Desquamative Gingivitis in Cicatricial Pemphigoid: A Diagnostic Paradox!

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Abstract

Desquamative gingivitis or gingival desquamation represents a clinical sign seen in many diseases, especially of muco-cutaneous nature. One of the common diseases, in which gingival desquamation is present is Mucous membrane pemphigoid (MMP) and represents a variety of chronic auto-immune sub-epithelial bullous diseases affecting the mucous membranes and sometimes the skin. MMP not only affects the oral mucosa but ocular involvement is also quite common. The severity of the disease is variable, and the gingival desquamation may be the first sign of this otherwise indolent disease and early diagnosis can reduce the morbidity especially ocular changes which could be irreversible leading to blindness can be prevented. We describe a case for documentation while describing the differential diagnosis and management of the disease.

Keywords: *Gingival desquamation, muco-cutaneous disorder, ocular involvement, sub-epidermal bulla.*

Introduction

The word "desquamation" taken from the Latin word 'Desquamare', meaning scraping fish flakes. In the literal sense, desquamation implies loss of epithelium, peeling off skin or mucosa. Desquamative gingivitis was first described by Tomes in 1894.[1] It is not a disease by itself rather a manifestation of some disease. It can progress to vesicle formation, erythema, erosion and ulceration of the attached gingiva.[2] Glickman and Smulowin 1964 stated that desquamative gingivitis could be a clinical manifestation of many diseases.[3] Dermatologic diseases, hypersensitivity, endocrine disturbances and chronic bacterial infections are common causes.[4]

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MMP or cicatricial pemphigoid accounts for the maximum number of cases of desquamative gingivitis and sometimes the gingival lesions are the sole manifestation of the disease, hence it requires meticulous history taking and careful examination by the general dentists as they have an important role in the diagnosis of this chronic muco-cutaneous disease.[5]

Case Report: A 53-year-old female patient reported with a complaint of redness and burning sensation in the gingiva. History revealed she had pain and bleeding while cleaning her teeth. Her medical, surgical, personal and family history were non-contributory. There were no lesions anywhere else in the body.

On clinical examination, the left sub mandibular lymph node was slightly palpable, movable and tender. The attached gingiva in the mandibular anterior region and the entire maxillary arch was erythematous and tender on palpation. (figure 1-5) There was desquamation and mild bleeding on probing. On applying compressed air to the apparently normal gingiva, there was desquamation. This elicited a positive Nikolsky sign in the oral cavity.

Since the patient had got gingival desquamation and

an erythematous area the lesion clinically was thought of as desquamation due to hormonal disturbances, lichen planus, pemphigus and lupus erythematosus. But the absence of white striae in the lesion, absence of any menstrual irregularities, and any other systemic signs and symptoms with a positive Nikolsky sign indicated towards MMP.

The patient was advised for an incisional biopsy and H & E stained section revealed sub-epithelial cleft and basal cell degeneration, with haemorrhagic areas, suggestive of mucous membrane pemphigoid. (figure 6) The patient was advised to apply a combination of Clobetasol propionate 0.05% and Miconazole 1% cream, thrice a day for a month and was to be reviewed. The patient was lost to follow-up.

Discussion

Mucous membrane pemphigoid is a chronic sub-epithelial bullous disorder mainly affecting elderly ladies.[6] However, there is no racial or geographic predilection. It has a rather complex aetiopathogenesis as it is a heterogenous disorder with the involvement of multiple antigens. The antibodies produced in MMP have been shown both in vitro as well as in vivo. Generally, there are IgG and IgA antibodies circulating in the blood directed against the parts of the basement membrane zone (BMZ) which show that humoral response against the antigens occurs in MMP.[7] Mislaying of immunologic tolerance to morphologic proteins in the BMZ leads to the formation of autoantibodies. Using immune-precipitation and immune-blotting mechanisms, many antigens have been discovered. The bullous pemphigoid antigen 2 (BPAg2) (a 180-kDa protein, BP180) is the most commonly targeted autoantigen in MMP.[8] When there is reaction between autoantibodies against antigens it results in detachment of the epithelium within the BMZ.

