CASE REPORT

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Desquamative Gingivitis as the First Clinical Sign of Pemphigus Vulgaris: A Case Report

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ABSTRACT

Desquamative gingivitis is characterised by desquamation of the gingiva with painful erosion and ulceration. It is predominantly a manifestation of several vesiculobullous diseases. Delayed diagnosis or misdiagnosis often led to disease progression. Pemphigus vulgaris is a chronic, life-threatening autoimmune disease resulting in blistering of the mucosa and skin. Oral lesions normally preceded skin lesions. Early diagnosis and treatment are important to prevent involvement of the skin, as the treatment and prognosis varies with extraoral involvement. Clinical, histopathological examination and direct immunofluorescent are necessary for the diagnosis of pemphigus vulgaris. Treatment of desquamative gingivitis involves improving oral hygiene, reduce irritation to the lesions and specific therapy to the underlying disease. This paper describes a case of a patient with desquamative gingivitis for one year, whom is ultimately diagnosed as having pemphigus vulgaris.

Keywords: *Desquamative gingivitis; direct immunofluorescent; oral lesions; pemphigus vulgaris; vesiculobullous diseases*

INTRODUCTION

Desquamative gingivitis is a clinical condition characterised by an intense erythema, blistering of the marginal and attached gingiva with epithelial desquamation or erosion that can lead to painful ulceration which may be localised or widespread, and may extend extra orally (Endo *et al.*, 2017), with women predilection (80%) and occurs in middle-ages (Maderal *et al.*, 2018; Shaqman *et al.*, 2020). It is not a specific diagnosis, but rather a descriptive term of gingival conditions (Odell, 2017) and is recognised to be a nonspecific manifestation of several diseases, ranging from vesiculobullous diseases to chemicals or allergies reactions. The most commonly diseases involved are lichen planus (24% to 80%), mucous membrane pemphigoid (14% to 48%) and pemphigus vulgaris (3% to 15%) (Shaqman *et al.*, 2020). Desquamative gingivitis can be the first or only clinical manifestation of associated diseases in 7% to 22% of cases (Endo *et al.*, 2017). Patients having desquamative gingivitis may show mild to severe presentation. In severe presentation, desquamation,

blistering, bleeding, erosions and ulceration are seen. Patients generally complain of oral discomfort, gingival soreness, or burning sensation (Scully & Porter, 1997; Endo *et al.*, 2017; Popescu *et al.*, 2019). These symptoms resulting in painful brushing and thus poor oral hygiene. The accumulation of dental plaque further aggravates gingival inflammation and possibly lead to periodontal breakdown and alveolar bone destruction (Maderal *et al.*, 2018).

Pemphigus vulgaris is a rare, chronic, life-threatening autoimmune disease characterised by acantholysis in the epithelium, forming multiple bullae or erosion that show little tendency to heal, on the skin or mucosal membrane (Neville *et al.*, 2008; Endo *et al.*, 2017; Schmidt *et al.*, 2019). Most patients with pemphigus vulgaris are in their fourth and fifth decade of life, and the disease is equal in men and women (Endo *et al.*, 2017). In most cases the disease started with oral lesions. Skin lesions may develop concomitantly or appear later during the course of disease (Popescu *et al.*, 2019; Schmidt *et al.*, 2019). Oral lesions are characterised by bullae that rupture rapidly, leaving erosion and shallow ulceration (Popescu *et al.*, 2019). At times, the gingiva is the only site involved in early lesions and manifests as desquamative gingivitis (Endo *et al.*, 2017; 2018). Delayed diagnosis and inappropriate treatment of pemphigus vulgaris should be minimised to avoid morbidity and sometimes fatal complications thus detection in early stage can prevent the progression to skin involvement. In this case report, we describe a patient presenting with persistent desquamative gingivitis of one year, whom is finally diagnosed as having pemphigus vulgaris.

