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A Unique Presentation of Excitatory Catatonia Alongside Anti-Nmda Receptor Encephalitis: A Case Report

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ABSTRACT

Introduction: Anti-NMDA receptor encephalitis, first described in 2007, is a prominent cause of autoimmune encephalitis, predominantly affecting females. Its clinical presentation includes a prodromal phase with flu-like symptoms, followed by psychosis, unresponsiveness, hyperkinesis, and recovery [1]. Catatonia can coexist sounds better than cooccur in patients with anti-NMDA receptor encephalitis, complicating diagnosis and effective treatment.

Case Summary: This case report discusses a 41-year-old African American female with a history of seizure disorder, schizoaffective disorder with catatonia, developmental delay, Catatonia in itself is not the diagnosis as per dsm 5. We could use it as a specifier with other diagnosis, be it schizoaffective disorder or seizure disorder here. So we could mention as schizoaffective disorder with catatonia. and recent bereavement. Her symptoms included variable motor hyperactivity, alternating between restlessness and sedation, with an initial Busch Francis Rating Scale score of 18. Initially diagnosed with excitatory catatonia secondary to schizoaffective disorder and treated with benzodiazepines, her condition failed to improve, prompting further investigation. Despite additional treatment for a urinary tract infection and medication adjustment, which included adding valproic acid and amantadine, her condition remained uncontrolled. Subsequent lumbar puncture revealed inflammatory markers, and her anti-NMDA receptor IgG antibody assay was positive at 1:1280, confirming the diagnosis of anti-NMDA receptor encephalitis. Treatment was escalated to include methylprednisolone and intravenous immunoglobulin, alongside six sessions of ECT for management of her catatonia. The patient showed significant improvement, achieving functional recovery and independence in daily activities before discharge.

Conclusion: This case emphasizes the importance of considering anti-NMDA receptor encephalitis in patients with unexplained, refractory neuropsychiatric symptoms, such as catatonia, and highlights the value of a multidisciplinary approach in managing and rehabilitating affected individuals. Prompt diagnosis and treatment are crucial to improving outcomes and reducing the risk of persistent cognitive deficits.

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Introduction

Anti-NMDA receptor (NMDAR) encephalitis, officially identified in 2007, has become increasingly recognized as a major cause of autoimmune and paraneoplastic encephalitis, much more commonly presented in females [1]. Catatonia has been increasingly presenting within the context of anti-NMDA receptor encephalitis, and frequently mistaken as part of another psychiatric process, delaying proper diagnosis and treatment [2]. While anti-NMDA receptor encephalitis is not a common disease, according to the California Encephalitis Project, it has become the most common cause of encephalitis compared to any viral encephalitis [3].

Typically, anti-NMDA receptor encephalitis presents in five phases : prodromal phase, psychotic phase, unresponsive phase, hyperkinetic phase, and recovery phase [4]. During the prodromal phase, an individual may experience viral upper respiratory infection symptoms. Patients may then enter the psychotic phase, characterized by psychosis, positive or negative symptoms, depression, mania, and other psychiatric symptoms. Following this phase, there is the unresponsive phase characterized by mutism or catatonia. Next, patients enter the hyperkinetic phase where they experience autonomic instability and movement disorder. Finally, patients enter the recovery phase after receiving months of treatment [4].

Treatment for anti-NMDA receptor encephalitis involves a course of early immunotherapy, typically a course of glucocorticoids, intravenous immunoglobulins (IVIG), and plasma exchange (PLEX) either concurrently or sequentially, as well as removal of any concurrent neoplasm [5]. Despite treatment, cognitive deficits can persist for up to years following treatment and are influenced by several factors, including delayed treatment, older age, and higher disease severity [4]. Electroconvulsive therapy has been noted as an adjunctive therapy in anti-NMDA receptor encephalitis

notably when the initial presentation includes catatonia with favorable outcomes [6]. We intend to present an interesting case where the patient, who initially presented with excitatory catatonia and had risk factors which indicated alternative diagnoses, was ultimately diagnosed with anti-NMDA receptor encephalitis and treated with early immunotherapy and ECT.

Clinical Vignette

The patient is a 41-year-old African American female with a history of seizure disorder from a left ventricle structural cause, schizoaffective disorder with catatonia (8 years ago), and developmental delay, who recently suffered the loss of her son three weeks prior. She was hospitalized for the past few weeks at three state psychiatric hospitals before admission to Wellstar MCG Health hospital, where she was diagnosed with excitatory catatonia secondary to schizoaffective disorder. On intake, she was hyperactive, non-verbal though non-violent, and had episodes which alternated between sprinting about the unit and self-harming (banging head on the wall, broke glass window with head). At the hospital, her urinalysis was indicative of a UTI, for which she received treatment, though she continued to display bizarre behavior. She had a Busch Francis Rating Scale (BFCS) of 18 (3 Excitement, 3 Mutism, 3 Grimacing, 3 Impulsivity, 3 Combativeness, 2 Autonomic Abnormality) which was dropped to 11 (3 Immobility, 3 Mutism, 3 Staring, 2 Rigidity) following Midazolam challenge.