MMP affects different mucosal sites, rarely there is cutaneous involvement. It is a chronic, indolent disease which commonly involves the oral mucosa followed by the conjunctiva, nasal mucosa, genital mucosa, gastro-oesophageal mucosa and the lesions always heal with scarring.[9] The disease differs in severity ranging from localized to a diffuse disseminated involvement. Initially the disease may be localized and later there is extensive involvement. Since the lesions heal by scarring, it leads to significant morbidity especially in those patients with ocular involvement.[10]

Oral lesions frequently occur on the gingiva and the palate, followed by labial mucosa, tongueand buccal mucosa.[11] These lesions can occur as erythema, erosions, pseudo-membrane, and rarely intact bullae. The gingival involvement manifests as desquamative gingivitis, which is not specific for MMP but may also be seen in oral lichen planus and pemphigus vulgaris.[12] Ocular lesions manifest as conjunctival erosions and erosions, keratinization, symblepharon, enteropion, trichiasis, ankyloblepharon, ankylosis and even blindness.[13]

The diagnosis needs histopathologic evaluation of the tissue samples which needs to be taken with utmost care as the epithelium easily dislodges from the underlying connective tissue. H & E sections characteristically reveals sub-epithelial split with an inflammatory infiltrate of chronic inflammatory cells.[14] Another specimen may also be taken from perilesional tissue and taken in buffered hypertonic saline solution and submitted for direct immunofluorescence. A linear IgG and C_3 is seen along the BMZ.[8,15]

Therapy for MMP comprises of early treatment of the lesions and aims at preventing complications especially when there is ocular involvement. The scarring can be avoided by early therapeutic interventions. Treatment is tailor made for individual patients based on the severity of the disease considering the age, general wellbeing, medical issues and contra-indications to the use of systemic drugs. Multidisciplinary treatment with a team of experts consisting of oral medicine specialist, dermatologist, ophthalmologist, gastroenterologist typically improve outcomes.[16]

High-potency topical glucocorticoids remain the mainstay of therapy. Fluocinonide, clobetasol propionate and betamethasone dipropionate are the prescribed drugs. Gingival desquamation alone can be well managed using topical glucocorticoids alone.[17] Use of a splint or custom-made tray to hold the gel on to the gingiva may be more effective. However due to long term use of these steroids leading to possible adverse effects like candidiasis, concomitant of topical anti-fungal drugs also is prescribed as in the present case a combination of clobetasol and miconazole was used.[18,19]

Intralesionaland systemic glucocorticoids are used to treat recalcitrant lesions or as an adjunct totopical steroid delivery. For patients not responding to steroids, calcineurin inhibitors, which are potent

immunosuppressants like Tacrolimus is prescribed with good efficacy and safety.[19]

Palliative care is also required to alleviate the masticatory discomfortusing topical anaesthetics-analgesic-antihistaminic mouthwashes or gels and improve patient's capability to eat, swallow and perform oral hygiene. [17-19]

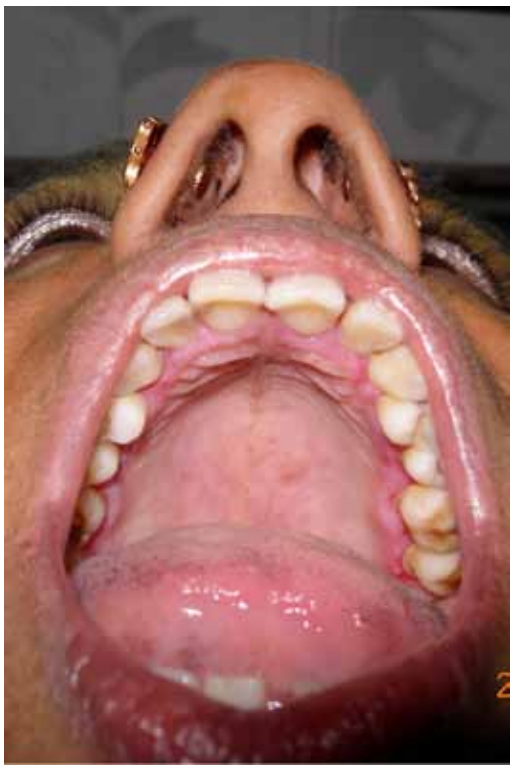

Figure 1: Gingival desquamation in the maxillary palatal gingiva.

