Original Research Article

Blistering mucocutaneous disease of oral cavity Pemphigus vulgaris – 8 year study in Nalgonda population

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Abstract

Background: Autoimmune disorders are conditions in which autoantibodies are directed against a single organ or tissue resulting in localized tissue damage. Pemphigus includes a group of autoimmune blistering diseases of skin and mucous membranes characterized by intra dermal blisters and immunologically by finding of circulating immunoglobulin G antibody directed against the cell surface of keratinocytes.

Objectives: To analyze age distribution of Pemphigus vulgaris, prevalence among males and females, predominant oral site and clinical presentation.

Materials and methods: A retrospective study of 31 cases of Pemphigus vulgaris obtained over a period of 8 years from January 2008 to September 2015 in the Department of Oral Pathology, Kamineni Institute of Dental Sciences was designed. Clinical details of age, sex, intraoral distribution and oral presentation were noted.

Results: Age distribution of Pemphigus vulgaris was 30 – 70 years with a mean age of 49.12 years. Mean age of presentation in males was 45.5 years and in females 46.76 years. Females are more commonly affected than males with a ratio of 2:1. Most commonly affected sites were buccal mucosa, lips and palate, tongue, floor of mouth and skin. Erosions and encrustations were the most common clinical presentation.

Conclusion: Pemphigus vulgaris is a fatal disease if left untreated. The skin and the mucosa are majorly involved and oral mucosa is often affected first. The study elucidates the characterization of Pemphigus vulgaris so that early diagnosis can be made. As oral lesions precede, oral health care professionals can play a major role in early diagnosis and managing oral lesions.

Key words

Autoantibodies, Desmogleins, Females, Pemphigus vulgaris.

Introduction

Pemphigus is a group of diseases associated with intraepithelial blistering [1]. In pemphigus there is damage to desmosomes by antibodies against the extracellular domains of the desmogleins with immune deposits intraepithelially [2]. Pemphigus vulgaris (PV) and Pemphigus foliaceous (PF) are the classically recognized clinical variants, but others are now known, such as IgA pemphigus, paraneoplastic pemphigus and herpetiform pemphigus. Oral lesions are commonly seen with Pemphigus vulgaris and Paraneoplastic Pemphigus [3]. Patients suffering from mucosal-dominant PV typically have antibodies against Dsg3 but not Dsg1, whereas mucocutaneous PV is characterised by the presence of antibodies to both Dsg 3 and Dsg 1.1 In contrast, PF patients usually possess auto-antibodies against Dsg1, but not Dsg3 [4].

Pemphigus vulgaris is the most common form of pemphigus. It is a chronic, recurrent, potentially life threatening bullous dermatological disease characterized by the appearance of intra epidermal blisters (bullae) and erosions on skin and mucous membranes. Incidence is rare 0.5 – 3.2 cases per 100,000 and occurs mainly in the adults in the age range of 30 to 60 years.

Skin bisters in Pemphigus vulgaris vary in diameter, tension, fragility. They rupture easily resulting in painful erosions. Oral mucosal lesions are common and predominantly involve the buccal mucosa followed by the palate, lips, gingival and tongue. Other mucosal surfaces are also affected like conjunctiva, pharynx, larynx, esophagus, bronchi, stomach and genitalia [5].

Diagnosis of Pemphigus vulgaris is made by the presence of active blisters and firm sliding pressure with a finger separates normal-looking epithelium (Nikolsky sign) but this is neither completely sensitive nor specific [6]. A smear taken from the base of a blister often contains acantholytic cells (Tzanck preparation), but this is rarely used as it is neither specific nor sensitive [7].

Biopsy of perilesional tissue with histological examination and immunostaining is valuable in the diagnosis of Pemphigus vulgaris. Immune deposits (IgG and C3) intercellularly precede the appearance of acantholysis in the suprabasal epithelium, and so it is more sensitive than conventional histopathology [8].

Corticosteroids are the main stay in the treatment of Pemphigus vulgaris. Topical corticosteroid rinses or creams can be used for mild localized lesions of oral mucous membrane pemphigus in patients with low titres of circulating autoantibodies. Systemic corticosteroids are used in patients with multifocal involvement and severe localized lesions [9]. (Mild and severe forms of oral involvement of PV are shown in **Figure - 1** and **Figure - 2** respectively).

