DERMATOLOGICAL MANIFESTATIONS-ARE THEY SPECIFIC MARKERS FOR SYSTEMIC INVOLVEMENT IN SLE?

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ABSTRACT: BACKGROUND: SLE is a multisystem autoimmune disease with protean manifestations; emphasizing upon the need for a thorough cutaneous and systemic workup. Certain cutaneous features act as forerunners of systemic involvement.
AIM: To analyse the association between skin manifestations and systemic involvement and disease activity.
METHODS AND MATERIALS: This is an institution based retrospective study conducted over a period of two years with a sample of 30 patients (2 males and 28 females; mean age 22.83 ± 8.53 years) with Cutaneous lupus erythematoses who satisfied atleast 4 out of 11 revised 1982 ARA criteria of SLE in a tertiary care centre. A thorough clinical, investigative and mortality data has been collected from the records. The diagnosis has been confirmed using histopathological and immunological analysis in all the patients. The disease activity and severity have been determined by using SLEDAI-2K score. The statistical evaluation has been carried out by determining the frequency of occurrence of systemic involvement in relation to cutaneous manifestations and p value by Fischer's exact test of significance has been calculated.
RESULTS: Cutaneous manifestations like malar rash, photosensitivity, noncicatricial alopecia, oral ulcers and vasculitis are associated with systemic involvement.
CONCLUSIONS: Cutaneous manifestations in SLE can be considered as markers of systemic involvement and indicate a prima facie suspicion which can guide in instituting appropriate management.

KEYWORDS: CLE-Cutaneous Lupus Erythematoses, SLE-Systemic Lupus Erythematoses, SLEDAI-2K- SLE Disease Activity Index 2K, SD-Standard Deviation, SE- Standard Error.

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INTRODUCTION: Lupus erythematoses (LE) is a multisystem autoimmune disease presenting with cutaneous manifestations and severe organ involvement. Skin being the second commonest organ to be affected after the joints in LE, presents with diverse manifestations- those specific to the disease and those which are nonspecific according to Gilliam's classification.The cutaneous features of LE may manifest as a spectrum with simple maculopapular rash and photosensitivity at one end and disfiguring and disabling lesions like lupus profundus, alopecia, scarring lesions, TEN like lesions and cutaneous vasculitis on the other end[1]. The current concepts on Lupus indicate that LE specific skin manifestations may be considered as important diagnostic clues and LE nonspecific lesions may point towards more activity of the disease measured by SLEDAI 2K score. The above manifestations prompt us to institute appropriate disease monitoring and intensive therapy.

The current study was undertaken to analyse the association between cutaneous manifestations of LE which account for 4 out of 11 Revised American Rheumatism Association criteria (ARA) and systemic involvement; assuming that skin manifestations can be considered as the forerunners and markers of the forthcoming systemic complications.

MATERIALS AND METHODS: This is an institution (tertiary care centre) based retrospective study conducted over a period of two years with a sample of 30 patients suffering with Cutaneous Lupus Erythematoses (2 males and 28 females of mean age 22.83± 8.53yrs) who satisfied atleast 4 out of 11 revised 1982 ARA criteria of SLE. A thorough clinical, investigative and mortality data was collected from the records of the patients. The diagnosis of SLE was confirmed by histopathological examination of skin and immunological analysis in all the sample patients at the time of admission.
The disease activity and severity was assessed using SLEDAI 2K scoring at the time of admission [Table 1].In the glossary of the original SLEDAI, certain descriptors like rash, alopecia and mucosal ulcers had been scored as active only if they were new or recurrent, and in the case of proteinuria, if new onset or a recent increase of more than 0.5 grams in 24 hours is present. SLEDAI- 2 K was introduced in 2002 and validated with an aim to lead to an apparent improvement; that did not occur. SLEDAI-2 K

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was subsequently modified to allow the documentation of ongoing disease activity in the descriptors- to include the presence of any inflammatory rash, alopecia or mucosal ulcers, and new, recurrent or persistent proteinuria greater than 0.5 grams in 24 hours. As in the original SLEDAI, all the descriptors inSLEDAI–2 K have been attributed to lupus activity.[2] Hence SLEDAI 2K has been preferred in the current study instead of SLEDAI as this study is on "Dermatological Manifestations of SLE" [Table 1]. The statistical evaluation was carried out by determining the frequency of occurrence of systemic involvement in correspondence to cutaneous manifestations and by computing p value using Fischer's exact test of significance of statistical association.

