

**Pemphigus vulgaris indices and management – A case report**

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**Citation of this Article:** Dr. Lalitha Basappa Shiggaon, Dr. Priyanka Sudamrao Ghule, Dr. Alka Sanjay Waghmare, Dr. Gunderao Kulkarni, “Pemphigus vulgaris indices and management – A case report”, IJDSIR- April - 2022, Vol. – 5, Issue - 2, P. No. 595 - 602.

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**Type of Publication:** Case Report

**Conflicts of Interest:** Nil

**Abstract**

Pemphigus is a rare chronic mucocutaneous autoimmune disease with the clinical feature of blisters that primarily appear with in the oral cavity and then in the skin. The dental professionals play a crucial role in diagnosing the disease. Early diagnosis and treatment determine the course and prognosis of the diseases. Management of Pemphigus vulgaris (PV) involves prolonged use of steroids and immunosuppressants to regulate the disease and prevent relapses, but associated adverse events constantly remain an excellent challenge. This case of 34year old women was evaluated for severity and management of pemphigus using indices autoimmune bullous skin disorder intensity score (ABSIS), pemphigus disease area index (PDAI), pemphigus oral lesion intensity score (POLIS) and treated for the same.

**Keywords:** Pemphigus, oral lesions, mucous membrane, steroids and immuno suppressants

**Introduction**

Pemphigus Vulgaris (PV) is an autoimmune intraepithelial blistering disease includes the skin and mucous membranes which is characterized by acantholysis with in the epithelium<sup>1</sup>

A variety type of dermatological diseases may present gingival manifestations with desquamative lesions of the gingiva or gingival ulcerations. McCarthy and colleagues suggested that desquamative gingivitis was not a specific disease entity but rather a gingival response associated with a variety of conditions. Desquamative gingivitis (DG) denotes a particular clinical picture and it is not a diagnosis in itself. The term desquamative gingivitis describe a condition

characterized by intense erythema, desquamation and ulceration of free and attached gingiva.

The common disease with relevance of these diseases includes Lichen planus, Pemphigoid, Pemphigus, Erythema multiforme, and Lupus erythematosus<sup>2</sup>. Studies and literature of desquamative gingivitis have shown that 10-20% of the cases representing pemphigus vulgaris<sup>3</sup>.

The oral mucous membrane is usually affected in PV patients; most of patients present with oral lesions as the first sign of PV<sup>4</sup>. Lesions may occur anywhere on the oral mucosa, but the buccal mucosa is the most typically affected site, followed by involvement of the palatal, lingual, and labial mucosae. The gingiva is that the least commonly affected site, and desquamative gingivitis (DG) is a common manifestation of the disease<sup>5</sup>.

This case report presents a case of 34year old women who was evaluated for severity and management of pemphigus using indices autoimmune bullous skin disorder intensity score (ABSIS), pemphigus disease area index (PDAI), pemphigus oral lesion intensity score (POLIS) and treated for the same.

### **Case report**

A 34year old patient reported to the department of Periodontology with chief complaint of burning sensation on having spicy food and peeling of gums on brushing teeth followed by oral ulceration in oral cavity Since 3months. The oral lesion occurred with repeated cycles of remission and exacerbations. Previous medical history and record was of skin lesions 9-10 months back and reported to dermatologist where she was diagnosed with pemphigus vulgaris and treated for the identical.

Clinical examination showed post inflammatory hyperpigmented lesions on posterior trunk region and limbs (Figure1,2,). On intraoral examination gingiva was erythematous involving upper and lower jaws. The

gingiva was tender, with yellowish white pseudo membrane that peeled off leaving an erythematous gingiva that bled on palpation (Figure 3,4).

Based on previous medical case history and clinical features of desquamative gingivitis with positive nikolsky's sign the provisional diagnosis of desquamative gingivitis due to pemphigus vulgaris was made.

There was no significant finding in blood investigation [Table I]. Perilesional incisional biopsy was carried out on the gingiva and histopathological investigation was carried out, which showed parabasal acantholysis near the tips of two adjacent rete pegs were recognized suggestive of pemphigus vulgaris and the following indices were recorded for evaluating the severity of pemphigus vulgaris.

