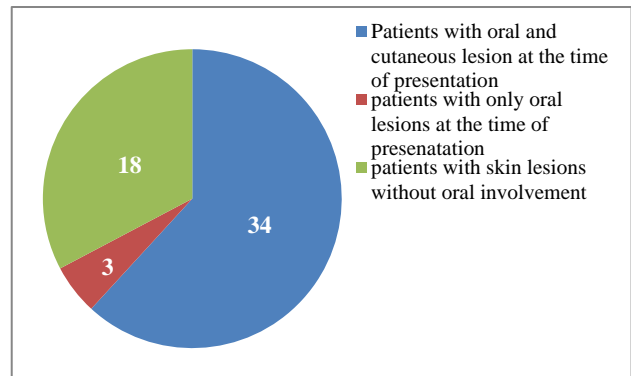


Figure 2: Frequency of oral lesions at the time of presentation (n=55).

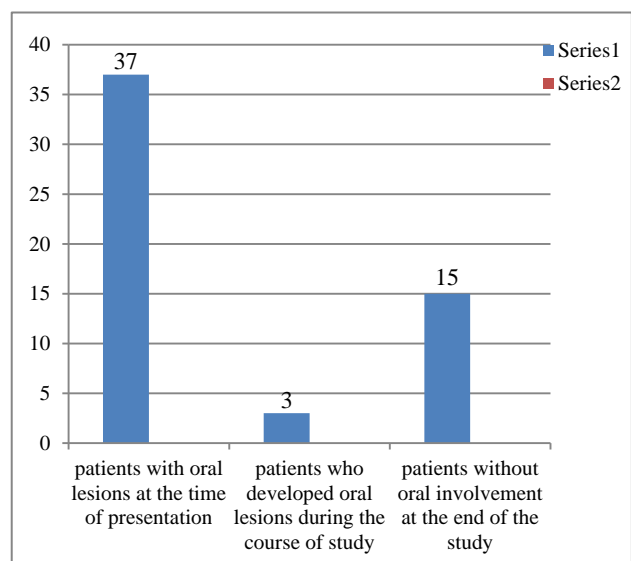


Figure 3: Frequency of oral lesions at the end of the study (n=55).

Table 1: Total number of oral lesions (n=55).

Oral lesions	N
At presentation	37
During course of treatment	3
Total	40

Out of 37 patients with oral involvement, 34 (91.89%) patients had both oral lesions and skin lesions at the time of presentation and 3 (8.10%) patients had only oral lesions at the time of presentation (Figure 2).

Out of the remaining 18 patients with no oral lesions at the time of presentation, 3 (16.66%) patients developed oral lesions within 3 months of start of treatment. Rest of the 15 patients did not develop oral lesions during entire course of treatment.

Total number of patients with oral lesions at the end of the study were 40 (72.7%) out of 55 patients (Table 1) (Figure 3).

DISCUSSION

Pemphigus vulgaris is an autoimmune condition characterised by presence of bullae and erosions over the skin, and mucous membrane involvement. In our study pemphigus vulgaris presented most commonly in 30–50 years of life with average age group of 42.5 years.

Babu et al conducted a study on 7 cases of pemphigus vulgaris, majority of the patients fell in the age group of 30-40 years.⁷

Similar findings were noted in a study conducted by Kumar et al.⁸ They reported incidence of pemphigus was 4.4/million in Kerala population of India and disease exposure was high between 40 and 50 years age. The average age of PV patients was 58 years in males (42 to 82 years) and 37 years in females (25 to 57 years).

These results are consistent with the previous reports of Shamim et al where the age group affected frequently was 15 to 70 years with a mean age of 42.73 years.⁹ Iamaroon et al also noticed preponderance of pemphigus vulgaris around 18 to 55 years.¹⁰ Camacho-Alonso et al found the data to be around 21-87 years.¹¹ In our study population more number of cases recorded were in females, with a female to male ratio of 3.5:1. 78.1% were females and 21.8% were males. Same results were noted by Babu et al, 71.4% of their cases were females.⁷

In our study out of 55 patients, oral lesions were noted in 37 (67.2%) patients as first manifestations. Out of which 3 (8.10%) patients had only oral lesions as there manifestation. Out of the remaining 18 (32.72%) patients with no oral lesions at the time of presentation, 3 (16.66%) out of 18 patients developed oral lesions within

3 months of start of treatment. Out of 55 patients total of 40 (72.7%) patients had oral lesions.

In a study conducted by Mahajan et al, out of 35 patients, 17 (48.57%) were males and 18 (51.42%) were females.¹² Male to female ratio was 1:1.1. The majority of the patients (12, 34.29%) were in the 40-49 year age group. Out of 27 cases, oral cavity was the primary site of involvement in 20 cases (57.14%). 2 (5.71%) cases presented with skin lesions initially and 5 (14.29%) cases presented with simultaneous involvement of the skin and the oral mucosa.

In a study conducted by Santosh et al, 11 out of 14 patients had oral mucosal manifestation as a primary site of disease process.¹³

In the published reports of Shamim et al, it was revealed that pemphigus vulgaris is the most common subtype, with the primary site being oral mucosa in 53.52% of cases.⁹

However, Bascones-Martinez noted that oral lesions were the first manifestation in only 18% of outpatients at dermatology clinics.¹⁴

Some studies have found major differences in the prevalence of oral lesions as initial manifestation of PV among distinct geographic areas, e.g., 66% in Bulgaria, 83% in Italia, and 92% in Israel.¹⁵

The oral cavity was the site of the first evidence of vesicles and bullae in 25 per cent of all cases of pemphigus as reported by Lever.¹⁶

In a study by Eversole et al, oral lesions become more prominent as the clinical course of pemphigus vulgaris develops, with reports of 74 of 80 patients eventually having oral lesions.¹⁷

CONCLUSION

Oral presentation as the initial manifestation of pemphigus vulgaris varies in different studies. In our study out of 55 patients, 34 (61.8%) presented with oral and skin manifestations at the time of initial presentation. 3 (5.4%) out of 55 patients presented with only oral lesions without skin involvement. 18 (32.7%) patients out of 55 patients presented with only skin lesions without oral involvement. Our study shows, oral lesions as initial manifestation of pemphigus vulgaris in 37 (67.2%) patients, 3 (5.4%) patients with only oral lesions at the time of presentation. This is in contrast to available literature which mentions oral lesions as the initial presentation in majority of patients.

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Ethical approval: The study was approved by the institutional ethics committee

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