RESULTS: Of the 30 sample patients with CLE, 28 (93.3%) were females and two (6.7%) were males i.e., in a ratio of 14:1. The range of patients' age under study was 8-40yrs. The mean and median ages at presentation to the hospital were 22.83 ± 8.53yrs SD and 19±1.58 yrs SE respectively. Majority of the sample suffered from childhood onset LE [late childhood and adolescence i.e., 15 patients (50%)] who also suffered from increased incidence (50-80%) of systemic manifestations later [Figure 1].

The sample population suffered from the symptoms of LE for a mean duration of 7.04± 11.63 months, shortest being fifteen days and longest being five years.Dermatological manifestations showed avid differences in their nature, distribution and frequency of occurrence.[3] The cutaneous findings were grouped into two–LE specific features and LE nonspecific features [Table 2]. All the patients belonging to our study showed a dimorphic picture i.e., atleast one each of LE specific and LE nonspecific features at the same time.

The LE specific manifestations noted were as follows: Malar rash presented more commonly as erythematous edematous rash accompanied by facial edema on the butterfly area of the face and bridge of the nose, rarely on the forehead, chin and V area of neck in 24 patients (80%). However, it spared the nasolabial folds. Discoid rash occurred in the form of sharply demarcated, coin shaped erythematous, hyper pigmented plaques with atrophy, telangiectasia and adherent scale extending into follicular orifices on head and neck in 11 patients (36%). 29 patients (96%) were found to be photosensitive.

Other LE specific features like papulosquamous rash starting initially as macules or papules on predominantly sun exposed areas occurred in 15 patients (50%), lupus profundus in one patient (3.34%) and Toxic epidermal necrolysis like lesions in one patient (3.34%). The lesions of lupus profundus were found to be tender subcutaneous nodules with deep saucerized depressions showing lymphocytic lobular panniculitis on biopsy. TEN like lesions presented as vast areas of denudation secondary to a diffuse rash on photoexposed areas and epidermal necrosis on biopsy.

The LE nonspecific features noted were noncicatricial alopecia in 17 patients (56.7%), painless oral ulcers mostly involving the hard palate and buccal mucosa in 28patients (93%), cutaneous vasculitis presenting as punctate lesions, palpable purpura, ulcers or erythematous plaques on lower limbs or diffusely in 24 patients (80%), bullous SLE like blisters in two patients (6.7%) and Raynaud's phenomenon in four patients (13%).

Occurrence of systemic manifestations like non erosive arthritis, hematological and renal manifestations and infections was more common compared to other features.[4] The occurrence of various manifestations was- non erosive arthritis in 24 patients (73.3%), infections in ten cases (33%), pleurisy in five (16%), lupus nephritis in twelve (40%), hematological manifestations in 28 patients (93%), cardiovascular manifestations in one (3.34%), gastrointestinal and hepatic manifestations in six (20%), neuropsychiatric in six (20%), obstetric in three (10%) and gynecological manifestations in seven (21%). In addition, myalgias occurred in 16% of the patients.

