CASE REPORT

Anti-N-methyl-D-aspartate Receptor Antibody Encephalitis: A Psychiatric Presentation

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Abstract

Anti-N-Methyl D-aspartate receptor (anti-NMDA-R) encephalitis is an underdiagnosed autoimmune condition that can present with "flu" like illness that progresses through stages of psychotic symptoms and movement disorders that further deteriorates to autonomic instability and even death if not treated promptly. The case condition often presents to psychiatrists who may find the condition not responding to standard treatments posing significant diagnostic challenges. Often it can present as a paraneoplastic syndrome, but recent data shows the occurrence of idiopathic presentations. Here we present of a 40-year-old male with one such typical clinical presentation of psychotic symptoms along with movement abnormalities and progression to further neurological deterioration who after systemic evaluation was diagnosed to be a case of anti-NMDA-R encephalitis. He responded well to IVIG and plasma exchange. There is a need to recognize this condition more vigilantly in practice and the factors favoring prognosis and outcomes are further discussed.

Keywords : Acute psychosis, Anti-NMDA receptor antibody encephalitis, Catatonia.

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Introduction

Anti-N-methyl-D-aspartate receptor (anti-NMDA-R) encephalitis was first characterized in 2007 by Dalmau et al.[1] It is defined as a paraneoplastic syndrome affecting young women with ovarian teratomas.[2] It is also occurred in association with mediastinal teratomas, sex cord-stromal tumors, small cell lung cancer, and testicular teratomas.[3,4] It is common among females with the ratio of male:female is 4:1. The clinical presentation involves multistage process. It starts with "flu"-like illness followed by psychotic symptoms. Next stage presents with movement disorders including dystonia, chorea, rigidity, and catatonia. The syndrome classically has autonomic instability (cardiac dysrhythmia, hyperthermia, unstable blood pressure, hyperhidrosis, and sialorrhea), reduced conscious levels, and can even lead to death. Anti-N-methyl-D-aspartate receptor Ab encephalitis has good response to treatment if diagnosed and treated promptly.[5]

This case report describes the psychiatric presentation of anti-NMDA-R encephalitis, in a male without any paraneoplastic syndrome.

Case Report

A 40-year-old male psychologist living alone abroad had fever with ear pain and headache for 4 days. A week later, he stopped going to work and was spending most of his time watching movies and listening to music. At times, he was noticed to be talking to self. He was not sleeping, his food intake was reduced, and he was not interacting with his family through social network like before. Three weeks later, he had decided to come to India. In the flight, he did not speak to anyone and kept staring ahead. At the airport, he could identify his brother and finish all emigration procedures on his own, and from there he was bought to the hospital. He consumed alcohol occasionally.

On examination, he had fluctuating febrile spikes. Blood pressure and pulse rate were also fluctuating. Gait was broad based, had tremors and cogwheel rigidity, plantar responses were extensor on right side, mild neck stiffness and myoclonic jerks were present.

On Serial Mental Status Examination

The patient had a staring look, did not answer to the questions asked, and was smiling inappropriately at times. Initially he was walking with difficulty, was oriented, and had intact memory. He had persecutory delusion. Initial provisional diagnosis of organic psychosis was made and neurologic evaluation was obtained for his atypical presentation.

Three to four days later, he had fever spikes with autonomic instability and gradually his cognitive and neurological status deteriorated significantly. He was transferred to neurology ward. He was disoriented to time, place, and person; became bedridden; progressively stopped taking diet; and became incontinent of bowel and bladder. Mental status waxed and waned. At times, he spoke, identified family members, and at other times he remained mute.

He began to develop symptoms consistent with worsening catatonia, waxy flexibility, posturing, active negativism, and blank staring. He received lorazepam injection 1 mg IV for catatonia, he became drowsier, and no response was noted, hence stopped.

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Routine blood investigations were normal. Other investigations including serum electrolytes; cerebrospinal fluid (CSF) analysis; CSF cytology; and adenosine deaminase (ADA), vitamin B12, and folate levels were within normal limits. Thyroid profile, liver function test, computed tomography (CT) and magnetic resonance imaging (MRI) brain were normal. Trop-T and procalcitonin came as negative. Urine toxicology and toxin profiles were within normal limits. Full body positron emission tomography (PET) scan is negative for malignancy. Less significant results found were Measles IgG-148 (? Significance) and ultrasonography (USG) abdomen showed healed granuloma in liver.

He was also empirically started on acyclovir injection to cover for presumed viral encephalitis of unknown etiology and T. doxycyclin as anti-parasitic. He developed cough, difficulty in breathing, and was shifted to medical intensive care unit (MICU) in view of aspiration pneumonia, intubated to protect airways.

Patient's serum and CSF samples were sent for autoimmune panel. Eight weeks later his CSF was found to be positive for anti-NMDA antibodies, and he was administered IVIG for autoimmune disorder. Plasma exchange was done. Patient gradually showed improvement and significant improvement noticed in 3 months.

Discussion

This patient presented with psychiatric presentation, with the idiopathic cause. Though it is a serious disorder, its prognosis is good with early treatment. Static or persistently elevated levels of antibodies in CSF may signify a recalcitrant type of N-methyl-D-aspartate receptor encephalitis (NMDARE), which will require aggressive immunotherapy.[6] Generally, recovery is more favorable in patients with an underlying tumor than in the idiopathic cases. Some idiopathic NMDARE may take up to 3 years for complete recovery.[7]

Conclusion

Though NMDA-receptor antibody encephalitis is a rare disorder, psychiatrist should be aware of this condition, its complex presentation, and it requires a multidisciplinary approach. Its early diagnosis and treatment gives a good prognosis.

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