

CNS Lupus Presenting as Acute Psychosis**Dr Nilesh Kumar¹, Dr. Nikhil srivastva², Dr Vijay Parashar³, Dr Ravindu⁴**¹assistant professor, department of General Medicine, IMS, BHU, Varanasi, Uttar Pradesh²Junior resident, IMS, BHU, Varanasi, Uttar Pradesh³Junior resident, IMS, BHU, Varanasi, Uttar Pradesh⁴Junior IMS, BHU, Varanasi, Uttar Pradesh***Corresponding author**

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Abstract: Systemic Lupus Erythematosus (SLE) is a connective-tissue disorder commonly affecting females of reproductive age group. Though any organ can be involved by the immunological attack with musculoskeletal, cutaneous and hematological systems being the most common and comprise the majority of presenting manifestations, neurological features as an initial complaints has also been very rarely reported in the literature. High index of suspicion is required to diagnose such cases as other lupus manifestations are lacking and there is no single laboratory test or radiological features that confirm the diagnosis. CNS Lupus should be included in the provisional diagnosis of a female patient of reproductive age group, who presents with complicated neurological manifestations and with no clear-cut clinical, pathological, or image finding. Psychosis and seizures are the only neurological lupus manifestations that are included in American college of rheumatology (ACR) diagnostic criteria for SLE. The ‘soft’ symptoms of SLE such as cognitive dysfunction and depression, though the most common, are not included.**Keywords:** Systemic Lupus Erythematosus, CNS Lupus.

INTRODUCTION

Lupus cerebritis in the form of psychosis, seizures, parkinsonian like features are rare and more so, as an initial presentation. We present a case in which patient presenting with acute psychosis, mutism and seizures in the absence of offending drugs or metabolic complications was found to be due to lupus cerebritis and was successfully treated over a course of 1 year. The treatment helped a young girl to recover from an initial completely dependent life and lead a near normal life over a period of time.

CASE REPORT

A previously healthy pubertal girl aged 16 yrs presented with low grade non-documented fever and global headache since last 1 month and episodes of laughing /crying and hearing voices since last 3 days. The episodes terminated after the patient suffered 2 episodes of generalized tonic clonic seizures (status epilepticus) followed by, on regaining consciousness, choreiform movements of the extremities and mutism. No similar past-history or drug abuse present.

On examination patient was disoriented to time, place and person, vitals were stable. Neurological examination revealed no meningeal signs and focal neurological deficit. Bilateral plantar were downgoing. There were marked choreiform movements of the left upper and lower limb. Other system examination was

normal. A provisional diagnosis of inflammatory brain disease was kept and patient was started on empirical treatment. Routine investigations were Hb- 8.8g/dl, MCV-86, TLC-5100/ul, Plat-1.48 lac/ul. Renal and liver functions were normal. CT of brain was normal and her CSF picture (total cells-05, L100%, protein-32mg/dl) ruled out any infective pathology. Slit-lamp examination of the cornea also did not find KF-ring. 24 hour urinary copper was also normal. Patient condition deteriorated over next few days and a neuroleptic was added for the symptomatic relief for the psychosis and choreiform movements. Subsequently MRI Brain was done which was also normal. Her autoimmune profile was advised finally which came out to be positive in high titres (ANA - 8.7 IU/ml (<1.0), Anti-dsDNA - >150 IU/ml (<30), Lupus Anticoagulant- positive, IgG ACLA-29.0G/PLU/ml (<10)). In the absence of other etiological factors, patient was diagnosed as isolated CNS lupus and was started on high dose systemic steroids. EEG demonstrated diffuse slow waves suggestive of generalized brain disturbance. However, patient didn't show any improvement after 5 days of steroids. Patient was considered for immunosuppressive treatment and then started on monthly pulse cyclophosphamide therapy. Other conservative measures were continued and first dose of cyclophosphamide (500mg/m² BSA) was administered.

Four months into the pulse Cyclophosphamide(CYC) therapy , patient gradually began to show improvement in her daily activities and communication like asking for food, recognizing family people, showing emotions, going to toilet alone. She showed gradual improvement so that at the end of sixth cycle patient showed marked improvement and return of her functional activities to near-normal . In view of lack of literature, her immunosuppression was continued for an indefinite period and switched after six CYC cycles to azathioprine.

One year from the initial episode, the girl now studies in 8th standard with a below average performance in an elementary school with MMSE on neurological examination found to be 26.

DISCUSSION

CNS is affected in about 30-50% of SLE patients but isolated involvement of CNS without any evidence of systemic lupus activity (also called as CNS lupus, lupus cerebritis) is extremely rare [1]. Prompt identification of CNS Lupus is extremely difficult and challenging. There is no definitive laboratory or radiological test to confirm a possible diagnosis. Assessment of the clinical features and absence of other etiological agents along with presence of antibodies in the serum and/ or CSF are necessary to conclude diagnosis.

ACR includes 19 different forms of neuropsychiatric involvement in SLE but only two has been used for the diagnostic criterion- seizures and psychosis. The most common neuropsychiatric manifestations in SLE are: cognitive deficit (memory disturbance), headache, psychoses, seizures, and cerebrovascular events [2].

Loss of the normal control mechanism of the immune system is the basic pathological response in SLE, resulting in loss of inhibition on body's autoimmune response. Circulating auto-antibodies are formed due to the recruitment of B-lymphocytes by these immune complexes. There are basic five mechanisms underlying CNS manifestations in SLE: Ischemia , hemorrhage , white matter damage, neuronal dysfunctions induced by antibodies and deficient psychological reactions[3]. Ischemia, the most prominent mechanism, manifests as cerebrovascular events, strongly associated with the presence of antiphospholipid antibodies but also occurring as a consequence of premature atherosclerosis, vasculitis,

vessel spasm, small vessel angiopathy, emboli etc. Hemorrhage, in the form of intracerebral, epidural, subdural are found in increased incidence in SLE. White matter damage in the form of demyelinating plaques in brain and brainstem, white matter degeneration in optic nerves and spinal cord and leuco-encephalopathy. Some NPSLE manifestations like psychosis, cognitive disorder, chorea and other movement disorders might be explained by antibody induced neuronal dysfunction. Mood disorders are more likely a reaction to the disease process and constitute deficient psychological reactions.

Serum antibody levels are highly valuable in the diagnosis of CNS lupus, more so when there is an isolated CNS involvement as the diagnostic criteria by ACR would less likely be fulfilled. The antiphospholipid antibodies, lupus-anticoagulant antibodies (i.e., IgG, IgA, IgM), antineuronal antibodies, brain-lymphocyte cross-reactive antibodies, anti-ribosomal P antibodies, commonly detected in the serum of such patients [4]. Complement components (C3 and C4) of a coagulation cascade show low serum and CSF concentrations. Determination of an immunological marker in the CSF is more specific of CNS involvement than that of the serum [1] .

Treatment involves steroids and immunosuppressive agents like cyclophosphamide and azathioprine. Symptomatic management should be used for neurological symptoms like chorea, psychosis (with neuroleptics) or parkinsonian symptoms like rigidity (with dopamine agonists). Seizures should be treated and AED continued till the disease activity gets controlled. Anti-thrombotic therapy should be considered in the presence of high APL titres or positive LA test in those who presents with seizures or demonstrate ischemia on MRI brain.

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