Hematological manifestations were noticed in the form of anemia (Hb<12g/dl), leucopenia (<4000cells/mm^3), lymphopenia (<1500 cells/mm^3), thrombocytopenia (<1 lakh/mm^3) and bleeding tendencies in 93%, 20%, 30%, 50% and 16% of the patients respectively.[5] Anemias being the commonest hematological dyscrasia in the sample, the sufferers were grouped into three grades based on their levels of hemoglobin in blood. The results indicated that 32% of the patients presented with milder grade (10-12g/dl of Hb), 32% with moderate grade (8-10g/dl of Hb) and 35.7% with severe grade (<8 g/dl of Hb) of anemia.

The hepatic manifestations (20%) in the form of lupus hepatitis (33%), fatty liver (50%), cholilithiasis (16%) and ascites contributed to a significant case load in the present study.[6] The gynecological manifestations among the sample women were characterized by irregularities of menstruation; contributed by amenorrhea with a negative UPT (11%), oligomenorrhea (21%), menorrhagia (11%) and premature menopause (11%). The chief obstetric complaints constituted abortions (13%) and infertility (6%) among the 15 married females. Autoimmune thyroiditis was found in one patient (3%), and three patients (10%) had a history of prior intake of isoniazid with antihistone Ab positivity in only one among these three.An attempt to study the frequency of systemic involvement in relation to various dermatological manifestations has been made in the present sample.[7] and the p values derived using Fisher's exact test of significance were depicted in [Figure 2] in an increasing order. Statistically significant association (p value<0.05within 95% confidence limits) was found between malar rash and arthritis (p=0.0021), malar rash and renal involvement (p=0.0313) and papulosquamous rash and arthritis (p=0.0176).[8]

The relationship between cutaneous manifestations and systemic involvement indicated that there is an absolute association between malar rash and systemic involvement and a relative association between cutaneous manifestations like photosensitivity, vasculitis, oral ulcers and hair loss, and increased systemic involvement. Discoid rash and Raynaud's phenomenon were not found to be associated with increased systemic involvement [Figure 3].[9]

Based upon the findings from [Figure 2] and [Figure 3] it was observed that patients with certain cutaneous manifestations like malar rash, photosensitivity, vasculitis, alopecia and oral ulcers presented with increased systemic involvement.[10] Patients with SLEDAI 2K score ≥16 also presented with increased systemic involvement [Figure 4].

Out of the five patients dead, three succumbed due to renal failure and two due to infections. Among the above five, three suffered from childhood onset of LE with high disease activity noted as increased SLEDAI-2K scores (≥20) [Table 3] who had significant association with malar rash, photosensitivity, oral ulcers, hair loss and vasculitis [Table 2].

Patients with systemic involvement showed differential positivity for autoantibodies. Autoantibodies like anti Sm Ab,anti-ds DNA Ab, anti-histone Ab and anti-Rib P Ab were more frequently found in patients suffering from childhood onset SLE. In patients with renal involvement, the frequencies of anti Sm Ab and anti-ds DNA Ab positivity were found to be 50% and 41% respectively. In patients with CNS involvement the frequency of anti-Rib P Ab positivity was found to be 83%. Anti-histone Ab positivity was found in ten patients of the current sample (30%). Among them two (20%) succumbed to death–one due to infection of lung and another due to renal involvement.

DISCUSSION: The present study showed preponderance of the disease in females compared to males (14:1) which is in consonance with the findings of Alakes Kumar Kole, et al, but not with Malaviya, et al which reported a ratio of 8:1. The mean age of presentation of the disease was 22.83 ± 8.53 years and median age was 19±1.58 years [Table 1]. These findings differed from those of Alakes Kumar Kole, et al and Malaviya, et al who reported 25years and 24 years as median age of disease onset respectively. Majority of the sample suffered from acute or subacute or chronic forms of childhood onset LE (≤18years) [Figure 1].[11]

The findings related to occurrence of LE specific features like malar rash, discoid rash and lupus profundus lesions, and LE nonspecific features like bullous lesions and Raynaud's phenomenon were found to be almost in consonance with those of Alakes Kumar Kole, et al study. But in the case of other manifestations like photosensitivity (96%), papulosquamous rash (50%), noncicatricial alopecia (56.7%), oral ulcers (93%) and cutaneous vasculitis (80%), the findings in the present study did not concur with those reported by Alakes Kumar Kole, et al (50%, 26.67%, 86.67%, 56.67% and 33.34% respectively).

