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Review Article

Desquamative gingivitis as a sole presentation in mucous membrane pemphigoid: A rare case report with a brief review of literature

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ABSTRACT

Desquamative gingivitis refers to an erythematous, ulcerated, or atrophic appearance of the gingiva. This clinical appearance is common to a number of pathologies, most frequently accounting for mucocutaneous autoimmune entities (Oral lichen planus and Mucous membrane pemphigoid). The oral physician must be well versed with its varied clinical presentations, and a prompt diagnosis is essential for early remission from this debilitating state.

1. Introduction

Desquamative gingivitis manifests as fiery red, glazed, atrophic or ulcerated appearance of gingiva. Loss of stippling is noticed and minor trauma results in gingival desquamation. In distinction to plaque triggered gingivitis, desquamative gingivitis is frequently seen in middle aged to elderly females, exhibits tenderness, primarily affects the buccal / labial gingiva, and may involve the entire attached gingival tissue. Conventional oral hygiene procedures or periodontal treatment alone does not result in alteration of the clinical appearance. Desquamative gingivitis lesions are best managed by oral hygiene maintenance, management of the underlying disease and the intake of local or systemic immunosuppressive drugs.[1,2] Tomes and Tomes (1894) were the pioneer in describing chronic desquamative gingivitis.[3] However, Prinz[4] and Merrit[5] suggested the nomenclature "chronic diffuse desquamative gingivitis" and first endeavored to explain the disease progression.

The oral health practitioner should be aware with the diverse clinical appearances of desquamative gingivitis and should be able to frame an accurate differential diagnosis. The etio-pathogenesis of Desquamative gingivitis associated entities may be classified in 2 major classes- a) Cell-mediated (e.g., Oral Lichen Planus, and lupus erythematosus), b) auto-antibody-mediated (e.g., Pemphigus Vulgaris, Mucous membrane pemphigoid, and erythema multiforme). The former usually causes amendments in the epithelial thickness/hyperkeratosis, while the latter results in blistering.[6]

Desquamative gingivitis represents a clinical presentation and is not a diagnosis per se. Glickman and Smulow[7] (1964) proposed that the entity may be a mutual clinical manifestation occurring in several disorders. Based on its etiology, coupled with histopathological and immunofluorescence features, desquamative gingivitis can be broadly classified into.[8]

1.1. A. Mucocutaneous diseases

1. Mucous membrane pemphigoid (MMP)
2. Oral lichen planus (OLP)
3. Pemphigus vulgaris (PV)
4. Psoriasis
5. Bullous pemphigoid
6. Epidermolysis bullosa acquisita

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7. Lupus erythematosus
8. Contact stomatitis
9. Chronic ulcerative stomatitis
10. Dermatitis herpetiformis
11. Linear IgA disease
12. Kindler's syndrome

1.2. B. Endocrine disturbances

1. Oophorectomy and menopause resulting in estrogen deficiency
2. Testosterone imbalance
3. Hypothyroidism

1.3. C. Local hypersensitivity reactions to various allergens

1. Plasma cell gingivitis
2. Mouth washes
3. Dental materials
4. Drugs
5. Cosmetics
6. Sodium lauryl sulphate in Toothpastes
7. Chewing gums

1.4. D. Unusual bacterial plaque response

1.5. E. Aging

1.6. F. Chronic granulomatous diseases

1. Tuberculosis
2. Sarcoidosis
3. Histoplasmosis
4. Crohn's disease
5. Wegener's granulomatosis

Overall, OLP, MMP and PV constitutes the majority of desquamative gingivitis cases, with OLP and MMP contributing for 80% of the causes.[1]

