Research Article

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A Case Report of Systemic Lupus Erythematosus with Oral Manifestations

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ABSTRACT

Background: The oral cavity is the window to the body and is often the area where systemic disease presents first. Systemic lupus erythematosus (SLE) is a multisystem disease characterized by various cutaneous and oral manifestations. It is an autoimmune disorder affecting various organs with diverse clinical manifestations. Here in the present study, we reported a case of 60-year female patient with systemic lupus erythematosus with oral lesions.

Keywords: Systemic lupus erythematosus (SLE), Autoimmune, Cutaneous, Oral manifestations.

INTRODUCTION

The oral cavity serves as an indicator of overall health as the oral cavity may exhibit manifestations of underlying systemic disease.[1] Systemic lupus erythematosus (SLE) is a chronic autoimmune condition mainly affecting various joints, internal organs and the skin.[2] It is characterized by autoantibodies, immune complexes and immune dysregulation resulting in tissue damage due to direct binding and/or immune complex deposition in tissues. Although the exact etiology is unknown, genetic predisposition may be important factor, along with other risk factors including hormonal factors, environmental factors such as infectious agents, stress, diet and toxins, and physical agents such as sunlight.[3]

It commonly peaks in 4th decade and women are affected nearly 8 to 10 times more frequently than men. Common findings may include fever, weight loss, arthritis, fatigue and generalized malaise.[4] The cutaneous lesions consist of erythematous patches on the face which coalesce to form a roughly symmetrical pattern over the cheeks and across the bridge of nose in a butterfly pattern. Other lesions may appear on neck, upper arms, shoulders and fingers which may present as itching or burning sensations or hyperpigmented areas. Acute erythematous patches may either arise de novo or develop in old chronic lesions, the severity of which is intensified by sunlight.[3]

CASE REPORT

A 60 year old female patient reported to the oral medicine and radiology department with a chief complaint of oral ulceration and burning sensation since 15 days. Before 15 days, she noticed ulceration in right posterior buccal mucosa along with difficulty in eating food and associated burning sensation. Her past medical history revealed that she had skin lesions present on face, back, upper limbs, scalp with alopecia since 2 last years and patient was under medication for the same. She had joint pain for the past 1 year and she has frequent respiratory tract infections. There was no relevant drug history or significant family history. Clinical examination revealed facial cutaneous depigmented erythematous malar rash (Fig.1). On extraoral

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examination, there was no facial asymmetry and no lymphadenopathy. Intraoral examination revealed brown– black hyperpigmented areas present over the left buccal mucosa (Fig.2). An erythematous area of approx. 3 x 2.5 cm^2 size was present over the posterior part of right buccal mucosa, however bleeding was absent (Fig.3). No blisters were found in the oral cavity.

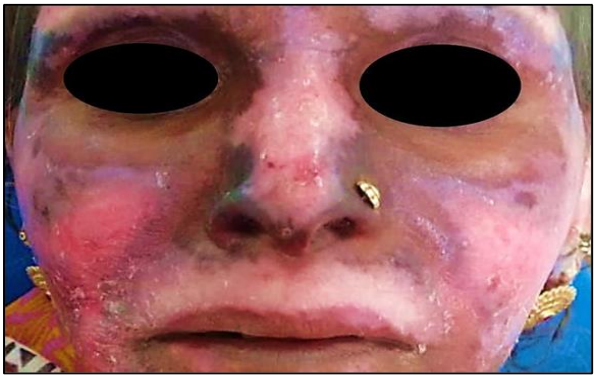

Fig 1: Photograph showing erythematous malar rash present on face.

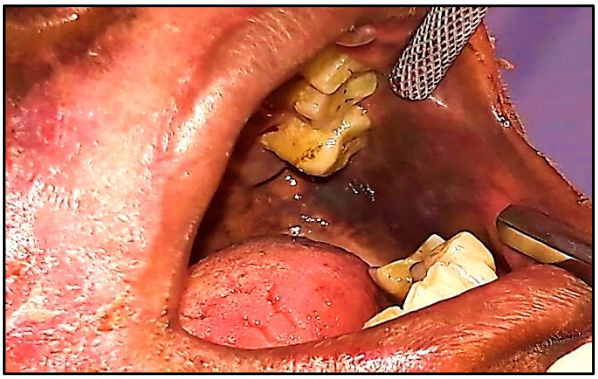

Fig 2: Photograph showing brown-black pigmented areas over the left buccal mucosa.

