Original Research Paper

Volume - 11 | Issue - 05 | May - 2021 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Dermatology

CLINICAL PROFILE OF LUPUS ERYTHEMATOSUS: A HOSPITAL BASED PROSPECTIVE STUDY.

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ABSTRACT **Objectives:** This present study was to evaluate the prevalence and clinical profile of various age group of patients with lupus erythematosus. **Methods:** A detail history, clinical examinations and relevant investigations were performed to all patients. The ACR criteria were used to diagnose the SLE patients. Investigations were performed like as including routine blood with platelet count, urine examination, 24-hour urine protein, renal and liver function tests, serum electrolytes, random blood sugar, HIV, HbsAg, chest x-ray and ECG. Peripheral smear and LE cell demonstration were done in all patients. Immunological tests like ANA, ANA profile, VDRL, APLA, C3 and C4 were also done. Skin biopsy from the uninvolved covered area (buttock/inner aspect of thigh) was done in all patients who were willing and is sent in Michel's medium for Lupus Band Test (LBT). **Results:** Most common age group of LE patients22(55%) was 31-45 years. Majorities of 25(62.5%) patients were females. Male and female ratio was 5:3. Most of the patients 23(57.5%) of lupus erythematosus had the features of arthalgia/arthritis followed by 20(50%) oral ulcer, 13(32.5%) malar rash, 11(27.5%) DLE localised, 9(22.5%) photosensitivity, 7(17.5%) pedal edema and 5(12.5%) DDLE. SLE patients had erosions in the hard plate (27.14%), lips (10.2%) and under surface of tongue (3.23). In the DLE patients, (4.78%) patients presented with erosions in the hard plate and (2.5%) patient with white lacy plaque in the buccal mucosa. **Conclusions:** Lupus erythematosus was commonly seen in middle age group population. Females was more preponderance than males. SLE features was more common in LE patients. Clinical presentations in most of the patients had arthalgia/arthritis and Oral ulcer. DLE palque was the most common skin lesion. Renal involvement was commonly seen in with positive lupus band test. Most of the patients were seen anaemia and raised ESR as haemotological abnormality.

KEYWORDS : Lupus erythematosus, SLE. DLE, Clinical profile, Gender

INTRODUCTION

Systemic Lupus Erythematosus (SLE) is a complex autoimmune systemic disease with a worldwide distribution and an unknown etiology [1]. SLE is up to 10 times more common in women than men, and typically has a predilection for women in their childbearing years [2]. It is characterized by a great clinical polymorphism and female predominance [3, 4]. The appearance, progression, and outcome of SLE are influenced by genetic, immunological, and environmental factors [5, 6]. Ethnicity also seems to contribute to the expression and heterogeneity of the clinical and immunological features of disease.

The risk for SLE may be influenced by epigenetic effects such as DNA methylation and post-translational modifications of histones, which can be either inherited or environmentally modified. Epigenetics refers to inherited changes in gene expression caused by mechanisms other than DNA base sequence changes. Th e most well understood type of epigenetic factor is DNA methylation, which plays a role in a variety of human processes, such as X chromosome inactivation and certain cancers. Previous research has also implicated the importance of DNA methylation in SLE. Differences in the methylation status of genes may explain, at least in part, the discordance observed in some identical twins that are discordant for SLE. Epigenetic mechanisms may represent the missing link between genetic and environmental risk factors [7]. Objective of our study was to evaluate the prevalence and clinical profile of systemic lupus erythematosus patients.

MATERIAL & METHODS

This present study was conducted in Department of Dermatology, Autonomous State Medical College, Shahjahanpur, Uttarpradesh, India during a period from March 2020 to January 2021.

A total 40 patients of lupus erythematosus with age group 15 to 60 years attending Dermatology OPD were enrolled in this study. Entire subjects signed an informed consent approved by institutional ethical committee was sought.

Methods

A detail history, clinical examinations and relevant investigations were performed to all patients. History was taken like as onset of disease, duration, past history, any triggering factors and family history. The ACR criteria were used to diagnose the SLE patients. Investigations were performed like as including routine blood with platelet count, urine examination, 24-hour urine protein, renal and liver function tests, serum electrolytes, random blood sugar, HIV, HbsAg, chest x-ray and ECG. Peripheral smear and LE cell demonstration were done in all patients. Immunological tests like ANA, ANA profile, VDRL, APLA, C3 and C4 were also done. Skin biopsy from the uninvolved covered area (buttock/inner aspect of thigh) was done in all patients who were willing and is sent in Michel's medium for Lupus Band Test (LBT).