Non erosive intermittent polyarthritis presenting as erythema, soft tissue swelling and tenderness of small joints of hands, wrist, ankle and knee joints in decreasing order of frequency, ranging from extremes of milder form to great physical disability was found to be less in the present study (73%) than that reported by Alakes Kumar Kole, et al (90%).

Various opportunistic infections like respiratory tract infections,tuberculosis,urinary tract infections, scabies, candidiasis, staphylococcal and herpes infections. affected 33% of the sample, probably the main reason being immunosuppression due to active disease induced leucopenia and usage of high doses of steroids and immunosuppressants as part of long term therapy.

The findings related to occurrence of pleurisy tallied with other Indian studies like Alakes Kumar Kole, et al (13.34%), Malaviya, et al (17%) and Vaidya, et al (15.5%) but not with Dubois, et al (45%).

Lupus nephritis, a life threatening systemic manifestations occurring in 40% of the present sample was found to be in tune with Alakes Kumar Kole, et al (46.67%) and Dubois, et al (46.1%) but differed from those of Malaviya, et al (73%) and Vaidya, et al (35%).

In 70 % of the sample, the chronology of progression of the disease was found to be a prodrome of arthritis followed by skin manifestations and later by renal manifestations, implying the probable role of cutaneous manifestations of LE as forerunners of renal involvement in majority of the patients.Hematological manifestations presenting as anemia or leucopenia or lymphopenia or thrombocytopenia coupled with bleeding tendencies together accounted for 93%; concurring with the findings of Alakes Kumar Kole, et al (90%).

These findings necessitated the need to perform peripheral smear, as majority of them presented with severe anemia (35.7%), and hemolytic anemia occurring as part of the disease process needs to be differentiated from microcytic anemia.[12] Cardiovascular involvement in the form of early onset myocardial infarction in 3.34% of patients in the current study concurred with the findings of Malaviya, et al (5%) but not with that of Alakes Kumar Kole, et al (13.34%), Vaidya et al (11.8%) and Dubois, et al (30.5%).

Patients with gastrointestinal involvement presented with complaints of nausea, vomiting and epigastric discomfort. The hepatic damage manifested as lupus hepatitis, fatty liver, cholilithiasis or ascites.Patients suffering from neuropsychiatric involvement presented with seizures, psychosis, loss of cognition, lupus headache mimicking migraine not responding to narcotic analgesia and depression. Their occurrence in our study (20%) almost matched the findings of Malaviya, et al (15%), Vaidya, et al (25.5%) and Dubois, et al (25.5%) but not with Alakes Kumar Kole, et al which showed a larger figure (73.34%).

The occurrence of autoimmune thyroiditis in a patient with LE supports the well-known immunological basis of SLE.

Statistically significant association (p value<0.05 within 95% confidence limits) was found between malar rash and arthritis (p=0.0021), malar rash and renal involvement (p=0.0313) and papulosquamous rash and arthritis (p=0.0176) on applying Fisher's exact test of significance that supported the hypothesis of cutaneous manifestations as markers of systemic involvement [Figure 2].

This was also supported by a line diagram which suggested an absolute association of malar rash and relative association of manifestations like photosensitivity, vasculitis, oral ulcers and hair loss with increased systemic involvement.[13] However, discoid rash and Raynaud's phenomenon were found to be not associated with increased systemic involvement [Figure 3]. In addition increased systemic involvement was observed in patients with SLEDAI 2K score ≥16 [Table 1and Figure 4].

