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## Short Communication

# Oral mucosal involvement and frequency of Pemphigus vulgaris inpatients at Khartoum

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## ABSTRACT

**Background:** Pemphigus Vulgaris(PV) is a potentially life-threatening autoimmune disease with a high mortality rate and oral manifestations often being the earliest symptoms. The aim of this study is to describe the frequency of Pemphigus Vulgaris in terms of Age, gender, residence and geographic origin, to study its frequency, type, and site distribution of Oral mucosal lesions (OMLs), analyze its association with skin lesions and to correlate it with systemic diseases and mortality rate.

**Materials and Methods:** A retrospective hospital based study was done using in-patient records at Khartoum Dermatology and Venereal diseases Teaching Hospital from Jan 2010 to Dec 2019. Clinical evaluation and histopathological records were the method of diagnosis. Study variable such as socio-demographic data, oral mucosal lesions (type, site, distribution), presence of skin lesions, association with chronic systemic diseases, mortality were used in the study. A structural data collection sheet questionnaire, modified from the standard (WHO) questionnaire of OMLs, was used.

**Results:** In this study, 321 (47.3%) patients out of 678 patients were diagnosed with PV, of which the most common age group reported was 31-50 years (41.8%), females (68.6%) were more affected. Out of 321 PV cases, OML reported in 227 (70.7%) patients. The majority of OMLs (81.9%) were reported in more than one site, among which the most common reported site was Lip (23.5%) and (32.4%) of PV cases were associated with chronic systemic diseases, (10.9%) with mortality, of which, 29 (82.9%) out of 35 cases, reported with OMLs and peak age group of mortality was (41-50 years) (41.5%) among which female had higher mortality (78.9%).

**Conclusion:** PV was seen to be the most commonly manifesting mucocutaneous disease among the study population. Majority of the cases had oral mucosal lesions in concordance with previous studies and literature.

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## 1. Introduction

Pemphigus Vulgaris (PV) is a potentially fatal autoimmune mucocutaneous disease (MCD).<sup>1</sup> Pemphigus Vulgaris is considered the most common type of pemphigus and comprises approximately (80%) of the reported cases among literature worldwide<sup>2</sup> The most common affected

sites include cheeks, lips, palate.<sup>3</sup> It develops most commonly during the 5th to 7th decade of life<sup>4</sup> affecting female more than males.<sup>5</sup> The estimated worldwide mortality rate is approaching 5-10%.<sup>6</sup>

## 2. Objectives

To study the Socio - demographic factors and Oral mucosal involvement of PV inpatients at Khartoum Dermatology and Venereal Diseases Teaching Hospital KDVTTH.

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### 3. Materials and Methods

A retrospective - hospital based study was conducted at KDVTB, Sudan from January 2010 to December 2019. From a total of (817) records of MCDs, (678) were included in the survey, of which (321) were selected on basis of the following criteria:

#### 3.1. Inclusion criteria

Records of inpatients at KDVTB diagnosed with Pemphigus Vulgaris.

#### 3.2. Exclusion criteria

1. Records of patients diagnosed with other types of Pemphigus.
2. Incomplete records.

All data were entered into version 26 Statistical Package for Social Sciences (SPSS) program. For all tests a P - value was set at (0.05) level, with 95% confidence interval.

### 4. Results

In the survey, (321) records were found reported with PV (47.3%), as the most common MCD.

Most of cases were adults falling between 20-60 years. Patients between 1 and 20 years constituted only (9.7%) of the sample, with only 10% of them over 60 years, as shown in Table 1. Females to male ratio was (2:1).

Among patients, OMLs were reported in 227 (70.7%) case, with only 5 cases reported with OMLs alone, and compared to 222 patients with both OMLs and skin lesions. The majority of OMLs (81.9%) were reported in more than one site, with lips as the most common reported OMLs site (23.5%), as shown in Table 2. As shown in Table 3, the most common recorded OMLs were erosions (42.8%), then bulla (21.6%), followed by Crustations (14.4%).

About One third (32.1%) of cases were associated with chronic systemic diseases. The overall frequency of mortality reported was 35 cases (10.9%), with 29 cases (82.9%) reported with OMLs, compared to only 6 cases (17.1%) without OMLs.

### 5. Discussion

Few studies have been conducted to describe OMLs involvement and epidemiology of PV in Sudan, with limited research available worldwide as well.