STATISTICALANALYSIS

Data was analysed by using simple statistical methods with help of MS-Office software. All data was tabulated and percentages were calculated.

OBSERVATIONS

This present study was included a total of 40 patients of lupus erythematosus. Most of the patients 22(55%) were in age group of 31-45 years. Majorities of 25(62.5%) patients were females. Male and female ratio was 5:3.

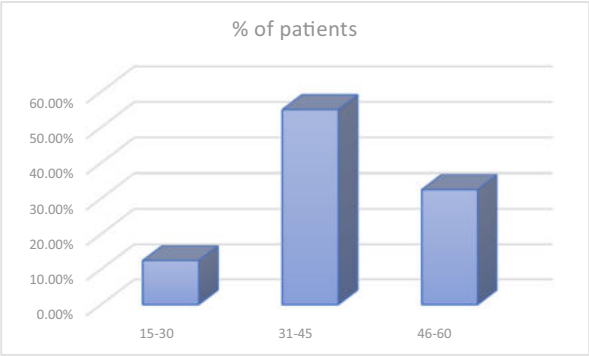

Figure.1. Age wise distributions of patients with lupus erythematosus.

Table.1. Clinical features of SLE and DLE patients.

Presentations	SLE	DLE	Total
Generalised rash	5(12.5%)	0	5(12.5%)
Malar rash	12(30%)	1(2.5%)	13(32.5%)
TEN-like rash	2(5%)	0	2(5%)
Bullous lesions-generalised	2(5%)	0	2(5%)
DLE localised	2(5%)	9(22.5%)	11(27.5%)
DDLE	1(2.5%)	4(10%)	5(12.5%)
Arthralgia/arthritis	20(50%)	3(7.5%)	23(57.5%)
Oral ulcer	17(42.5%)	3(7.5%)	20(50%)
Photosensitivity	7(17.5%)	2(5%)	9(22.5%)
Alopecia-diffuse	3(7.5%)	0	3(7.5%)
Alopecia-scarring	0	2(5%)	2(5%)

Raynaud's phenomenon	2(5%)	0	2(5%)
Fever	3(7.5%)	0	3(7.5%)
Pedal oedema	7(17.5%)	0	7(17.5%)
Purpura	4(10%)	0	4(10%)
Lichen planus - oral	1(2.5%)	0	1(2.5%)
Lichen planus - else where	1(2.5%)	1(2.5%)	2(5%)
Fungating plaque on DLE	1(2.5%)	0	1(2.5%)
Leg ulcer	2(5%)	1(2.5%)	3(7.5%)
Urticarial vasculitis	2(5%)	0	2(5%)
Sweet syndrome	0	1(2.5%)	1(2.5%)
Verrucous plaque	0	1(2.5%)	1(2.5%)
Chilblains	1(2.5%)	0	1(2.5%)

In this present study, most of the patients 23(57.5%) of lupus erythematosus had the features of arthalgia/arthritis followed by 20(50%) oral ulcer, 13(32.5%) malar rash, 11(27.5%) DLE localised, 9(22.5%) photosensitivity, 7(17.5%) pedal edema, 5(12.5%) DDLE, 3(7.5%) fever and leg ulcer, 2(5%) TEN-like rash, bullous lesions-generalised, Raynaud's phenomenon and urticarial vasaculitis and 1(2.5%) lichen planus, fungating plaque on DLE, sweet syndrome, verrucous plaque and chilblains.

Table.2. Type of LE lesion in LE patients

Type of lesions	Number of patients
1. ACLE	
A) General rash	5(12.5%)
B) Malar rash	12(30%)
C) Ten-like rash	2(5%)
2. SCLE	0
3. CCLE	
Classic DLE-	
Localised	11(27.5%)
Generalised (DDLE)	5(12.5%)
Chilblains	1(2.5%)
LE/LP overlap	2(5%)
Mucosal	2(5%)
Total	40

In the patients of ACLE, most of the patients 12(30%) had malar rash, and 5(12.5%) general rash, 2(5%) ten-like rashes. In CCLE cases had localised 11(27.5%), generalised (DDLE) 5(12.5%), LE/LP overlap and mucosal 2(5%) and chilblains 1(2.5%).

