CASE REPORT

Neuropsychiatric Presentation of Systemic Lupus Erythematosus—When to Attribute: A Case Report

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Abstract

Systemic lupus erythematosus (SLE) is an autoimmune disease with the involvement of various systems. Neuropsychiatric symptoms of SLE (NPSLE) are nonspecific. The attribution of NP symptoms is difficult. We report a case with NP symptoms followed up for more than 20 years which tested positive for SLE.

Introduction: SLE is a chronic autoimmune disease that is known to affect different organs in the body.[1] The diagnostic criteria of the disease have evolved over time, and the prevalence of the illness has also gradually increased. Nervous system involvement is common and can manifest as neurological or NP symptoms. The American College of Rheumatology has set definitions for 19 NP syndromes [12 central nervous system (CNS) and 7 peripheral nervous system (PNS)] deemed to occur in SLE, which is widely used in clinical practice and for research.[1] These NP syndromes are classified into frequent, common, infrequent, and rare based on the frequency of the symptoms.[2] None of the NP syndromes is specific for SLE; however, one-third of the NP syndromes are related to SLE autoimmunity.[2] Determination and attribution of NP syndromes to SLE in patients is a challenging but critical step in treatment.[2] Among psychiatric syndromes, mood disorder and anxiety have been found to be frequent compared with psychosis, which is infrequent.[3]

Keywords: NP syndromes, Psychosis, SLE.

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Case Description

M, 54-years-old, married female, homemaker, educated up to 5th standard, belonging to lower socioeconomic status from urban Bengaluru, was brought by her son with a past history of acute psychotic episode of 3 days in 2000 for which conservative management was done and depressive episode with psychotic symptoms in year 2002 for which antipsychotic and antidepressant medication were started with significant improvement. There was relapse of symptoms in 2003 because of poor adherence to medication. The depressive symptoms improved however, and the patient had complaints of pulling sensation in the head, giddiness and imbalance in gait, and blurring of vision. She lost the follow-up and stopped taking medications until 2009, when she presented with a relapse of depressive symptoms and medication were restarted and improvement was seen. She had a manic episode in 2010, when diagnosis was revised to bipolar disorder and mood stabilizer lithium was started along with antipsychotics. Later in 2010, the patient developed pain in the lower limbs and swelling and numbness of both hands along with stiffness of joint. A diagnosis of early rheumatoid and Raynaud phenomenon was made by the treating physician and medication was started. She continued to have leg pain and joint pain of hands with poor improvement with medications. She had a mixed mood episode in 2013 because of poor compliance with medication, which improved with treatment. However, her joint pain worsened over time and was diagnosed as mixed connective tissue disorder in 2015. She continued her treatment for medical illness and lost the psychiatric follow-up. In 2018 and 2020, the patient had a relapse with mixed episode, which was managed with restarting and titration of early medicines. Currently, she presented with severe body pain for 20 days and drowsiness along with dysarthria and hemiparesis for one day. She was thoroughly investigated during her stay in hospital, which revealed strongly positive titres for anti-ds DNA, Nrnp/Sm, and SS-A/Ro-52, and was positive for Sm/dsDNA/nucleosomes/histone. Magnetic resonance imaging of the brain revealed an acute infarct on the right side of the mid pons and subacute infarct on the left side of the upper pons along with gliosis of pons. Routine blood investigation was normal. So a primary diagnosis of cerebrovascular accident and SLE associated with neuropsychiatric syndrome was made. The patient was managed conservatively with significant improvement.

Discussion

SLE has a diagnostic classification according to SLICC and EULAR/ACR criteria published in 2012 and 2019, respectively.[4] NPSLE is common but is a nonspecific presentation of SLE. NPSLE is associated with morbidity, mortality, and poor quality of life of the patients. In the

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absence of any lab investigation and radiological finding correlating NP syndromes of SLE, attribution is mostly guided by the expert opinion based on assessment of symptoms, regular follow-up and clinical experience. The management guidelines for most NP syndromes are lacking in view of the absence of randomized trials on the topic. The indexed case has been followed up for more than 20 years. The contact presentation in this case had been mood symptoms which gradually evolved into mood associated with psychotic symptoms and then mono/polyneuropathy. Gradually, the presentation involved the CNS (stroke) which prompted for autoimmune work-up revealing strongly positive results for SLE. Poor response to medication, loss of follow-up, and suicidality have been associated with NP syndromes of SLE and have been positive in this case. Furthermore, autoimmune treatment supplemented the NP syndromal conservative management done earlier. The frequent NP syndromes such as mood disorder, anxiety, and headache were present along with the infrequent symptoms such as mono/polyneuropathy and psychosis initially and later had presentation with a common syndrome like cerebrovascular stroke. The current case gives an insight that neurological symptoms not related to general psychiatric mood symptoms or psychosis that do not respond to regular prescribed medication should be searched for general medical etiology and a liaison team can be involved. Moreover, the investigation protocol should be exhaustive if recovery is not attained with appropriate protocol medication.

Conclusion

The present case gives a valuable insight that NP syndromes were present as initial symptoms which varied as the disease progressed. The core SLE symptoms were absent. However, the investigation results revealed strongly positive for SLE. Therefore, if autoimmune illness had been diagnosed early, the severe presentation like stroke could have been prevented.

Future Direction

SLE is a multisystemic disease. However, neuropsychiatric research is at its infancy as diagnosis or management is concerned. Attribution of NP syndromes to SLE is an area for further detailed investigation.

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