

# Serious Blister Outbreak - Pemphigus Vulgaris A Clinicopathologic Study

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## INTRODUCTION

Pemphigus is a group of rare autoimmune blistering diseases characterised by widespread epithelial blistering affecting cutaneous and or mucosal surfaces. Pemphigus vulgaris (PV) is the most common variant. Affects only 1-5 patients per million populations per year. Peak incidence occurs between the fourth and sixth decades of life with a male to female ratio of 1:2. The lesion is characterized by intraepidermal vesicles

## ABSTRACT:

Pemphigus is a chronic autoimmune bullous lesion affecting skin and mucosa and oral manifestation can be the first clinical sign.

**Aim:** The present study aims a clinicopathological review of pemphigus vulgaris patients reported in our clinic during a time period of 15 months.

**Methodology:** A retrospective study of oral pemphigus vulgaris documented over a period of 15 months in the Department of Oral medicine and radiology, Government Dental College Thiruvananthapuram, including referral case from Dermatology Department Medical College was analysed. Sociodemographic features and detailed clinical characteristics with relevant histopathological and immunofluorescent features were noted. Statistical analysis was done using SPSS software.

**Results:** 13 cases of oral pemphigus vulgaris were reported. 3 patients showed mucosal manifestation while all the rest showed mucocutaneous features. Buccal mucosal involvement was highest (76.9%) in oral cavity whereas floor of mouth least affected (30.7%). Cutaneous involvement was generalised with facial skin involvement most common (69.2%). Oral /skin biopsy was diagnostic in 11 cases while confirmation was possible after DIF in 2 cases. All patients were treated with systemic steroids and adjuvant drugs in Dermatology Department. Topical steroids were given to patients with oral mucosal lesions in which 77.9% patients responded well.

**Conclusion:** Early diagnosis and treatment of oral isolated pemphigus is of great importance for dentist since it inhibit progressive course of disease, decrease mortality rates, and improve prognosis.

**Key words:** Oral, pemphigus vulgaris, autoimmune disease.

with acantholysis and an intact basal layer. Oral lesions of pemphigus are seen in up to 18% of patients at dermatology out-patient clinics.<sup>1</sup> Pemphigus affects the skin and may also affect the mucosa of the mouth, nose, conjunctivae, genitals, oesophagus, pharynx, and larynx; it is found mainly in middle-aged and elderly patients. Oral manifestations are the first clinical signs in 60% of cases. Diagnosis depends on biopsy confirmation of intraepithelial vesicle formation, acantholysis and the presence of tzanck cells. Demonstration of immunoglobulins especially IgG and complement in the intercellular space by direct immunofluorescence is a very reliable test. If left untreated, PV is frequently fatal with a mortality rate ranging from 60% to 90%. While systemic corticosteroid use and other therapeutic advances have reduced this mortality rate to approximately 10%, complications from treatment are now the primary cause of morbidity and mortality.<sup>2</sup> It is generally managed with topical, oral or parenteral corticosteroids. Immunosuppressants, anti-inflammatory drugs and antibiotics are used as adjuvant; other options include plasmapheresis and intravenous immunoglobulin coupled with cytotoxic drugs. Though the majority of patients respond to conventional therapies, few will develop recalcitrant disease. The present study aims a clinicopathological review of pemphigus vulgaris patients reported in our clinic during a time period of 15 months.

## METHODOLOGY

A retrospective study of 13 cases of pemphigus vulgaris obtained over a period of 15 months from January 2011 to March 2012 in the Department of Oral Medicine and Radiology, Government Dental College, Thiruvananthapuram was done. Study subjects were histopathologically confirmed cases of oral pemphigus, including referral case from Dermatology Department Medical College. Only cases with adequate treatment records were included. Clinical details were noted from the patient files. Histopathologic examination with or without direct immunofluorescence was the method of diagnosis in all cases. Statistical analysis was executed using SPSS software.

