Pemphigus Vulgaris – Role of Corticosteroids in its Management

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ABSTRACT

Pemphigus vulgaris is a chronic autoimmune mucocutaneous disease that initially manifests in the form of intra oral lesions, which spread to other mucous membrane and skin. The etiology of pemphigus vulgaris is still unknown. Pemphigus is derived from Greek word Pemphix meaning bubble or blister. Blistering is due to production of auto-antibodies against desmoglein 1 and 3. Dental professionals must be efficient to recognize the clinical features of pemphigus vulgaris to ensure early diagnosis and treatment, so that it determines the favorable prognosis and course of the disease. This paper reports a case of pemphigus vulgaris and role of corticosteroids in its management.

KEY WORDS: Pemphigus, Autoimmune, Supra Basal Split, Corticosteroids.

INTRODUCTION:

Pemphigus is a group of potentially life threatening autoimmune disease. Pemphigus Vulgaris is a chronic mucocutaneous autoimmune blistering disease which primarily affects the oral mucosa and later may spread to skin and other mucus membrane.[1,2,3] Histologically, it is characterized by Intra-epidermal blister with tombstone appearance of basal cells and immunopathologically, circulating immunoglobulin G (IgG) antibody directed against the cell surface of keratinocytes. Here, we report, case of Pemphigus vulgaris, lesions in oral mucosa without any skin lesions. It is especially important for the dentist to recognize not only that some dermatoses exhibit concomitant lesions of the oral mucous membranes, but also that manifestation of some diseases may be preceded by oral lesions.[4,5]

Case report

A 45 year male patient Mr. Manoj Dighe reported to the department of Oral Medicine and Radiology with the chief complaint of pain and burning sensation in his mouth since 2 year. Patient was apparently asymptomatic a year back after which he started experiencing roughness on his cheeks and lips from inside which was associated with burning sensation while having food. He also noticed ulcerations on cheeks and lips from inside. With this chief complaint he visited Sasoon Hospital, Pune; where routine blood investigations with HIV and Hepatitis were carried out. All the values were under normal limit and he was advised topical anesthetic Mucopain Gel and Vitamin B Complex, 1 Capsules daily orally at bed time. Patient was not relieved of symptoms and he again consulted a local Dentist where patient was advised topical steroid Kenacort Ointment four times a day on the affected areas along with 'A-Z 1Tablets orally. Patient got symptomatic relief for a few days, but lesions did not heal completely. After this the Patient visited a Dental Institute where he was advised to apply topical Antifungal Drugs after which symptoms still got exaggerated. So after all this, the patient came to our Department of Oral Medicine And Radiology with same complaint.

On general examination, Patient appeared moderately healthy and vital signs were within normal limits. There were no signs of cutaneous lesions. Patient is not suffering from any systemic ailment. Intra oral examination revealed mild occlusal caries in relation to 17, 26, 47 and root stumps with 15. On soft tissue examination, the labial and right & left buccal mucosa revealed, a multiple ill defined irregular shaped ulcers having tissue tags, which were surrounded by erythematous halo and the base of each was covered with yellowish pseudomembrane. On palpation all the ulcers were soft in consistency, non indurated but tender on palpation (shown in Figure 1a, 1b and 1c). The tongue was reddish pink in color with areas of depapillation on its dorsal surface and lateral borders. The gingival was reddish pink in color, soft in consistency and scalloped contour with blunt papillae. Stippling was absent with peeling of epithelium from papillary and attached gingiva. There was generalized bleeding on probing.

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Patient was subjected for further investigations; Routine blood investigations were done which were under normal limits. The biopsy was taken from buccal mucosa and was sent to Histopathological examination. The H&E section showed hyperplastic and hypertropic stratified squamous orthokeratinised epithelium which showed acantholysis in spinous layer. There was supra basal split seen and within the split few large, round epithelial cells with prominent nucleus were present suggestive of Tzank cells (Figure 3). The connective tissue showed inflammatory cells of infiltrate chiefly of Lymphocytes and Polymorphonuclear neutrophils (PMN). Based on the clinical and histopathologic features, lesion was suggestive of Pemphigus Vulgaris. Patient was advised Tablet Wysolone (Prednisolone) 20 mg twice daily for two weeks and the dose was tapered. After the lesions were healed completely, the maintenance dose of 5 mg alternate daily for a month was advised. The patient was also advised Squash and spit method in which patient was asked to crush Wysolone tablet in water and rinse the mouth with the same and spit it out for 1 week. Patient was follow up for a period of 1 year without any recurrence (Figure 2a, 2b and 2c).