Major contributors to mortality were found to be renal failure (60%) and infections (40%). Among the patients who succumbed to death, majority suffered from childhood onset LE (age ≤18yrs) and a higher disease activity (SLEDAI 2K≥20) [Table 3] and had significant association with malar rash, photosensitivity, oral ulcers, hair loss and vasculitis [Table 2]. It may be noted that the above cutaneous manifestations could indirectly help in predicting the prognosis by contributing individually to SLEDAI 2K score even in the absence of significant systemic involvement [Figure 4].

Autoantibodies like anti Sm Ab, anti-ds DNA Ab, anti-histone Ab and anti-Rib P Ab were more frequently found in patients suffering from childhood onset SLE. It was also observed that

an increased frequency of renaldisease occurred in those with anti Sm Ab and anti-ds DNA Ab positivity.

An increased neuropsychiatric involvement was seen in those with anti-Rib P Ab positivity. Among those with anti-histone Ab positivity prior history of intake of isoniazid in only one patient and death in two suggested its less specific nature in diagnosis.Though the sample in the present study was proportionally higher in comparison to the local population and duration of the study, conducting a larger study with similar objectives in near future may be necessary for a more meaningful interpretation of data to overcome the limitations of this study.[14]

CONCLUSIONS: Patients who presented with childhood onset LE (age ≤18yrs) have increased probability of developing early systemic involvement.Statistically significant association between malar rash and arthritis, malar rash and renal involvement and papulosquamous rash and arthritis, and relative association between photosensitivity, vasculitis, oral ulcers, hair loss andoccurrence of systemic damage suggests that these features could be considered as markers of systemic involvement, i.e., mostly forerunners of renal involvement. Discoid rash and Raynaud's phenomenon could be considered as non-indicators of any systemic involvement in SLE.

The above cutaneous manifestations indirectly help in predicting the prognosis by contributing individually to SLEDAI 2K scores even in the absence of systemic involvement and in alerting the physician regarding the forthcoming events.[15] Mortality tends to be more among the patients with SLEDAI 2K≥20 belonging to younger age group (≤18yrs) especially those who suffered from either lupus nephritis or infections.Hence, our study agrees with the earlier observations that LE specific features are of diagnostic importanceand LE nonspecific features are important predictors of disease activity, indicating that their identification pavesway for early suspicion of systemic involvement andbetter management of the disease.

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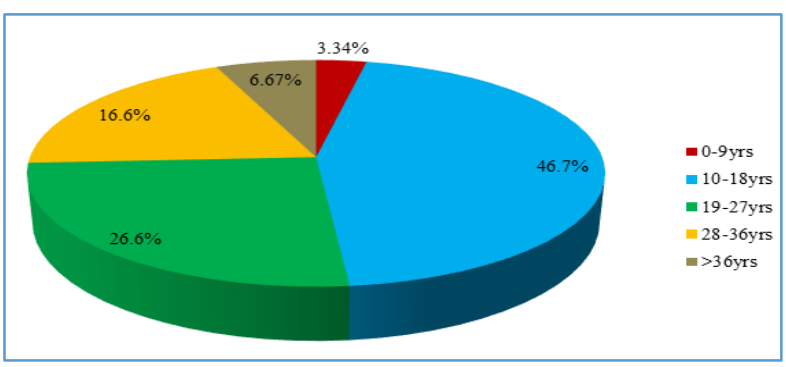

Fig.1: Age Distribution

Majority of the sample population who suffered from morbidity and mortality under present study suffered from childhood onset SLE (age ≤18yrs).

