Oral Manifestation in Systemic Lupus Erythematosus: A Case Report

Wan NurHazirah WAKamil[1,*], and *Nor Nazaliza Basri*[2]

[1]Faculty of Dentistry Universiti Teknologi MARA Sungai Buloh Campus, 47000 Sungai Buloh, Selangor, Malaysia

[2]Oral Medicine Clinic, Departmenr of Oral and Maxillofacial Surgery, Hospital Kuala Lumpur, 50586 Jalan Pahang, Kuala Lumpur

Abstract. Systemic lupus erythematosus (SLE) is a chronic autoimmune inflammatory disease with diverse clinical manifestations affecting several organs such as the brain, lungs, kidneys, heart, blood vessels, muscles and skin. Oral manifestations might be the first sign and symptoms of SLE. Hence, this paper describes a 58-year old lady whom oral lesions were the first manifestation of SLE disease. The oral manifestations and management of this patient was discussed.

1. Introduction

Systemic lupus erythematosus is chronic autoimmune disease in which the body's own immune system attacks its own tissue causes inflammation that leads to cell death and tissue destruction (1). However, the etiology of this disease remains unknown. The pathogenesis of SLE is thought to develop when a T-lymphocyte to an antigen-presenting cell (APC) is initiated. The T-cell receptor binds to the major histocompatibility complex (MHC) portion of the APC, in which lead to cytokine release, inflammation, and B-cell stimulation (4).

SLE is commonly in women and peak onset of age is between third and fourth decade of life (2, 3). This increased incidence may be attributed to hormones, namely oestrogen, as studies have shown women who used oral contraceptives or hormonal therapies had an increased risk of SLE (5). Oral manifestation of SLE are frequently encountered (6) and may include oral ulceration, honeycomb plaque, raised keratotic plaque, nonspecific erythema, purpura, petechiae and cheilitis (7).

2. Case report

A 59-year-old Malay lady was admitted to Hospital Kuala Lumpur presenting with multiple painful ulcers in the oral cavity and on the lips for one week. It was a sudden onset. She also complained of burning mouth, bleeding gums, and joint pains at hand (wrist), ankle, knee and shoulders. Her medical history included treatment for valvular heart disease at National Heart Institute Malaysia and was previously investigated for rheumatoid arthritis. However, patient was discharged from rheumatology.

At initial presentation, she complaint of multiple painful oral ulceration in the mouth and lips with associated burning mouth, joint pains and causes decrease of oral intake. Extraoral examination revealed, crusted bleeding ulcerated lesion seen on vermillion border of upper and lower lip (Figure 1).

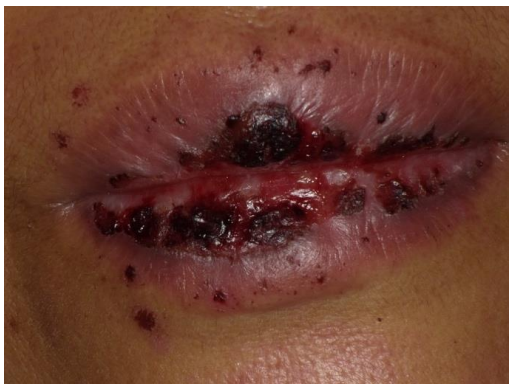

Fig 1. Crusted bleeding and erosive areas on vermillion border of upper and lower lip.

There is no obvious facial swelling, no palpable submandibular lymph node and no skin lesion noted on examination. Intraorally, ulcers are present at both buccal mucosa, tongue and hard palate. The tongue revealed whitish coated with ulcers and white patches with mild erythema. (Figure 2). Acute ulcerations and erythema seen on the hard palate (Figure 3a and 3b).

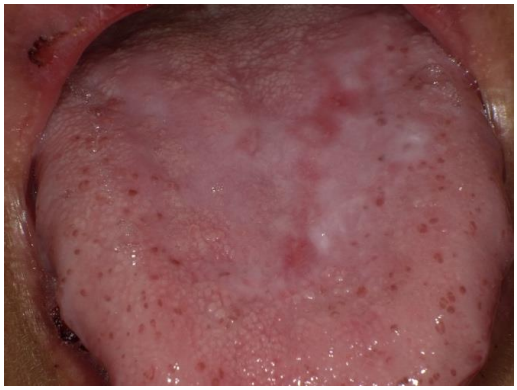

Fig 2. Erythematous area surrounded by keratoticpatch on the dorsum of tongue.