Figure - 1: Mild form of PV with encrustations.

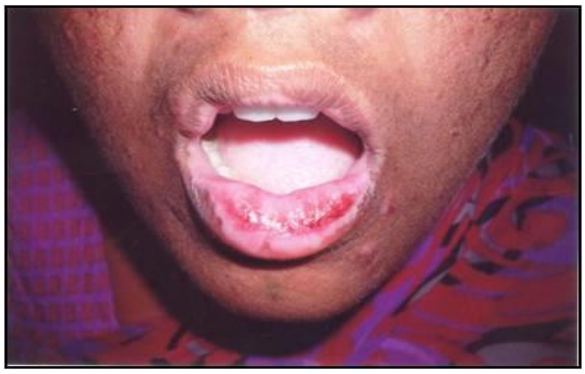
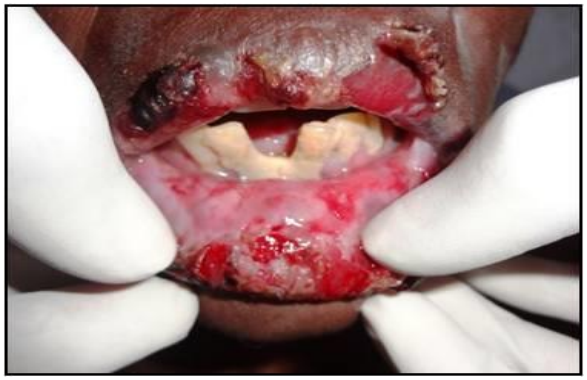


Figure - 2: severe form of PV with erosions, encrustations and ulceration.



Materials and methods

A retrospective study of 31 cases of Pemphigus vulgaris obtained over a period of 8 years from January 2008 to September 2015 in the Department of Oral Pathology & Microbiology, Kamineni Institute of Dental Sciences was designed. Clinical details like age, sex, intraoral distribution of oral lesions, oral presentation were noted. Histopathologic examination was the method of diagnosis in all cases. Statistical analysis was executed using Microsoft Excel computer software.

Inclusion criteria

All age groups, histopathologically confirmed cases of oral Pemphigus vulgaris and reports with adequate case histories were included.

Results

Age distribution of pemphigus vulgaris was 30 – 70 yrs with a mean age of 49.12 years. Mean age of presentation in males was 45.5 years and in females 46.76 years. Majority of the patients affected were in the 5^{th} decade of life (48.31%) followed by 6^{th} decade (25.8%), 4^{th} decade (19.35%) and 7^{th} decade (6.45%).

Females were more commonly affected than males with a ratio of 2:1. In all the cases Nikolsky sign was positive. In only 4 cases there was simultaneous mucosal and skin involvement. In one case there was involvement of conjunctiva.

Buccal mucosa was the most commonly affected oral site 90.32% followed by lips and palate 58.06%, tongue 38.7%, floor of mouth 16% and skin involvement was seen in 12.9% of cases. In 80.6% of the cases, erosions and encrustations were the predominant clinical presentations followed by vesicles and erosions in 12.9% and ulcerations and erosions in 9.6% of cases presented.

Discussion

Pemphigus vulgaris (PV) is one of the classic autoantibody mediated disease that can manifest as lesions on the skin and mucous membranes [10]. It is the commonest type of pemphigus which accounts for approximately 70% of pemphigus cases.

In the present study, pemphigus vulgaris (PV) most frequently occurred in patients in the fifth decade. These results are consistent with the previous reports that the peak incidence of PV occurs between the fourth and sixth decades of life. A mean age of 56.5 years and 56.1 ± standard deviation of 14.9 years was reported [11-13]. Iamaroon, et al. in their study reported a mean age of 37.7 $\pm$ 10.6 years [10]. PV is least commonly seen in young age. Robinson, et al. reported PV at a very young age of 3 years in a child [14].

In the present study, females are affected more frequently with a female to male ratio of 2:1. Equal predilection among males and females is reported in few studies (Davenport, et al.). A female to male ratio of 9:3 has been reported by

Robinson, et al. [14]. This difference in ratios may be due to differences in geographic and ethnic nature of patients studied. Autoimmune diseases are more common among women when compared to men and this could be due to hormonal make-up which renders women more likely to develop Pemphigus than men [15, 16].

