Latent Systemic Lupus Erythematosus: A Rare Case Report

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Abstract

Systemic lupus erythematosus (SLE) is a chronic, autoimmune, connective tissue disorder which has multi-organ involvement. In many cases oral manifestations might be first one to appear and these manifestations might bring patient to oral physicians for treatment. Responsibility lies on Oral Physician to identify such oral manifestations as a part of systemic manifestation and initiate appropriate treatment required for their oral symptoms. Management of this disease should be individualized and should include both pharmacological and non-pharmacological modalities for symptom relief and resolution for improved quality of life. Various treatments like use of occlusal splint have been implicated for relief of oral symptoms and have been found to be very effective.

Keywords: Autoimmune disease, Corticosteroids, Oral manifestations, Occlusal splints, Systemic Lupus Erythematosus

INTRODUCTION

Systemic Lupus Erythematosus is a clinically heterogeneous autoimmune disease[1] of complex aetiology, with protein manifestations, having a variable course and prognosis. It effects primarily women in their childbearing years (20-40 years).[2] Prevalence of SLE is 1:1000.[3] Etiologic mechanism of SLE remains unknown, but multiple factors like genetic, hormonal, immunological and environmental have been found to play a role in its development. Presentation of SLE is complex, as disease has multi-organ involvement (Table 1). Patients experience flare-ups to varying degrees as well as periods of disease recurrence. Certain clinical features are common in every patient but with a unique set of identifiers. A Minimum of four out of eleven criteria should be met in order to confirm the diagnosis of SLE patient (Table 2). A rheumatologist or nephrologist may diagnose a patient if the patient meets only three of the criteria (1 must be clinical and 1 must be serologic) and has other clinical manifestations such as alopecia, skin vasculitis, Reynaud's phenomenon, or lung fibrosis.[4] Some patients may have only one organ involved or only have some of the manifestations and will, therefore, not be diagnosed under the ACR criteria. Such patients are classified as having "Incomplete" or "Latent" lupus.[5] The disease activity usually occurs in three phases i.e flare, chronic, and long quiescence. A flare or relapsing remission is an exacerbation that occurs suddenly and unpredictably; patients are usually in good health between flares. Chronic SLE has persistent activity of some type such as chronic synovitis and chronic cytopenias.Patients with long quiescence have a long remission period before having additional flare –up.[6,7] As the prevalence of SLE is low, most of oral physicians have insufficient experience for diagnosis and management of "Latent SLE".[8] The authors present a case report of a female patient suffering from systemic lupus erythematosus with associated oral manifestations treated with mild corticosteroids and occlusal splint to reduce flare-up with regular follow up visits and subsequent symptomatic relief and focuses on the identification and management of such patients.

CASE REPORT

A 45 year old female reported to the Oral Medicine Department, Jaipur Dental College with chief complaint of generalized redness of gums with associated pain, burning sensation, difficulty in chewing food and drinking water since 8 months.Medical history revealed presence of extra-oral eruptions (Figure 1a-c) on forehead, vermilion border of lips and upper extremities peripheral joints which

Table 1: Clinical features of systemic lupus erythematosus

System	Features
Constitutional	Fatigue
	Fever (in absence of infection)
	Weight loss
Musculoskeletal	Arthritis, arthralgia
	Myositis
Skin	Butterfly Rash
	Photosensitivity
	Mucous membrane lesion
	Alopecia
	Raynaud's phenomenon
	Purpura, Urticaria
	Vasculitis
Renal	Hematuria, Proteinuria
	Casts
	Nephrotic syndrome
Gastrointestinal	Nausea, vomiting
	Abdominal pain
Pulmonary	Pleurisy
	Pulmonary parenchyma
	Pulmonary hypertension
Cardiac	Pericarditis, Endocarditis
	Myocarditis
Reticuloendothelial	Lymphadenopathy
	Splenomegaly
	Hepatomegaly
Hematologic	Anemia
	Thrombocytopenia
	Leukopenia
Neuropsychiatric	Psychosis, Seizures
	Organic brain syndrome
	Transverse myelitis
	Cranial neuropathies
	Peripheral neuropathies

(Guidelines for referral and management of SLE in adults. Arthritis Rheumatol. 1999 September; 42(9):1785-1796)

