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Review Article

Neuropsychiatric Manifestations in Patients with Systemic Lupus Erythematosus Presenting at a Tertiary Care Teaching Hospital of North India

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Abstract

Background: Systemic Lupus Erythematosus (SLE) is a multisystem autoimmune disease caused by an aberrant immune response.

Aims & Objectives: To study the neuropsychiatric manifestations in patients with Systemic Lupus Erythematosus presenting at a Tertiary Care Teaching Hospital.

Material and Methods: A review of records of SLE patients admitted or evaluated in OPD from March 2010 to June 2017 was carried out.

Results: Over all 85 SLE patients out of 269 which constituted 31.6% of all SLE study group had neuropsychiatric manifestations.

Keywords: Neuropsychiatric Manifestations; Systemic Lupus Erythematosus

Introduction

Systemic Lupus Erythematosus (SLE) is an autoimmune disease that targeted cytoplasm and nucleous of body cells. It causes wide manifestations from the most outer organ to the internal organs. Organ damages may varies, from mild to severe. About 5,000,000 people around the world are affected by SLE. United Kingdom reported the prevalence of 97 cases per 100,000 people in 2012.Signs and symptoms of SLE can be varied. Genetic is predisposition factor which plays role in SLE pathogenesis. Asian's reported more severe clinical manifestations and higher mortality rate compared to the American and European. SLE can be found in all groups age and both male and female, but it is more common found in females during their fertile period [1]. Systemic Lupus Erythematosus (SLE) is a multisystem autoimmune disease of unknown etiology characterized by the production of non-organ specific auto-antibodies directed to nuclear, cytoplasmic and cell surface antigens, which may lead to a wide range of tissue injuries [2].

The involvement of vital organs and tissues such as the brain, blood, and the kidney in most patients, the vast majority of whom are women of childbearing age. The prevalence ranges from 20 to 150 cases per 100,000 population, with the highest prevalence reported in Brazil, and appears to be increasing as the disease is recognized more readily and survival increases. In the United States, people of African, Hispanic, or Asian ancestry, as compared with those of other racial or ethnic groups, tend to have an increased prevalence of SLE and greater involvement of vital organs. The 10-year survival rate is about 70% [3].

Patients with systemic Lupus Erythematous (SLE) that suffer from one or more of several neuropsychiatric symptoms represent

a subcategory termed 'neuropsychiatric lupus' (NPSLE). Cohorts of SLE patients suggest that nearly half will suffer from NPSLE during their disease course. The definition of NPSLE is a tough challenge owing to the broad spectrum of neuropsychiatric symptoms that it encompasses, most of which are non- specific (for example, headache, cognitive dysfunction, etc.). The most accepted effort, so far, to classify NPSLE was made by an American College of Rheumatology (ACR) expert-committee, in 1999 . This committee identified 19 neuropsychiatric conditions, termed 'case definitions', in NPSLE patients, including 12 central nervous system (CNS) and 7 peripheral nervous system ones. It should be noted that a few population-based studies, aimed at validating these ACR-NPSLE case definitions, did not find them to be effective in differentiating NPSLE patients from those with neuropsychiatric manifestations not associated with SLE [4]. The present study was conducted to study the neuropsychiatric manifestations of systemic lupus erythematosus presenting to a tertiary care teaching hospital.

Objectives

To study the Neuropsychiatric manifestations in patients with Systemic Lupus Erythematosus presenting at a Tertiary Care Teaching Hospital.

Material and Methods

a) Study Design: Review of records(retrospective and prospective).

b) Study Duration: Seven Years, 2010-2017

c) Study Setting: A hospital-based study: Rheumatology Division and Medical record section, SKIMS

d) Sample size: All the patients admitted or evaluated during the study period.

e) Criteria for classification of the SLE: American College of Rheumatology (ACR) classification criteria revised in 1982 updated in 1997 and the Systemic Lupus Erythematosus Collaborating Clinic (SLICC) criteria.

f) Statistical Analysis: The data was entered in Ms excel sheets and was analysed by SPSS 23.0.The data was represented in the form of graphs and tables.

Results

A total of 269 patients were studied. Data of patients with systemic lupus erythematosus patients over a period of seven years from March 2010 to July 2017 was collected. Over all 85 SLE patients out of 269 which constituted 31.6% of all SLE study group had neuropsychiatric manifestations (Table 1). In our study, 54(20.1%) had headache, 18(6.7%) had seizures, 17(16.3%) had cerebrovascular disease, 11 (4.1%) had confusion, 9(3.3%) had anxiety disorder, 7(2.6%) and 8(2.9%) had mood disorder and psychosis respectively. Peripheral neuropathy and BIH each was present in 2(0.7%) patients. 5(1.9%) patients had movement disorder (Figure 1).

