

Anti-NMDA Receptor Encephalitis in a Young Girl with Altered Behaviour and Abnormal Movements

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ABSTRACT

Autoimmune encephalitis is a rare central nervous system disorder in which the patient presents with neuropsychiatric symptoms. We herein present a case of anti-NMDA receptor encephalitis in which the patient initially presented in the psychiatric department. A 14-year girl was brought with complains of irritability, altered behaviour, abnormal movements, self biting, and decreased sleep for the past 10 days. Her condition deteriorated during the admission and she became mute, immobile, and drowsy. Her baseline investigations and CT scan brain were normal. CPK was high and the CSF showed pleocytosis. Autoimmune encephalitis profile showed presence of antibodies against the NMDA receptors. Improvement in the symptoms was noted after treatment with steroids and plasmapheresis. This case report highlights the importance of multidisciplinary approach involving the neurologist, psychiatrist, and immunologist in accurately diagnosing and managing a rare neurological disorder presenting mainly with psychiatric symptoms.

Key Words: *Anti-N-Methyl-D-Aspartate receptor encephalitis. Altered behaviour. Young girl. Psychiatric symptoms. Auto-immune encephalitis. CSF pleocytosis.*

INTRODUCTION

The incidence of autoimmune encephalitis is on the rise due to improvement in diagnostic facilities.¹ Anti-NMDA receptor encephalitis is the commonest autoimmune encephalitis and has been associated with ovarian teratoma and other malignancies.² Hallucinations, behavioural problems, memory loss, psychosis, and personality changes are the earliest symptoms with which most of the patients present, so psychiatric team is approached first in majority of the cases.³

Early recognition and treatment of this disease has been associated with good prognosis and protection from permanent brain damage.⁴ We present a case of anti-NMDA receptor encephalitis in a young female who presented with behavioural problems and abnormal movements in the psychiatry OPD; and combined neuropsychiatry approach led towards an accurate diagnosis and prompt treatment.

CASE REPORT

A 14-year girl from Lahore was brought by her parents with complains of abnormal movements, irritability, altered behaviour, decreased sleep, and continuous biting of the lower lip for the past 10 days. They consulted the local GP at Lahore and also visited the church priest for prayer but no improvement occurred;

so they brought the patient to the psychiatry department. History and mental state examination did not reveal any psychosocial stressor. Her clinical picture did not fit in any psychiatric diagnosis, so neurology department was involved to look for any organic cause of her illness. Meanwhile, her condition worsened. She became mute, immobile, and started voiding the urine in front of everybody in the room. Tone was increased and abnormal movements consisting of catatonic movements with stereotypies became more frequent. Her lip biting became so severe that her lower lip swelled. The patient's family did not report any unusual behaviours or difficulties in daily functioning prior to the onset of current episode of illness. There was no history of unconsciousness, fits, fever, headache, vomiting, visual disturbances, head injury or illicit drug use.

Physicians from the neurology department examined her in detail and ordered her baseline laboratory examinations, CT scan, EEG and the CSF routine examination. Her baseline investigations were within normal limits except a very high CPK. CT scan brain was normal, EEG showed diffuse slowing and the CSF examination showed pleocytosis. She was suspected as a case of viral encephalitis initially and was put on acyclovir. PCR for *Herpes simplex* virus DNA was sent. Her condition did not improve with the current medication and PCR turned out to be negative. This raised the suspicion of autoimmune encephalitis. Immunology department of a private hospital was involved and profile for autoimmune encephalitis was performed. MRI brain was also ordered which turned out to be normal and anti-NMDA receptor antibodies were present on the autoimmune encephalitis profile.

Acyclovir was stopped and she was put on high pulse methylprednisolone 1000 mg daily for five days.

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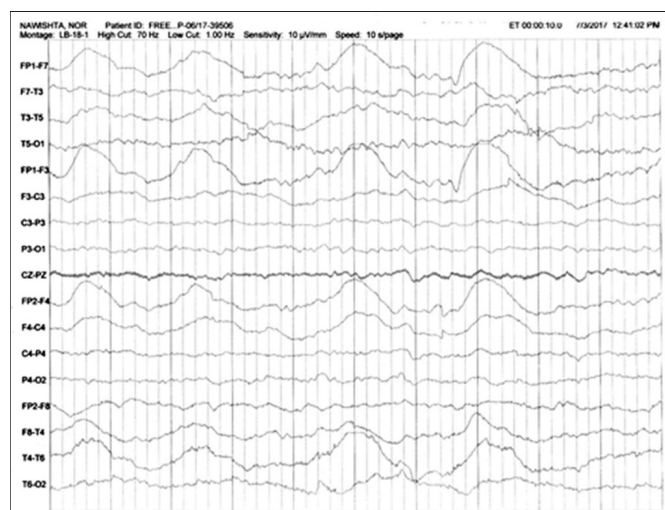


Figure 1: EEG showing diffuse slowing

Bromocriptine and amantadine were given to cater for the rigidity. Sessions of plasmapheresis were started soon after the steroid course. After two weeks, she showed improvement in her symptoms. Her abnormal movements reduced. Her speech and cognition also improved and behavioural problems settled, too. Five sessions of plasmapheresis were done.

Due to association of this disease with malignancies, her CT scan neck, chest, abdomen and pelvis was performed along with the tumor markers. CT scan did not show the presence of any neoplastic lesion and tumor markers were also negative.

The final diagnosis was primary anti-NMDA receptor encephalitis. After the appropriate treatment, all the neuropsychiatric symptoms settled to an extent that she was discharged after five sessions of plasmapheresis. Follow-up visit was planned after one week to decide for the requirement of any further immunosuppressive treatment.

DISCUSSION

Patients suffering from autoimmune encephalitis usually present with irritability, abnormal movements and other neuropsychiatric symptoms, making it difficult to differentiate them from primary psychiatric patients.⁵ This patient had similar presentation, as reported in the few case reports involving the local as well as foreign patients.^{5,6}

Psychiatric manifestations have been reported among patients suffering from various neurological disorders including dementia, Parkinson disease, encephalitis, multiple sclerosis, epilepsy and many others.⁷ However, in most cases the neurological symptoms appear first or distinct pattern of disease does not allow the psychiatric

symptoms to mask the original disease; but in the case of anti-NMDA receptor encephalitis, neurological symptoms are not distinct enough to reach the diagnosis at the early phase of presentation.

This case presented a diagnostic challenge given the limitations of the psychiatric team to diagnose such a disorder and normal appearance of the brain on basic neuro-radiology. Presence of anti-NMDA receptor antibodies in CSF or serum has high sensitivity and specificity in the diagnosis of anti-NMDA receptor encephalitis.⁸ A similar case was reported from this city two years ago, but that was from a private hospital. At that time, autoimmune screening for encephalitis was not available in our country so they got it done from the UK. The patient could afford, so IVIG was preferred as the treatment instead of plasmapheresis.⁶

The diagnosis of anti-NMDA receptor encephalitis rests on the clinical criteria published in the *Lancet Neurology*.⁹ Detailed investigation by the neuropsychiatric team and the multidisciplinary approach involving the neurologist, psychiatrist, and immunologist served as a key to make this difficult diagnosis in time and saved the patient from prolonged misery.

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