

Oral Manifestations of Systemic Lupus Erythematosus: A Case Report

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Abstract- Systemic lupus erythematosus (SLE) is a multifactorial autoimmune disease. The hallmark feature of SLE is chronic inflammation that affects the skin, joints, kidneys, lungs, nervous system, serous membranes such as the pleura and pericardium, mucous membranes and other organs of the body. Oral lesions of lupus erythematosus show an array of clinical aspects and most affected sites are lips and buccal mucosa [1, 2]. This case report presents a clinical profile of a patient with SLE along with different oral lesions. This article highlights methods to recognize and manage the oral manifestations of this systemic disease.

Index Terms- Systemic lupus erythematosus, SLE, Autoantibodies

I. INTRODUCTION

Systemic lupus erythematosus (SLE) is the prototype of systemic autoimmune diseases. The etiology is not known as yet and the pathogenesis is complex, involving immunological, genetic, hormonal and environmental factors. Damage to tissues and cells results from pathogenic autoantibodies and immune complexes. Classically, Lupus Erythematosus has been subdivided into a systemic and a cutaneous form where in, Systemic lupus erythematosus (SLE) is a multiorgan disease with variable prognosis and Cutaneous lupus erythematosus (CLE) is a more benign condition limited to skin and/or mucosal surfaces. This case report highlights oral lesions seen in an SLE patient along with clinical presentation and management of oral lesion with general therapeutic approach.

II. CASE REPORT

A 36-year-old female patient with the complaint of burning sensation in the left inner cheek which was intermittent since 1 year came to this dental centre.

On eliciting her medical history it was known that she is a known case of diabetes mellitus, hypertension, hypothyroidism and arthralgia. She had no family history of similar illness. She also complained of photo-sensitivity for the past 1 year. She had diffuse hair loss for past 4 months. She complained of severe pain in both knee joints which was more in the morning and reduced as the day progressed. She had visited a local dentist for routine examination and undergone restorations during her pregnancy.

Extra oral examination revealed butterfly shaped malar rash [Fig: 1A] and multiple erythematous patches in face and

extremities. Dry, scaling and itchy lesions were seen in bilateral ear lobes. Occular examination, chest X-ray and cardiovascular system were found to be normal by respective specialist consultants on referral. During the period of investigation patient complained of episodes of memory loss for which she was referred to neurologist where she was advised to write down whatever she practices or about to practice.

Intra oral examination revealed multiple ulcers on the buccal mucosa. Multiple erythematous lesions were found only in the palate, buccal and vestibular mucosa [Fig: 1B, 1C, 1D & 1E]. Left buccal mucosa showed erythematous lesion with white irregular radiating lines in region of mandibular and maxillary third molar. The white lines were non-scrapable. She also added that these lesions appeared after child birth.

Laboratory investigations showed Haemoglobin level 10mg%, ESR level 45mm fall in 1st hour by Westergren's method, platelet count was 365000 cells per cubic mm, serology for antinuclear antibody (ANA) was positive with a homogenous pattern. Double-stranded deoxyribonucleic acid antibody (Anti-ds DNA) was raised to 37 IU/ml (normal 0-6). C3 complement fraction was lower than normal range of 0.738-1.80 GL. Her C-reactive protein and Rheumatoid Factor were negative. MRI scan showed normal brain study and diffuse disk bulge at C5-6 level with minimal impingement on bilateral exiting nerve roots. Her Mantoux test was 7mm and TSH was 4.00 which were within the normal range.

Based on the typical skin lesion, intra oral findings, systemic examinations, special investigations especially ANA test positivity, raised Anti-ds DNA, depleting C3 complement fraction contributed to the diagnosis of this case as a case of Systemic Lupus Erythematosus. Presently the patient is on, Triamcinolone with Orabase topical steroid along with Prednisolone (5mg) tablet twice daily.

III. DISCUSSION

Patients with Systemic Lupus Erythematosus (SLE), a prototypic autoantibody disease, develop auto antibodies to nuclear molecules in many of their cells, cell components and tissues. Clinical manifestations appear to result from deposition of antigen antibody complex in the tissues. Cytokines are thought to play a key role in SLE; however, the extent to which they affect progression of lupus is not clear.