2. Case Report

A 53-year-old postmenopausal female patient was referred to our Outpatient department for persistent burning sensations and gum tenderness on eating hot and spicy food for the past 2 years. She has taken multiple treatments for the same without much of a relief. History revealed that the patient had observed on and off gum blister formation which would rupture spontaneously. The medical history was unremarkable, and general physical examination did not reveal any associated cutaneous, ocular, or genital lesions. Intraoral assessment was suggestive of extensive inflamed and erythematous labial and buccal gingiva, and desquamative lesion was noticed on the buccal aspect of free, marginal, and attached gingiva irt[9–11] and with mild sulcus suppuration. On close inspection, a pin-point vesicle 0.3x0.2 mm was also appreciable involving the marginal gingiva of.[9,10] [Figure 1 a]. Gentle handling of the normal mucosal tissue resulted in a positive Nikolsky's sign. An elliptical erosion 1.5x1cm in diameter covered by fibrin-coated pseudo membrane was seen irt 25 involving the attached and marginal. [Figure 1 b]. The patient had a fair oral hygiene and gingival bleeding on probing was seen without attachment loss. Correlating a history of gum tenderness and burning sensation with occasional blister formation, and clinical assessment showing positive Nikolsky's sign, desquamative gingivitis, and an isolated erosion secondary to bullae formation, the condition was provisionally diagnosed as Desquamative gingivitis secondary to vesiculo-bullous disorder. Mucous membrane pemphigoid, pemphigus vulgaris, bullous pemphigoid, and bullous lichen planus were given a place in the differential diagnosis. Incisional biopsy from the perilesional gingival tissue was made after an informed written consent, and histopathology revealed variably thickened parakeratinised stratified squamous epithelium with distinguishing subepithelial clefting and disintegration of the basal cell layer. Chronic inflammatory cells (predominantly plasma cells) with interspersed hemorrhage and vascularity were seen in the connective tissue [Figure 2 a & 2b]. Direct immunofluorescence was suggestive of C3 and IgG linear deposits at the dermo-epidermal intersection [Figure 2 c]. Based on clinical, histopathological and immunofluorescence studies, a final diagnosis of Mucous membrane pemphigoid was arrived at. After a thorough oral prophylaxis and oral hygiene instructions, triamcinolone acetonide 0.1% (Kenacort 0.1%) was prescribed for topical application 2-3 times daily for a month and chewable vit. C tablets (Celine). The patient was advised regular follow up for the first month. Significant healing was achieved with topical steroids therapy [Figure 3 a & 3b].

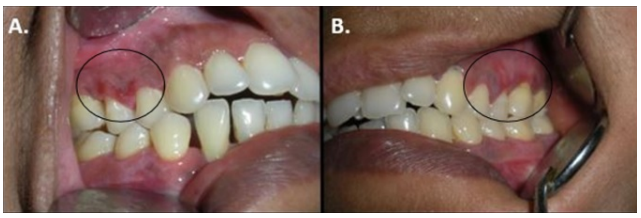

Fig. 1: a: & b: Desquamative gingivitis with erosive lesion.

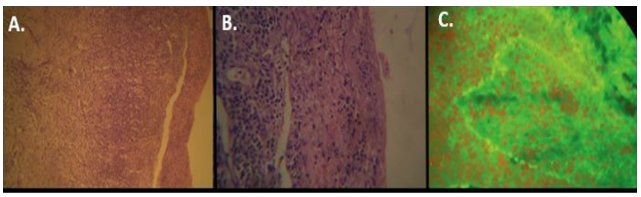

Fig. 2: a:- c: Histopathology & DIF with typical MMP features.

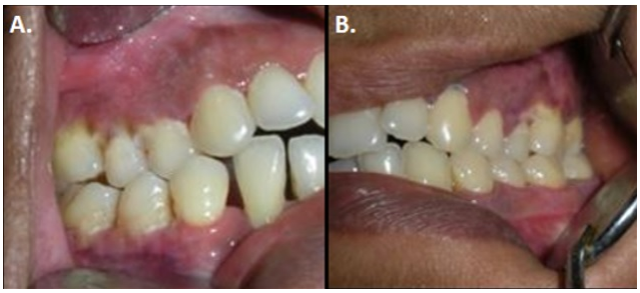

Fig. 3: a: & b: Healed lesions.

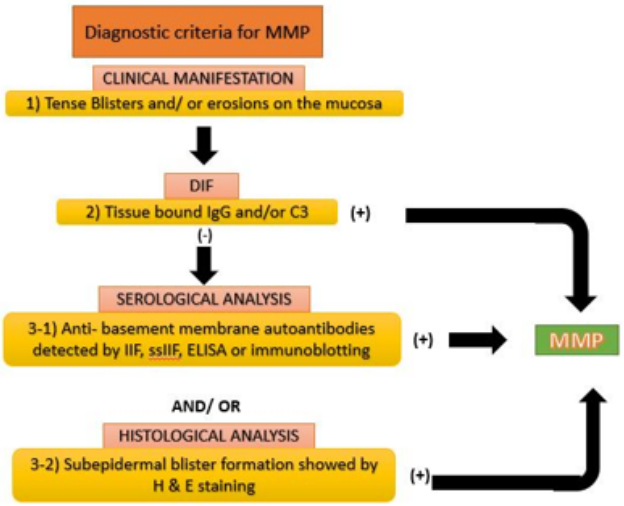

Fig. 4: Diagnostic algorithm for MMP.