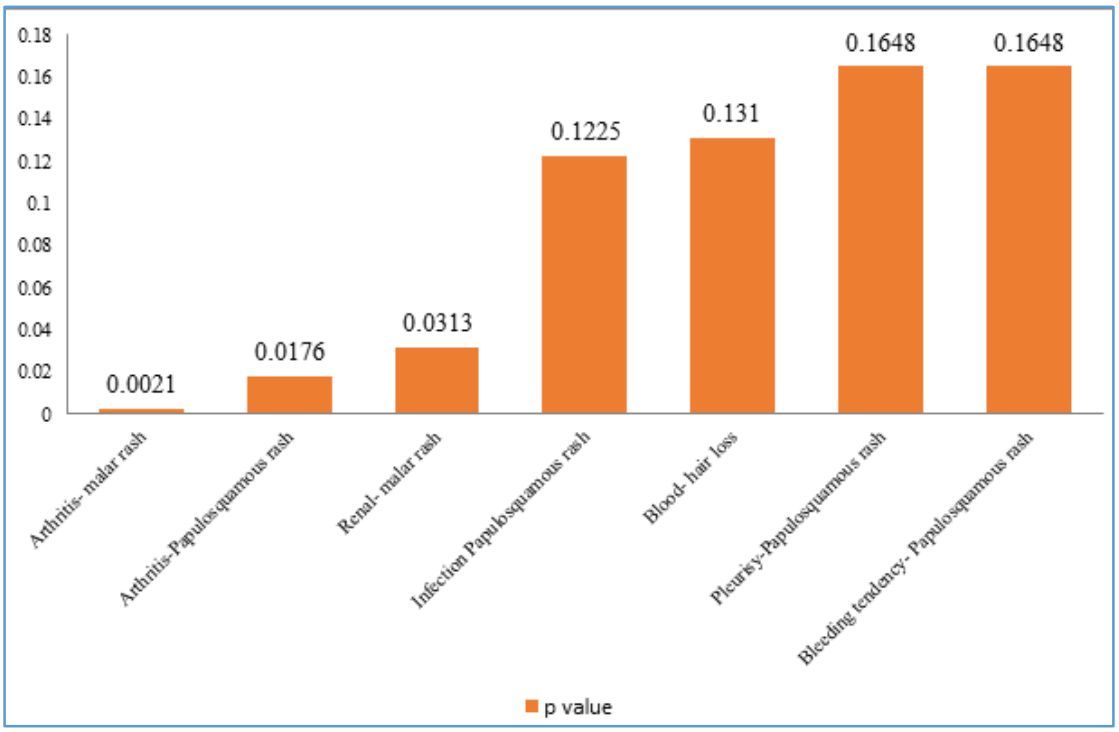

Fig. 2: Statistical Association between Skin Manifestations and Systemic Involvement

p values- plotted in an increasing order.
Statistically significant association within 95% confidence limits was found between malar rash and arthritis (p=0.0021), malar rash and renal involvement (p=0.0313) and papulosquamous rash and arthritis (p=0.0176).

x axis- type of systemic involvement;
y axis-percentage of cutaneous manifestations in relation to systemic involvement

INFERENCE:

- The study suggests an evidence of absolute association of malar rash and relative association of photosensitivity, vasculitis, oral ulcers and hair loss with systemic involvement.
- Discoid rash and Raynaud's phenomenon are not associated with increased systemic involvement.