The patient failed initial medical management with benzodiazepines and continued to exhibit hyperactivity. Thus, she was restrained, sedated with Dexmedetomidine, and transferred to the Medical Intensive Care Unit (MICU). Olanzapine, valproate acid and amantadine were added to her regimen with no notable change to her behaviors. A thorough investigation for an organic cause of her condition was launched. Her diagnostics resulted in an unremarkable EEG and laboratory results, as well as CT Head with no evidence of an acute intracranial process and a CT Abdomen Pelvis with no evidence of a primary malignancy, metastases, or adnexal lesions. However, a lumbar puncture was positive for neutrophils, lymphocytes, elevated glucose, normal protein, and with no organisms on a gram stain. While the anti-NMDAR IgG antibody results were pending, excitatory catatonia was also high on her differential and hence ECT (Electro Convulsive Therapy) was initiated 3 times a week.

On her 10th day of admission, the anti-NMDAR IgG antibody assay was positive at 1:1280. The patient was subsequently started on a 5-day course of 1000 mg Methylprednisolone and a 4-day course of IVIG. The patient completed 6 ECT therapies and her Methylprednisolone taper after 30 number of days in the hospital. She gradually demonstrated independence with ADLs and returned to her functional baseline prior to her discharge.

Conclusion

This case demonstrates an atypical presentation compared with other cases of anti-NMDA receptor encephalitis. By the time the patient came into our care, she had already been experiencing symptoms of psychosis and excited catatonia without signs or evidence of undergoing a prodromal phase. Given her failed management with benzodiazepines, organic causes of catatonia were pursued. In cases of anti-NMDA receptor encephalitis, EEG results are typically abnormal, with the presence of extreme delta brushes [7]. Although her EEG was normal, her lumbar puncture as well as her anti-NMDAR IgG antibody assay were positive. Her diagnosis of anti-NMDA receptor encephalitis was confirmed as she fit the diagnostic criteria proposed by Graus - having a subacute onset (<3 months), having pleocytosis on lumbar puncture, and excluding all alternative neuropsychiatric diagnoses [8]. Although first line treatments such as steroids, IVIG, or plasmapheresis are often used for anti-NMDA receptor encephalitis, these are only useful in about half of patients [9, 10] . Delayed onset of treatment greater than four weeks can worsen prognosis and lead to a higher risk of long-term cognitive deficit[11]; those who do not respond within four weeks can be switched to another treatment such as ECT which was employed in the patient to work synergistically with her immunotherapy.

While the mechanism of how ECT provides therapeutic response in catatonia is still debated, Singh and Kar have proposed three hypotheses: neurophysiologic, neurobiochemical, and neuroplasticity [9, 12]. The neurophysiologic theory suggests stimulus delivery may alter cerebral blood flow and glucose consumption leading to ictal phase changes in ictal pressures and the induction of brain-derived neurotropic factor (BDNF). The neurobiochemical theory suggests stimulus delivery may directly affect synthesis, release, and reuptake of neurotransmitters which may directly upregulate NMDA receptors. The neuroplasticity theory suggests stimulus delivery may reduce arborization of dendrites and excitatory synapses in the hippocampus and amygdala, as seen in animal models, which may lead to recovery [9, 12]. Tanguturi provides that management in patients with anti-NMDAR encephalitis who also present with catatonia should include ECT in addition to immunotherapy based on previous case reports as it may lead to faster recovery [9].

Given that 20% of patients can die from anti-NMDA receptor encephalitis, there is no doubt that prompt evaluation and treatment should be prioritized, especially in individuals who develop unexplained, complex, and refractory neuropsychiatric symptoms [10]. In the case of this patient, her catatonia within the setting of anti-NMDA encephalitis highlights a unique presentation that should be considered by providers to not delay treatment.

Although patients with anti-NMDA receptor encephalitis are expected to recover over the course of several years, many patients have persistent cognitive deficits, and there is a 10% relapse rate within the first two years, though this is often less severe than the initial presentation[10]. In addition to prompt treatment and evaluation for anti-NMDA receptor encephalitis, w e recommend the involvement of a multidisciplinary team early in the disease course. This team may include neurology, psychiatry, medicine, PT/OT, and other rehabilitation specialists who can create individualized plans for these patients, and who should be knowledgeable about recovery given the longevity of disease sequelae and potential for relapse [13].

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