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ISSN 1462 2815

A RARE CASE OF ANTI-NMDA RECEPTOR POSITIVE CEREBELLITIS: A COMPREHENSIVE CASE REPORT

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DOI: 10.5281/zenodo.11203401

Abstract

Autoantibodies directed against the NMDA receptors in neuronal cells cause anti-N-methyl-D-aspartate (NMDA) receptor encephalitis, an autoimmune inflammation of the brain parenchyma. In pediatric patients, it is the most prevalent type of autoimmune encephalitides. This case report describes the diagnosis of anti-NMDA receptor positive cerebellitis in a 1-year-old male infant. This case study highlights how crucial it is to identify this illness early and treat it appropriately in order to have the best results.

INTRODUCTION

A potentially fatal inflammatory brain disease with neurological and behavioral symptoms is encephalitis. There are a number of potential causes for it, including as immune system mediation and direct infection of the central nervous system (usually by viral infections). Autoantibodies against NMDA receptors cause anti-N-methyl-D-aspartate (NMDA) receptor encephalitis, an autoimmune encephalitis. The most well-characterized subtype of ionotropic glutamate receptors (GluR) in neuronal cells is the NMDA receptor [1]. They are crucial for the development of healthy neural networks, brain functions like memory and learning, and synaptic plasticity [1,2]. This syndrome has emerged as a major cause of autoimmune encephalitis in children and adolescents, with 40% of patients under the age of 18 [3].

Case Presentation: A one year old male child presented to the emergency department with a GCS of 12, baby also had complaints of fever, loose stools, vomiting and two episodes of generalized tonic clonic seizures. No prior investigations done, baby was immunized up to date. Neonatal period was uneventful. Milestones achieved appropriate for age.

Diagnostic Evaluation: Investigations revealed dyselectronemia with hypocalcemia was present and elevated total count and CRP. Lumbar puncture was done which was found to be normal, through the course of time baby was started on antibiotics and electrolyte correction was done. Child developed seizures on first day after which sensorium didn't improve hence MRI brain was performed and it showed bilateral

symmetrical diffusion restriction with T2 hyperintensity in deep cerebellar nuclei and cerebral hemisphere. Since the child condition didn't improve and presented with irritability, no spontaneous eye opening or verbal response to mother voice, suspected Encephalopathy and repeat MRI was done which showed Diffuse swelling of both cerebellar hemispheres which appears hyperintense on both T2 and flair sequence and showed diffuse restriction and raised possibility of cerebellitis . Started on injection methyl prednisolone as child neurological condition started worsening Repeat Lumbar puncture was done in view of CSF analysis for Autoimmune panel which showed positive for anti NMDA receptor antibody.

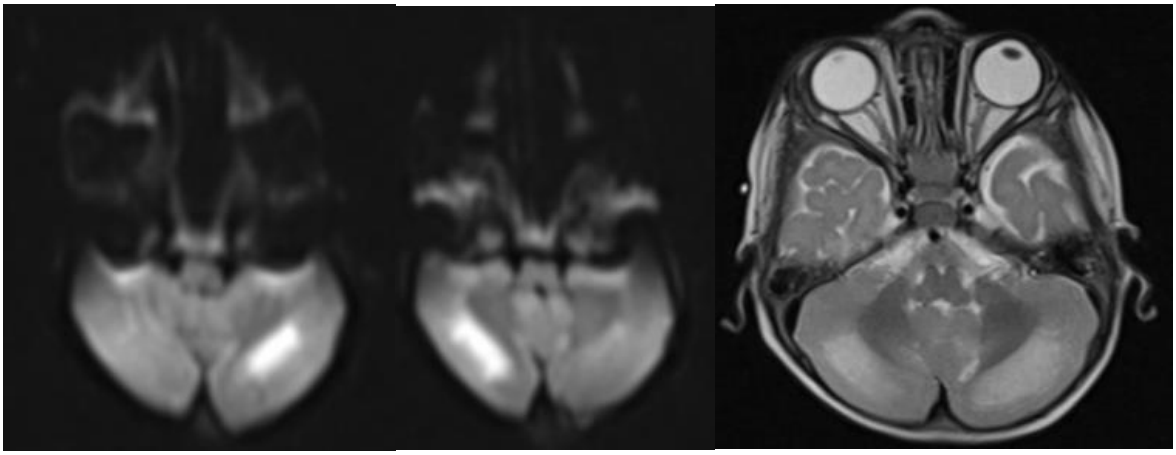

Figure: A) and B) Diffusion Weighted Imaging Showing Symmetrical Restriction in Bilateral Cerebral Hemispheres. C) Axial T2 weighted Image Showing Hyperintensity Involving Bilateral Cerebral Hemispheres

Treatment and Outcome: patient was started on steroids but no response with increase in the involvement in further follow up MRI. Treatment for acute cerebellitis may include steroids, acyclovir, and decompressive craniectomy if necessary.Early treatment with immunomodulatory therapies, such as corticosteroids, IVIG, and rituximab, played a crucial role in the patient's recovery. This case highlights the importance of prompt recognition and appropriate management of anti-NMDA receptor encephalitis in paediatric patients.

DISCUSSION

After acute demyelinating encephalomyelitis, anti-NMDAR encephalitis is known to be the most common autoimmune encephalitis in children [5]. It is an illness brought on by antibodies against the NR1 subunit of the NMDAR. The development of auto-antibodies against NMDAR, a protein important in synaptic plasticity and memory, is linked to encephalitis. In actuality, the antibodies from the patients have harmful effects on the NMDAR. Both neurological and psychological signs are indicative with anti-NMDAR encephalitis. In youngsters, isolated psychiatric symptoms are uncommon when the illness first manifests. In adults, they occur more frequently, and during relapses, they are more common [1]. Two to four weeks after the prodromal phase of the illness, patients experience neurological and/or psychiatric symptoms (typically fever, upper respiratory tract symptoms, fever, nausea, headache, vomiting, or diarrhea) [2, 6]. In contrast, children present more frequently with irritability, bouts of inappropriate laughing or crying, hyperactivity, temper tantrums, and violent behaviors, in addition to insomnia and anxiety. Deterioration in speech is frequently

reported in both in pediatric and adult ages as an initial symptom [1]. In the 12 years and under age group, the initial symptoms are usually neurological, consisting of dystonia, dyskinesia, and/or seizures (usually partial seizures)

CONCLUSION

Imaging results for pediatric patients with anti-NMDA receptor encephalitis vary widely and may be influenced by the underlying etiology of the condition. In contrast to adult patients, pediatric children may have different imaging characteristics, and over half of patients may have a normal initial MRI. We think that underage cases of anti-NMDA encephalitis are likely underreported [12]. This case illustrates the difficulties with diagnosis that this age group faces.

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