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Case Report

ORAL PEMPHIGUS - A CASE REPORT

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ABSTRACT

PEMPHIGUS is a serious chronic skin disease characterized by the appearance of vesicles & bullae, small or large fluid filled blisters that develop in cycles. The term pemphigus refers to a group of autoimmune blistering diseases of skin and mucous membranes that are characterized histologically by intraepidermal blisters due to acantholysis and immunopathologically by an in vivo bound & circulating IgG directed against the cell surface of keratinocytes. Pemphigus vulgaris is the most common form and frequently affects oral cavity. Main antigen is Dsg 3 (desmoglein) but 50% of patients also have autoantibodies to Dsg 1. Dsg 3: Dsg 1 is directly proportional to severity. We present a case of pemphigus occurring in the oral cavity of a 45-year-old male. Treatment with oral prednisolone and topical steroid resulted in remission of the disease. Clinical features, histological features and treatment of pemphigus vulgaris will be discussed.

KEYWORDS: Pemphigus, blisters, acantholysis, autoantibodies.

INTRODUCTION

Pemphigus vulgaris is a chronic autoimmune mucocutaneous disease that initially manifests as intraoral lesions later spreads to other mucocutaneous surfaces and the skin. The pemphigus and other mucocutaneous diseases are characterized by the production of autoantibodies against intercellular substances and so are named as autoimmune diseases. The etiology of pemphigus vulgaris is still unknown, although the disease has attracted considerable interest. Most patients are initially misdiagnosed and improperly treated for many months and even years. Dental professionals must be sufficiently familiar with the clinical manifestations of pemphigus vulgaris to ensure early diagnosis and proper treatment.[1]

The term pemphigus named by Wichman- 1791, is derived from the Greek pemphix (bubble or blister).[2] Oral lesions are common, causing blisters, erosions and ulcers. Vesiculobullous lesions occurring orally usually rupture later new bullae developing upon the older ones rupture and ulcerate and thus erosions and ulcers are main features and are seen primarily in the buccal mucosa, palate and lips.[2] Ulcers heal with or without scarring. Desquamative gingivitis (DG) may be seen, but it is not a diagnosis in itself.[2]

CASE REPORT

A 45 year-old man was referred to our department with a two month history of painful oral ulcers and erosions. The patient reported that the lesions caused considerable discomfort and affected his normal oral function. On intraoral examination, ulcers were observed on the buccal mucosa, vestibule and palatal mucosa (Figure 1 & 2) and ventral surface of tongue, floor of the mouth (Figure 3) superimposed by white scrapable curdy material. On scraping of curdy white material, erythematous erosive underlying areas were evident. No skin lesions were seen on extra oral examination. A diagnosis of pemphigus vulgaris superimposed with candidal infection was made after evaluating the biopsy samples. Histopathological examination of present case revealed characteristic suprabasal acantholytic split, intercellular edema and disappearance of the intercellular bridge in the lower one third of the epithelium (Figure 4).

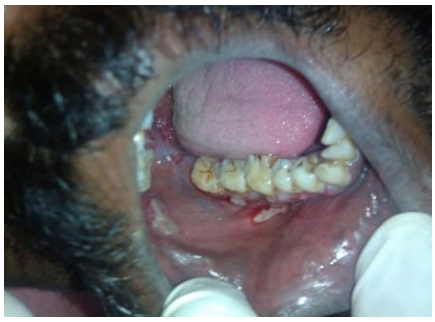

Figure 1. Intra oral pictures of patient with erosive ulcerated lesions on buccal mucosa.

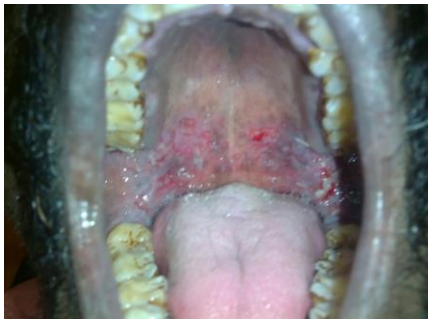

Figure 2. Intra oral pictures of patient with erosive ulcerated lesions on buccal mucosa.

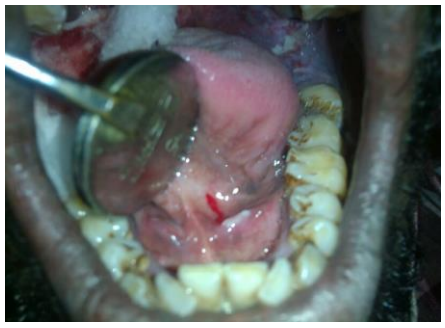

Figure 3. Intra oral pictures of patient with erosive ulcerated lesions on floor of the mouth and tongue.