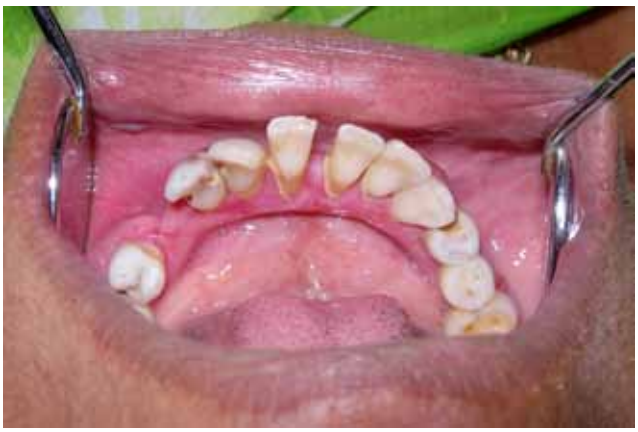

Figure 2: Gingival desquamation in the mandibular lingual gingiva.

Figure 3: Gingival desquamation in the attached gingiva labially and buccally.

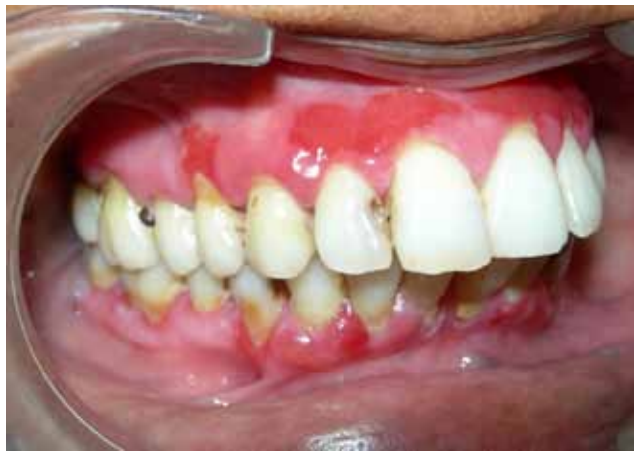

Figure 4: Gingival desquamation in the attached gingiva labially and buccally.

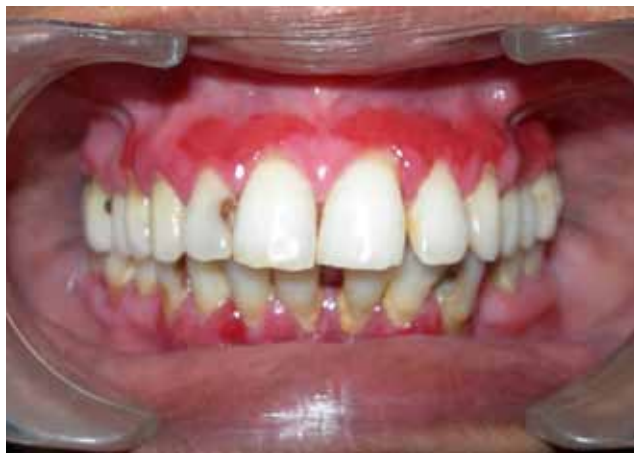

Figure 5: Gingival desquamation in the attached gingiva labially and buccally.

