

Cutaneous Manifestations of Systemic Lupus Erythematosus (SLE)

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Abstract: Background: Systemic lupus erythematosus (SLE) is an autoimmune disease. (SLE) has many different skin lesions due to SLE can produce serious range of morbidity resulting from painful skin lesions, Some of the cutaneous manifestations for SLE, such as photosensitivity reactions, oral ulcers, alopecia, urticaria, vasculitis, vesiculo-bullous lesions, cutaneous mucinoses, and cutaneous calcinosis, they maybe specific to SLE but also they could be associated with different dermatological diseases such Eczema. **Objective:** There are several studies that had already discussed a very important point concerning this topic but the aim of this paper is to evaluate the symptoms on the skin caused by SLE based on a previous different studies. **Methodology:** the methods of this study will be based on a systemic analysis and literature search through US national library Midline (pubmed) all these studies which discusses the cutaneous or skin symptoms of SLE will be included in this study. **Results:** Skin involvement, including the rare variant of TEN-like acute cutaneous SLE, is very common among SLE patients. The acute syndrome of pan-epidermolysis or apoptotic pan-epidermolysis may become a useful designation when considering a clinical diagnosis of drug-induced TEN or SLE. Further studies are required to verify our findings. **Conclusion:** Skin disease in patients with lupus erythematosus may be subdivided into two broad categories - those lesions that when biopsied demonstrate interface dermatitis and those that do not demonstrate interface dermatitis. The skin lesions that are represented by the interface dermatitis include discoid lupus erythematosus, subacute cutaneous lupus erythematosus and acute cutaneous lupus erythematosus. Patients with these 'specific' manifestations have varying degrees of systemic involvement from rare systemic disease in patients with localized discoid lupus erythematosus to common and often severe involvement in patients with acute cutaneous lupus erythematosus.

Keywords: cutaneous mucinoses, Systemic Lupus, erythematosus (SLE), Skin involvement.

1. INTRODUCTION

Systemic lupus erythematosus (SLE) is a chronic prototypic autoimmune disease of unknown etiology with many clinical manifestations. that can affect almost any organ system but The skin is one of the target organs most variably affected by the SLE disease; it is a disease with a large varieties of cutaneous and systemic manifestations that has been the subject of clinical research for more than a century. The term “lupus” originated in ancient Greece. Usually the skin and/or mucous membranes are involved in up to 85% of systemic lupus erythematosus (SLE).

The determination of the cutaneous manifestations of LE depends on clinical, histopathology, and immunohistology of skin lesions. Moreover, serum autoantibodies are viewed as immunologic markers for particular clinical sorts of the ailment. The Cutaneous Lupus Erythematosus Disease Area and Severity Index (CLASI) is utilized as a clinical device that institutionalizes the way sickness action is portrayed and gives rules to recognizing a clinical change. This clinical apparatus measures ailment movement and harm in cutaneous lupus erythematosus. The action score depends on the erythema, scale, mucous layer lesions, and nonscarring alopecia. A late study gives us an establishment for the handy utilization of the CLASI in clinical trials as a device to gauge illness seriousness and responsiveness to treatment (R. Klein, et al.2011).

recently there is The Systemic Lupus International Collaborating Clinics (SLICC) is a universal gathering committed to SLE clinical exploration. This gathering delivered instruments that form the premise of result studies in SLE today, for example, the SLICC-ACR Damage Index (A. Kuhn, et al.2006). In the present study SLICC embraced a correction of the SLE classification criteria to address various worries that have emerged subsequent to the 1982 criteria were produced.

The SLICC formal evaluation of the imperative clinical manifestations of SLE and constraints of the 1982 ACR criteria is abridged in the diary: Lupus (I. Baumann, et al.2002). Worries about the clinical criteria in the current ACR classification including: conceivable duplication of profoundly corresponded cutaneous lupus terms, (for example, malar rash and photosensitivity) and the nonattendance of consideration of numerous different lupus cutaneous manifestations; exclusion of numerous SLE neurologic manifestations; and the need to use new norms in the measurement of pee protein. Worries about the immunologic basis incorporated the oversight of low supplement, and the need to incorporate new learning on antiphospholipid antibodies. The vast majority of all, there were worries about patients with no immunologic criteria being delegated SLE (an autoantibody-intervened illness). In fact clinical trials have needed to include the necessity for the vicinity of a SLE autoantibody while enlisting patients to streamline the probability of reaction to immunosuppressive treatment

2. LITERATURE REVIEW

The various skin manifestations of LE are divided by a classification system proposed by James N. Gilliam (1936–1984), who distinguished between LE-specific and LE nonspecific cutaneous manifestations based on histological criteria as seen in **Table1**.

