

## A Clinical Study on NPSLE in a Tertiary Care Hospital in Northeast India

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### ABSTRACT

**Introduction:** Neuropsychiatric Systemic Lupus Erythematosus (NPSLE) is a disease with central and peripheral nervous system manifestations. The diagnosis of NPSLE is often a diagnostic challenge though the criteria for NPSLE is well-established.

**Objective:** To study the prevalence and pattern of neuropsychiatric manifestations in SLE.

**Materials and Methods:** This hospital based observational study was carried out from Nov 2017 to June 2018. A total of 58 cases of SLE were assessed clinically and investigated accordingly. Special emphasis was given to look for Neurological involvement. MMSE, HAM A and HAM D scales were used for assessment of cognitive dysfunction, anxiety and depression respectively. The disease activity was measured by the SLEDAI.

**Results:** In a total of 58 patients with SLE evaluated, 92% were female. The most common age group was 21 to 30 years. Mean age was 25.6 years. Nervous system involvement was found in 37 (63.79%) patients. Headache was the most frequent manifestation, present in 32 (55.17%) patients followed by Cognitive dysfunction in 8 patients (13.79%). Seizure disorder was present in 6 (10.34%), acute confusional state in 2 (3.44%), depression in 4 (6.89%), anxiety in 2 (3.44%) and psychosis in 2 (26.31%) patients. Aseptic meningitis and peripheral neuropathy was found in 2 (3.44%) and 1 (1.72%) patients respectively. Many of the patients had

more than one neurological involvement. SLEDAI score was high in SLE patients with neurological manifestations.

**Conclusion:** The frequency of Neuropsychiatric involvement was found in the majority of the patients with SLE and headache was the most common manifestation. Patients with NPSLE showed high disease activity.

**Keywords:** SLE, NPSLE, SLEDAI.

### Abbreviations:

**SLE:** Systemic Lupus Erythematosus; **MMSE:** Mini Mental Status Examination; **MCTD:** Mixed Connective Tissue Disorder; **SLEDAI:** SLE Disease Activity Index.


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### INTRODUCTION

Neuropsychiatric involvement is common in patients with SLE and include a wide variety of central and peripheral nervous system manifestations. Although the criteria for NPSLE is well established, the diagnosis of NPSLE still poses a challenge. Therefore the study was undertaken to evaluate the pattern of neuropsychiatric involvement in SLE and its correlation with disease activity.

### OBJECTIVE

To determine the prevalence of NPSLE and pattern of Neuropsychiatric manifestations among a sample of SLE patients from a tertiary care hospital in Northeast India.

### MATERIALS AND METHODS

The study was carried out from Nov 2017 to June 2018 at Gauhati Medical College and Hospital. It was a hospital based observational study. A total of 58 cases were studied. Ethical clearance was taken from the institutional ethics committee, GMCH and informed consent was taken from each patient. Statistical analysis was done using Microsoft Excel 2017. The patients were classified according to the Systemic Lupus International Collaborative Clinics Criteria (SLICC) 2012.<sup>1</sup>

### Inclusion Criteria

Cases of either gender aged  $\geq 12$  years fulfilling the SLICC criteria 2012.

**Exclusion Criteria**

- Patients <12 years.
- Patients with Overlap Syndrome or MCTD
- Patients with Drug Induced SLE
- Patients with Neuropsychiatric manifestations due to etiologies other than SLE
- Patients who denied consent