## RESULTS

The study characteristics are described in table-1. Age distribution of pemphigus vulgaris patients was from 20 to 65 years with a mean age of 42.73 years. The majority of the patients were in the 30 - 60 year age group (69.2%), with a male to female ratio of 1:1.6.

3 patients showed only mucosal manifestation while all the rest showed mucocutaneous features. The mean duration of symptoms was 15.08 weeks. In 51.8 % patients lesions first appeared in the oral cavity. 10 patients showed cutaneous manifestations. Among them, all patients showed hyperpigmented macule or patch over skin. Intact vesicle was seen in only 2 patients.

Intra orally, pain was the presenting symptom in the majority of cases (53.8%) followed by burning sensation (23.1%). Intra oral manifestations were seen as ulcers or erosions in all patients. Blistering was evident only in 2 cases. The buccal mucosa was the most commonly affected site followed by labial mucosa and palate. This was followed by tongue, gingiva and floor of mouth. Tzanck smear was done in 5 patients with skin lesion and all were positive. Oral/skin biopsy was diagnostic in 11 of 13 cases while confirmation was possible after DIF in 2 cases. Histopathological examination showed intra epithelial split and acantholysis. DIF was done in 6 cases in which IgG deposits were strong in 1 patient and moderately strong in 5 patients. C3 was focally strong in 3 and weak in 3 patients. Inga, I'm and fibrinogen was negative in all cases.

All patients were treated with systemic steroids and adjuvant drugs in Dermatology Department. Topical steroids were given to patients with oral mucosal lesions in which 77.9% patients responded well.

## DISCUSSION

Pemphigus vulgaris is a potentially life-threatening autoimmune mucocutaneous disease. The overall incidence of pemphigus vulgaris has been estimated to be 0.5 to 3.2 per 100,000 persons,<sup>3</sup> affecting both genders equally. In the present study, (PV) most frequently occurred in patients in the

30-60 year age group. In our study, the mean age was 42.73 years. Almost similar findings are obtained in several other studies.<sup>4, 5</sup> There was a patient aged 20 years which was the least reported age. In our study females were affected more frequently with a male to female ratio of 1:1.6. This was in agreement with the published literature.<sup>4, 6</sup> But contradictory reports of equal gender predilection have also come out.<sup>7</sup>

The duration of complaints varied from 2 weeks to 1 year, with a mean duration of 15.08 weeks which was short compared to other reports.<sup>4, 5, 8</sup> The comparatively short duration of oral presentation is may be due to the fact that in most of our cases, the presenting symptom was pain so that the patient would seek treatment as soon as possible. The other presenting symptoms noted were burning sensation, followed by peeling mucosa. In our study 51.8 % patients the lesions first appeared in oral cavity which is in accordance with other studies. The oral mucosa is affected in nearly all cases and, the oral mucosa is the site of the first lesion in the majority of cases.<sup>9</sup> Earlier results showed that the oral cavity was the sole initial site of PV lesions in 56 percent of the cases, and that 88 percent of patients had primary lesions in the mouth alone or in combination with other sites.<sup>10</sup> 76.9 % patients showed mucocutaneous features.

Majority of cutaneous lesions occurred over face (69.2%) followed by thorax. The least involved was skin over back and abdomen. With regard to the intraoral distribution of PV, buccal mucosa (76.9%) was the most common site followed by labial mucosa (69.2%) while floor of mouth was least involved (30.7%). This was in line with the reports of various authors.<sup>4, 7, 11</sup> However some studies had found majority of oral lesions occurring in gingival<sup>12</sup> while some others in palate.<sup>13</sup>

Although PV accounts for only 2 percent of intraoral ulcerative lesions, the serious nature of the disease justifies its consideration in nearly any situation in which multiple chronic oral ulcerations or desquamative gingivitis is present. The most common clinical presentation of oral pemphigus is multiple, chronic ulcerations which is same as in

our study.<sup>7</sup> Clinically it is different to differentiate PV from other vesiculobullous disorders.