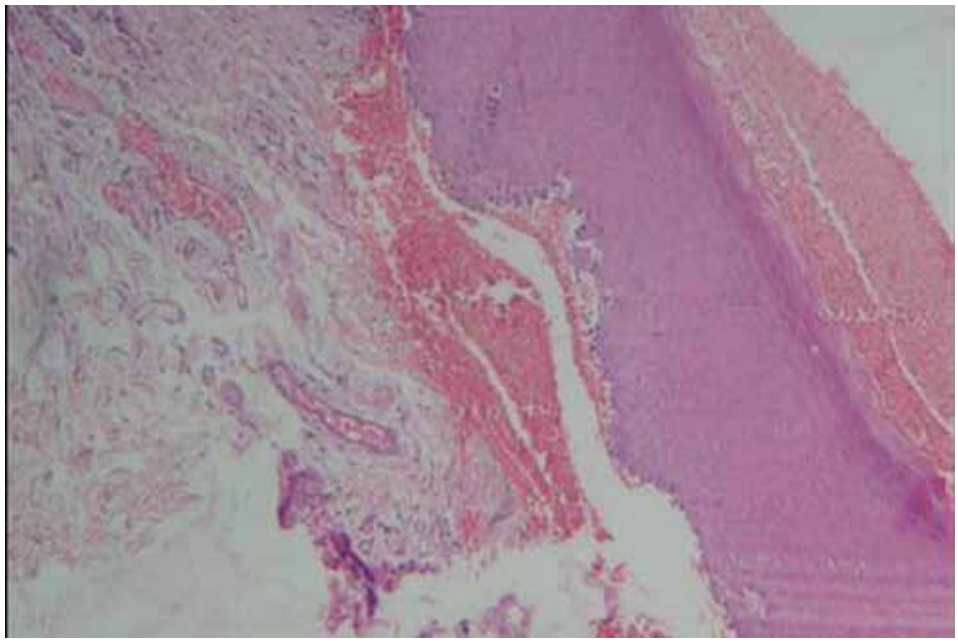

Figure 6: H & E (10 X original magnification) stained section revealed sub-epithelial cleft and basal cell degeneration, with haemorrhagic areas.

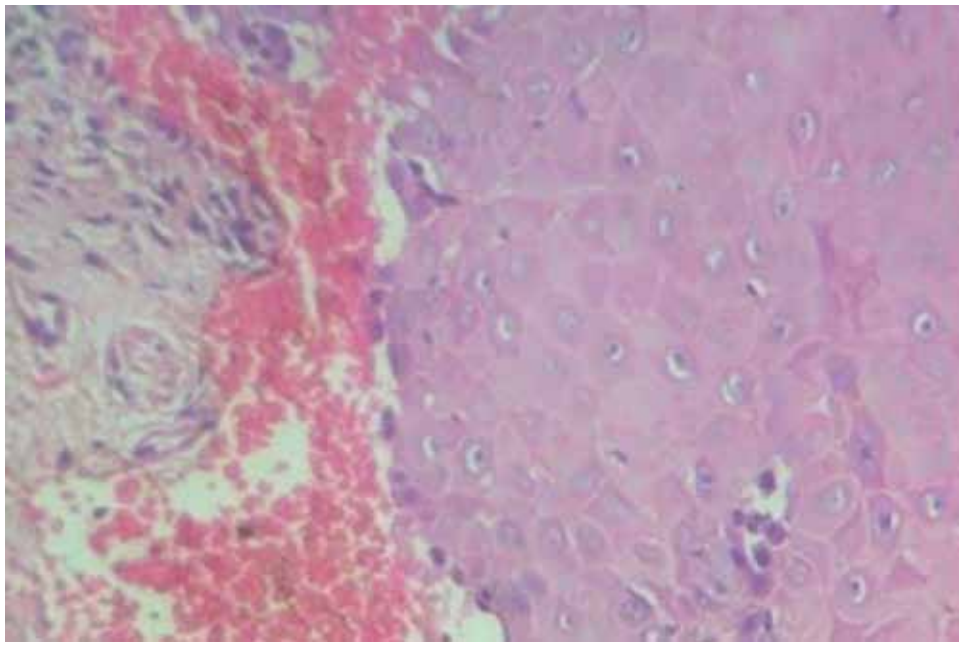

Figure 7: H & E (40 X original magnification) stained section revealed sub-epithelial cleft and basal cell degeneration, with haemorrhagic areas.

Conclusion

Dentists are often the first people to examine and diagnose this mucosal disease. Prompt diagnosis of mucous membrane pemphigoid requires taking a careful meticulous history together with a detailed oral mucosal examination along with checking the possible involvement of other mucosae and using histopathologic and immunofluorescent studies. Desquamative gingivitis may be the sole manifestation of the disease and usually heals uneventfully by improving the oral hygienic measures with topical glucocorticoidand antifungal therapy.

Conflict of Interests: None

Ethical Permission: Approved

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