In our study the lesions are seen commonly effecting buccal mucosa followed by lips and palate. This could be due to, the epithelium in buccal mucosa demonstrates less intercellular substance and fewer intercellular junctions making the area more susceptible to acantholysis. These findings are in line with other studies [17]. Iamaroon, et al. and Robinson, et al. in their study reported gingiva to be the most commonly affected oral site followed by buccal mucosa whereas Laskaris, et al. reported palate to be the most commonly affected site followed by lips and buccal mucosa. (clinical presentation of PV on skin, tongue and conjunctiva are shown in **Figures - 3, 4, 5** respectively).

Figure - 3: lesions of PV on the skin.

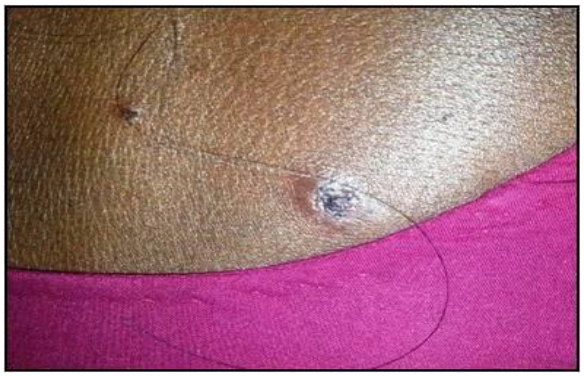


Figure - 4: lesions of PV on the tongue.

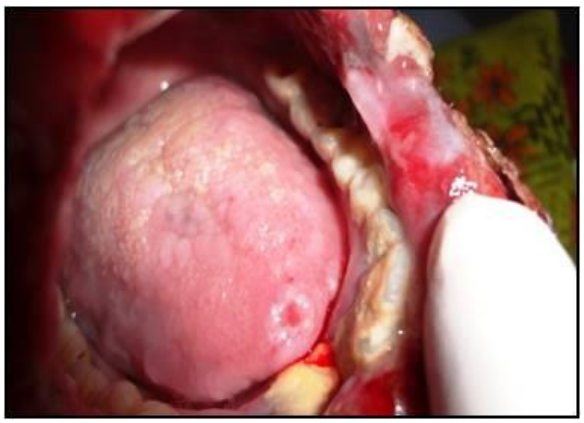


Figure - 5: lesions of PV on the conjunctiva.

In the present study erosions and encrustations were the most common presenting symptoms. This is because of the vesicles are intraepithelial in nature and so rupture easily resulting in formation of erosions [3]. Moreover as the oral mucosa is always subjected to minimal trauma and as the mucosal covering is thin, it forms an extreme area of erosion and ulceration. The clinical characterization of Pemphigus vulgaris in various studies is shown in **Table - 1**.

Conclusion

Pemphigus vulgaris is a fatal disease if left untreated. The skin and the mucosa are majorly involved and oral mucosa is often affected first. The present study evaluated the clinical parameters of age, sex, oral distribution and clinical presentation of Pemphigus vulgaris. Early diagnosis and management are crucial in reducing the morbidity associated with Pemphigus. As mucosal lesions are seen earlier, oral health care professionals play a major role in early diagnosis and managing oral lesions.

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Table - 1: Clinical characterization of Pemphigus vulgaris in various studies.

Parameters	**Present study**	**Shamim, et al. [17]**	**Iamaroon, et al. [10]**	**Davenport, et al. [11]**	**Sirois, et al. [9]**
No. of cases	31	20	18	33	42
Age group (years)	20-70	20-69	18-55	27-79	27-68
Average age	49.12	42.3	37.7	56.5	56.1 ± standard deviation of 14.9 years
Females	21	12	12	25	30
Males	10	8	6	8	12
M: F	1:2	2:3	1:2	1:1	2.5:1
Buccal mucosa	28	18	11	-	18
Lips	18	11	3	-	0
Palate	18	12	6	-	3
Tongue	12	6	4	-	2
Gingiva	0	1	17	-	13
Floor of mouth	5	2	3	-	0