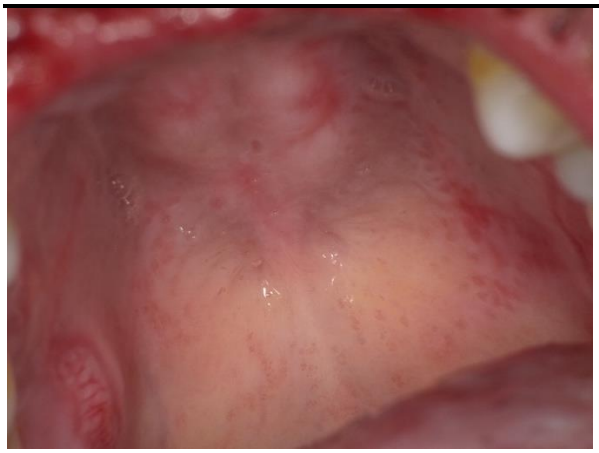

Fig 3a. Erythematous patches seen at hard palate.

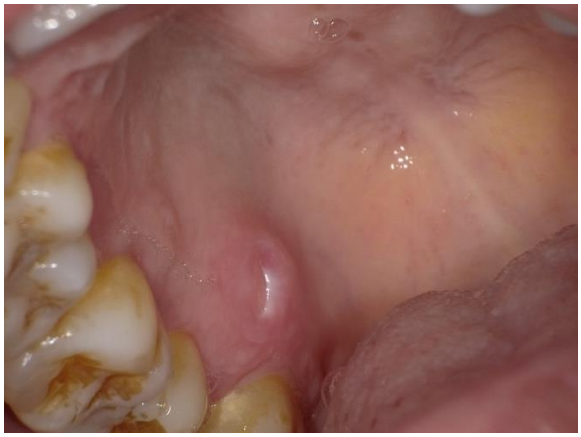

Fig 3b. Well demarcated shallow ulcer with hyperkeratotic border and grey base at hard palate.

A complete full blood count (FBC) investigation revealed, positive for antinuclear antibody (ANA), anti-double standard DNA (anti-ds DNA) and ENA SSA. Meanwhile, C3 showed 1:10, C4 0.28 and rheumatoid factor is within normal ratio. On the basis of clinical and examination and investigations, a diagnosis of multiple ulceration secondary to systemic lupus erythematosus was made. The patient was given IV Hydrocortisone 100mg in ward and followed by steroid mouthwash used at home for a month (Tab Dexamethasone 0.5mg in 10ml H20). Patient was also prescribed with Gengigel mouthwash and gel. Oral hygiene reinforcement and dietary advice was also given to the patient for homecare management and maintaining oral hygiene. She was referred to Rheumatology for the management of SLE. A total resolution of mucosal manifestations was achieved in one month of topical and systemic steroid therapy.

3. Discussion

Systemic lupus erythematosus (SLE) is a serious multisystem condition with variety of cutaneous and oral manifestations. Oral ulceration is reported to be the most common oral manifestation present among SLE patients with a prevalence rate ranging between 7 and 41% observed to be more severe as the disease (9). The commonest site for oral manifestation of SLE are buccal mucosa, palate and vermillion border of lips. In the new Systemic Lupus International Collaborating Clinics Classification Criteria (SLICC) group classification criteria, oral ulcers remain as one the clinical features of SLE (10). Oral ulcers of SLE presented in various types such as palatal erythematous ulcers, aphthous ulcers and lupus cheilitis (11). A palatal erythematous ulcer is described as painless and single lesion at the hard palate. The lupus cheilitis occurs on the buccal lips (especially lower lip) with small or diffuse erythematous and oedematous or crusty painful ulcers (11). Here in our case, patient had multiple ulcers similar presentation of ulcers in SLE.

In this patient, blood investigation showed positive for ANA, anti-ds DNA and ENA SSA. The presence of large number of autoantibodies specific to self-antigens mainly of nuclear origin (double-stranded DNA (dsDNA), Smith antigen and ribonucleoproteins (Sm/RNP), anti-Sjogren's syndrome related antigen A and B (SSA/Ro and SSB/La, respectively) is the hallmark of the disease. The anti-nuclear antibodies (ANAs) are considered markers of diagnosis and prognosis of the disease (12). ANA present in 98% of patient and most sensitive test. Meanwhile anti-double stranded (anti-dsDNA) present in 50-60% of SLE patient and highly specific.