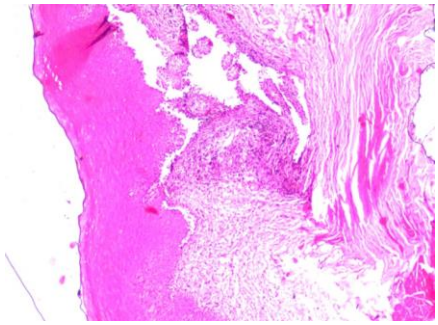

Figure 4. H & E stained histopathological picture with suprabasal acantholytic split, intercellular edema and disappearance of the intercellular bridge in the lower one third of the epithelium.(10x)

The patient was referred to dermatology department of the medical college at Kurnool. Initially for 14 days prednisolone 80 mg, azathioprine 50 mg and topical antifungal was prescribed and later increased to prednisolone 100 mg for other 14 days. Prednisolone was decreased by 10 mg in the following week. At the end of five weeks, prednisolone was decreased to 30 mg. In the follow up examination after two months of treatment, ventral surfaces of tongue, buccal mucosa and palatal mucosa healed without scar formation.

DISCUSSION

The role of environmental factors like medicines, diet (garlic) and physical and viral agents that may trigger the disease is unclear but there is genetic basis to many cases .Pemphigus vulgaris is an autoimmune disease caused by antibodies directed against desmoglein 1 and desmoglein 3 present in basement membrane resulting in the loss of desmosomes and therefore the loss of cohesion between keratinocytes in the epidermis, type II hypersensitivity reaction.[3] When autoantibodies attack desmogleins, the cells become separated from each other a by phenomenon called acantholysis leading to epidermis becomes broken.[4] This causes blisters that slough off and turn into sores, ulcers. There are several types of pemphigus which vary in severity: pemphigus vulgaris, pemphigus foliaceus, intraepidermal neutrophilic IgA dermatosis, and paraneoplastic pemphigus.

Pemphigus vulgaris is characterized by extensive flaccid blisters and mucocutaneous erosions. The severity of the disease, as well as the mucosal lesions, is believed to be directly proportional to the levels of desmoglein 3. Milder forms of pemphigus (like foliaceus and erythematoses) are associated more with desmoglein 1. It arises most often in middle-aged or older people, usually starting with a blister that ruptures easily.[5] The lesions can become quite extensive. The pathogenesis of the disease involves autoantibodies against desmosome proteins, separating keratinocytes from the basal layer of the epidermis. On histology, the basal keratinocytes are usually still attached to the basement membrane leading to the appearance and thus the term, "tombstoning".[6]

Transudative fluid accumulates in between the keratinocytes and basement membrane (suprabasal split), forming a blister and

resulting in what is known as a positive Nikolsky's sign. This is a contrasting feature from bullous pemphigoid, which is thought to be due to anti-hemidesmosome antibodies, and where the detachment occurs between the epidermis and dermis (sub epidermal bullae)[7] Pemphigus is a potentially chronic dermatological condition which was misdiagnosed in its earliest stage as stomatitis.

Prognosis is worse in patients with extensive disease and in older patients. Before the introduction of corticosteroids 75% of patients died within the first year. Currently, less than 10% of patients die, usually due to secondary effects of the treatment.[8] Pemphigus should be suspected in cases of persistent gingivostomatitis, persistent and multiple oral erosions or severe desquamative or erosive gingivitis as they mimic clinically.[9] The most frequent diagnoses in cases of oral lesions are recurrent aphthous stomatitis, Behcet's disease, erythema multiforme, erosive lichen planus and oral candidiasis. ELISA for the detection of antibodies to Dsg1 and Dsg 3 can be used for diagnosis of Pemphigus.[10] Assay of serum antibody titers by indirect immunofluorescence (IIF) may also help to guide procrastination and therapy.[11] Lesions of oral mucosa in patients with low antibody titers may be controlled with mouth washes or topical creams containing corticosteroid. Intralesional injection of triamcinoclone acetonide or paramethasone can be used in refractory oral lesions. The wellbeing of patients may be improved by analgesics, a strict oral hygiene with diluted antiseptic mouth washes, and a soft diet without irritants, correct prosthetic restorations and anti-candida therapy.[12] Traumatisations may trigger or exacerbate Pemphigus; therefore some authors recommend the prophylactic use of prednisolone (20mg/day) for 5-7 days before dental procedure involving gums.

The age of the patient and initial Dsg 3 antibody levels have a significant impact on the prognosis of Pemphigus vulgaris. The initial aim of treatment should be to induce disease remission. This should be followed by a period of maintenance treatment using the minimum drug doses required to control disease in order to minimize their side effects. Pulse therapy using Dexamethasone and Cyclophosphamide has been used for management of pemphigus in India and abroad for the past 30 years.[13]

CONCLUSION

Pemphigus is a rare cause of chronic mouth ulcers. Oral manifestations are the first presenting symptoms of the disease in Pemphigus patients. Dermatological lesions usually occur 2-6 months after the appearance of oral mucosal hemorrhagic lesions. This can lead to misdiagnosis, delayed diagnosis and inappropriate treatment of a potentially fatal disorder. In the present case the patient had previously undergone treatment based on misdiagnosis and whose complaints were not relieved even after a long period of treatment by various specialists. The importance of the roles of dentists in early diagnosis and treatment of oral pemphigus vulgaris and the lack of awareness about oral manifestations of Pemphigus among general practitioners was evident here. These oral features help in early diagnosis of disease and prompt treatment procedures to be followed in initial stages of the disease itself.

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