Indian Journal of Medical Case Reports ISSN: 2319–3832(Online)
An Open Access, Online International Journal Available at http://www.cibtech.org/jcr.htm
2017 Vol.6 (2) April-June, pp. 10-12/Hashmi et al.
Case Report

N-METHYL-D-ASPARTATE ANTIBODY (ANTI NMDA RECEPTOR) ENCEPHALITIS PRESENTING AS COMPLEX PARTIAL NON CONVULSIVE STATUS EPILEPTICUS (NCSE)

Nabeel Hashmi[1], Sourya Acharya[1], Yogesh Khitani[1] and Samarth Shukla[2]
[1]*Department of Medicine, J.N Medical College, DMIMS University, Sawangi (Meghe), Wardha- 442001, Maharashtra*
[2]*Department of Pathology, J.N Medical College, DMIMS University, Sawangi (Meghe), Wardha- 442001, Maharashtra*
**Author for Correspondence*

ABSTRACT
Encephalitis is the inflammation of the brain parenchyma. The commonest etiology is usually viral. Rarer etiologies especially anti NMDA receptor encephalitis usually present as myriad of bizarre chronic neuropschychiatric manifestations. Anti NMDA receptor encephalitis is autoimmune disorder. Few studies are available describing neuropschychiatric changes of this entity. We present a case of a 32 year old male who presented to us with chronic abnormal psychotic behavior and complex partial non convulsive status epilepticus (NCSE) finally diagnosed as anti NMDA receptor encephalitis.

Keywords: *Encephalitis, Anti NMDA Receptor, Brain Parenchyma, Autoimmune, Complex Partial NCSE*

INTRODUCTION
Anti NMDA receptor encephalitis is a rare autoimmune variant of chronic encephalitis which can present as abnormal psychiatric features like anxiety, bizarre behavior, paranoid delusions, agitations and cognitive disturbances. Epileptic seizures, dyskinesias and dysautonomias are known features (Dalmau *et al.,* 2008; Irani *et al.,* 2010).
Sometimes it may be a sole paraneoplastic manifestation of occult malignancies (Vitaliani *et al.,* 2005). The cognitive features usually occur in latter course of the disease. Attention span, memory and executive functions are impaired, because of this, patient is usually diverted for psychiatry consultation. Despite a growing body of literature the condition still remains under recognized.

CASES
A 32 year old male presented to us with history of irrelevant talking, not recognizing relatives, episodes of aggressive and bizarre behavior and abnormal movements since 1 month. There was no history of fever, headaches, vomiting, generalized tonic clonic seizures, weakness, dysphagia, breathlessness.
General examination revealed that the patient was conscious but was not responding to any questions. He was continuously talking irrelevant with inappropriate words. His vitals and other general examination findings were normal.
CNS examination: Patient was disoriented yet conscious, speech lacked comprehension. There was no dysarthria. The score in MMSE examination was 0 out of 30. The patient did not cooperate for cranial nerve examination but it was evident that there was no dysarthria and ocular nerve abnormality.
Motor nerve examination: power was grade 5 in all limbs, there were abnormal complex automatisms bilaterally present in the form of puckering of mouth intermittently and picking movements by the fingers, intermittent moaning and throat clearing was also present, which suggested complex partial seizures.
The automatisms were present throughout continuously suggesting complex status epilepticus. Patient was not cooperative for sensory examination but pain sensation was intact, there were no signs of meningeal irritation.
Investigations: His complete blood count, kidney function test, liver function test, blood sugar, electrolyte profile was within normal limits. MRI brain revealed features of encephalitis (Figure 1).

Case Report

CSF study revealed total cell count of 350 cells with 80% lymphocytes. CSF proteins: 165 mg%, sugar: 80 mg/dl. Electrophoresis revealed oligoclonal bands, suggesting immune up regulation of intra the cal antibody production. CSF PCR for HSV 1 and 2 was negative. CSF was sent for ANTI NMDA receptor antibodies which came positive. EEG showed sustained rhythmic spikes and spike and wave complexes. Anti nuclear antibody, antiphospholipid antibodies, IgM-EBV and CMV was negative. ELISA for HIV was negative.

Patient was treated with diazepam, anti pschychotic medications, like risperidone 0.5 mg tds for pschychosis. Tablet carbamezipine 200 mg tid was given for suspected complex partial seizures. Intravenous immunoglobulin 0.4 gm /kg/day was given for 5 days along with iv methylprednisolone 500 mg for 3 days with oral prednisolone with tapering dose was prescribed for one month. The abnormal movements responded well and mild improvement occurred in dyscognitive features. Episodes of agitation decreased. Risperidone and diazepam was withdrawn in 2 weeks. The patient took voluntary discharge and is awaiting follow up.