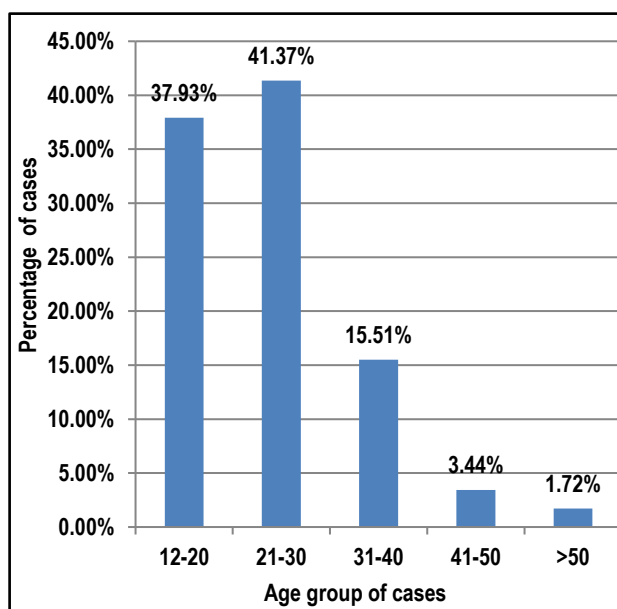
A diagnosis of Neuropsychiatric Lupus syndrome was made as per American College of Rheumatology (ACR) case definitions for NPSLE.<sup>2</sup> Cognitive dysfunction was assessed using MMSE.<sup>3</sup> Hamilton Rating Scale for Anxiety (HAM-A)<sup>4</sup> and Hamilton Rating Scale for Depression (HAM-D)<sup>5</sup> was used to assess the prevalence of Anxiety and Depression respectively. The disease activity was measured using SLEDAI.

**RESULTS**

Out of the 58 cases of SLE, 53 (91.38%) were females and 5 (8.62 %) were males with a mean age of 25.6 years and mean disease duration of 23.15 months. NPSLE was diagnosed in 37(63.79%) patients. Headache (55.17%) was the most frequent neuropsychiatric manifestation followed by cognitive dysfunction (13.7 9%), seizures (10.34%), depression (6.89%), anxiety (3.44%), psychosis (3.44%), aseptic meningitis (3.44%) and peripheral neuropathy (1.7 2%). Many of the patients had more than one neurological involvement. Mean SLEDAI Score was higher in NPSLE patients (29.67) as compared to non- NPSLE patients (16.94)

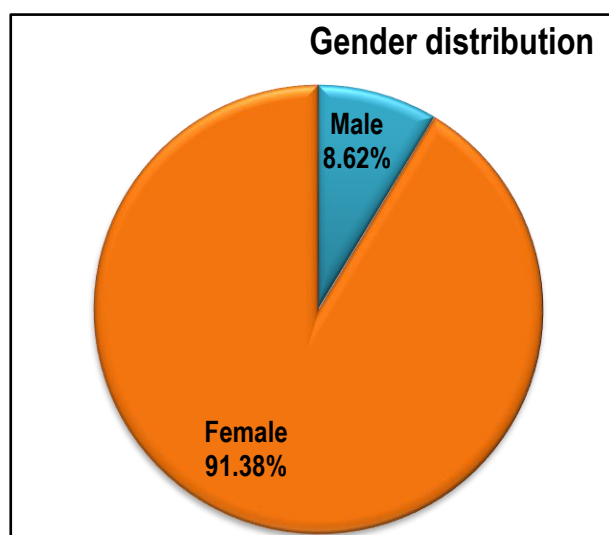
**Table I: Age distribution of cases**

Age group (in years)	Number of cases (n=58)	%
12-20	22	37.93
21-30	24	41.37
31-40	9	15.51
41-50	2	3.44
>50	1	1.72



**Table 2: Gender distribution of cases**

SEX	Number of cases (n=58)	%
Male	5	8.62
Female	53	91.38



**Table 3: Clinical presentation of cases**

Clinical presentation	Number of cases	%
Cutaneous Malar rash	24	41.37%
Rash Discoid rash	6	10.34%
Photosensitivity	34	58.62%
Oral ulcers	12	20.68%
Edema	18	31.03%
Pallor	39	67.24%
Arthritis	19	32.75%
Alopecia	5	8.62%
Fatigue	43	74.13%
Fever	16	27.58%
Weight loss	36	62.06%
Raynaud's Phenomenon	2	3.44%
Digital Gangrene	4	6.88%
Neurological manifestations	37	63.8%