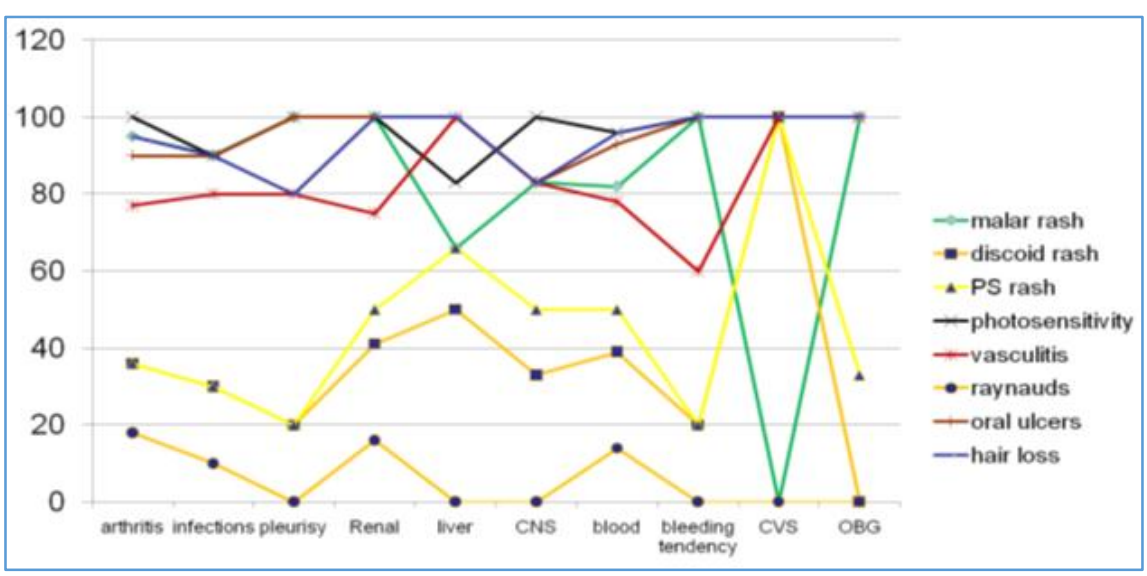

Fig. 3: Relationship between Cutaneous Manifestations & Systemic Involvement

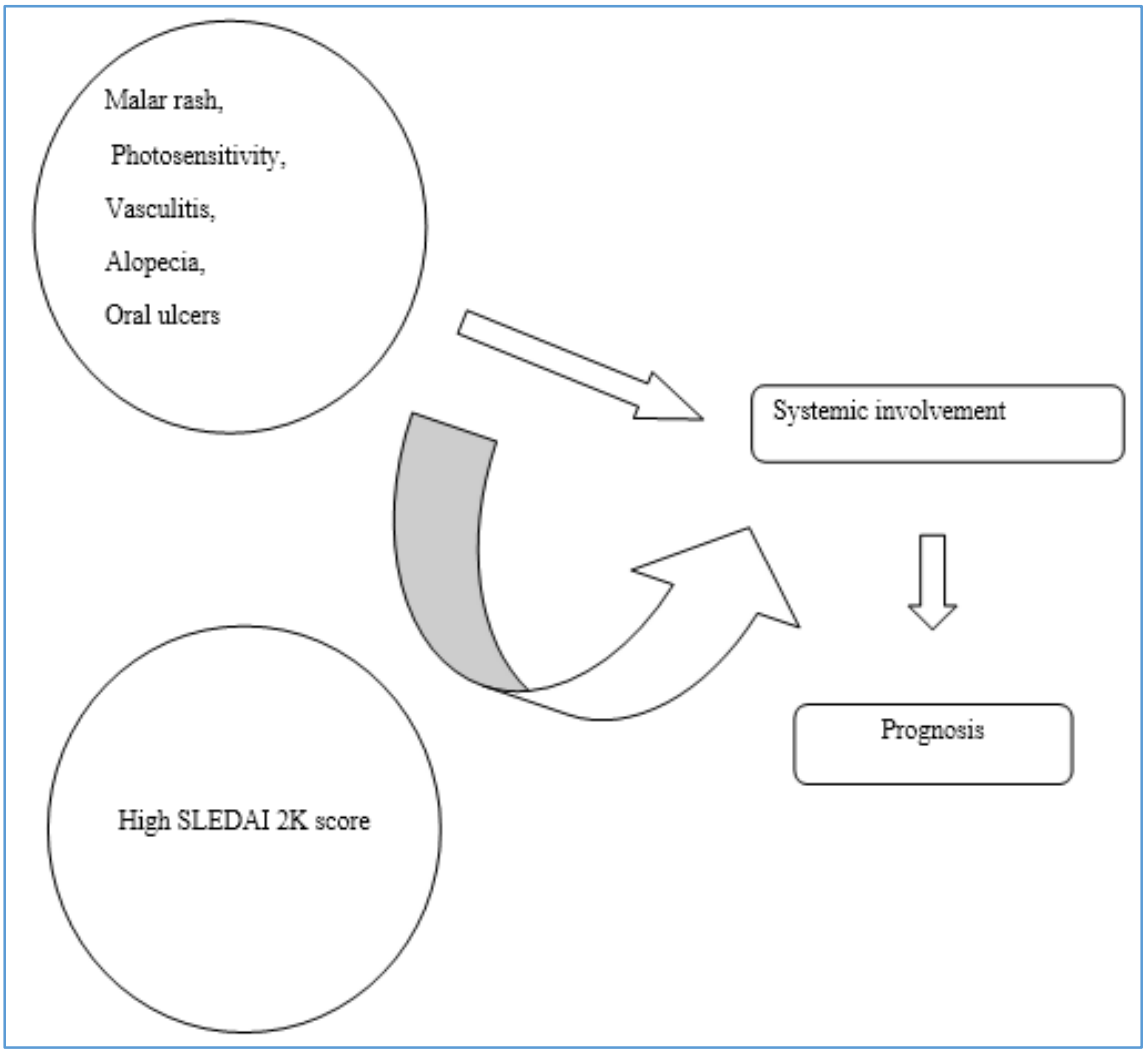

Fig. 4: Relationship between Sledai 2k Score and Prognosis

Patients with LE specific lesions like malar rash and photosensitivity and LE nonspecific lesions like vasculitis, alopecia and oral ulcers presented with increased systemic involvement. Patients with SLEDAI 2K score≥16 presented with increased systemic involvement.

- It has been observed that the above cutaneous manifestations can indirectly help in predicting future systemic involvement and thus prognosis by contributing individually to SLEDAI 2K score even in the absence of systemic involvement.

8-each	4-each	2-each	1-each
Seizures Psychosis Organic brain syndrome Cranial nerve disorder Lupus headache Cerebrovascular accident Visual disturbances Vasculitis	Arthritis≥2 joints Myositis Hematuria Proteinuria>0.5g/24h Pyuria Urinary casts	Inflammatory type rash Mucosal ulcers Alopecia Pleurisy Pericarditis Low complement Increased DNA binding	Fever Thrombocytopenia Leucopenia

Table 1: SLEDAI-2K SCORE- [SLE DISEASE ACTIVITY INDEX-2K]