The current study supported evidence from similar reports [Suliman *et al.* 2013; Vlckova-Laskoska *et al.* 2007; Seo *et al.* 2003; Uzun *et al.* 2006; Zaraa *et al.* 2011] that PV was the most common reported MCD in this survey (47.3%).<sup>7-11</sup>

The most common age groups reported in this survey were (31-40), (41 – 50) years, each with (20.9%) with

a mean age of (40.3 ± 12.5) years. It is encouraging to compare these outcomes with that found by Seo *et al.* 2003; Daneshpazhooh *et al.* 2009; Shamim *et al.* 2008.<sup>9,12,13</sup>

Consistent with literature [Lamey *et al.* 1992; Firth *et al.* 1991], the current survey indicated that Females were more affected than males with a ratio of (2:1).<sup>14,15</sup>

The current survey reported a high frequency of OMLs (70.7%), this is consistent with what was reported by Mignogna *et al.* 2009 (66%).<sup>16</sup>

The results of this survey indicated high association between OMLs and skin lesions (69.2%). This outcome can be compared with those obtained by Saraf. 2016 (85%); who reported high association (more than 50%).<sup>17</sup>

The most obvious finding to emerge from the analysis of OMLs involvement is that more than one site was affected (81.9%), and Lips were the most common reported OMLs site (23.5%), this is in agreement with Kapoor *et al.* 2013, in which more than half of cases, multiple sites were recorded with OMLs, and with Munhoz *et al.* 2011, in which Lips were the most common reported OMLs site.<sup>6,18</sup>

The current survey indicated that the most common reported OMLs were erosions (42.8%). This outcome is in agreement with Suliman *et al.* 2013; Saraf. 2016; Shamim *et al.* 2008; Uzun *et al.* 2006, Firth *et al.* 1991; Kapoor S *et al.* 2013, who suggested erosions as the leading OMLs.<sup>7,10,13,15,17</sup>

The current survey claimed that about 1/3 (32.4%) of PV cases were associated with chronic systemic diseases [Significant]. This outcome can be compared conveniently with Uzun *et al.* 2006 (28.1%); and less conveniently with Seo *et al.* 2003 (21.1%).<sup>9,10</sup>

The most obvious finding to emerge from the analysis is that the frequency of mortality reported in this survey was (10.9%), which is high, but still can be compared with the estimated world wide range (5 - 10%), which is well supported by evidence from Chi *et al.* 2006; Kulthanan *et al.* 2011; Abualgasim & Yousif. 2020.<sup>5,19,20</sup> Among which (82.9%) were associated with OMLs and the peak age group reported was (41 – 50) years by (41.5%), the majority of which were females by (78.9%). This is in agreement with Uzun *et al.* 2006 in regard of peak age of mortality (50 ± 10) years and females predilection.<sup>10</sup>

### 6. Conclusion

Pemphigus Vulgaris was the most frequent MCD (47.3%), well-supported by wide range of literature. Most of the ages of patients fall between 31 and 50 years (41.8%), and females were twice as affected as males. The frequency of OMLs was high (70.7%) (Significant), with high skin lesions association (69.2%). Most of the OMLs were presented at multiple sites (81.9%), and the Lips reported as the most frequent site (23.5%). One third of cases (32.4%), were associated with chronic diseases (Significant). Although, the frequency of mortality with PV

**Table 1:** Shows age distribution among study population

PV	1 to 10 year	11 to 20 year	21 to 30 year	31 to 40 year	41 to 50 year	51 to 60 year	61 and above	Total
(n)	4	27	64	67	67	60	32	321
%	1.3	8.4	19.9	20.9	20.9	18.6	10.0	100

P-Value (0.000)

**Table 2:** Shows detailed OMLs site distribution (n, %) among pemphigus vulgaris patients

PV	Cheek	Tongue	Lip	Palate	Gum	Others	Total
(n)	23	59	99	88	18	134	421
%	5.5	14.0	23.5	20.9	4.3	31.8	100

**Table 3:** Shows types (n, %) of OMLs among target study population

PV	Bullae	Erosion	Curst.	Vesicle	Ulcer	Other	Total
(n)	45	89	30	10	18	16	208
%	21.6	42.8	14.4	4.8	8.7	7.7	100

in this survey was high (10.9%) (Significant), but still can be compared to the estimated worldwide range 5-10%.


## 7. Conflict of Interest


None.

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