Tzanck smear from skin lesions done in 5 patients were positive. The diagnosis of pemphigus vulgaris is based on 3 independent sets of criteria: clinical features, histology and immunological tests.<sup>14</sup> Histologically, there is an intraepidermal blister associated with acantholytic cells. DIF is diagnostic in almost 100% cases.

The current therapeutic regimen of pemphigus vulgaris is largely based on systemic immunosuppressants such as systemic corticosteroids along with other adjuvant like methotrexate, cyclophosphamide, mycophenolate mofetil and intravenous immunoglobulins. Rarely, patients with mild disease, particularly if confined to the mucosal surfaces, can be managed on topical therapy alone. Huilgol and Black have reviewed topical therapy for pemphigus and pemphigoid in detail.<sup>15</sup> For oral pemphigus, measures such as soft diets and soft toothbrushes help minimize local trauma. Local corticosteroid therapy is used in cases where the PV is not extensive and lesions are limited to the oral cavity. Topical analgesics or anaesthetics, for example benzydamine hydrochloride, are useful in alleviating oral pain. Oral hygiene maintenance is crucial. Patients are susceptible to oral candidiasis, which should be treated. Topical corticosteroid therapy may help reduce the requirement for systemic agents.

## CONCLUSION

Early diagnosis and treatment of oral isolated pemphigus is of great importance for dentist since it inhibit progressive course of disease, decrease mortality rates, and improve prognosis. Probably this condition is best managed by a team approach involving both the dentist and physician.

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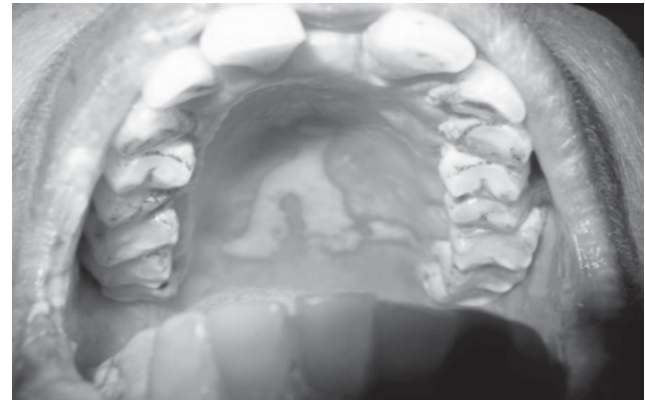
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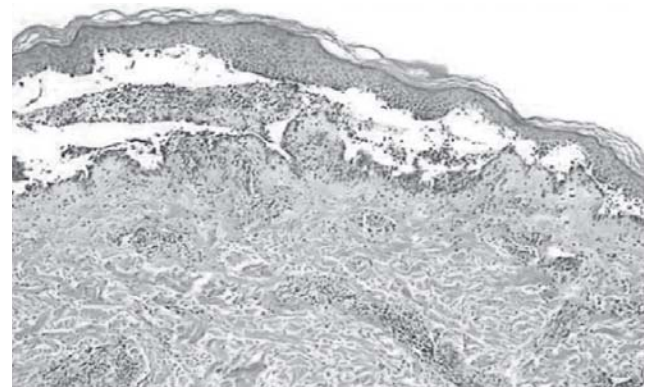
**Conflict of interest:** nil

