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Simultaneous involvement of Brain and Peripheral nerves in a Child with Systemic Lupus Erythematosus (SLE): A Case Report

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Abstract

Systemic lupus erythematosus (SLE) is a chronic autoimmune disorder that affects almost every system in the body. Neuropsychiatric symptoms are common in SLE, but diagnosis often remains difficult. In this case, we report a girl of 16 years who presented with an acute confusional state and features of peripheral neuropathy. A detailed clinical evaluation with a review of the medical history and appropriate laboratory analyses allows this diagnosis to be made. Treatment with immune-modulating therapy, cyclophosphamide resulted in significant improvement of her symptoms. Neuropsychiatric SLE should be considered a potential differential diagnosis for patients presenting with acute confusional state and peripheral neuropathy.

Keywords: Systemic lupus erythematosus, peripheral neuropathy, acute confusional state, NPSLE

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Introduction:

Systemic lupus erythematosus (SLE) is a multi-system connective tissue disorder with variable presentation. Women are affected nine times more frequently than men. The global prevalence is estimated to be 43.7 per 100 000 persons with marked regional variation.[1] Involvement of the nervous system is not uncommon and is associated with a poor prognosis. Neuropsychiatric lupus (NPSLE) includes the neurologic syndromes of the central, peripheral, and autonomic nervous system and the psychiatric syndromes observed in patients with SLE in which other causes have been excluded.[2] These manifestations can precede the onset of lupus or occur at any time during its course. [3,4] These can occur in the setting of active SLE or during quiescent periods, and may present as single or multiple neurologic events in the same individual. The American College of Rheumatology (ACR) established case definitions for 19 central and peripheral nervous system syndromes, some of which are very rare while some may occur in diseases other than SLE.[2] Here we report a case of SLE presenting with simultaneous involvement of brain and peripheral nerves.

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Case Report:

A 16-year-old, right-handed girl presented with the complaints of fever, headache, and vomiting for 10 days, repeated convulsions, and an acute confusional state for 5 days. On admission, she was labeled as a case of viral encephalitis and was treated with antivirals, antibiotics, steroids, and antiseizure medications. On the second day of admission, the patient noticed symmetrical weakness in both lower limbs with subsequent involvement of upper limbs in the next 24 hours. The patient had no history of sensory symptoms, bladder-bowel involvement, dysphagia, dyspnea, fatigability, or diurnal variation of weakness. On query, she gave a history of occasional painful oral ulcer, joint pain, and hair fall for the previous 6 months. There was no family history of such type of illness.

On examination, the patient was moderately anemic with an ulcer on the tip of the tongue and alopecia with normal vitals. Her higher psychic functions including speech, and cranial nerves were intact. Power in the upper limbs was 4/5 while in the lower limbs, it was 3/5 proximally and 2/5 distally. Deep tendon reflexes were absent with equivocal plantar reflexes. Sensory and cerebellar signs were intact. Gait could not be assessed. The left knee joint was tender although there was no joint swelling or deformity. The hemoglobin was found to be 8.1 gm/dl, ESR 80 mm in the first hour, CRP 56.34, S creatinine 1.78 mg/dl. ANA

(on Hep-2 cells) and anti-ds DNA were positive. C3 and C4 were low. Routine urine examination was unremarkable. Magnetic resonance imaging (MRI) of the brain was reported as bitemporal encephalitis based on the findings of multifocal T2WI and FLAIR hyperintensity in both temporal and left frontal cortico-subcortical areas. DWI showed mild patchy increased signal intensity signifying minimal restricted diffusion. After IV Contrast no enhancement was seen (Fig 1). MRI of the spine was normal. Electroencephalography (EEG) revealed bitemporal intermittent rhythmic delta waves on a slow background. The nerve conduction study (NCS) of the right upper and left lower limbs revealed reduced CMAP amplitude in right Median, Ulnar and left Tibial and Peroneal nerves. The findings were compatible with motor axonal polyneuropathy (Table 2). Cerebrospinal fluid (CSF) examination found a normal cell count (04/mm^3) and elevated protein (115.4 mg/dl). Based on the clinical features, laboratory examination results, imaging, and NCS findings a final diagnosis of lupus encephalitis and peripheral neuropathy was made. The patient made a significant recovery with intravenous methylprednisolone and pulse cyclophosphamide.