Table 2: Diagnostic criteria of SLE. Adapted from Tan et al, 1982 [1]. A person is said to have SLE if he/she meets any 4 of these 11 criteria simultaneously or in succession

Criterion	Definition/examples
1. Malar rash	Fixed erythema over the malar eminences, tending to spare the nasolabial folds
2. Discoid rash	Erythematosus raised patches, may scar
3. Photosensitivity	Skin rash as a result of unusual reaction to sunlight
4. Oral ulcers	Usually painless
5. Arthritis	Non-erosive: Jaccoud's arthropathy
6. Serositis	a) Pleuritis-pleuritic pain, pleural rub, pleural effusion b) Pericarditis-ECG changes, rub, pericardial effusion
7. Renal disorder	a) Proteinuria (> 3+ or 0.5 g/day) b) Cellular casts in urine
8. Neurological disorder	a) Seizures b) Psychosis
9. Haematological disorder	a) Haemolytic anaemia b) Leukopaenia c) Lymphopaenia d) Thrombocytopaenia
10. Immunological disorder	a) Anti-DNA antibodies b) Anti-Sm antibodies c) Anti-phospholipid antibodies
11. Anti-nuclear antibody	Exclude drug causes

(Manson and Rahman. Orphanet journal of rare diseases. 2006 1:6 doi: 10.1186/1750-1172-1-6)

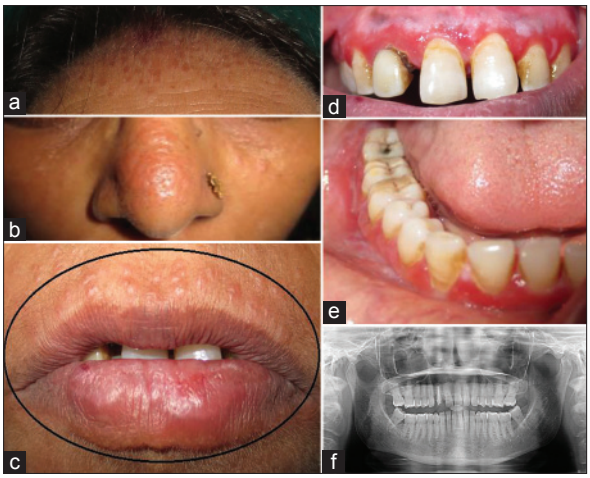

Figure 1:Extraoral and intraoral manifestations (I visit). (a) Pigmented papules on forehead. (b) Butterfly rash. (c) Vesicles involving perioral area and vermilion border. (d) Desquamative gingivitis. (e) Multiple crops of oral ulceration. (f) Orthopantomograph

aggravated on exposure to sun rays with previous history of pain and inflammation in upper and lower peripheral joints and constitutional symptoms of fatigue, headache, fever, malaise and unsatisfying sleep since last 8 months. Patient also complained of reduced vision and dryness of eyes since last 1 year. Medical consultation sought by the patient from a physician 8 months back revealed a suspected case of "systemic lupus erythematosus" (SLE). Drug History revealed consumption of Tab.Bioquin 200 mg BD *15 days, Digene gel TDS * 15 days, Derantox capsules OD *10 days, Tacrorotor 0.1% ointment to be applied topically on affected area, Melagard 50+ TDS * 15 days. Patient discontinued medication four months back as there was only mild relief. In family history, there was no record of presence of any such signs, symptoms or disease in any of patient's distant family relatives/cousins. Patient had no deleterious habit and kept insignificant oral hygiene. In general examination, the height and the weight of the patient was within the normal range for a female of given age. In Extra oral examination, facial profile was symmetrical, TMJ was clinically normal and lymph nodes were non-tender and non-palpable. Mild alopecia was present. Chronic Cutaneous manifestations involved presence of "butterfly rash-mask"[9]- (Figure 1a-c) shaped flat erythema over the malar eminences, bridge of nose and upper vermilion border of lips typically sparing the nasolabial folds. Eruptions aggravated on exposure to sun and hence a generalized photosensitivity was present along with cracking of lower lips. Intra oral examination revealed generalized inflammation and erythema of upper and lower attached gingiva showing typical characteristic features of desquamative gingivitis(Figure 1d).There were localised regions of atrophied mucosa with erythema and multiple crops of painful oral ulceration (Figure 1e) interspersed with small patches of white keratotic regions present in lower

left vestibule, right buccal vestibule and posterior region of right buccal mucosae. The oral lesions were tender, burning sensation was measured on VAS score of 3 with incidence of unpredictable irregular flare up pattern. Bleeding from mucosa was noted on wiping with gauze. Examination of hard tissue revealed dental caries wrt 36, 37, 38, 46, 47, 48 spacing between upper anteriors and midline diastema.

