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Case report

Catatonía – An unusual presenting clinical manifestation of systemic lupus erythematosus



Debasish Chaudhury^{a,*}, Arshna Qureshi^a, Shiv Prasad^b, Ravi Kumar Meena^a, Simran Sharma^a

^a Department of Medicine, Lady Hardinge Medical College (LHMC) and Associated Hospitals, New Delhi, India

^b Department of Psychiatry, Lady Hardinge Medical College (LHMC) and Associated Hospitals, New Delhi, India

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ABSTRACT

A 24-year-old female presented with catatonía and symptoms suggestive of Depressive Disorder. She also gave history of undocumented low grade irregular fever. The patient was worked up to rule out any organic cause or psychiatric illness. However, further investigations revealed immunological profile diagnostic of Systemic Lupus Erythematosus (SLE) with CNS involvement (CNS lupus). The diagnosis of SLE in this patient presenting with catatonía was of practical importance because catatonía as one of the manifestations of SLE or as a standalone presenting symptom is extremely rare. Hence, clinicians should be aware of this rarity so that diagnosis of Neuropsychiatric SLE (NPSLE) or catatonía as a presenting feature of SLE is never missed.

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Catatonía-manifestación clínica de presentación infrecuente del lupus eritematoso sistémico

RESUMEN

Mujer de 24 años que se presentó con catatonía y síntomas indicativos de trastorno depresivo. También presentó historia de febrícula discontinua no registrada. Se llevó a cabo evaluación diagnóstica para descartar cualquier causa orgánica o enfermedad psiquiátrica. Sin embargo, exploraciones complementarias posteriores revelaron un perfil inmunológico diagnóstico de lupus eritematoso sistémico (LES) con implicación del SNC (lupus del SNC). El diagnóstico de LES en esta paciente que se presenta con catatonía era de significado práctico ya que la catatonía, como una de las manifestaciones del LES o como un síntoma que se presenta de forma independiente, es extremadamente rara. Por ello, los médicos deben ser conscientes de esta rareza y no deben olvidar nunca el diagnóstico de LES neuropsiquiátrico (LESNP) o catatonía como rasgo presente en el LES.

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Introduction

SLE is a chronic autoimmune disease of unknown etiology, typically diagnosed on the basis of the SLICC criteria.¹ Its clinical presentation is variable with arthralgia, photosensitivity and rash being the most common symptoms. Although Central Nervous System (CNS) involvement in SLE varies from 14% to over 80%,^{2,3} initial

presentation with neurological features is considered rare, seen only in approximately 3% of the patients.⁴ In the absence of typical manifestations, patients presenting solely with neuropsychiatric symptoms of SLE can pose a diagnostic dilemma to the clinician. We report a case of catatonía as the presenting manifestation of SLE in a young female.

Case report

A 24-year-old young female patient presented with the complaints of behavioral abnormalities like decreased oral intake,

* Corresponding author.

E-mail address: debasishccc@yahoo.com (D. Chaudhury).

speech output and interaction with family members, inability to recognize relatives or communicate via eye contact, immobility, abnormal posturing, vacant staring, grimacing, repetitive utterances, poor self-care, loss of bowel and bladder control and low grade undocumented fever since last one month.

General physical examination revealed disoriented patient with stable vitals. CNS examination demonstrated decreased speech output, catatonic posturing, waxy flexibility but no focal neurological sign. Examination of other systems was unremarkable. The patient did not have any typical manifestations of SLE like photosensitivity, arthralgia, arthritis and malar rash. Fundus and slit lamp examinations were also normal.