CASE REPORT

A 33-year-old female patient of Dusun ethnicity was referred to Periodontology Unit, Putatan in Sabah, for persistent desquamative gingivitis. She complained of ulcers and soreness in mouth for the past one year which has led to discomfort and progressively affected her oral intake. It started with multiple ulcers in the mouth during second trimester of pregnancy which sometimes coalesce to form a bigger ulcer with ragged border. Burning sensation aggravated on taking hot, spicy and acidic food, and painful gingiva with greyish sloughs covering her gingiva from time to time were noted. Ulcer relieving gel and antifungal mouthwash were advised by different dentists to no avail. A physician informed the patient that her oral lesions were possibly a pregnancy allergy reaction. No genital or ocular involvement were experienced. She delivered a healthy neonate six months prior on normal term, with no improvement on the lesions post-delivery. Patient experienced stress in taking care of her baby and weight loss due to eating difficulty.

On general examination, multiple hyperpigmented patches were noted on upper and lower limbs noted. Intra-oral examination revealed generalised moderate to severe inflammation with ulceration of marginal gingiva and interdental papilla (Fig. 1A), marginal gingiva with epithelial detachment leaving erythematous area (Fig. 1B), diffuse shallow erosion with ulceration on the left and right aspect of hard palate extending to the soft palate (Figs. 1C and 1D). Multiple irregulars with erythematous surrounding were noted on buccal mucosa, buccal and labial sulcus and lateral borders of the tongue. This gingival lesion showed easily bleeding on manipulation. Mucosa over the lower lip peeled on lateral pressure suggesting positive Nikolsky's sign. The patient had poor oral hygiene and presented with multiple teeth with grade I and grade II mobility. Meanwhile, the highest score of Basic Periodontal Examination (BPE) was score of 3.

Based on the history and clinical findings, these clinical features were described as desquamative gingivitis which are commonly seen in pemphigus vulgaris, mucous membrane pemphigoid, lichen planus,

and recurrent aphthous ulcers. Routine haematological investigation showed reduced level of mean corpuscular volume (MCV) and mean corpuscular haemoglobin (MCH). Incisional biopsies were taken from the perilesional area at left buccal mucosa and right hard palate. The samples were submitted for histopathological investigation and direct immunofluorescence (DIF) microscopic in the Department of Oral Medicine and Pathology, Queen Elizabeth Hospital. Histopathological examination showed para keratinised stratified squamous epithelium exhibiting acantholysis and supra basal cleftings (Fig. 2A). In some areas, the epithelium was almost loss with remaining rows of basal and supra basal cells forming "tombstone" appearance (Fig. 2B). DIF microscopy revealed intercellular deposition of IgG (immunoglobulin G) and C3 (complement component 3) (Fig. 3). These