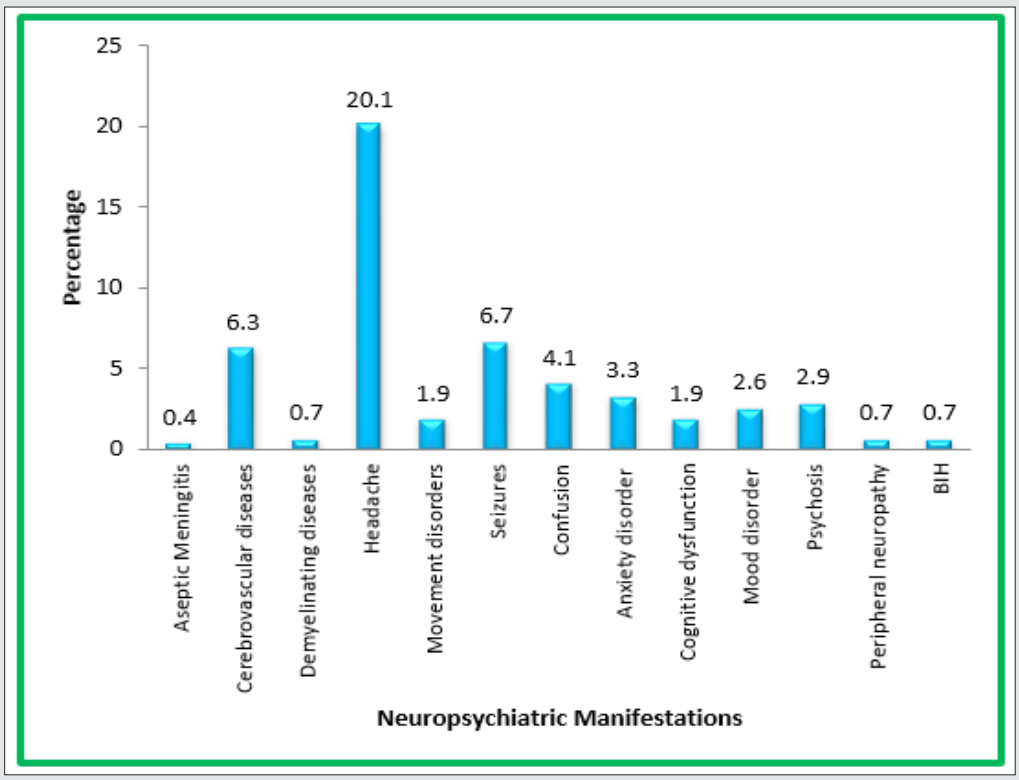

Figure 1: Distribution of Neuropsychiatric Manifestations in Study Patients (CNS and PNS).

Discussion

The onset of Systemic Lupus Erythematosus (SLE) is often marked by seemingly innocent musculoskeletal symptoms, and these may dominate its early course. Unless they are recognized, patients may go on for a long time with some rheumatic diagnosis until an acute febrile episode or visceral or cutaneous symptoms afford clues to the correct diagnosis. Systemic lupus erythematosus is protean in its manifestations and follows a relapsing and remitting course. Cumulative Neuropsychiatric manifestation in our study patients (e.g aseptic meningitis, cerebrovascular diseases, demyelinating diseases, headache, movement disorder, seizures, confusion, anxiety disorder, cognitive dysfunction, mood disorder, psychosis, peripheral neuropathy and benign intracranial hypertension) were present in 31.6% of SLE patients (85/269) which was in agreement with those of American study by Dubois et al, [5-9], Arabia by Al-attia et al, [10-13], and multicentric study in Tunisia by Monia Smiti Khanfir et al [7]. Although the incidence of neuropsychiatric systems were lower as was observed by Malaviya et al, [5], which can be because of ethnic, environmental and socioeconomial variations and because of variation in disease activity in genetically different populations. Estimates of the prevalence of neuropsychiatric lupus (NPSLE) have ranged from 14% to over 80% in adults. A retrospective study of NPSLE in 185 Chinese children over a 20-year period found that 11% had NPSLE manifestations at the time of diagnosis and an additional 16% developed them within one year [14]. Studies in adults using the ACR case definitions collectively have detected the presence of 14-17 of the 19 NPSLE syndromes and reported a fairly consistent prevalence of the following syndromes: total spectrum of headache (39%–61%), seizures (8%– 18%), cerebrovascular disease (2%–8%), psychosis (3%–5%), cranial neuropathy (1.5%–2.1%) [15]. Our results of NPSLE (Table 1) were consistent with the world literature (Table 2).

Table 1: Prevalence of Neuropsychiatric manifestations in SLE patients.

	Frequency	Percentage
Neuropsychiatric manifestations	85	31.5%
Total Cases	269	100.00
Prevalence	31.6%	

Table 2: Neuropsychiatric Manifestations in Study Patients (CNS and PNS). N=85 (31.6%)

Neuropsychiatric Manifestations	Number of patients (n)	Percentage (%)
Aseptic Meningitis	1	0.4
Cerebrovascular diseases*	17	6.3
Demyelinating diseases**	2 0.7	
Headache	54	20.1
Movement disorders	5	1.9
Seizures	18	6.7
Confusion	11	4.1
Anxiety disorder	9	3.3
Cognitive dysfunction	5	1.9
Mood disorder	7	2.6
Psychosis	8	2.9
Peripheral neuropathy	2	0.7
BIH	2	0.7

*Cerebrovascular diseases included are Stroke, PRES and CNS Vasculitis.

**Demyelinating diseases include are Optic Neuritis and Transverse myelitis.

Summary

Systemic Lupus Erythematosus (SLE) is a multisystem autoimmune disease of unknown etiology characterized by the production of non-organ- specific auto-antibodies directed to nuclear, cytoplasmic and cell surface antigens, which may lead to a wide range of tissue injuries. Over all 85 SLE patients out of 269 which constituted 31.6% of all SLE study group had neuropsychiatric manifestations. In our study out of total patients, 54 (20.1%) had headache, 18(6.7%) had seizures, 17(16.3%) had cerebrovascular disease, 11(4.1%) had confusion, 9 (3.3%) had anxiety disorder, 7(2.6%) and 8(2.9%) had mood disorder and psychosis respectively. Peripheral neuropathy and BIH each was present in 2(0.7%) patients. 5(1.9%) patients had movement disorder.

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