Table-I

Nineteen neuropsychiatric syndromes (NPSLE) associated with SLE divided into two groups[2]

Central NPSLE	Peripheral NPSLE
Aseptic meningitis	Guillain Barre syndrome
Cerebrovascular disease	Autonomic neuropathy
Demyelinating Syndrome	Mononeuropathy
Headache	Myasthenia gravis
Movement disorder	Cranial neuropathy
Myelopathy	Plexopathy
Seizure disorders	Polyneuropathy
Acute confusional state	
Anxiety disorder	
Cognitive dysfunction	
Mood disorder	
Psychosis	

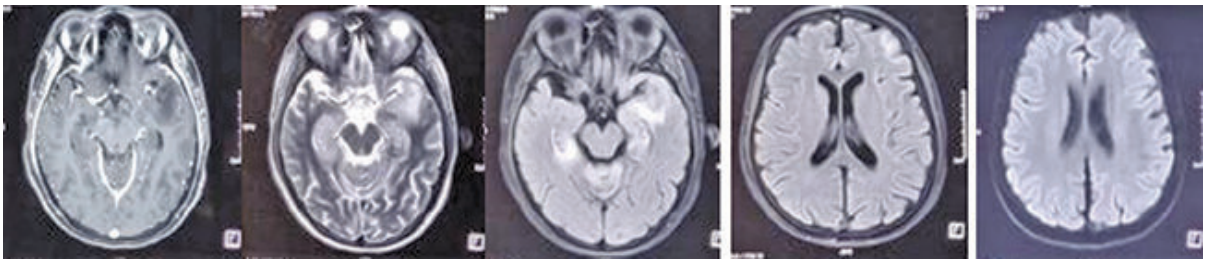

Fig 1: MRI of the brain showing T1 hypo intensity, T2WI, and FLAIR hyperintensity in both temporal and left frontal cortico-subcortical areas

Table-II

NCS findings of Motor nerves of right upper and left lower limbs

Name of nerve	Latency (ms)	Amplitude	NCV (m/s)
Right Median			
Wrist	2.48	3.71 mV	
Elbow	5.8	3.56 mV	66.3
Right Ulnar			
Wrist	2.06	2.88 mV	
Elbow	5.72	2.42 mV	62.8
Left Tibial			
Ankle	3.15	1.78 mV	
Popliteal	10.85	1.34 mV	44.2
Left Peroneal			
Ankle	3.5	240.00 uV	
Head of fibula	9.05	210.00 uV	52.3
Popliteal	10.05	210.00 uV	70.0

Discussion:

NPSLE, most of the time, remains under-diagnosed or there is a delay in the diagnosis because of the rarity of the disease and heterogeneous presentation. Sometimes neuropsychiatric symptoms may be the initial manifestation of SLE or at times, these may manifest during the course of SLE. Psychiatric manifestations include cognitive dysfunction, anxiety, mood and personality disorders, and psychosis.[5,6] The commonest neurological manifestations are headache, stroke, seizure, and peripheral neuropathy.[7-10] The less common ones include cranial neuropathies, transverse myelitis, meningitis, and various movement disorders like chorea, ataxia, choreoathetosis, dystonia, and hemiballismus.[11-13] In our case the diagnosis of SLE was established based on 2019 EULAR/ACR criteria for SLE (with a score of 29 points meeting the minimum requirement of e" 10 points).[14]

Benavente E et al. and also there are many reported cases of SLE, where the patient developed encephalitis during the course of the disease but encephalitis is reported to be rare as the initial manifestation of the disease.[15-18] SLE, presenting as encephalitis is relatively rare and many a time, mimics viral encephalitis in the early period. The studied case presented with features of encephalitis. These are various pathophysiological mechanism which cause encephalitic features. Vasculitis, noninflammatory vasculopathy, antiphospholipid (aPL) antibody syndrome (APS), and autoimmune-inflammatory process have been implicated as the mechanism of cerebral lupus.[19]

GBS as a presenting feature of SLE remains uncommon, with only a few cases reported in the last half-century. Korn-Lubetzki et al. reported a case in 1964 which was the first case of SLE presented as Guillain-Barre Syndrome (GBS).[20] GBS an acute-onset immune-mediated polyneuropathy, is a rare complication of SLE.[21-22] The prevalence of SLE with GBS has been reported to be between 0.6% and 1.7%.[20-23] In our case, it is motor axonal polyneuropathy (i.e AMAN variety of GBS). But in most previous studies, demyelinating subtype was the predominant variety. Although the pathogenesis of GBS in SLE is not clear, both cell-mediated and humoral processes have been implicated.[21]

Simultaneous involvement of the central nervous system (encephalitis) and peripheral nervous system (GBS) in SLE is very rare. To the best of our knowledge, this is one of the rare cases where the involvement of both the central nervous system (cerebral lupus) and peripheral nervous system (GBS) as presenting feature of SLE. NPSLE represents a major cause of morbidity and mortality in SLE patients. Therefore, recognition, early diagnosis and treatment of NPSLE are of great significance.

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Ethical consideration: The study was conducted after approval from the ethical review committee. The confidentiality and anonymity of the study participants were maintained

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