Table.3. Non-specific lesions

Type of Nonspecific Lesions	Number of Patients
1. Vascular	
A) Vasculitis	
I. Leukocytoclastic vasculitis	
a) Palpable purpura	4(10%)
b) Urticarial vasculitis	2(5%)
B) Periungual telangiectasia	1(2.5%)
C) Raynaud phenomenon	2(5%)
2. Non-scarring alopecia	
I. Lupus hair	3(7.5%)
2. Sclerodactyly	1(2,5%)
3. Le-nonspecific bullous lesion	2(5%)
4. Acanthosis nigricans	1(2.5%)
6. Lichen planus	2(5%)
5. Leg ulcers	2(5%)
Total	20

In this present study. Out of 40 patients, 20(50%) patients had nonspecific lesions. Out of total 40 patients of LE, leukocytoclastic vasculitis patients had palpable purpura 4(10%), urticarial vasculitis and raynaud phenomenon 2(5%) and periungual telangiectasia 1(2.5%). In non-scarring alopecia, lupus hair 3(7.5%), 2(5%) le-nonspecific bullous lesions, lichen planus and leg ulcers, 1(2.5%) sclerodactyly.

SLE patients had erosions in the hard plate (27.14%), lips (10.2%) and under surface of tongue (3.23). In the DLE patients, (4.78%) patients presented with erosions in the hard plate and (2.5%) patient with white lacy plaque in the buccal mucosa.

Of the 40 patients, major involvement was noted in renal system. 20% of patients had albuminuria and elevated 24-hours urine protein. 5% patients each had altered renal function. In other systems, 5% patients each had seizures, CVA, retinal vasculitis, hepatomegaly and raised liver enzymes. 2.5% patient each had pulmonary artery hypertension. Patients with systemic involvement were all having SLE except one patient (2.5%) with retinal vasculitis having DDLE.

Most common haematological abnormalities among SLE patients were anaemia and raised ESR occurred in 25% patients. Leucopenia occurred in 20% patients and lymphopenia in 10% patients. 2.5% patients had thrombocytopenia. Most common immunological abnormality among SLE patients was raised ANA titre in 52.5% patients followed by raised anti-dsDNA titre in 55%. Anti-Sm antibody was positive in 17.5% and SSA was positive in 16% patients. 12.5% patients each presented with AMA m2, nucleosome and antiphospholipid antibody.

10% patients each had complement C3 deficiency and ribosomal P protein. 5% patients each had RNP and antihistone. 2.5% patients each had PCNA and complement C4 deficiency. Among DLE patients, 5% patients presented with positive ANA and 2.5% patients each presented with SSA and AMA M2.

25% patients of SLE who were having positive lupus band had renal involvement and 12.5% patients with negative lupus band had no renal involvement. Lupus band was negative in 2.5% patient with renal involvement. Among SLE patients, 17.5% had hypothyroidism and 10% had hypertension. 2.5% patient had diabetes. One patient with DDLE had vitiligo. Among SLE patients, 7.5% patients each had cerebrovascular accidents, seizures, squamous cell carcinoma developing on DLE plaque and mortality. 5%% patient each had fracture hip, retinal vasculitis, pulmonary embolism and pulmonary artery hypertension.

DISCUSSIONS

Systemic lupus erythematosus (SLE) is the prototypic multisystem autoimmune disorder with a broad spectrum of clinical presentations encompassing almost all organs and tissues. The extreme heterogeneity of the disease has led some investigators to propose that SLE represents a syndrome rather than a single disease [7].

In this present study, a total of 40 patients of lupus erythematosus were included. Most of the patients 22(55%) were in age group of 31-45 years. Females 25(62.5%) was more preponderance than males. Male and female ratio was 5:3.

Immune complexes and complement activation pathways mediate effector function and tissue injury. In healthy individuals, immune complexes are cleared by Fc and complement receptors; failure to clear immune complexes results in tissue deposition and tissue injury at sites. Tissue damage is mediated by recruitment of inflammatory cells, reactive oxygen intermediates, production of inflammatory cytokines, and modulation of the coagulation cascade.