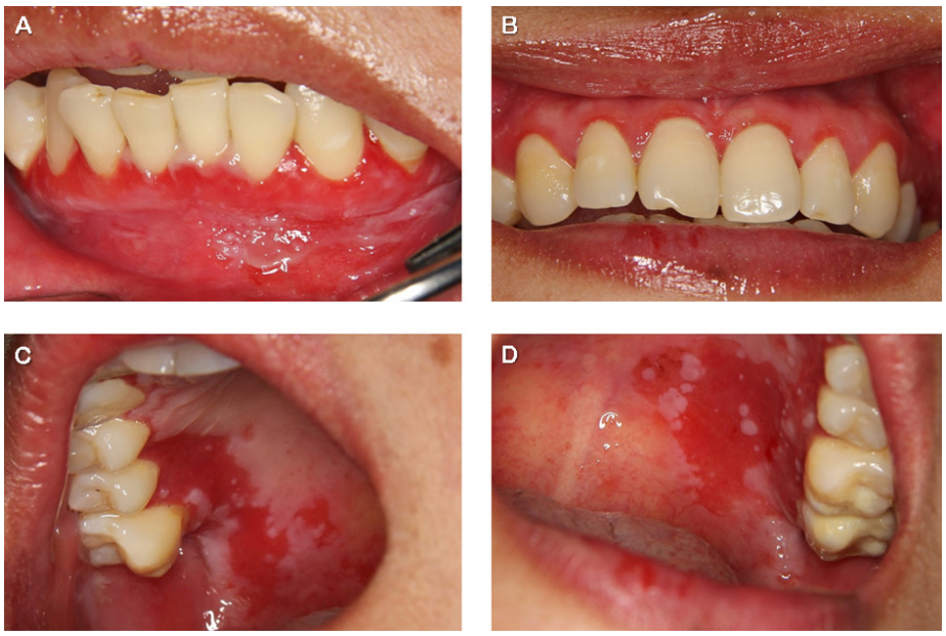

Fig. 1 (A) Generalised erythematous marginal gingiva on upper teeth; (B) Marginal gingiva with epithelial detachment leaving erythematous area; (C) Erosion on hard and soft palate right side; (D) Erosion on left hard palate and soft palate and peeling on the upper and lower lips.

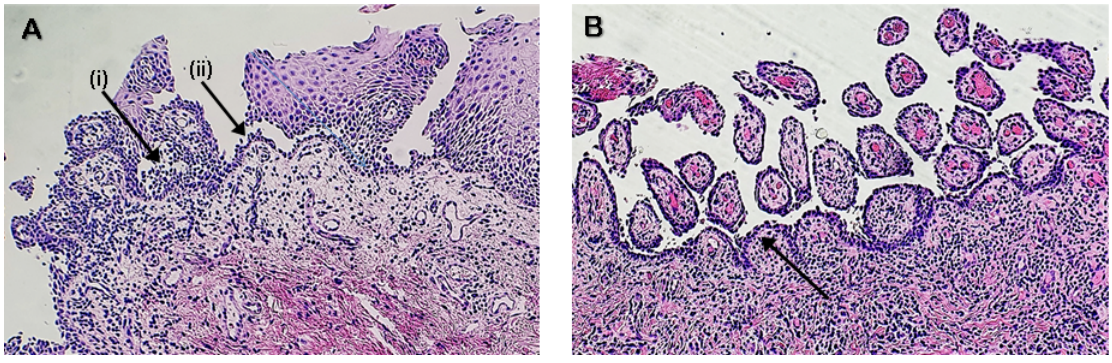

Fig. 2 (A) (i) Photomicrograph shows loss of intercellular adherence of suprabasal spinous cells (acantholysis) and (ii) formation of clefts by separation of epithelium just above the basal cell; (B) Photomicrograph shows characteristics feature of a layer of basal cells remains (arrow) covering the mucosal papillae (original magnification: 40×; H&E stain).

evaluations gave a definitive diagnosis of pemphigus vulgaris.

Supragingival scaling and proper oral hygiene instructions were given before patient was referred to the Department of Oral Medicine and Pathology for further management. Patient initially started with topical steroid mouthwash (dexamethasone 2 mg in 10 ml water) for 10 days. Followed with topical steroid paste (clobetasol propionate 0.05% + triamcinolone acetonide [1:1]) three times daily (TDS). Patient was also advised to change to lower potency steroid such as betamethasone valerate 0.1% + triamcinolone acetonide [1:1] TDS once condition improves. Three weeks after the initiation of topical steroids, patient claimed there was reduce in pain and better oral intake (Fig. 4A). However, slow improvement was noted on the palatal area upon re-examination (Fig. 4B). On subsequent visit three weeks later, patient presented with multiple skin lesions on her body, where she was then referred to a dermatology clinic. Skin biopsy was carried out and patient was prescribed with systemic steroids with tapering dosage of prednisolone. The oral lesions and skin lesions have ultimately shown improvements with systemic steroids therapy. Patient is currently under monthly follow up with the Department of Oral Medicine and Pathology, and the Dermatology Clinic for Management of Pemphigus Vulgaris. As for periodontal managements, oral hygiene review and root surface debridement on teeth with pocket depths (PD) ≥ 4 mm (only PD of 4 mm with bleeding on probing) were planned for every 6 to 8 weeks intervals with the aim to stabilise her periodontal condition.