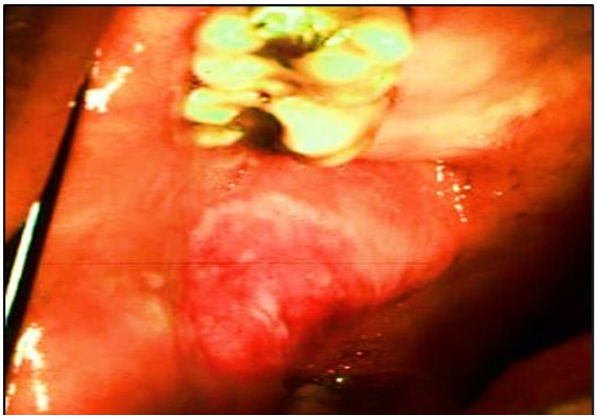

Fig 3: Photograph showing erythematous area over the posterior part of right buccal mucosa.

On laboratory examination, her anti-nuclear antibody (ANA) test was positive. Based on history, clinical and laboratory findings, a provisional diagnosis of systemic lupus erythematosus was made. Differential diagnosis included oral lichen planus. After taking patient's informed consent, incisional biopsy was performed under local anesthesia involving lesional mucosa along with surrounding clinically healthy mucosa. A single soft tissue specimen was obtained which was of approximately 0.4 x 0.8 cm^2 size, creamish brown in colour, firm in consistency with irregular surface (Fig. 4).

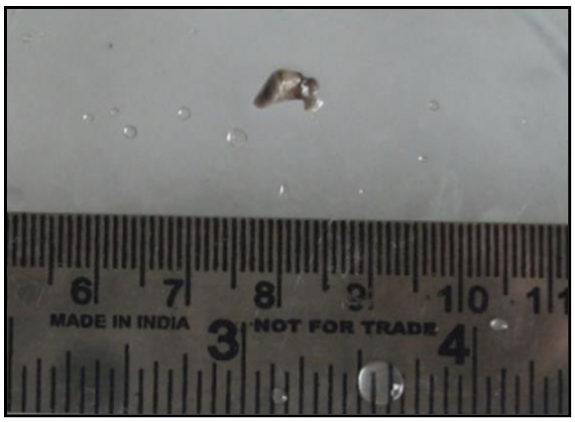

Fig 4: Photograph showing gross appearance of incised specimen.

Histological examination of H&E stained sections revealed hyperparakeratinized stratified squamous epithelium which showed basal cell degeneration in some of the cells and suprabasilar split in one area. The basement membrane was thickened. There was hyalinization of subepithelial connective tissue along with presence of brown pigments. The underlying connective tissue showed moderate inflammation consisting of lymphocytes, plasma cells and few neutrophils which were diffusely scattered. The deeper area showed transverse and longitudinal sections of muscle tissue and adipose tissue (Fig. 5 & 6). All the above findings were consistent with Systemic lupus erythematosus (SLE).

DISCUSSION

Signs of systemic disease are often manifested in the oral cavity before the systemic disease itself is suspected. Some changes seen in the oral cavity are disease specific while others may simply increase the clinician's level of suspicion. A well-planned

pattern for examination of the oral cavity will help ensure that no areas are unexamined. Systemic lupus erythematosus (SLE) presents with oral manifestations in up to 45% of the patients.[5]

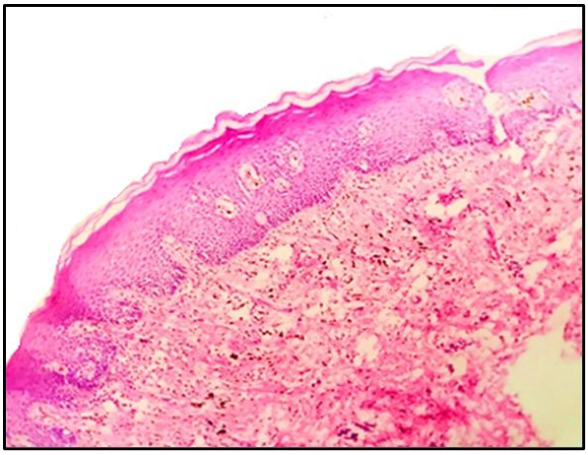

Fig 5: Photomicrograph showing hyperparakeratinized stratified squamous epithelium with suprabasilar split in one area along with the presence of brown pigments in the subepithelial connective tissue (H&E stain, 4X).