Autoantibody-mediated tissue injury has been implicated in neuropsychiatric SLE (NPSLE), where antibodies reacting with both DNA and glutamate receptors on neuronal cells can mediate excitotoxic neuronal cell death or dysfunction. Locally produced cytokines, such as IFNα and tumour necrosis factor (TNF), contribute to affected tissue injury and inflammation. Th ese mediators, together with the cells producing them (macrophages, leucocytes, dendritic cells and lymphocytes), are the subject of investigation as potential therapeutic targets in lupus. Recent studies have also highlighted the role of locally expressed factors for the protection of tissues under immune attack. For example, defects in kallikreins may jeopardise the ability of lupus kidneys to protect themselves from injury, PD-1-ligand down-regulates the activity of the infiltrating lymphocytes, and impaired regulation of complement amplifies vascular injury [7]. Vascular damage in SLE has received increased attention in view of its relationship with accelerated atherosclerosis. Homocysteine and proinfl amatory cytokines, such as IFNα, impair endothelial function and decrease the availability of endothelial precursor cells to repair endothelial injury. Pro-infl ammatory high density lipoproteins (HDL) and a dysfunction of HDL mediated by antibodies have also been implicated in defective repair of endothelium. Moreover, pathogenic variants of ITAM (immuno-tyrosine activation motif) alter its binding to ICAM-1 (intercellular adhesion molecule 1) and may increase the adherence of leucocytes to activated endothelial cells. Impaired DNA degradation as a result of mutations of the 3' repair exonuclease 1 (TREX1), and increased accumulation of single stranded DNA derived

from endogenous retro-elements in endothelial cells, may activate the IFN-stimulatory DNA response and direct immune-mediated injury to the vasculature [7].

In this present study, common features of lupus erythematosus were 23(57.5%) arthalgia/arthritis followed by 20(50%) oral ulcer, 13(32.5%) malar rash, 11(27.5%) DLE localised, 9(22.5%) photosensitivity. In the patients of ACLE, most of the patients 12(30%) had malar rash, and 5(12.5%) general rash.

The diagnosis of SLE is based on a combination of clinical manifestations, laboratory findings, serology and histology of affected organs (usually skin and kidney). Classification criteria for SLE are used mainly to ensure that patients are comparable in research studies, rather than as diagnostic criteria in routine clinical care. This has evolved from the American Rheumatism Association 1982 criteria [8] and the ACR 1997 criteria [9] to the SLICC 2012 criteria [10]. The SLICC 2012 criteria set has been shown to be more sensitive than the ACR 1997 criteria, to be applicable in childhood-onset SLE and in those with early disease and to be usable in clinical practice [11]. The classification of LN evolved from the World Health Organization 1995 classification [12] to the International Society of Nephrology/Renal Pathology Group 2003 classification [13]. In the SLICC 2012 classification for SLE, biopsy-proven LN plus positive ANA or anti-dsDNA is sufficient to fulfil SLE classification criteria. In 1999, the ACR developed a standardized nomenclature for NPSLE [14], which was subsequently validated. However, the prevalence of NPSLE has been difficult to establish. The 19 syndromes in the ACR list include common problems, such as headache, which have a high likelihood of being unrelated to the underlying disease. Furthermore, the pathogenesis of most NPSLE syndromes remains obscure and may be multifactorial, so associations with autoantibodies or other putative biomarkers are not well established [15]. In addition, NPSLE syndromes may mimic those seen in APS [16] and SS [17]. Cutaneous lesions occur in up to 85% of patients with SLE and are the first sign in up to 28%. Cutaneous lupus erythematosus is classified into acute, subacute, chronic and intermittent lupus erythematosus [18]. In 2004, the European Society of Cutaneous Lupus Erythematosus was founded to achieve a general consensus on evidence based clinical standards for disease assessment [19]. The Revised Cutaneous Lupus Erythematosus Disease Area and Severity Index is a validated instrument used in clinical practice and clinical trials to score activity and damage [20].

In this present study, Out of 40 patients, among them, leukocytoclastic vasculitis patients had palpable purpura 4(10%), urticarial vasculitis and raynaud phenomenon 2(5%) and periungual telangiectasia 1(2.5%). In non-scarring alopecia, lupus hair 3(7.5%), 2(5%) le-nonspecific bullous lesions, lichen planus and leg ulcers, 1(2.5%) sclerodactyly.

CONCLUSIONS

This present study concluded that the lupus erythematosus was commonly seen in middle age group population. Females was more preponderance than males. SLE features was more common in LE patients. Clinical presentations in most of the patients had arthalgia/arthritis and Oral ulcer. DLE palque was the most common skin lesion. Renal involvement was commonly seen in with positive lupus band test. Most of the patients were seen anaemia and raised ESR as haematological abnormality.

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