(Fries, et al. 1987) in his methodology paper has reviewed the basic methodology in creating classification criteria for SLE to keep away from circularity, that is, the evasion of criteria that are formed to the test information and not as a matter of course generalizable. The basic strategies incorporate utilization of a "gold" standard which should be set up by very experienced clinicians. Sequentially treated patients and numerous establishments should be utilized to minimize choice inclination. Control populaces ought to be spoken to a reasonable range of related maladies that imitate the symptomatic issues that emerge, in actuality. The variables must be characterized with exactness, in light of the fact that a little change in the definition for a paradigm could prompt a vast change in affectability and specificity. At long last the proposed criteria should be accepted on another populace (since criteria dependably function admirably in the populace from which they were produced)

(Callen JP, et al.2006) stated in his study that skin disease in patients with lupus erythematosus might be subdivided into two general classes - those lesions that when biopsied exhibit interface dermatitis and those that don't show interface dermatitis. The skin lesions that are spoken to by the interface dermatitis incorporate discoid lupus erythematosus, subacute cutaneous lupus erythematosus and intense cutaneous lupus erythematosus

Patients with these "particular" manifestations have differing degrees of systemic inclusion from uncommon systemic sickness in patients with limited discoid lupus erythematosus to regular and frequently serious association in patients with intense cutaneous lupus erythematosus. Patients who don't exhibit interface dermatitis likewise might have systemic malady and in a few occasions the skin manifestations are connected to a portion of the more extreme systemic manifestations. Inflammatory periorbital edema is uncommon in patients with SLE, which is useful as a distinguishing feature from dermatomyositis. Patients with localized DLE, hypertrophic LE, LE panniculitis, and lupus tumidus tend to have skin disease only; however, progression to systemic disease is possible (Durosaro O, et al. 2009).

(R. Jonsson, et al.1984) stated that The buccal mucosa, hard sense of taste, and vermilion outskirts are the area's most habitually included by lesions, which can be three sorts (discoid lesions, erythematosus lesions, and ulcers) and might exist together, prompting oedema and petechiae. Discoid lesions show up as focal regions of erythema with white spots encompassed by transmitting white striae and telangiectasia at the fringe. Erythematosus lesions are frequently joined by oedema and petechial blushing on the hard sense of taste, despite the fact that they are generally discovered unexpectedly as level macular ranges with ineffectively characterized outskirts. Ulcers have a tendency to happen in products and are shallow. They are normally 1-2 cm in measurement and in around 33% of patients might stretch out into the pharynx.

Table.1: LE-specific skin disease – subtypes of cutaneous lupus erythematosus (CLE).

1. Acute cutaneous lupus erythematosus (ACLE)
Localized form
Generalized form
2. Subacute cutaneous lupus erythematosus (SCLE)
Annular form
Papulosquamous form
3. Chronic cutaneous lupus erythematosus (CCLE)

Discoid lupus erythematosus (DLE)
Localized form
Disseminated form
Lupus erythematosus profundus (LEP; Synonym: LE panniculitis)
Chilblain lupus erythematosus (CHLE)
4. Intermittent cutaneous lupus erythematosus (ICLE)
Lupus erythematosus tumidus (LET)

3. OBJECTIVES

This study aimed as to evaluate the different patterns and manifestations on the skin that are caused by systemic lupus erythematosus. so our study comes to strengthen the evidence in this topic and highlights the main points as we performed a systematic literature review and meta-analysis to assess the clinical differences in cutaneous manifestations of SLE.

4. METHODOLOGY

This study is systematic review and meta- analysis of the literature, so we performed a comprehensive search was undertaken by searching through the US National Library of Medicine (Pubmed), The following criteria had to be met for the publication to be selected topic and all these studies which were discussing the **systemic lupus erythematosus** was included all studies that were conducted up to December 2015, our search terms were as following, SLE, cutaneous manifestation, skin symptoms of systemic lupus erythematosus.

Then we finally analysis the data and results of each included study to come out with the main and useful summarized results about the Cutaneous manifestation in SLE.

5. RESULTS

(Alakes Kumar Kole, et al. 2009) found out that out of the 150 patients, 140 (88%) were female and 10 (12%) were male. The female to male ratio was 14:1. The mean age at presentation was 30 years. All patients (100%) developed skin lesions during their follow-up period; although, at the time of presentation only 45 patients (30%) had cutaneous lesions and one third of the patients had acute presentation. The lupus erythematosus-specific lesions were noted as malar rash in 120 patients (80%) [Figure 1], photosensitive dermatitis in 75 patients (50%), generalized maculopapular rash in 40 patients (26.67%), discoid rash in 30 patients (20%) [Figures 2], subacute cutaneous lupus in 5 patients (3.34%), and lupus profundus in 5 patients (3.34%) but mucosal DLE, lichenoid DLE, and chilblain lupus were not detected.