Another theory might be that SLE autoantibodies are a sequelae of cross reactions to exogenous antigens e.g. RNA

retroviruses. Extrinsic factors such as exposure to sunshine, infections and drugs may trigger SLE reactions in some patients. No matter the etiology, genetic predisposition is a probability, as expressed in associations with specific HLA/MHC antigenic profiles. Dysfunction of B-lymphocytes is one of the main defects in SLE. In addition, suppressor T-lymphocytes are also reduced in number, permitting a considerable increase in autoantibodies [3, 4].

SLE is rare in India. A prevalence study in India (carried out in a rural population near Delhi) found a point prevalence of 3 per 100,000 [6]. This disease commonly affects young women of child bearing age with a female : male ratio of 10:1 (90% of cases are in women), more predominantly in young women due to higher levels of estrogen, while at premenstrual and post-menopausal women, ratio with males decreases to 3:1 [1,2].

Cutaneous manifestations are seen in the form of erythematous patches on the face that coalesce to form a symmetrical pattern over the cheeks across the bridge of the nose in a butterfly pattern of distribution which is known as malar rash [Fig: 1A]. Skin around the neck, arms, shoulders and fingers are also affected. They may also be associated with itching or burning sensation, and areas of hyper pigmentation. Macules or papules occur on the face, or a generalized rash occurring on the body, which may or may not be sun induced.

Diffuse alopecia can generally occur when the disease is active and is usually reversible during remission. Patchy alopecia, on the other hand, may lead to scarring and can become permanent [5, 6]. Cardiovascular and respiratory symptoms are also common and include chest pain on inspiration due to pleurisy or pericarditis. Renal complications (glomerulonephritis and microvascular thrombosis) and neuropsychiatric complications (seizures, psychosis, neuropathies, stroke, and depression) are common as well [7]. Ophthalmic and gastrointestinal manifestations are usually uncommon but can be serious, including kerato-conjunctivitis sicca, pancreatitis, hepatitis, and subacute bowel obstruction. Pain in the joint and arthritis are common manifestations [5]. Present case showed similar cutaneous involvement and joint pains in the form of arthralgia.

In 20-50% of patients' oral lesion of SLE are seen. These manifestations of the oral cavity may be seen either prior to or following the development of skin lesions or even in the absence of skin manifestations [8]. In a decreasing order, locations more frequently affected are buccal mucosa, hard palate and lower lips in the form of ulcers, erythema or hyperkeratosis. Oral lesions appear as erythematous areas, without induration and with white spots. The margins of these lesions are not sharply demarcated but quite often show the formation of narrow zone of keratinization. Hyperemia with edema is a common occurrence and there may be a tendency for bleeding [8]. The main clinical differential diagnoses are lichen planus, leukoplakia, squamous cell carcinoma and even vesico-bullous diseases [9].

Although investigation plan for a case of SLE will depend on the clinical picture, the minimum laboratory workup should include [10]:

- Haemoglobin, WBC, Differential count, ESR
- Urine routine (preferably a fresh sample examined) and microscopy, and 24 hour protein and creatinine estimation if necessary
- Serum chemistry (urea, creatinine, liver function tests, lipid profile)
- Chest x-ray
- ANA, anti-dsDNA, C3, C4

Histological appearance of SLE is not pathognomic but is suggestive of the disease. Histopathological features include, changes at the dermo-epidermal junction that include thickening of the basement membrane (best demonstrated by periodic acid-Schiff staining) and vacuolar degeneration of the basal cells along with perivascular and peri-appendageal inflammatory cell infiltration of a variable degree in the reticular dermis. Hyperkeratosis is more evident and follicular plugging may be seen in more mature lesions [11].

Direct immunofluorescence testing is often used to detect the presence of IgG, IgM and IgA at the basement membrane. The antinuclear antibodies (ANA) test is highly specific with a positive result in >95% of SLE patients. The anti-dsDNA antibody test is positive in 60% of SLE patients and is considered the best marker for disease activity, with a specificity of almost 100%, except in elderly patients who have a lower prevalence of anti-dsDNA. Complement levels (C3 and C4) are negatively correlated with lupus activity and their levels deplete because of consumption [10].