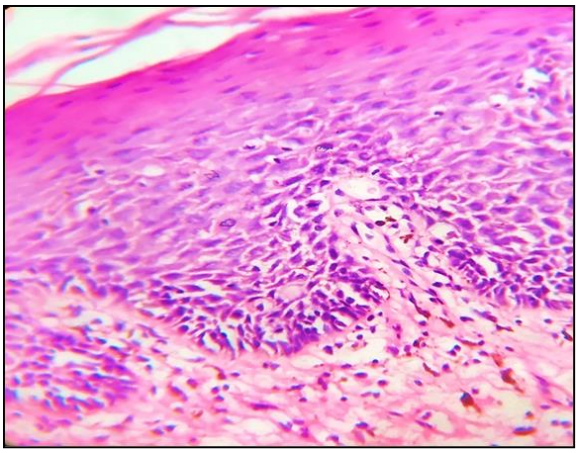

Fig 6: Photomicrograph showing hyperparakeratinized stratified squamous epithelium with hydropic basal cell degeneration in some of the cells. The underlying connective tissue showed moderate inflammation consisting of lymphocytes, plasma cells and few neutrophils which were diffusely scattered (H&E stain, 10X).

Systemic lupus erythematosus (SLE) is a serious cutaneous-systemic disorder which characteristically manifests repeated remissions and exacerbations. Patients develop autoantibodies against DNA, other nuclear antigens, ribosomes, platelets, erythrocytes, leukocytes and other tissue-specific antigens. Thus, the resulting immune complexes result in widespread tissue damage.[3] In the presence of appropriate antigens, SLE develops as a result of the formation of soluble immune complexes that are mainly composed of IgG and IgM. It is a type III hypersensitivity reaction triggered by an endogenous antigen that may be generalized or organ-specific. Because of the affinity of the antibody and the size of the immune complexes, the kidneys, lungs and joints are frequently involved. Tissue damage is caused by platelets and neutrophils. The lesions contain primarily neutrophils and deposits of immune complexes and complement, namely C3a, C4a and C5a. Macrophages infiltrating in later stages are involved in the healing process.[6]

The oral mucosa may be involved either prior to or following the development of skin lesions or even in absence of skin manifestations.[3] Oral counterpart may manifest as oral ulceration, honeycomb plaque, raised keratotic plaque, nonspecific erythema, purpura, petechiae, and cheilitis.[7] Superimposed oral candidiasis as well as xerostomia has also been reported.[3] An unusual presentation of severely crusted cheilitis as a presenting feature of systemic lupus erythematosus has also been reported.[8] Oral ulcers have also been reported in a case of Juvenile-Onset Systemic Lupus Erythematosus.[9] Among SLE patients, a high prevalence of oral lesions including angular cheilitis, ulcers, mucositis and glossitis along with oral complaints such as dysphagia, dysgeusia and glossodynia was found.[10] In a pilot study among SLE patients in Qatar, high rates of gingivitis, periodontal disease, dental caries, and missing teeth was observed.[11] Immunological pathways and predisposing genetic factors common to periodontal disease and rheumatic diseases, including systemic lupus erythematosus have been studied. Mangla et al reported an association between SLE and periodontal disease as desquamative gingivitis. Fabbri *et al*. showed that treatment of periodontal disease among SLE patients on immunosuppressive therapy is beneficial in controlling disease activity.[12] Oral lesions such as desquamative gingivitis, marginal gingivitis or erosive mucosal lesions have been seen in almost 40% of patients. In advanced cases of SLE, patients may have features of Sjogren's syndrome, such as xerostomia, dry eyes and skin.[6]