The Pemphigus Disease Area Index (PDAI), has 3 components relating to the skin, scalp, and mucous membranes. The skin has activity and damage scores. The activity score is a value given to the number of erosions, blisters, or new erythema at the time of examination and damage score is 1 of post inflammatory hyper pigmentation or erythema from a resolving lesion and 0 otherwise. For skin and mucous membrane summation of individual scores provides a final activity score, which is out of 250 and 13 for damage score with a total maximum score is 263<sup>13</sup>. In this case report the skin score was 37/120 for activity and 8/12 for damage. [table 3b] and mucous score 27/120 as activity score and 5/12 as damage score [table 3a] and scalp score is 1/10 for activity and 1/[table 3c] for damage with a total maximum score of 79 /263.

The Autoimmune Bullous Skin Disorder Intensity Score (ABSIS), a scoring system with a maximum score of 206 and uses the rule of 9s, which is used in burns measurement, to assess the percentage of involvement of

blisters and erosions on the skin with a weighting factor for the stage of the blistering and erosions. The scoring for skin in this case report was 81/150 [table 4A]. The maximum scores for oral involvement are 11 for extent and 45 for severity<sup>13</sup>. In this case report the oral involvement extend score was 6/11 [table 4B] and severity score was 35/45[table 4C].

According to suggested quartile and model based categorizing groups for pemphigus PDAI score of skin is 37 and mucosal is 27 which is > 19 and ABSIS score of skin involvement is 81, mucous membrane is 6 mucocutaneous score is 87 which is > 46.52 suggestive of severe pemphigus vulgaris<sup>19</sup> (Table 5) and also According to S2k guidelines for the treatment of pemphigus vulgaris/foliaceus and bullous pemphigoid: 2019 update PDAI score > 15 is considered as severe pemphigus vulgaris<sup>18</sup>. Therefore the present case was diagnosed as extensive case of pemphigus vulgaris with desquamative gingivitis.

Treatment plan included was scaling and root planning along with local and systemic corticosteroid therapy, immunomodulators and antibiotics.

### **Treatment and management**

Treatment for PV and management of patient was coordinated with dermatologist and treatment run in 2 phases: a loading phase, to control the disease and maintenance phase, which is further divided into consolidation and treatment tapering. Local treatment consists of a paste, ointment and mouthwash administered alone or in conjunction with systemic treatment.

Within the present case report in initial phase the patient was commenced on systemic corticosteroids (prednisolone) at an initial dose of 0.5 mg /kg/ day (40 mg), vitamin D, Calcium supplements at initiation of steroid treatment to prevent osteoporosis and Tess

ointment (triamcinolone 0.1%) or topical application twice with adjuvant Azathioprine 50 mg once daily and ciprofloxacin 500mg twice daily, is prescribed for 15 days, in addition patient was given betadine gargles. After initial 15 days for consolidation phase the dose of prednisone is tapered to 30mg whereas other medicaments are continued as per the previous dosage for another 15 days [Figure 5]. For treatment tapering phase prednisolone was tapered to 25% for every 2 weeks after consolidation phase and 5mg reduction every 4 weeks when the dose is reduced to < 20mg.

### **Discussion**

Pemphigus is defined as a group of life-threatening blistering disorder of skin and mucous membrane characterized by acantholysis<sup>7</sup>. In PV, autoantibodies are produced against desmosomes (adhesion proteins), chiefly Desmogleins 3 (Dsg 3) and Desmogleins 1 (Dsg 1). The latter is that the target of autoantibody formation in pemphigus foliaceus which affects cutaneous sites only. Dsg1 and Dsg 3 are components of desmosomal cadherin accountable for holding the cells of the epithelium together. The loss of adhesive function among the spinous cells is because of anti-Dsg 3 antibodies which leads to bulla formation immediately supra basal in pemphigus vulgaris. Skin integrity is maintained by Dsg 1.<sup>8-11</sup> As immuno histochemistry was not possible histo pathological examination of lesion along with clinical features the case was confirmed as pemphigus vulgaris with desquamative gingivitis.

In most cases (70-90%), the first sign of the disease appears on the oral mucosa. While the lesions can be located anywhere within the oral cavity and then precedes to cutaneous lesions<sup>12</sup>. The present case there were severe cutaneous lesion preceded to gingival lesions which is rare.