DISCUSSION:

Pemphigus refers to a group of potentially life-threatening autoimmune diseases of the skin and mucous membranes, characterized by the formation of blisters and erosions of the skin. An autoimmune process, directed against keratinocytes desmosomal cadherins, interferes with the adhesive function of these molecules.[3,4] This results in the separation of keratinocytes and clinical manifestation of blistering. The disease appears to affect both sexes with equal frequency.[1,2] In present case, it was a male patient. It is most common in the fifth and sixth decades and extremely rare in children and adolescents.[5] In present case, age of patient was 45 years which is consistent with the literature. The mouth is the first site to be involved in up to 70% of cases. Oral lesions are the predominant feature with skin lesions occurring in an only small proportion of cases and then only when disease is longstanding. In present case, oral lesions were present without skin lesions although it was longstanding. There is damage to desmosomes by antibodies directed against the extracellular domains of cadherin-type epithelial cell adhesion molecule–the Dsg with immune deposits intra-epithelially, and loss of cell-cell contact (acantholysis), leading to intraepithelial vesiculation.[6,7,8] In present case, histopathological examination revealed hyperplastic and hypertropic stratified squamous orthokeratinised epithelium which showed acantholysis in spinous layer. There was supra basal split seen and within the split few large, round epithelial cells with prominent nucleus were present suggestive of Tzank cells.

The main aim of treatment is to reduce inflammatory response and autoantibody production, thereby achieving disease remission. Systemic corticosteroids remain the mainstay of therapy for patients with oral lesions, transforming invariably fatal disease into one whose mortality is now below 10%.

The adjuvant drugs are immune-suppressants and intravenous immunoglobins, which are commonly, used incombination with steroid-sparing effect in order to increase efficacy and reduce side effects of systemic corticosteroids.[9,10]

Patients with mild disease are treated with initial prednisolone doses of 40 to 60 mg per day and in more severe cases, 60 to 100 mg per day. If there is no response within 5 to 7 days, the dose should be increased in 50 to 100% increments until there is disease control, i.e. no new lesions and healing of existing ones. If doses above 100 mg per day are required, pulsed intravenous CS should be considered. Once remission is induced and maintained with healing of the majority of lesions, the dose of CS should be cautiously tapered. A 50% reduction every 2 weeks has been suggested.[9]

In present case, Tablet Wysolone (Prednisolone) 20 mg twice daily for two weeks and the dose was tapered. After the lesions were healed completely, the maintenance dose of 5 mg alternate daily for a month was advised. The patient was also advised Squash and spit method in which patient was asked to crush Wysolone tablet in water and rinse the mouth with the same and spit it out for 1 week. Patient was follow up for a period of 1 year without any recurrence.

The theoretical aims of pulse therapy is methyl prednisolone 250-1,000 mg are to achieve more rapid and effective disease control compared with conventional oral dosing, thus allowing a reduction in long-term maintenance. The azathioprine is a commonly prescribed adjuvant drug.

Oral cyclophosphamide could be considered as an alternative to azathioprine. Mycophenolate mofetil (MMF) is a relatively newer agent in PV therapy. It is considered in recalcitrant cases or when azathioprine and cyclophosphamide cannot be used.[10]

CONCLUSION

Chronic mucocutaneous disease with formation of painful ulcers is the first manifestation of Pemphigus Vulgaris, at least initially. Early detection of the lesion may prevent delayed diagnosis and inappropriate treatment of a potentially chronic determatological condition.