Various musculoskeletal signs and symptoms such as arthralgia predominate in SLE and is often the earliest manifestation. The joints of the hands are most frequently affected. The arthritis is moderately painful and non-destructive. Generalized sun-induced skin rashes and discoid lesions secondary to epithelial atrophy and subsequent scarring are quite common. Among them, the classic malar or butterfly rash is most common. Hair loss caused by hair follicles plugged with keratin is also reported. Kidneys and heart are most frequently affected organs. Renal disease or lupus nephritis is a fatal complication of SLE affecting 30% of patients. Pericarditis and endocarditis are two predominate cardiac features. Advanced SLE-induced vasculitis can lead to multiorgan dysfunction.[6] Sverzut et al reported a case of a young female with lupus nephritis having oral manifestations.[13]

ANA are antibodies detectable in the blood that have the capability of binding to certain structures within the nuclei of cells. ANA's are directed against several nuclear antigens and can be grouped into four categories, namely antibodies to DNA, antibodies to histones, antibodies to non-histone proteins bound to RNA and antibodies to nucleolar antigens. The most commonly utilized method is indirect immunofluorescence, which detects a variety of nuclear antigens including DNA, RNA and proteins (generic ANA). ANAs detected by this technique are present not only in SLE but also in other auto immune conditions like Hashimoto's thyroiditis, rheumatoid arthritis and Sjogren's syndrome. The immuno-fluorescence test for ANA is sensitive for the detection of SLE, but it is 95% specific for SLE.[14]

For purpose of identifying patients in clinical studies, American Rheumatology Association (ARA) revised criteria (1982) for classification of lupus was used. A person was said to have SLE if any 4 or more of the 11 criteria were present, serially or simultaneously during any interval of observation. In 2012, The Systemic Lupus International Collaborating Clinics (SLICC) criteria for SLE classification has emerged which requires: 1) Fulfillment of at least four criteria, with at least one clinical criterion AND one immunologic criterion OR 2) Lupus nephritis as the sole clinical criterion in the presence of ANA or anti-dsDNA antibodies.[16]

Clinical Criteria		**Immunological Criteria**	
1	Acute cutaneous lupus	1	ANA above laboratory reference range
2	Chronic cutaneous lupus	2	Anti-dsDNA above laboratory reference range, except ELISA: twice above laboratory
3	Oral ulcers: palate	3	Anti-Sm
4	Nonscarring alopecia (diffuse thinning or hair fragility with visible broken hairs)	4	Antiphospholipid antibody: any of the following • lupus anticoagulant • false-positive RPR • medium or high titer anticardiolipin (IgA, IgG or IgM) • anti-β2 glycoprotein I (IgA, IgG or IgM)
5	Serositis	5	Low complement
6	Synovitis involving 2 or more joints, characterized by swelling or effusion OR tenderness in 2 or more joints and 30 minutes or more of morning stiffness.	6	Direct Coombs test in the absence of hemolytic anemia
7	Renal		
8	Neurologic		
9	Hemolytic anemia		
10	Leukopenia (< 4000/mm3 at least once)		
11	Thrombocytopenia (<100,000/mm3) at least once		

In the present case, the medical history of patient revealed the presence of skin lesions, joint pains and frequent respiratory tract infections. Along with medical history, the clinical appearance of oral lesions, histopathology and special investigations especially ANA, contributed to the diagnosis of systemic lupus erythematosus.

Although a potentially debilitating disease, survival rates for SLE have doubled since the 1950s. Death due to SLE was more than 5 times higher in women than in men and more than 3 times higher in black than white women. The major cause of death in the first few years of the onset of SLE is active disease that involves multiorgan systems or infection from drug-induced or lupus-related immunosuppression. Delayed deaths are caused by SLE or associated with end-stage renal disease or complications of treatment, including infection, coronary disease and malignancy. Poor prognostic factors are being young at onset, being male, having a poor

socioeconomic status and having positive titres of antiphospholipid antibodies.[6]

The treatment of SLE is based on preventive measures such as reducing inflammation, preventing organ damage and relief of symptoms. The most common therapies are immunosuppression, cytotoxic treatment and immunoglobulin therapy. Recently, new immunotherapy strategies that act on specific molecules or immune cells have emerged, resulting in lower toxicity and higher selectivity.[15] Patient is advised to avoid excessive sunlight exposure because ultraviolet light may precipitate disease activity. For oral lesions, topical corticosteroids are effective and along with that systemic corticosteroids are prescribed.[14]

CONFLICTS OF INTEREST

The authors declare they have no potential conflict of interests regarding this article.

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