Validated scoring systems like standardized laboratory values are essential for objective and accurate assessment of clinical severity, deciding therapeutic options, prognostication of disease and maintaining the homogeneity of outcome measures in clinical trials. The severity scoring indexes utilized in present case report are Autoimmune Bullous Skin Disorder Intensity Score (ABSIS), Pemphigus oral lesion intensity score (POLIS), and also the Pemphigus Disease Area Index (PDAI). They have been validated and used already in the evaluation of pemphigus and in clinical trials<sup>13</sup>. Evaluation with these indices helped us to assess severity and management of the case.

The dose topical corticosteroid ointment can control limited oral disease. In most cases, however, disease control or remission is achieved using systemic corticosteroids alone or along with immuno suppressive medications<sup>15</sup> because of more severity of the disease and no improvement after topical corticosteroid ointment, systemic corticosteroid and immuno suppressive were advised, reduction in burning sensation and peeling of gums was seen at 1 week of treatment and no complaints and recurrence at 3 and 6 month follow up.

Periodontal clinical parameters like plaque score and full mouth gingival bleeding score were severe. Probing depth and clinical attachment level. These findings were in accordance with Manoj Kumar s. throat et al and Akeman et al implicational that poor oral hygiene (high plaque score) and long term immunosuppression therapy alters the host defense in pemphigus vulgaris might contribute to the development and/or progression of periodontitis.<sup>16,17</sup>

In contrast the probing depth and clinical attachment loss were not significantly greater this might be because

of initial stage of pemphigus vulgaris as compare to chronic cases.

An important aspect of patient management is early diagnosis when lower doses of medication can be used for shorter periods of time to manage the disease<sup>14</sup> the initial and extremely important aspect of management is improving oral health and minimizing the irritation of lesions. This alteration was noticed when patient was followed up after scaling and root planning therapy.

### **Conclusion**

It is of paramount importance to establish the identity of the disease answerable for desquamative gingivitis to establish the appropriate therapeutic approach and management. Pemphigus vulgaris is a life-threatening blistering disorder can manifest as oral mucosal and cutaneous lesion together resulted into the painful ulceration. It should be diagnosed as early as possible and managed to boost the periodontal health and quality of patient's life.

### **References**

1. Dagistan S, Goregen M, Milogluo, Lakur B. Oral Pemphigus Vulgaris: A Case Report with review of literature. *J Oral Sci* 2008;80: 359- 62.
2. Scully C, Parter SR. The clinical spectrum of desquamative gingivitis. *Sem Cutan Med Surg.* 1997; 16:308–13.
3. Robinson JC, Lozada-Nur F, Friden I. Oral pemphigus vulgaris; a review of 12 cases. *Oral Surg, Oral Med, Oral Pathol, Oral Radiol Endod.* 1997; 84:349–52.
4. Mignogna MD, Lo Muziol, Bucci E. Clinical features of gingival pemphigus vulgaris. 2001; 28:489–501.
5. C. Scully, O Paes De Almeida, SR Porter and JJH Crilkes. Pemphigus Vulgaris, manifestations and long-

term management of patient with oral lesions. British Journal of Dermatology 1999;140 (1):84-89.

6. Holmstrup P, Plemons J, Meyle J. Non-plaque-induced gingival diseases. Journal of clinical periodontology. 2018 Jun;45: S28-43.

7. Michael H, Carrian S. Pathogenesis, Clinical Manifestation and diagnosis of Pemphigus. url: <http://www.uptodate.com/store> July 7, 2013.

8. Udey MC, Stanley JR. Pemphigus-disease of anti desmosomal autoimmunity. JAMA 1999; 282:572-6.

9. Amagai M, Klaus-Kovtun V, Stanley JR. Autoantibodies against a novel epithelial cadherin in pemphigus vulgaris, a disease of cell adhesion. Cell 1991; 67:869-77. 9

10. Shirakata Y, Amagai M, Hanakawa Y, Nishikawa T, Hashimoto K. Lack of mucosal involvement in pemphigus foliaceus may be due to low expression of desmoglein 1. J Invest Dermatol 1998; 110:76-8.

11. Amagai M, Koch PJ, Nishikawa T, Stanley JR. Pemphigus vulgaris antigen (Desmogleins 3) is localised in the lower epidermis, the site of blister formation in patients. J Invest Dermatol 1996; 106:351-5.