With the patient's consent, Investigations were done which included a histopathological smear of the offending regions intraorally, revealing presence of sub-epithelial and perivascular infiltrates and disturbed keritinaztion cellular atypia. Haematological investigations revealed Glomerular filtration rate (GFR-28), TLC count 9500/mm^3(normal 4000-11000/mm^3), haemoglobin 9.0 gm % (normal 12-15.5 %) indicating anaemia,haematocrit 33%(normal 45 %), ESR 15 mm 1st hr (normal westgreen 0-20 mm), platelets 2,10,000/mm^3(normal 150,000-400,000/ul), total RBC 3.6 ml/mm^3(normal 3.8-5.8 * 10^{12}/l), DLC count was done and all the leucocytes were found to be within normal range, Anti Nuclear Antibody (ANA) test was negative,C-reactive protein was found to be negative and rheumatoid factor was Positive.Radiographic examination included an Orthopantomograph (OPG) which revealed a completely normal maxillary, mandibular jaw bone and a normal permanent dentition (Figure 1f).

Examining the medical history and analysing Constitutional, Musculoskeletal, Cutaneous and Haematological manifestations of patient, but a negative ANA, 4 of 11 criteria were documented specific for diagnosis of SLE. Thus, it was concluded to confer a confirmed diagnosis of "Latent or Passive Systemic Lupus Erythematosus" under the *1999 Revised American College of Rheumatology (ACR) criteria.*

Patient consent was seeked prior to initiation of treatment. Treatment plan included providing supportive and symptomatic care. Patient was educated, counseled and reassured about the relapsing, remission pattern and unpredictable course of her disease. Major part of treatment involved administration of topical and mild oral corticosteroids. Local anesthetic gel was applied in offending regions intraorally (2% lignocaine) to provide on the spot symptomatic relief to the patient from burning sensation, also patient was instructed for topical application of triamcinolone acetonide 0.1% BD * 7 days loaded on occlusal splint fabricated as a part of treatment and worn for 2 hrs 3 times daily for 7 days. Given the significant side effects of steroids, Oral Prednisolone was started at a mild dose of 30mg OD * 7 Days and gradually tapered to 20 mg and 10mg subsequently in 1st and 2nd follow up. Protection from sun by wearing of protective clothing and application of sunscreen protection factor of 15 was emphasized and dietary counseling was also done. Oral prophylaxis and oral hygiene was recommended. Periodic recall was done after 7 days.

1st follow up was after 7 days. On enquiring Patient reported reduction in burning sensation measured on VAS score of 1 and ease in chewing and drinking habits. Patient had complete symptomatic relief from pain. Extra oral examination revealed reduction in number of eruptions in the vermilion regions of lips, malar regions and forehead (Figure 2a, b). Intraorally erythema was reduced both in the upper and lower attached gingivae (Figure 2c). Consumption of prednisolone was tapered to a dose of 20 mg OD* 7 days, topical application of triamcinolone acetonide was continued. Patient was asked to follow previous instructions. Second Periodic Recall was done again after 7 days.

In 2nd follow up, patient reported complete elimination of burning sensation measured on VAS score of 0. Joint pain in upper and lower extremities still persisted but in a milder form. Extra orally patient reported a complete cessation of eruptions on forehead, malar region and upper vermilion borders of lips (Figure 3a, b). Intraorally there was marked reduction in erythema both in upper and lower attached gingivae (Figure 3c). No side-effect or toxicity from steroid dose was reported. Prednisolone was tapered to 10 mg OD * 7 days. As the prognosis was good, all previous instructions were re-advised and patient was kept on a regular follow up and monitoring.