The diagnosis of SLE typically needs four out of 17 criteria (including at least one clinical criteria and one immunologic criteria) or lupus nephritis with presence of antinuclear antibody (ANA) or anti-double stranded (anti-ds DNA) antibodies based on the Systemic Lupus International Collaborating Clinics Classification Criteria (SLICC) group classification criteria (10).

The goals of SLE management are based on prevention, reversal of inflammation, maintaining conditions of remission and alleviation of symptoms (13). Topical corticosteroids (example: 0.1% Triamcinolone oral paste) are one of the most frequently used medications in SLE. The duration of the steroid usage depends on the severity of the symptoms. Local treatment oral ulcers such as using Gengigel gel which contain hyaluronic acid (HA). The main function of Hyaluronic acid includes tissue healing including activation and moderation of the inflammatory response, promotion of cell proliferation, migration, and angiogenesis. A study reported on the efficacy of topical 0.2% HA on recurrent aphthous ulcer which immediately reduced discomfort and also reduced ulcer duration. The occurrence of new ulcers was lower when patients are treated with Gengigel (13). Steroid mouthwash maybe beneficial for multiple oral ulcers in the oral cavity. Other treatment such as chlorhexidine mouthwashes help to prevent periodontal disease and infection.

4. Conclusion

In conclusion, this case report serves to illustrate that importance of early diagnosis of oral lesions to recognize patients with SLE. Most patients have an episodic relapsing and remitting course that may be managed with

high-dose steroids during severe flare- ups. Thus, for patients with SLE, emphasis is focused on the dental team's continuous reinforcement of good oral hygiene and provision of close monitoring

5. Reference

1. Berbert AL, Mantese SA. *Cutaneous erythematosus – clinical and laboratory aspects.* An Bras Dermatol **80**: 119–131 (2005)
2. Albilia JB, Lam DK, Clokie CM, et al. *Systemic lupus erythematosus: a review for dentists. J Can Dent Assoc* **73**: 823–828 (2007)
3. Neville BW, Damm DD, Allen CM, et al. *Oral and Maxillofacial Pathology. Rio de Janeiro: Guanabara Koogan*, (2009)
4. Rahman A, Isenberg D. *Systemic lupus erythematosus. N Engl J Med*. **358**:929–939 (2008)
5. Costenbader KH, Feskanich D, Stampfer MJ, Karlson EW. *Reproductive and menopausal factors and risk of systemic lupus erythematosus in women*. Arthritis Rheum. **56**:1251–1262 (2007)
6. Z. Rutter-Locher, T. O. Smith, I. Giles, and N. Sofat, *"Association between Systemic Lupus Erythematosus and Periodontitis: A Systematic Review and Meta-analysis,"* Frontiers in Immunology **8** (2017)
7. A. C. Chi, B. W. Neville, J. W. Krayer, and W. C. Gonsalves, *"Oral manifestations of systemic disease," American Family Physician* **82**,**11**: 1381–1388 (2010)
8. Jayakumar ND, Jaiganesh R, Padmalatha O, Sheeja V. *Systemic lupus erythematosus.* Indian J Dent Res **17**:19(2):91-93 (2006)
9. G. C. Tsokos, *"Systemic lupus erythematosus,"* The New England Journal of Medicine **365, 22**:2110–2121 (2011)
10. Petri M, Orbai AM, Alarcón GS, et al *Derivation and validation of the Systemic Lupus International Collaborating Clinics classification criteria for systemic lupus erythematosus.* Arthritis Rheum **64**:2677–86 (2012)
11. Nico MM, Vilela MA, Rivitti EA, Lourenco SV. *Oral lesions in lupus erythematosus: correlation with cutaneous lesions*. EJD. **18**(4):376–381 (2008)
12. Tan, E.M. Antinuclear antibodies: *Diagnostic markers for autoimmune diseases and probes for cell biology*. Adv. Immunol. **44**:93–151 (1989)
13. Nolan A, Baillie C, Badminton J, Rudralingham M, Seymour RA. *The efficacy of topical hyaluronic acid in the management of recurrent aphthous ulceration*. J Oral Pathol Med **35**: 461–465 (2006)