12. Dagistan S, Goregen M, Milogluo, Lakur B. Oral Pemphigus Vulgaris: A Case Report with review of literature. J Oral Sci 2008;80: 359- 62.

13. Benjamin S. Daniel, MBBS, BA, B Coma, b, Michael Hertl, MDc, Victoria P. Werth, MDd, Severity score indexes for blistering diseases

14. Fellner MJ, Sapadin AN. Current therapy of pemphigus vulgaris. Mt Sinai J Med 2001; 68(4-5):268–78.

15. H. Endo, T. D. Rees, W. W. Hallmon et al., “Disease progression from mucosal to mucocutaneous involvement in a patient with desquamative gingivitis associated with pemphigus vulgaris,” Journal of Periodontology, vol. 79, no. 2, pp. 369–375, 2008.

16. Akman A, KacarogluH, YILMAZ E, Alp soy E(2008) periodontal status in patient with pemphigus vulgaris .oral disease 14,640-643.

17. Thorat MS, Raju A, Pradeep AR. Pemphigus vulgaris: effects on periodontal health. Journal of oral science. 2010;52(3):449-54.

18. Schmidt E, Sticherling M, Sárdy M, Eming R, Gobbler M, Hertl M, Hofmann SC, Hunzelmann N, Kern JS, Kramer H, Nast A. S2k guidelines for the treatment of pemphigus vulgaris/foliaceus and bullous pemphigoid: 2019 update.

19. Boulard C, Duvert Lehembre S, Picard-Dahan C, Kern JS, Zambruno G, Feliciani C, Marinovic B, Vabres P, Borra Dori L, Prost-Squarcioni C, Labeille B. Calculation of cut-off values based on the Autoimmune Bullous Skin Disorder Intensity Score (ABSIS) and Pemphigus Disease Area Index (PDAI) pemphigus scoring systems for defining moderate, significant and extensive types of pemphigus. British journal of dermatology. 2016 Jul;175(1):142-9.

### Legend Tables

Table 1: Routine blood investigation

Hb	10.2 gm%
BP	120/80
TLC	6,600/ mm3

Table 2:

Polymorph's	68%
Lymphocytes	25%
Eosinophils	2%
Monocytes	5%
Basophils	0%

Indices

**Pemphigus oral lesion intensity score (POLIS) [Table 2]**

Items	Points				
	0	1	2	3	4
<b>Symptoms related to oral cavity</b>					
Difficulty in eating normal food <sup>a</sup>	None	Mild	Moderate	Severe	Very severe
Difficulty in eating food according to consistency <sup>b</sup>	None	Raw solids	Cooked solids	semisolid	Liquids
Difficulty in brushing	None	Mild	Moderate	Severe	Very severe
Difficulty in mouth opening	None	Mild	Moderate	Severe	Very severe
<b>Symptoms related to other mucosae</b>					
Difficulty in swallowing	None	Mild	Moderate	Severe	Very severe
Difficulty in speaking	None	Mild	Moderate	Severe	Very severe
<b>Oral cavity examination</b>					
Number of sites involved in the oral cavity (maximum 11) <sup>c</sup>	0	1-2	3-5	6-8	9-11
Overall size of the erosions/ulcers <sup>d</sup>	0	Up to 10 cm	>10-20 cm	>20-30 cm	>30 cm
Depth of the erosions <sup>e</sup>	0	1-10 superficial erosions	11-20 superficial erosions	20-30 superficial	>30 superficial erosions/any number of deep erosions

Pemphigus disease area index (PDAI) [Table 3]

Table 3 A: mucous score

Site	Activity score	Damage
Eyes	0	0
Nose	0	0
Buccal mucosa	0	0
Hard palate	5	1
Soft palate	1	1
Upper gingiva	10	1
Lower gingiva	10	1
Tongue	1	1
Floor of mouth	0	0
Labial mucosa	0	0
Posterior pharynx	0	0
Total	27	5/12

Mucous score activity is 27 and damage score is 5

Table 3 B: cutaneous score

Site	Score	Damage
Ears	3	1
Nose	3	1

Rest of face	5	1
Neck	5	1
Chest	5	1
Abdomen back / buttock	5	1
Arms	5	1
Hands	3	1
Legs	3	0
Feet	0	0
Genitals	0	0
Total	37/120	8/12