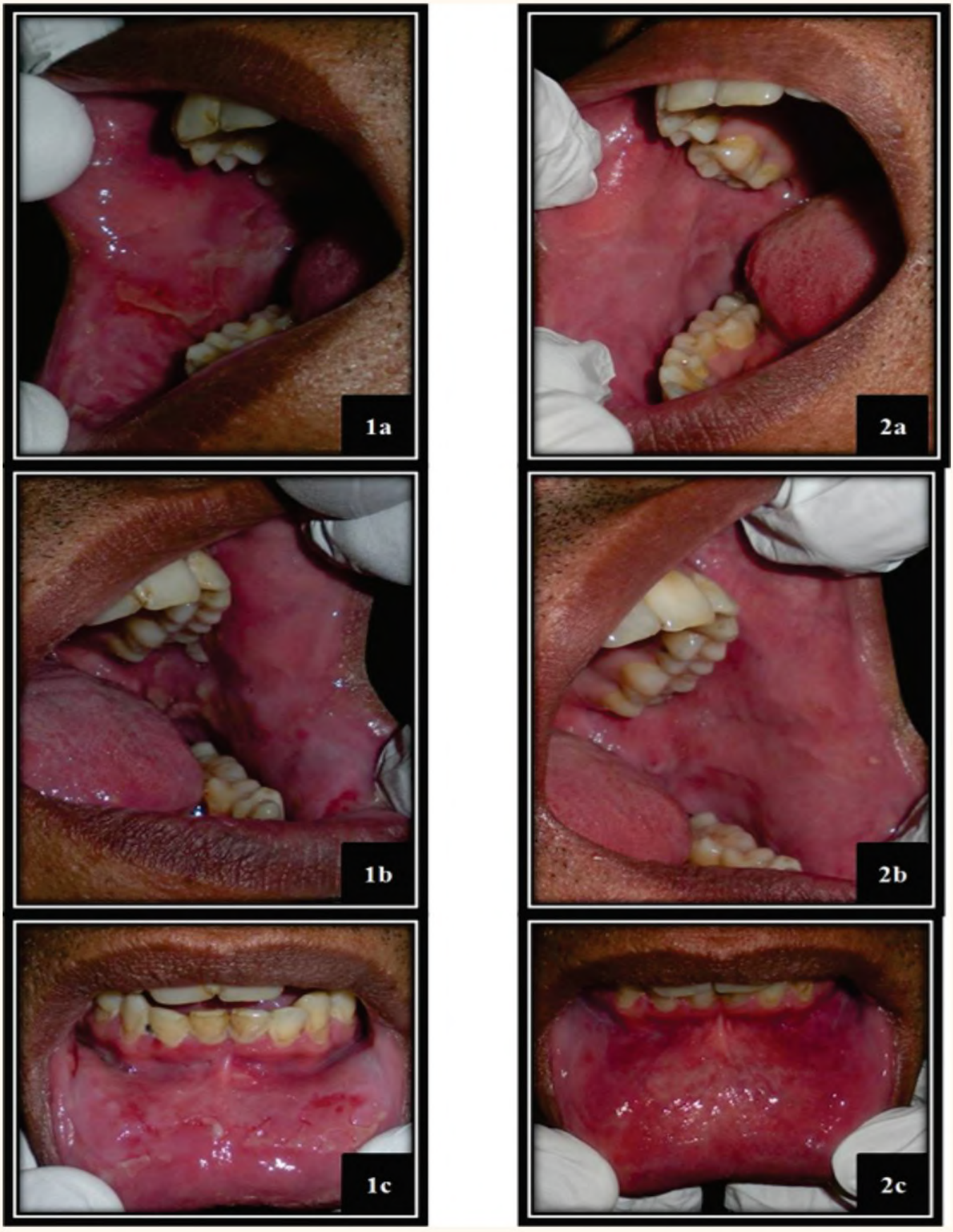

Figure 1 a and 1bRight & left buccal mucosa shows multiple ill defined irregular shaped ulcers with yellowish pseudomembrane at the base. 1c Labial mucosa shows multiple ulcers with erythematous halo and tissue tags present. 2a, 2b and 2c show complete remission of lesion.