**Table 4: Neurological manifestation of cases**

Neurological manifestations	Number of cases n=37	%
Seizures	6	(10.34%)
Acute confusional state	2	(3.44%)
Psychosis	2	(3.44%)
Depression	4	(6.90%)
Headache	32	(55.17%)
Cognitive dysfunction	8	(13.79%)
Aseptic meningitis	2	(3.44%)
Anxiety disorder	2	(3.44%)
Polyneuropathy	1	(1.72%)

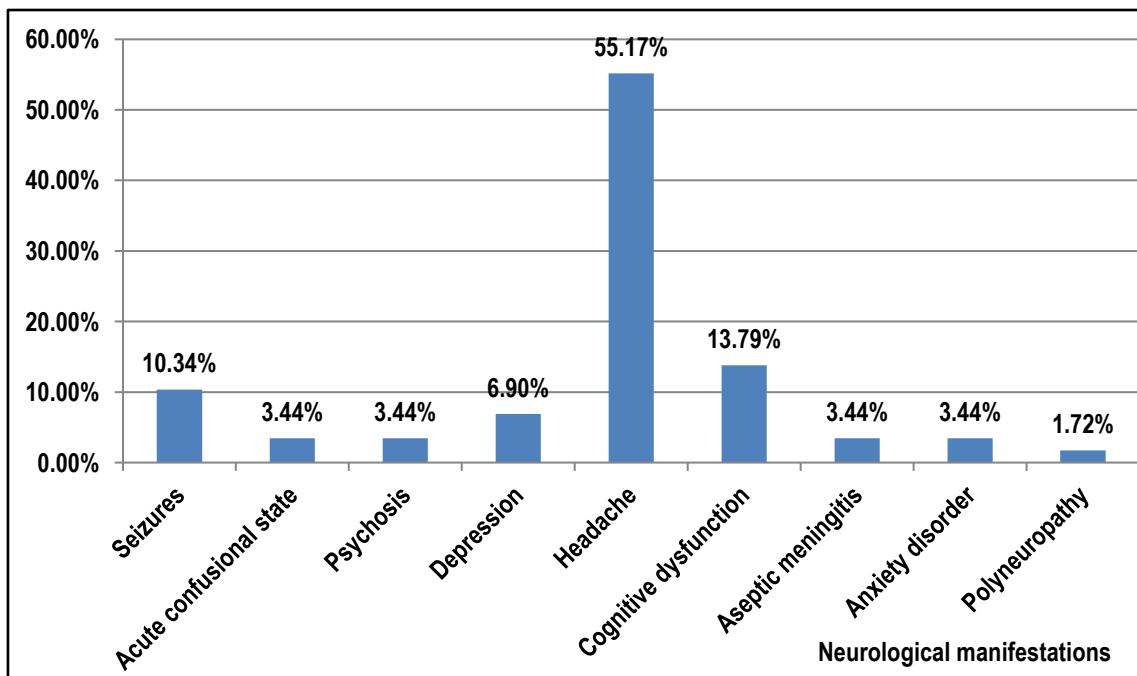
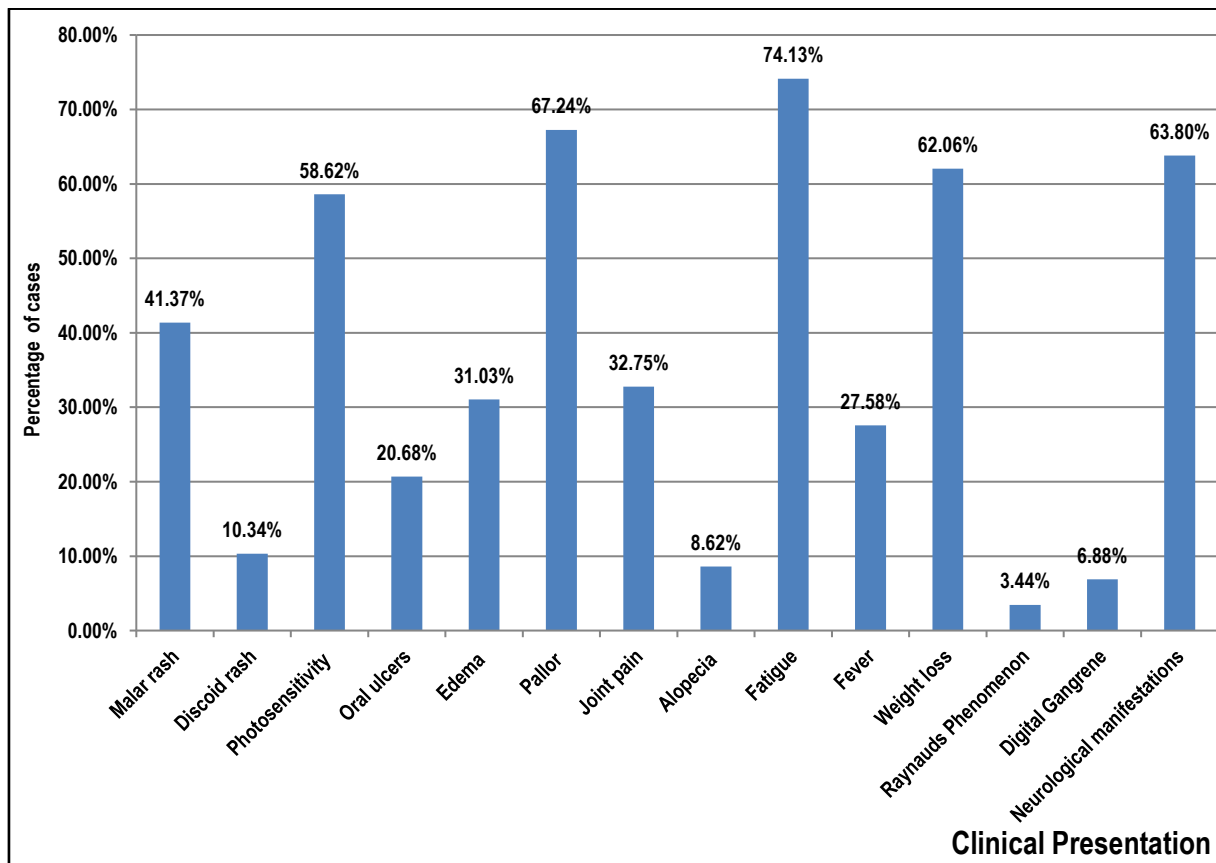
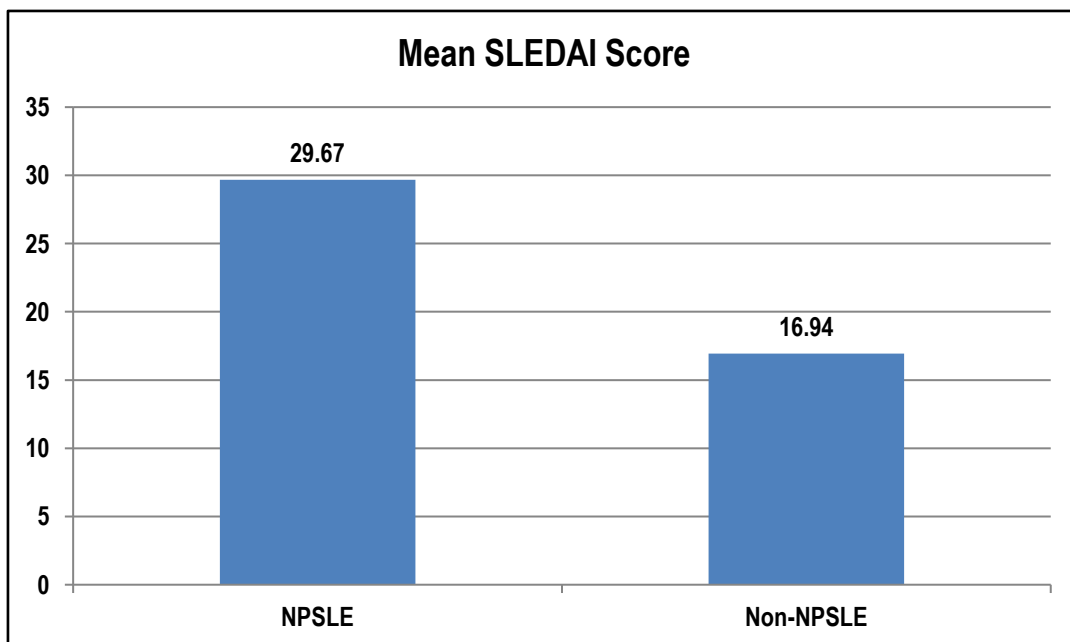
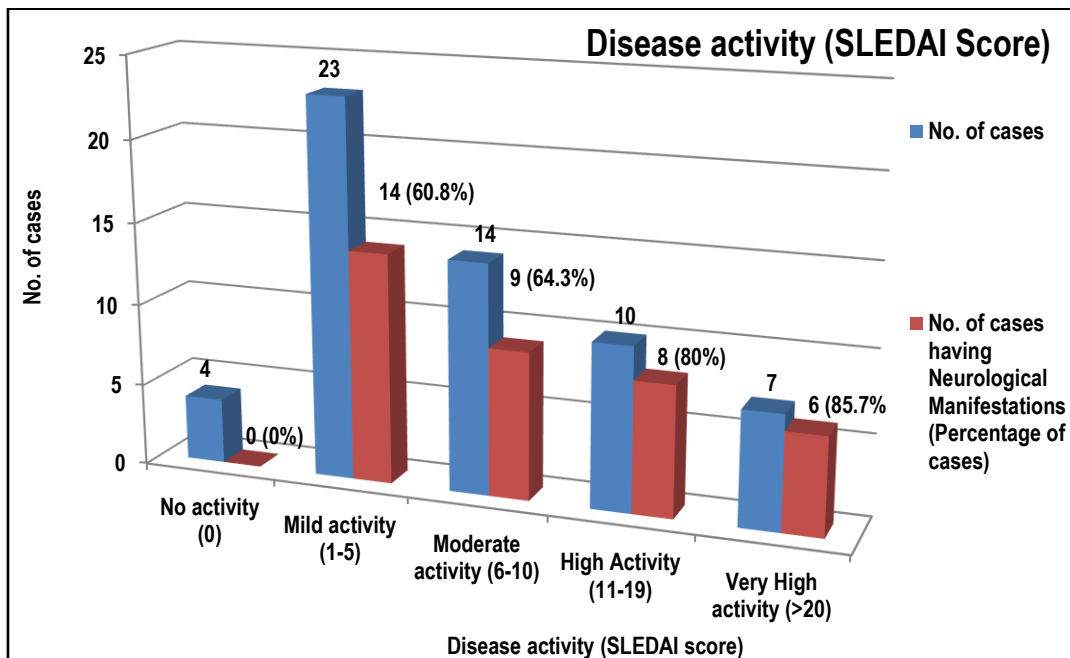