DISSCUSION AND MANAGEMENT OF LATENT SLE

SLE is a systemic illness with multiple end-organ involvement. In addition to the persistent risk of disease

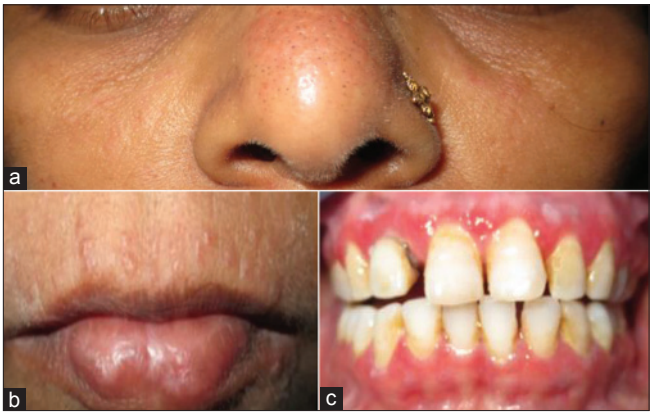

Figure 2: Periodic Recall (2nd visit) Cessation of burning sensation & pain wrt lesion intraorally and reduction in number of extra oral & intraoral manifestations. (a) Extraoral eruptions –malar region. (b) Perioral area and vermilion border of lips. (c) reduction in intraoral erythema

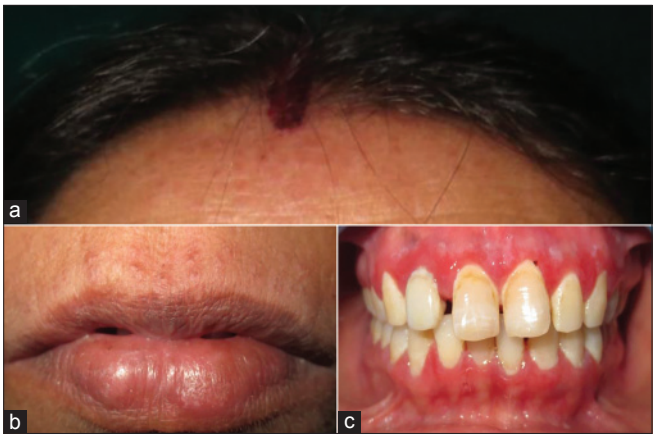

Figure 3: Periodic Recall (3rd visit) Marked reduction in erythema of attached gingivae and reduction in number of extra oral eruptions. (a) Forehead region (b) Perioral region (c) Intraoral region

flares more than one-half of SLE patients develop permanent organ system damage.[10] As such it challenges both the patient and their families. The general principle in management of SLE is analogous to that of other inflammatory disorders: suppression of inflammation in an attempt to prevent organ damage.[11] Patients with newly diagnosed SLE often has anxieties about a possibly fatal chronic illness with unpredictable flares and potential disability; these anxieties should be addressed. Patient should learn how to cope with and monitor their disease and to assist physician in distinguishing coincident unrelated symptoms from signs and symptoms of a flare. Psychological support by either the physician and appropriate health professional is essential.[12] SLE patients may need the expertise of professionals in the fields of social work, vocational counselling, psychology, physical and occupational therapy, ophthalmology, dermatology, nephrology, cardiology, orthopaedic surgery and oral physicians too play an important role in diagnosis and treatment of SLE. Not all of these are needed at any one time and their coordination is best done by a specialist, usually a rheumatologist, who has experience in following up patients with SLE and knows what value is added from these consultants. The four major tasks of the oral physician in diagnosis and management of "Latent or Mild SLE" are to 1) be alert to the possibility of SLE in their patients and to make diagnosis as early as possible 2) to manage and monitor patients with mild SLE 3) to recognize when referral to rheumatologist is indicated 4) collaboration with specialist in monitoring disease activity and treatment in latent SLE. In Latent SLE disease, routine health assessment including regular gynaecologic assessments, dental care and ophthalmologic examinations for females (in patients consuming glucocorticoid) is very important. With the experience and significant knowledge oral physicians in collaborating with specialist can play a key part in monitoring and management of this disease.

CONCLUSION

SLE is a very complex disease with multifactorial aetiology and multiple organ involvement making the diagnosis challenging.Clinical manifestations as well as immunological abnormalities assist in the diagnosis. Management of SLE depends on the level of disease activity and can include general measures, NSAIDs and steroids. Literature reveals an ongoing research to improve the quality of life and increase survival of patients affected by SLE.

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