Cutaneous activity score is 37/120 and damage score is 8/12

Table 3 c: scalp score

Skin	Activity	Damage
Pdai scalp	Erosion – blister or new erythma 0= absent 2=in one quadrant 3= three quadrant 4=whole skull 10=at least	Number of lesion if <3 Post inflammatory hyper pigmentation from resolving lesions 0= absent

Total scalp	One lesion 1/10	1= present 1/1
PDAI- mucous membrane anatomic location	Erosion blisters 0= absent 1= 1 lesion 2= 2-3 lesions 5=>3lesions 10=entire area	Number of lesions if <3

Autoimmune bullous skin disorder intensity score (ABSIS) [Table 4]

Table 4 A

Skin involvement total score - %BSA x weighting factor = 99% x 0.5= 81

Skin involvement	Patient BSA	Weighting factor
Head & neck (9%)	9%	0.5
L Arm including hands (9%)	9%	0.5
R Arm including hands (9%)	9%	0.5
Trunk (front and back)	36%	0.5
L leg (18%)	18%	0.5
R leg (18%)	18%	0.5
Genitals (1%)		
Total		81

Table 4 B: Oral involvement

1.Extent (1 for present and 0 for absent) [Table 4-B]

Lower gingival mucosa	1	Floor of mouth	1
Upper gingival mucosa	1	Hard palate	1
Upper lip mucosa	0	Tongue	1
Lower lip mucosa	0	Soft palate	1
Left buccal mucosa	0	Pharynx	0
Right buccal mucosa	0		

Oral involvement extend total score = 6/12

Table 4 C: Severity

Food	Level	Factor of discomfort	Severity score
Water	1	0	-
Soup	2	0	-
Yogurt	3	0	-
Custard	4	0	-
Mashed potato	5	1	5
Baked fish	6	1	6
White bread	7	1	7
Apple	8	1	8
Fried steak	9	1	9

Severity score = level x by factor of discomfort= 35/45

Table 5 A: quartile and model-based categorizing groups for pemphigus severity in each subtype based on Pemphigus Disease Area Index (PDAI) [A] [ Table V]

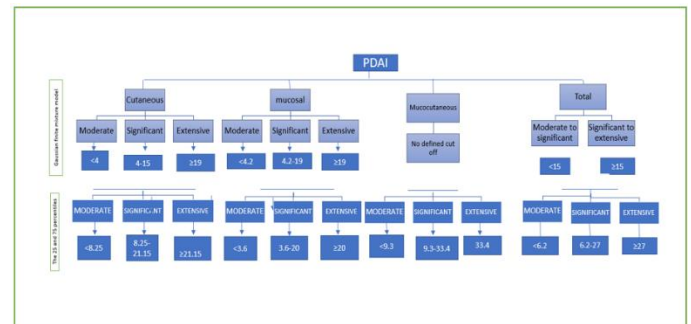


Table 5 B: Autoimmune Bullous Skin Disorder Intensity Score (ABSIS)

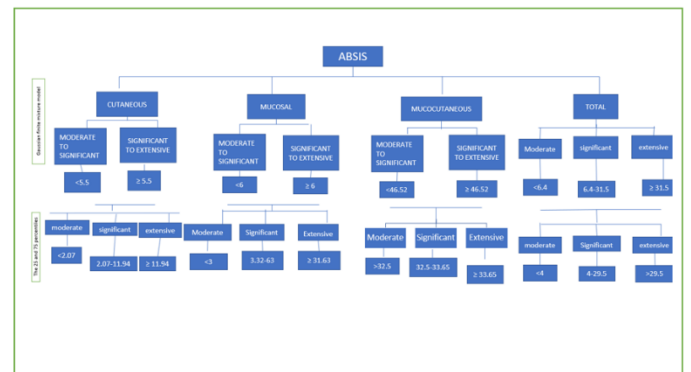




Figure 1: Healed lesion of pemphigus vulgaris on hand



Figure 2: Healed lesion of pemphigus vulgaris on trunk



Figure 3: The initial examination revealed a patchy erythematous labial gingiva.



Figure 4: Gentle palpation with a periodontal probe elicited some desquamation of the gingiva around tooth



Figure 5: 3 month follow up Intra-oral photograph of the patient showing almost complete healing of ulcerative lesions.