Table 5: Disease activity (SLEDAI Score)

Disease activity (SLEDAI Score)	Total No. of cases n=58 (%)	No. of cases having Neurological Manifestations (n=37)	%
No activity (0)	4 (6.9%)	0	0%
Mild activity (1-5)	23 (39.7%)	14	60.8%
Moderate activity (6-10)	14 (24.1%)	9	64.3%
High Activity (11-19)	10 (17.24%)	8	80%
Very High activity ( $\geq 20$ )	7 (12.06%)	6	85.7%



**DISCUSSION**

In our study, the mean age of patients was 25.6 years. Eissa M et al. (2018)<sup>6</sup> noted a mean age of 30.7 years. Muhammed H et al (2017)<sup>7</sup> found a mean age of 32.3 years in their patients Kakati S et al. (2017)<sup>9</sup> noted a mean age of 25.59 and Wei Fan et al (2014)<sup>8</sup> noted a mean age of 28 years in their study.

Out of 58 subjects studied, 53 (91.38%) cases were female and 5 (8.62%) cases were male with a female to male ratio of 10.6:1. The sex ratio in the study by Eissa M et al. (2018)<sup>6</sup> was 6.94:1. It was 10.2:1 in the study by Muhammed H et al (2017).<sup>7</sup> Kakati S et al. (2017)<sup>9</sup> noted a sex ratio of 12:1 and Wei Fan et al (2014)<sup>8</sup> found a sex ratio of 6.6:1. The mean disease duration in our study was 23.15 months. Whereas in studies by Eissa M et al. (2018)<sup>6</sup>, Muhammed H et al (2017)<sup>7</sup> and Wei Fan et al (2014)<sup>8</sup>; it was 72 months, 54 months and 12 months respectively.

In our study of the total 58 cases 37 (63.8%) patients had neurological manifestations. The most common neurological

manifestation was headache which was found in 32 (55.17%) cases followed by Cognitive dysfunction in 8 (13.79%) cases, Seizures in 6 (10.34%) cases, Depression in 4 (6.90%) cases, Anxiety disorder in 2 (3.44%) cases, Acute confusional state in 2 (3.44%) cases, Psychosis in 2 (3.44%) cases, Aseptic meningitis in 2 (3.44%) cases and Polyneuropathy in 1 (1.72%) case.

The disease activity was measured using SLE disease activity index (SLEDAI). We found 4 cases with no disease activity (SLEDAI SCORE = 0), 23 cases with mild Disease activity (SLEDAI score between 1 and 5), 14 cases with moderate disease activity (SLEDAI score between 6 and 10), 10 cases with high Disease activity (SLEDAI score between 11 and 19) and 7 cases with very high disease activity (SLEDAI score ≥20). The mean SLEDAI score in patients with NPSLE was 29.67, whereas it was 16.94 in patients with Non-NPSLE. Table 6 presents a short comparison of our study with other studies.

Table 6: Comparison of our study with other studies

Study features	Present study	Previous studies			
		Eissa M et al.(2018) <sup>6</sup>	Muhammed H et al (2017) <sup>7</sup>	Kakati et al.(2017) <sup>9</sup>	Wei Fan et al (2014) <sup>8</sup>
Number of cases included in the study	58	301	101	52	1772
Mean Age	26.6	30.7	32.3	25.59	28
Sex ratio	10.6:1	6.94:1	10.2:1	12:1	6.6:1
Mean Disease Duration	23.15	72	54	-	12
Nervous system involvement	37(63.79%)	101 (33.5%)	33(32.67%)	19(36.5%)	4.3%
Seizure (%)	6 (10.34%)	65(21.8%)	4 (3.9%)	8(15.4%)	36(47%)
Acute confusional state (%)	2 (3.44%)	-	2 (1.98%)	6(31.57%)	20(26%)
Anxiety	2 (3.44%)	-	5 (4.9%)	2 (3.84%)	2(3%)
Headache (%)	32(55.17%)	167(55.4%)	10 (9.9%)	5(26.3%)	14(18%)
Psychosis (%)	2 (3.44%)	101(33.7%)	0	4(21.05%)	10(13%)
Polyneuropathy (%)	1 (1.72%)	-	12(11.88%)	4(21.05%)	2(3%)
Cognitive dysfunction (%)	8 (13.79%)	-	4(3.9%)	21.2%	9(12%)
<b>SLEDAI</b>					
NPSLE	29.67	-	31.1	32.42	
Non-NPSLE	16.94	-	20.9	17.3	21.53

## CONCLUSION

Neuropsychiatric involvement was found in majority of the patients with SLE and headache was the most common neuropsychiatric manifestation. Patients with NPSLE showed higher disease activity than those without NPSLE.

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