If a patient fulfills more than 4 of 11 criteria made by American College of Rheumatology, then the diagnosis of SLE can be made with about 95% specificity and 85% sensitivity. Diagnosis of SLE is based on clinical judgment. SLE can be suspected whenever 2 or more organ systems listed in Table-1 are involved [10, 12].

There is no known cure for Systemic Lupus Erythematosus, hence treatment is based on relieving symptoms, suppressing inflammation, and preventing future pathology. Symptomatic treatment for the specific involved organ as per the severity of the disease: topical sunscreens, avoidance of UV light and estrogens, NSAID's for arthritis, antimalarials for dermatologic manifestations, topical steroids for rash, use of systemic steroids for prevention of end organ damage, Calcium and Vitamin D supplements to fight osteoporosis and corticosteroids as immunosuppressant drugs for serious organ involvement (e.g. Cerebritis, nephritis). All medications used to treat SLE require monitoring periodically for potential toxicities [13, 14].

Preventive dental hygiene care in Lupus patients is very important. Chlorhexidine mouthwashes helps contain periodontal disease by chemical plaque control. Mucous membrane ulcers can be managed with hydrogen peroxide gargle or steroid impregnated gel. Intralesional injections of corticosteroids are also effective modality. Bacterial, viral and fungal infection should be treated using conventional, proven

therapy specific for the infection. No Dental procedures should be undertaken on patients with active Lupus, and if necessary, antibiotic premedication is always advisable due to high incidence of bacterial endocarditis.

Adverse effects of Lupus therapy in oral cavity [20]

Long term use of medications to control Lupus can induce significant intra-oral pathology such as mentioned below:

- Corticosteroids: leads to root canal calcification, delay of tooth eruptions and root dilacerations.
- Steroids: causes necrotizing ulcerative gingivitis.
- NSAID's: induces gingival bleeding, but due to the inherent property of NSAID's to inhibit alveolar bone resorption, periodontal health in some patients with Lupus has been found to improve.
- Cyclosporine: causes gingival enlargement (hyperplasia).
- Immunosuppressive treatment: fights against intra-oral infections but promotes Candidiasis and Herpes Simplex Virus opportunistic infections.

IV. CONCLUSION

Systemic lupus erythematosus is an autoimmune disorder that can affect several organ systems, including the skin, kidneys, and CNS. Of patients with SLE, 25% have associated oral lesions, which are usually superficial ulcers with surrounding erythema. It is essential that dental practitioners should know these pathologies and diagnose them at an early stage of the disease to help assist in providing more effective treatment to improve survival rates. The intraoral examination should be incorporated as a part of dermatologic examination as the oral manifestations can represent preliminary signs or can coexist with the disease [15].

APPENDICES

Appendix 1: Table 1.

Appendix 2: Fig 1.

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Appendix 1

Table 1: Diagnostic criterion for systemic lupus erythematosus include any 4 of the 11 criteria given below simultaneously or in succession [14]

SLE criterion	Definition or examples
Malar (butterfly) rash	Fixed erythema over the malar eminences
Discoid rash	Erythematous raised patches, may scar
Photosensitivity	Skin rash as a result of unusual reaction to sunlight
Oral ulcers	Often painless sores
Arthritis	Non-erosive: Jaccoud's arthropathy
Serositis	Pleuritis — pleuritic pain, pleural rub, pleural effusion Pericarditis — ECG changes, pericardial rub, pericardial effusion
Renal disorder	Proteinuria (with 3+ or more protein noted in urinalysis specimen or 0.5 g of protein/day) Cellular casts in urine
Neurological disorder	Seizures Psychosis
Hematological disorder	Hemolytic anemia Leukopenia Lymphopenia Thrombocytopenia
Immunological disorder	Anti-DNA antibodies Anti-Sm antibodies Antiphospholipid antibodies
Antinuclear antibody	Antibodies to nuclear constituents

Appendix 2



Figure. 1. Clinical aspects of lupus erythematosus (A) Rash on bridge of nose (Malar Rash), forehead (B) Lesion on buccal mucosa and gingival (C) Erythematous lesion with irregular white lines on buccal mucosa (Left) (D) Erythematopurpuric lesion on hard palate (E) Erythemato-keratotic lesion on buccal mucosa (Right).