**REFERENCES**

1. Ramirez-Amador VA, Esquivel-Pedraza L, Orozco-Topete R. Frequency of oral conditions in a dermatology clinic. *Int J Dermatol* 2000; **39**:501-505.
2. Bystryn JC, Steinman NM. The adjuvant therapy of pemphigus.7.An update.*Arch Dermatol* 1996 Feb. **132**(2):203-212
3. Korman N. Pemphigus. *J Am Dermatopathol* 1988; **18**: 1219-1238.
4. Shamim T, Varghese VI, Shameena PM, Sudha S. Pemphigus vulgaris in oral cavity: Clinical analysis of 71 cases. *Med Oral Patol Oral Cir Bucal*.2008 Oct1; **13**(10):E622-626.
5. Iamaroon A, Boonyawong P, Klanrit P, Prasongtunskul S, Thongprasom K. Characterization of oral pemphigus vulgaris in Thai patients. *JOral Sci*. 2006 Mar; **48**(1):43-46.
6. Laskaris G, Sklavounou A, Stratigos J. Bullous pemphigoid, cicatricial pemphigoid, and pemphigus vulgaris. A comparative clinical survey of 278 cases. *Oral Surg Oral Med Oral Pathol*. 1982; **54**(6):656-662.
7. Sirois D, Leigh JE, Sollecito TP. Oral pemphigus vulgaris preceding cutaneous lesions: recognition and diagnosis. *J Am Dent Assoc*. 2000; **131**(8):1156-1160.
8. Camacho-Alonso F, López-Jornet P, Bermejo-Fenoll A. Pemphigus vulgaris. A presentation of 14 cases and review of the literature. *Med Oral Patol Oral Cir Bucal*. 2005; **10**(4):282-288.
9. Robinson JC, Lozada-Nur F, Frieden I. Oral pemphigus vulgaris: a review of the literature and a report on the management of 12 cases. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 1997;**84**(4):349-355.
10. Pisanti S, Sharav Y, Kaufman E, Posner LN. Pemphigus vulgaris: incidence in Jews of different ethnic groups, according to age, sex, and initial lesion. *Oral Surg Oral Med Oral Pathol* 1974;**38**(3):382-387.
11. Ata-Ali F, Ata-Ali J. Pemphigus vulgaris and mucous membrane pemphigoid: Update on etiopathogenesis, oral manifestations and management. *JClin Exp Dent*. 2011; **3**(3):e246-250.
12. Laskaris G, Sklavounou A, Stratigos J. Bullous pemphigoid, cicatricial pemphigoid, and pemphigus vulgaris. A comparative clinical survey of 278 cases. *Oral Surg Oral Med Oral Pathol*. 1982 Dec; **54**(6):656-662.
13. Robinson JC, Lozada-Nur F, Frieden I. Oral pemphigus vulgaris: a review of the literature and a report on the management of 12 cases. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod*. 1997; **84**(4):349-655.
14. Bystryn JC, Rudolph JL. Pemphigus. *Lancet*. 2005; 2-8; **366**(9479):61-73.
15. Huilgol SC, Black MM. Management of the immunobullous disorders. II. Pemphigus. *Clin Exp Dermatol* 1995; **20**: 283-293.



**Figure 1:** Intra oral pemphigus lesions



**Figure 2:** Histopathological appearance of pemphigus

**Table 1: Clinical charecteristics of study sample**

|   |          |   |       |
|---|----------|---|-------|
| <b>Age group:</b>                       |          |   |       |
| Less than 30                            | 3(23.0)  |   |       |
| 30-60                                   | 9(69.2)  |   |       |
| More than 60                            | 1(07.0)  |   |       |
| <b>Gender:</b>                          |          |   |       |
| Male                                    | 5 (38.4) |   |       |
| Female                                  | 8 (61.5) |   |       |
| <b>Distribution of cutaneous lesion</b> |          | <b>Distribution of intra oral lesions</b> |       |
| Scalp                                   | 30.8 %   | Buccal mucosa                             | 76.9% |
| Face                                    | 69.2 %   | Palate                                    | 61.5% |
| Thorax                                  | 61.5 %   | Gingiva                                   | 46.2% |
| Back                                    | 23.1 %   | Labial mucosa                             | 69.2% |
| Extremities                             | 30.8 %   | Tongue                                    | 61.5% |
| Abdomen                                 | 23.1 %   | Floor of mouth                            | 30.7% |
| Genitalia                               | 38.5 %   |   |       |
| <b>Diagnostic confirmation:</b>         |          |   |       |
| Skin or intra oral biopsy:              |          | 11  |       |
| Direct immunofluorescence only:         |          | 02  |       |

Data represents number of individuals(%)