

**CASE REPORT****OPEN ACCESS**

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New Onset Refractory Status Epilepticus (NORSE) as a Presentation of NMDA Encephalitis in a Pediatric Patient

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Anti-N-methyl D-aspartate (NMDA) receptor (anti-NMDAR) encephalitis is caused by immunoreactivity against the NR1 subunit of the NMDA receptor. We present the case of a thirteen years old boy with history of sudden onset of seizure for five days back. After a detailed history and a thorough clinical examination the autoimmune encephalitis antibodies profile were sent. This came positive for NMDA receptor and negative for CASPR-2, Type AMPA1/2, LGI-1, DPPX, and GABA_B receptor antibodies. He was started on sodium valproate, carbamazepine, phenobarbital, Clonazepam along with full dose corticosteroids but did not respond significantly. Patient was shifted to ICU and intubated. Anesthesia with ketamine and propofol was started. After twenty days the patient was seizure free. He was given IVIGs and immunosuppressive therapy. He was sent home and two weeks follow up was advised. New onset refractory status epilepticus preceded the diagnosis of NMDA Encephalitis. The diagnosis of autoimmune encephalitis should be considered a top differential in relapsing patients as it has a very wide range of presentation. Prompt diagnosis and early treatment are the need of the hour as the ramifications of a mis or missed diagnosis could be detrimental.

Keywords: Encephalitis; NORSE; NMDA

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INTRODUCTION

Anti-N-methyl D-aspartate (NMDA) receptor (anti-NMDAR) encephalitis is caused by immunoreactivity against the NR1 subunit of the NMDA receptor. It is beyond doubt one of the most common encephalitis's. The term was first explained and evaluated by Dalmau and his team. They described it as a psychiatric and neurologic condition found in women that had ovarian teratomas.¹ However recent advances and studies suggest that tumour involvement wasn't necessary to be a part of this condition. It can and may present with or without the tumour. Recent literature has shown an overlap of psychiatric and neurological pathology in association of autoimmune encephalitis.^{2,3} The disease progresses as Prodromal phase moving into the Psychotic phase followed by a Catatonic stage.⁴

CASE REPORT

A 13 years old boy with full term birth via SVD at hospital with immediate cry. Developmentally normal child. History of seizure, sudden onset 45 days back. GTC seizures with up rolling of eyes and neck retraction. Abnormal movements of hands and legs. Seizures were continuing for several days with short seizure free duration. No history of fever, trauma. No family history of seizure activity.

Bp: 100/60 mm Hg, Pulse 96/minute and temperature of 98F. GCS: 11/15 with no signs of meningeal irritation. Cranial nerves all normal with no evidence of focal neurological deficit. Rest of systemic examination was not significant.

Baseline investigation showed HB: 13g/dl, TLC: 8000/mm³, platelets: 170000/mm³, MCV: 81fl. Rest of baseline investigation were including RFTs, LFTs, serum electrolyte, Serum calcium, Serum magnesium and PO4. Urine R/E showed no significant activity.

EEG record showed encephalopathy picture with dominantly abnormal background delta and theta activity. MRI brain done which showed patchy swollen gyri with T2 weighted hyperintensity visualized along with both cerebral hemispheres feature due to encephalitis. CSF study showed normal opening pressure with WBC < 5/ul, no Polymorphs and RBCs. Biochemistry showed protein of 11mg/dl, sugar of 54 mg/dl excluding infectious etiology.

Autoimmune encephalitis antibodies profile was done which was positive for NMDA receptor antibodies and negative for CASPR 2, Type AMPA1/2, LGI 1 DPPX and GABA_B antibodies.

He was started on sodium valproate, carbamazepine, phenobarbital, Clonazepam along with full dose corticosteroids but not responding. He was shifted to neurology ICU care and was intubated. Ketamine and propofol anesthesia started. He was under ICU for 20 days. After 20 days he became fit free, but there were still focal limb fits. He was given IVIGs and proper immunosuppressive therapy was started. He was sent home and follow up was advised after 2 weeks.

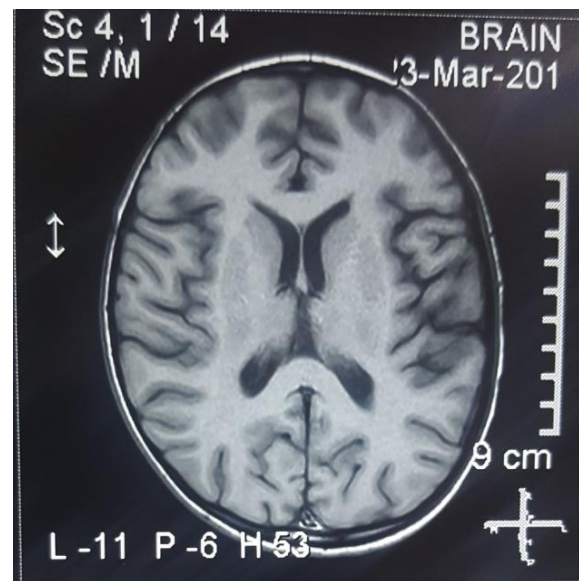


Fig.1 Axial T1W MR brain sequence of a thirteen-year-old boy at the level of lateral ventricles. This sequence is not done for determining signal changes.

DISCUSSION

Antibody testing to confirm the diagnosis should be requested in all suspected cases especially those presenting with psychiatric signs and symptoms. The current methods for testing antibody detection has a sensitivity and specificity of 100%.⁵

It is beyond doubt one of the most common encephalitis's. The term was first explained and evaluated by Dalmau and his team. They described it

as a psychiatric and neurologic condition found in women that had ovarian teratomas. ¹ However recent advances and studies suggest that tumor involvement wasn't necessary to be a part of this condition. It can and may present with or without the tumor.

We highlight and stress upon the atypical presentations, prompt diagnosis and early treatment of anti-NMDA-receptor encephalitis as the ramifications of a mis or missed diagnosis could be detrimental. The extensive investigations, detailed history and physical exam is ambiguous in giving us a definitive diagnosis early on, especially in case of overlapping neurological and psychiatric presentations having strong associations with the medical condition.

CONCLUSION

The diagnosis of autoimmune encephalitis should be considered a top differential in relapsing patients as it has a very wide range of presentation. Prompt diagnosis and early treatment are the need of the hour as the ramifications of a mis or missed diagnosis could be detrimental.

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