LE specific features like rash and photosensitivity; and LE nonspecific features like vasculitis, oral ulcers and alopecia can indirectly help in predicting systemic involvement and thus prognosis by contributing individually to SLEDAI-2K score even in the absence of significant systemic involvement.

Type of CLE	SLEDAI 2K score		
	Range (Min-Max)	Mean	Median
LE SPECIFIC FEATURE			
• Malar rash	14-38	25.2	26
• Discoid rash	12-38	23.36	23
• Photosensitivity	11-38	23.7	24
• Papulosquamous rash	11-31	22	21
• Lupus profundus	28	28	28
• TEN like lesions	26	26	26
LE NONSPECIFIC FEATURE			
• Noncicatricial alopecia	11-36	24.29	24
• Oral ulcers	11-38	23.25	23.5
• Cutaneous Vasculitis	12-38	24.1	23.5
• Bullae	18-19	18.5	18.5
• Raynaud's phenomenon	11-27	21.25	23.5

Table 2: Relationship between Dermatological Manifestations with Sledai 2k Score

High SLEDAI-2K scores were noted in the presence of malar rash, photosensitivity, oral ulcers, hair loss and vasculitis.

CAUSE OF DEATH	AGE IN YRS.	SLEDAI 2K score
Renal failure	13	24
Renal failure	15	28
Renal failure	25	27
Infection	18	26
Infection	25	20

Table 3: MORTALITY DATA

Out of the five patients, three died due to renal failure and two due to infections. Majority of them suffered from childhood onset SLE with SLEDAI 2Kscores ≥20.

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