Figure 1: A case of SLE with malar rash and lip DLE



Figure 2: A case of SLE on the scalp with scarring alopecia

SLE non-specific skin lesions noted were non scarring alopecia in 130 patients (86.67%); scarring alopecia in 10 patients (6.67%); oral ulcers in 85 patients (56.67%) [Figure 3], which were mostly painless; vasculitic lesions in 50 patients (33.34%); bullous lesions in 15 patients (10%) involving the retroauricular region, palms, upper trunk, etc. with frequent relapses; Raynaud's phenomenon in 10 patients (6.67%); erythema multiformae in 10 patients (6.67%); leg ulcers in 10 patients (6.67%); urticaria in 10 patients (6.67%); panniculitis in 2 patients (6.67%); periungual telangiectasia in 2 patients (1.34%); pyoderma gangrenosum in 2 patients (1.34%) and nail-fold infarct in 2 patients (1.34%); but lichen planus, sclerodactyly, livedo-reticularis, erythromelalgia, acanthosis nigricans, calcinosis, facial edema hyperpigmentation, and bluish pigmentation of the nails were not detected.



Figure 3: A case of SLE with bullous lesions

In some cases, skin lesions may be associated with the involvement of other organs. Vasculitic skin lesions in some cases are associated with neuropsychiatric manifestations of lupus but in this study, patients having such lesions were devoid of any overt neuropsychiatric features. In this study, patients having bullous skin lesions had systemic flares that were also reported previously by Malcangi, *et al.* An association between concomitant lupus nephritis and bullous lesions had been documented by Ng, *et al.* but no such association has been documented in this study.

In our study group, patients with LE non-specific skin lesions, specially generalized maculopapular vasculitic lesions, and diffuse non-scarring alopecia were associated with more active disease. This was also reported previously by Zeevi, *et al.* Another interesting finding in this study was that patients with cutaneous lesions had significantly more lymphadenopathy (20%); this was also previously reported by Arjeh, *et al.*

6. DISCUSSION

In several thinks about, cutaneous contribution was the most widely recognized element (73.34%) in the ailment range. Cutaneous lesions were the starting presentation in 25% of the cases as reported by Waston in 1989 ; while, Feng, *et al.* what's more, Malaviya, *et al.* reported joint inflammation as a beginning indication in 44% and 57% of the patients, separately. Established frameworks (e.g., fever, weight reduction, and so forth.) was the second most basic presentation in this study. Among the LE-specific cutaneous lesions, malar rash was the most widely recognized injury (80%) noted in this study; though Wysenbeek, *et al.*1992 and Vaidya, *et al.*1997 from western India reported malar rash in 49% and 53.18% of the patients, separately. Lesions of discoid lupus was impressively lower (20%) in this study and pretty much validated with the concentrates beforehand directed by Kapadia in 1996 and Wysenbeek. Diffuse maculopapular rash was noted in 26.67% of the cases rather than 59% of the cases as reported by Wysenbeekn, *et al.* Lesions of subacute cutaneous lupus were identified in this study in 3.34% of the cases; though, Wysenbeek, *et al.* reported the lesions in 13% of the cases. Mucosal DLE, chilblain lupus, and so forth were not recognized in this study.

Among LE non-specific, skin lesions non-scarring diffuse alopecia was more regular (86.67%) when contrasted with 57% noted by Wysenbeek in 1992 and 82% by Malaviya in 1988. Oral ulcers were found in this study in 56.67% of the cases when contrasted with 9.1% and 64% reported by Dubois in 1964 and Malaviya, individually. Raynaud's wonder is a less normal skin injury in SLE. In this study, we had seen this in 6.67% of the cases, though Malaviya, *et al.* from north India and Vaidya, *et al.* from Western India noticed Raynaud's marvel in 32% and 15.5% of the cases, individually

7. CONCLUSION

Cutaneous manifestations are seen in 72–85 % of patients with SLE, can occur at any stage of the disease, irrespective of disease activity, and indeed are the first sign of disease in 23–28 %. In addition to characteristic discoid lesions, which are included in the ACR criteria for diagnosing SLE, there are a variety of other LE-specific skin manifestations known as CLE and its subtypes ACLE, SCLE, CCLE, and ICLE. CCLE is further divided in the subtypes DLE, LEP, and CHLE. The subtypes of CLE are often not distinguished by any other medical specialty and thus not defined as distinct disorders. CLE is therefore a dermatological domain, even though close interdisciplinary cooperation is needed at the time of diagnosis as well as over the course of disease to exclude any progression to SLE.

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