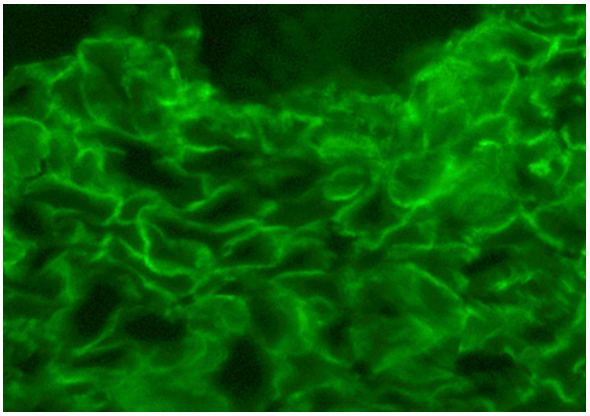

Fig. 3 Frozen tissue stained with fluorescent antibody IgG shows green fluorescence along the lines of intra-epithelial attachments of the keratinocytes typical of pemphigus (original magnification: 40×; H&E stain).

DISCUSSION

Most cases of desquamative gingivitis are of severe mucocutaneous diseases. Lichen planus, mucous membranous pemphigoid and pemphigus vulgaris account for more than 80% of cases. Although oral lesions of pemphigus vulgaris can occur anywhere in the oral cavity, the oral lesion noted to be

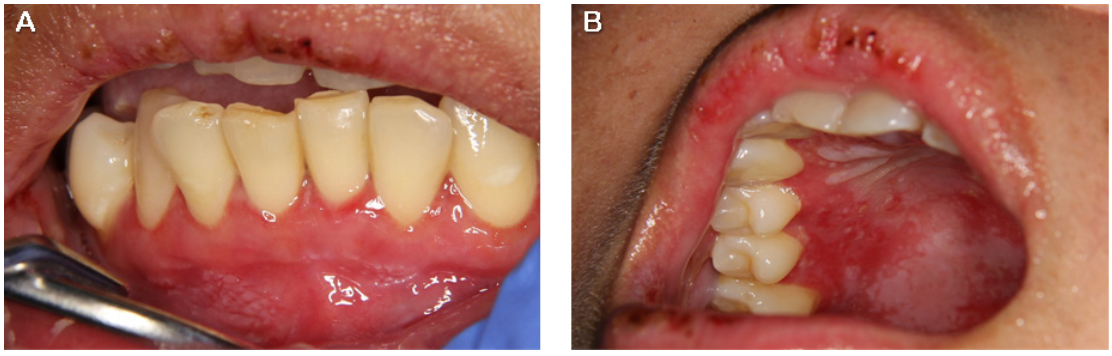

Fig. 4 (A) Less erythematous areas on the gingiva three weeks after treatment with topical steroids; (B) Slow improvement on hard palate three weeks after treatment with topical steroids. Erosion on anterior palate and toward midline, with superimposed candidiasis infection and crusting on lips.

limited to gingiva is between 3% and 30% (Endo *et al.*, 2018). In this case, patient had lesions in several sites of the oral cavity. However, she came to us with concern of painful and bleeding gingiva with burning sensation, which is the characteristic of desquamative gingivitis, in spite of having other oral lesions.

The clinical appearance of desquamative gingivitis sometimes mimics the commonly seen plaque-induced gingivitis hence misdiagnosis and delayed diagnosis are usually encountered by dental practitioners. In fact, diagnostic delays of more than 6 months were common in oral pemphigus vulgaris showing that early diagnosis of autoimmune bullous diseases intraorally was still a challenge (Endo *et al.*, 2018). In this patient, one year was taken for a definitive diagnosis of pemphigus vulgaris to be made. During this period, different misdiagnosis and medications were given before she was finally referred to us. Thus, dentists are obliged to thoroughly consider the differential diagnosis of oral lesions. Misdiagnosis prolong the disease progression leading to widespread of desquamation. This usually comes with chronic pain and reduced oral intake resulting in weight loss and malnutrition (Maderal *et al.*, 2018; Schmidt *et al.*, 2019), which was unfortunately experienced by our patient.