3. Discussion

Mucous membrane pemphigoid (MMP) is a rare, autoimmune, subepithelial blistering entity typified by a chronic course, primary mucosal involvement, and a propensity towards scarring of the involved region.[12]

MMP is an autoimmune disease with an obscure etiology. Inflammatory mucosal injuries,[13] drugs (indomethacin, clonidine, D-penicillamine),[14] ultraviolet radiation, viruses, and sporadic association with other autoimmune diseases are some of the predisposing factors.[15] Ethnic or geographic preponderances are not known, but there is a likelihood of an immunologic backdrop that is associated with HLA DQB1*0301.[9,16]

The probable pathogenesis of MMP includes an autoantibody-induced, complement-facilitated leukocytic sequestration with subsequent cytokines and leukocytes liberation, causing basal cell detachment from the basement membrane. This results in subepithelial slit formation which is the classical histopathological finding in MMP. IgG autoantibodies produced by MMP patients primarily attack the BP180 epidermal antigen.[10] The reported incidence of MMP is 1.5-9.5 cases/1 lakh individuals annually.[11] There is a preference for middle aged females (F:M=2.7:1), although a few cases have occurred in the elderly and children.[17] MMP is typified by a gradual onset intermingled by acute flare ups and recoveries.[18] MMP preferentially involve several mucosal areas, with infrequent skin involvement. It is a long-standing, gradually progressing disorder that primarily affects the oral mucosa (85%), followed by conjunctiva (65%), nasal mucosa (20-40%), cutaneous (25-305), anogenital area and/or pharynx (20%), larynx (5-15%), and esophagus (5-15%).[19]

The most frequently involved site in MMP is the oral cavity. Gingiva (80%) accounts for the most frequent affected intraoral site, followed by check mucosa (58%), palatal mucosa (26%), alveolar ridge (16%), tongue (15%), and lower lip (7%).[20] Desquamative gingivitis, vesicles/bullae formation, and ulcerations are the salient clinical manifestations,[21,22] and the lesions exhibit healing with minimal scarring. The cardinal manifestation of MMP is desquamative gingivitis[23] and it may be the only manifestation in some cases. Desquamative gingivitis may exhibit varied presentation ranging from mild trivial lesions to extensive erythema with a friable gingival surface. Flaccid vesicle or bullae may be seen, which eventually break leaving irregular erosions covered with a yellowish slough and a persistent adjacent inflammatory halo.[24] Minimal sliding pressure elicits a distinct epithelial peeling (positive Nikolsky's sign),[20] although, it may also be seen in pemphigus vulgaris, epidermolysis bullosa, and erythema multiforme.[25,26] Advanced lesions may result in adhesion development between the check mucosa and the alveolar process, peri-tonsillar region, and between the floor of the oral cavity and ventral tongue surface. Frenal involvement may result in restricted tongue activity or ankyloglssia.[27]

The clinical appearance and severity vary significantly among individuals with localized and widespread involvement. "Low-risk" MMP patients present with exclusive oral mucosal and/or cutaneous involvement, with less predisposition to scarring. This is in contrast to the "High-risk" patients where the disease occurs in one of the following areas: ocular, genital, esophageal, laryngeal, and nasopharyngeal mucosa. The lesions in these sites have a poor prognosis as they are more vulnerable to scarring.[19] Clinical diagnosis of MMP is quite problematic, primarily due to lower autoantibody titer in MMP. Thus, circulating autoantibodies are identified infrequently in MMP than in bullous pemphigoid resulting in diagnostic conflicts.[28] The definitive diagnosis can only be established based on the histopathological and immunofluorescence findings.[29] The biopsy for routine histology and direct immunofluorescence (DIF) must be preferred from the vesicle, ulcer edge, or erythematous tissue. Histopathology reveals subepithelial clefting with varied inflammation/mixed inflammatory infiltrate. The gold standard diagnostic test is positive direct immunofluorescence (DIF) studies for IgG, IgM, C3, and often IgA is seen in 50% to 80% of cases.[30]

The international consensus has established that the clinical features of mucosa-predominant lesions and DIF detecting tissue-bound IgG, IgA, and/or C3 are critical for the diagnosis.[28] [Figure 4]

Maintaining dental hygiene and avoiding local predisposing factors is essential in the treatment MMP.[31] Coordinated efforts between the oral and general physician is required for an efficient systemic therapeutic protocol. MMP therapy frequently entails topical corticosteroids either alone or as combination therapy with systemic corticosteroids.[32] Dapsone, an antimicrobial agent with immunosuppressive action, has proved to be promising.[33–35] However,dapsone usage should be regularly followed with periodic blood studies because of the associated complication of hemolytic anemia. Other systemic medications, including immunosuppressive agents such as azathioprine, methotrexate, cyclophosphamide, levamisole, and mycophenolate mofetil may also prove beneficial in the treatment of pemphigoid,[36–42] Studies have shown that MMP can be treated efficiently with tetracycline derivatives or combination therapy of tetracycline and niacinamide.[39–41] Alternately, intravenous immunoglobulins, plasmapheresis, and Low-level laser therapy (LLLT) can also be used as a therapeutic modality.[42,43]

4. Conclusion

Desquamative gingivitis should be considered in the differential diagnosis of vesiculo-bullous disorders, and a detailed history, clinical assessment coupled with histopathology and immunofluorescence is essential for a confirmatory diagnosis.

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None.

6. Conflict of Interest

None.

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