Psychiatric referral suggested a diagnosis of organic brain syndrome presenting as catatonia and depression. Routine and special laboratory investigations were carried out to rule out secondary causes of catatonia and psychiatric conditions. All laboratory tests including hemogram, coagulation, biochemical and thyroid profile, liver and kidney function tests, urine routine microscopy, culture sensitivity and 24 h urinary protein were found to be within normal limits. Peripheral blood smear and rapid kit test for malarial parasite (MP), direct and indirect Coomb's tests, viral markers for hepatitis B and C virus were all negative. VDRL test was non-reactive. Immunological profile was carried out for antinuclear antibodies (ANA) by indirect immunofluorescence on Hep 2 cells which showed diffuse positivity (2+) at a dilution of 1:320, positive anti-dsDNA antibodies with a titer of 52 IU/ml (positive, >25 IU/ml) by ELISA. Nuclear antigen line assay results were also positive for dsDNA. Anti-neutrophil cytoplasmic and perinuclear antibodies (c-ANCA and p-ANCA) were detected with a titer of 8 RU/ml and 2 RU/ml by ELISA (positive ≥ 20 RU/ml, negative <20 RU/ml). Cerebrospinal fluid (CSF) analysis revealed protein, glucose and cell count of 42 mg/dl, 56 mg/dl, 15 cells (all lymphocytes) per cubic mm, respectively, suggestive of lymphocytic pleocytosis with mildly elevated protein. Culture sensitivity of CSF was unremarkable. Lymphocytic pleocytosis and mildly raised protein levels with similar clinical features can be observed in viral, fungal, mycobacterial and paraneoplastic etiologies. In our patient tubercular meningitis was ruled out due to normal CSF levels of adenosine deaminase, negative PCR results for mycobacterium tuberculosis, and negative staining for acid fast bacillus. Absence of cryptococcal antigen in CSF also ruled out common fungal infection involving nervous system. Total IgG and IgG index in the CSF were also within normal limits. IgM and IgG levels specific for herpes virus were not detectable, absence of IgG anti-N-methyl-D-aspartate (NMDA) receptor antibodies in our patient helped to rule out NMDA encephalitis subsequent to any possible malignancy. Anti-phospholipid and anti-cardiolipin antibodies were negative, thus ruling out their contribution to development of cerebral vascular thrombotic events and consequent neurologic presentations. Serological tests for HIV antibody and western blot were non-reactive and it ruled out HIV associated neurocognitive (HAND) syndrome⁴ as an explanation for neurobehavioural manifestations associated with CSF pleocytosis. Neuroimaging using contrast MRI was normal. Ultrasound abdomen and pelvis was carried out to rule out any malignancy that can present with neuropsychiatric manifestations as part of paraneoplastic encephalitis. EEG findings were normal in our patient.

Thus, a final diagnosis of catatonia secondary to neuropsychiatric SLE (NPSLE) was made due to the presence of typical laboratory profile and by excluding other causes of catatonia. The patient was administered intravenous methylprednisolone for 3 days followed by oral prednisolone 40 mg daily. Following treatment, the patient showed marked improvement after 2 weeks and was later discharged with an advice to follow up in Medical outpatient clinic.

Discussion

NPSLE can present with various neurologic and psychiatric manifestations. The etiology behind NPSLE is postulated to be multifactorial involving up-regulation of the expression of adhesion proteins on endothelial cells due to production of proinflammatory cytokines and autoantibodies, facilitating lymphocyte entry into the central nervous system.³ Cataonia is defined by DSM-V⁵ and as a presentation of NPSLE has been described very rarely in literature; however, it is not clearly understood why it is rare. Case reports with such presentation have been documented sparsely.^{6,7} Presence of catatonia in any patient raises several diagnostic possibilities which can be psychiatric, neurological, substance abuse or medical illnesses.⁸ The diagnosis of NPSLE remains largely one of exclusion and can be made only after these secondary causes have been excluded. So, our patient presented with fever, abnormal neurologic manifestations like altered sensorium and behavioral manifestations with the presence of lymphocytic pleocytosis and mildly elevated protein in CSF raised several differentials including viral meningo-encephalitis, tubercular meningitis, fungal infection of brain, para-neoplastic encephalitis, etc. But the absence of any preceding history of infection, trauma, drug or substance abuse and personal or family history of similar catatonic illness as a part of psychiatric disorder helped us to rule out any primary psychiatric etiology. Similarly absence of clinical features suggestive of herpes, malignancy or primary neurologic illness with normal neuroimaging and electro-encephalographic studies excluded other causes of catatonia. Moreover absence of antibodies to viruses in CSF, anti-NMDA receptor antibody, antibodies to HIV and no metabolic abnormality also helped us to rule out the secondary causes of catatonia. Presence of specific autoantibodies - ANA and anti dsDNA in significant titers and exclusion of other causes of catatonia with CSF pleocytosis. She was diagnosed as a case of SLE with catatonia being the presenting manifestation. This patient had two atypicalities. First, she presented with only CNS lupus (neuropsychiatric manifestation) without any typical feature of SLE and second, out of all the described neuropsychiatric manifestations of SLE in literature she presented with catatonia which is an extremely rare kind of manifestation among all other presentations of NPSLE.

Conclusion

This case report gives a strong insight that patients of SLE like ours may not have any of the typical features of SLE and present with highly unusual neuropsychiatric symptoms like catatonia. This can be misleading to the clinician leading to erroneous diagnosis of a condition that is amenable to treatment. Catatonia with SLE is completely reversible with steroid and/or cytotoxic drug therapy. Hence it is highly essential to differentiate catatonia as a result of NPSLE from other causes of catatonia so that appropriate therapeutic intervention can be undertaken.

Ethical disclosures

Protection of human and animal subjects. The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors must have obtained the informed consent of the patients and/or subjects mentioned in the article. The author for correspondence must be in possession of this document.

Conflict of interest

The authors declare that they had no conflicts of interest.

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