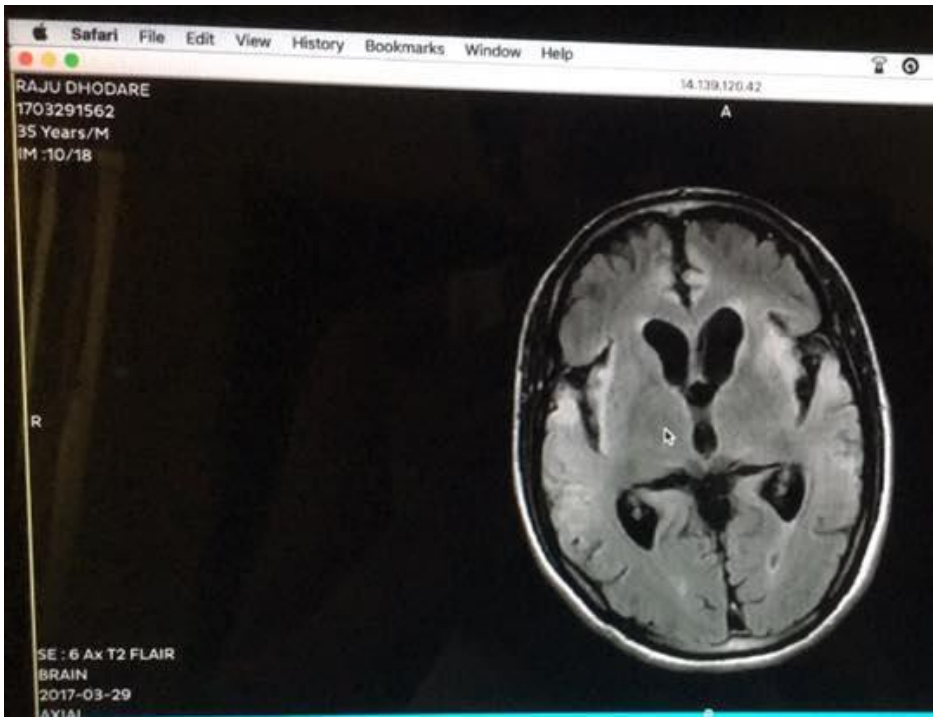

Figure 1: MRI Brain with Contrast Showing Altered Signal Intensity in Bilateral Temporal Lobes, Adjacent to Bilateral Sylvian Fissures and Singulate Gyrus with Mild Patchy Post Contrast Enhancement; Features Suggestive of Encephalitis

DISCUSSION

The above case signifies that ANTI NMDA encephalitis though rare should be kept in mind while treating patients with chronic encephalitis. It is important to rule out other common causes especially viral encephalitis. Neuropschychiatric manifestations usually pose confusion of a pschychiatric disease. Occult tumours should be sort for, as it may present as a paraneoplastic manifestation (Graus and Dalmau, 2007). With the advent of antibody markers diagnosis is becoming easy (Davies *et al.*, 2010; Prüss *et al.*, 2012). Presence of seizures early in the illness is a classical feature, complex and generalized seizures are reported in majority of cases.

Our case presented with typical complex seizures, which was continuous non convulsive complex partial status epilepticus (NCSE) since 1 month distinguishing this entity from most cases of viral encephalitis (Gable *et al.*, 2009). Encephalitis mimcs (table 1) should be excluded through investigations, in young

patients who present with ANTI NMDA receptor encephalitis as a paraneoplastic manifestation, ovarian and testicular tumours should be sorted (Graus and Dalmau, 2007). Combination of pschychiatric, neurologic and autonomic symptoms and signs should raise suspicion of autoimmune encephalitis.

Table 1: Encephalitis Mimics

1] Viral Encephalitis [HSV, HIV, CMV, EBV]
2] Paraneoplastic Encephalitis
3] Metabolic Encephalopathy
4] Autoimmune [Systemiclupus Erythematous, Vasculitic Encephalopathy]

Empirical treatment should be started, till date the treatment regimens which are used are intravenous corticosteroids, plasma exchange, intravenous immunoglobulins or both. Rituximab an anti CD -20 lymphocyte targeting monoclonal antibody may be considered in refractory cases (Irani *et al.*, 2010; Ishiura *et al*., 2008).

Conclusion

ANTI NMDA receptor encephalitis is a complex entity with variable symptomatology and response to treatment. The differential diagnosis is broad and should be ruled out appropriately. The diagnosis and treatment requires awareness and communication between physicians, neurologist, psychiatrists, microbiologists.

REFERENCES

Dalmau J, Gleichman AJ, Hughes EG, Rossi JE, Peng X, Lai M *et al.*, (2008). Anti-NMDA-receptor encephalitis: case series and analysis of the effects of antibodies. *Lancet Neurology* **12** 1091–8.

Davies G, Irani SR, Coltart C *et al.*, (2010). Anti-N-methyl-D-aspartate receptor antibodies: A potentially treatable cause of encephalitis in the intensive care unit. *Critical Care Medicine* **38** 679-82.

Gable MS, Gavali S, Radner A *et al.*, (2009). Anti-NMDA receptor encephalitis: report of ten cases and comparison with viral encephalitis. *European Journal of Clinical Microbiology & Infectious Diseases* **28** 1421-29.

Graus F and Dalmau J (2007). Paraneoplastic neurological syndromes: diagnosis and treatment. *Current Opinion in Neurology* **20** 732–737.

Irani SR, Bera K, Waters P, Zuliani L, Maxwell S, Zandi MS *et al.*, (2010). N-methyl-D-aspartate antibody encephalitis: temporal progression of clinical and paraclinical observations in a predominantly non paraneoplastic disorder of both sexes. *Brain* **133** 1655–67.

Ishiura H, Matsuda S, Higashihara M *et al.*, (2008). Response of anti-NMDA receptor encephalitis without tumor to immunotherapy including Rituximab. *Neurology* **71** 1921-23.

Prüss H, Dalmau J, Harms L *et al.*, (2012). Retrospective analysis of NMDA receptor antibodies in encephalitis of unknown origin. *Neurology* **75** 1735-39.

Vitaliani R, Mason W, Ances B, Zwerdling T, Jiang Z and Dalmau J (2005). Paraneoplastic encephalitis, psychiatric symptoms, and hypoventilation in ovarian teratoma. *Annals of Neurology* **58** 594–604.