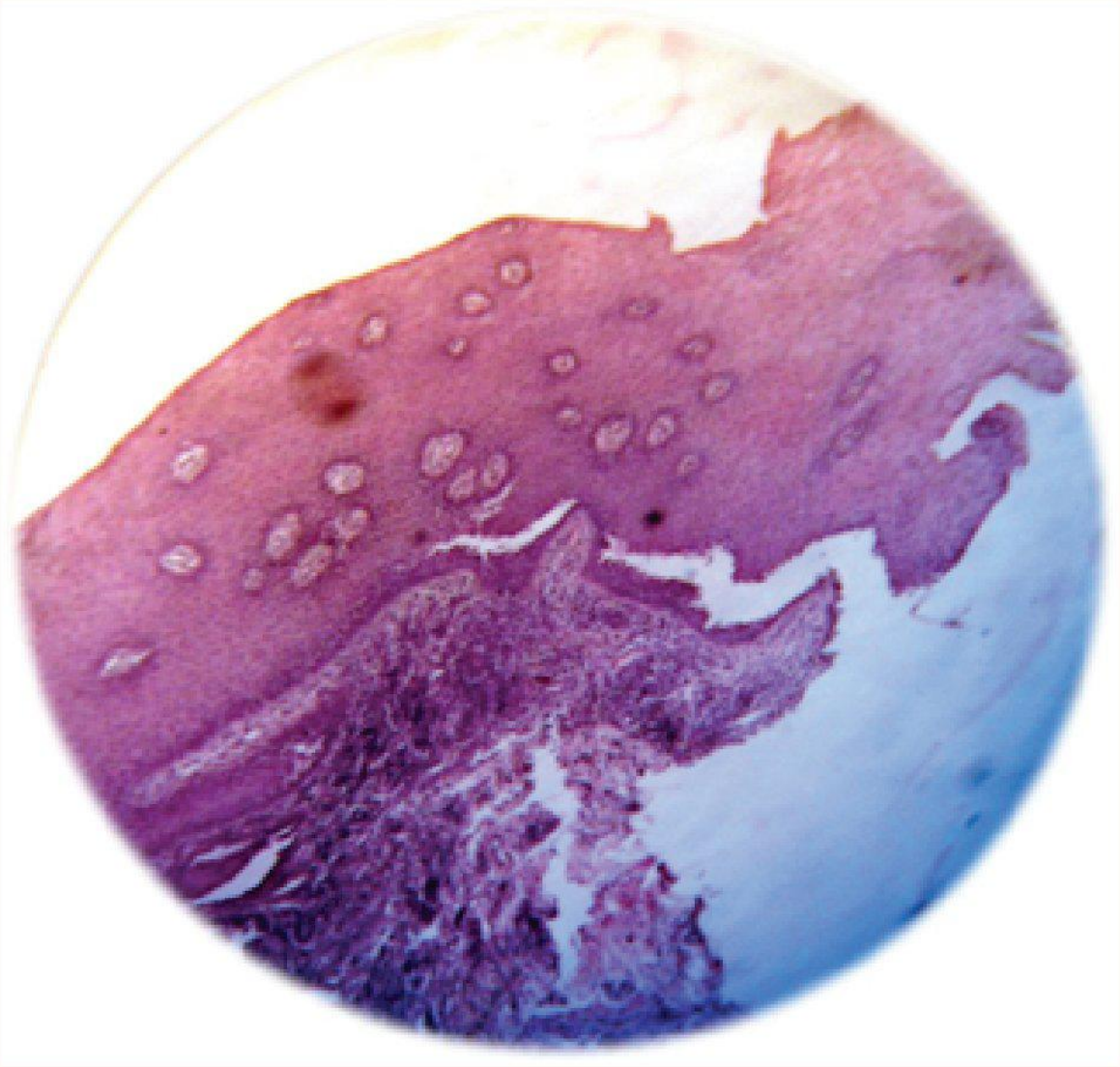

Figure 3: Histopathological picture showing

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