Accumulation of dental plaque aggravates inflammation of the periodontium, which led to periodontal attachment loss and bone destruction, and ultimately tooth loss (Maderal *et al.*, 2018; Shaqman *et al.*, 2020). This condition is consistent with our patient who presented with poor oral hygiene and multiple mobile teeth.

In histological examination, pemphigus vulgaris is characterised by acantholysis of the epithelium. This characteristic is caused by circulating pathogenic pemphigus vulgaris autoantibodies in the serum. These autoantibodies mostly target desmoglein, Dsg3 and Dsg1, which is a constituent of desmosomes on the keratinocytes. Desmosomes responsible for connecting neighbouring keratinocyte and important for the integrity of various tissues (Endo *et al.*, 2017; Popescu *et al.*, 2019; Schmidt *et al.*, 2019).

Oral involvement usually preceded skin lesions by several months as characterised by ragged erosion, painful ulceration, and desquamative gingivitis. Bullae are prone to rupture hence infrequently seen. Nikolsky's sign is not specific for pemphigus vulgaris, as it also can be seen in other diseases such as mucous membrane pemphigoid and erythema multiforme (Urbano, 2001). Most cases of pemphigus are idiopathic, but can be triggered by drugs, radiation, surgery, garlic, emotional stress, virus (HPV 8), pesticide exposure and pregnancy (Schmidt *et al.*, 2019). Our patient had onset of lesions during second trimester, and had delivered a healthy neonate.

Definitive diagnosis of disease causes desquamative gingivitis is not possible by merely depending on the clinical appearance. Histopathological examination and DIF are essential to establish the final diagnosis of desquamative gingivitis (Endo *et al.*, 2017; 2018). Histopathological examination showing acantholysis suggests pemphigus vulgaris. On the other hand, supra basilar split detected can be used to differentiate pemphigus vulgaris from other condition with subepithelial blistering such as mucous membrane pemphigoid. Direct immunofluorescent (DIF) microscopy is the gold standard to diagnose pemphigus vulgaris, which will show intercellular binding of IgG and/or C3 within the epithelium (Endo *et al.*, 2017; 2018; Popescu *et al.*, 2019; Schmidt *et al.*, 2019; Shaqman *et al.*, 2020).

The treatment of desquamative gingivitis involves improving oral hygiene, reducing irritation to the lesions and specific therapy to the underlying disease. Pemphigus vulgaris patients limited to oral lesions are usually treated with topical steroid and proper plaque control. Since plaque-induced

gingivitis is seen in almost all desquamative gingivitis, effective therapy should include non-surgical periodontal therapy. Care should be taken as vigorous scaling and debridement may further damage the existing desquamative gingivitis lesions (Endo *et al.*, 2017).

Progression of pemphigus vulgaris to extra-oral involvement may require systemic corticosteroid treatment. However, long term use of corticosteroids may lead to herpes virus and fungal infections (Schmidt *et al.*, 2019), as seen in the present patient who developed candidiasis intra-orally. As recurrence of pemphigus vulgaris is possible, patient should be monitored for a long period of time, or perhaps lifelong.

CONCLUSION

Diagnosing desquamative gingivitis early is important because the prognosis varies. Early detection of pemphigus vulgaris is paramount as it is life-threatening. Clinical appearance, histopathological examination and DIF microscopy are needed to confirm the definitive diagnosis. In our case, delayed diagnosis of pemphigus vulgaris after one year of onset led to progression of extra oral lesion. Dental practitioner should always be aware of the manifestations of desquamative gingivitis. As pemphigus vulgaris has multi systemic nature, a multidisciplinary approach for the management of disease is crucial.

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