A Case Report on Anti-NMDAR Encephalitis

Reshma Rej, Aloysius James, Sai Keerthana P.C, Nimmu Mani, Remya Reghu*
Department of Pharmacy Practice, Amrita School of Pharmacy, Amrita Vishwa Vidyapeetham, Amrita University, Kochi, India.
*Corresponding author’s E-mail: remyareghu@aims.amrita.edu

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ABSTRACT

Anti NMDAR encephalitis, an immune mediated disorder, which shows psychiatric and neurological manifestations, gained its first mention in the year 2007. This condition is most often found in young females and has an association with a number of malignancies, most commonly ovarian teratomas. It is often misdiagnosed as mental indisposition or viral encephalitis. Prompt identification and treatment of the disorder are essential. Here in we discuss a case where the patient was misdiagnosed to have psychotic syndrome and was treated for the same.

Keywords: Anti NMDAR Encephalitis, Neurological, Ovarian teratoma, Psychiatric syndrome.

INTRODUCTION

Anti NMDAR encephalitis is a serious yet treatable neuroautoimmune disease arising from the production of antibodies targeting synaptic proteins1. Clinical presentation include an initial phase with non-specific symptoms, psychiatric phase, unresponsive phase with catalepsy like symptoms, hyperkinetic phase with orofacial limb dyskinesia and orolingual dyskinesia. The diagnosis of NMDAR Encephalitis is confirmed by the presence of antibodies against N-methyl-D-aspartate receptor. Treatment includes definitive treatment and psychiatric symptom management.2 The following case report illustrates dramatic psychiatric presentation of N-methyl-D-aspartate receptor encephalitis and encompasses a study of the clinical signs and symptoms, diagnosis and treatment of this disorder.

CASE HISTORY

A female child, aged 10 years, came to our hospital with seizure episode. Earlier she was admitted in a local hospital for her psychiatric symptoms and was undergoing treatment with Anti Psychotics. Several investigations were done in our hospital. MRI was normal. Since EEG showed abnormalities, she was initiated on Tab Valparin (Sodium Valproate) 200 mg BD and her Anti Psychotics were also continued while awaiting for the Neuroimmunology reports (CSF NMDA Antibody). The results showed CSF NMDA type of glutamate receptor antibody positive and serum NMDA Antibody negative. She was diagnosed with Anti NMDAR encephalitis in Sept 2015. She was given one course of IVIg and corticosteroids during this period and was discharged with advice to review in Neurology OPD.

The patient was again admitted on 15 Jan 2016 with complaints of back pain and difficulty in writing. She was diagnosed with drug induced Hepatitis during this admission. After stopping Tab Valparin (Sodium Valproate), the LFTs showed falling trend. She was discharged with Tab Clonazepam 0.25 mg BD, Tab Carnisure (L Carnitine) 330mg BD, Tab. Pan(Pantoprazole) 20 mg 1-0-0 and Tab Udiliv (Ursodeoxycholic acid) 300mg BD (30 days).

She was admitted again on 27 Feb 2016 for break through seizures and initiated on Tab Levipil 250 mg ½-0-1. LFTs were found to be normal during this time.

She was admitted, for the third time, on 15 March 2016 for aggression, behavioral outbursts and her parents sought to stop Levipil in view of behavioral changes. Levipil dose was decreased. In this admission also a Neuroimmunology study was sent. The patient’s CSF and Serum sample tested positive for NMDA type of glutamate receptor antibody. Her discharge medications included Tab .Levipil (Leviteracetam) 250mg 1/2-0-1/2 Tab.Wysolone (Prednisolone) 30mg 1-0-0 Tab. Oleanzapine 2.5mg 1/2 -0-1 ,Tab Clonazepam 0.25mg BD, Tab.Serenace (Haloperidol) 0.5mg BD She was advised IVIG. It was not given due to financial constraints. The option of plasmapheresis was also discussed with parents and the patient was discharged. She was given IVlg from a Government hospital, 10gm for 5 days and 5gm on 6th day (55 gm total).

She was admitted for the fourth time as her clinical symptoms progressed to akinetic mute state with dystonia of left upper limb, walking with support and fluctuation in symptoms (catatonic features). During this admission her Liver enzymes were elevated. She was started on Rituximab, 600 mg in 500 ml Normal saline as per protocol. The patient was found to be gradually improving.

DISCUSSION

Anti N-methyl-D-aspartate receptor Encephalitis presents with a typical introductory phase that consists of fever,
fatigue and other flu-like symptoms followed days later by onset of mental illness (hallucinations, mood lability, agitation, aggression, irritability and delusional thought content). In the progression phase most patients experience sleep disturbances, seizures, dyskinesias and alternating periods of agitation and catatonia followed by autonomic instability. 2. The patient was brought to the hospital, with complaints of temper tantrums, anger outbursts and aggression (noted over 1-2 weeks) and an episode of seizures. She had abnormal feeling of tightness of fingers of left hand and staring episodes.

Diagnosis is made by clinical features, antibodies to NMDA receptor in serum or CSF, CSF pleocytosis and EEG showing diffuse delta activity with paroxysmal discharge. 4. MRI brain FLAIR (fluid-attenuated inversion recovery) sequence at presentation may be normal or it may demonstrate bilateral or medial temporal lobe hyper intense signals predominantly involving hippocampus. 5. The patient's CSF and Serum sample tested positive for NMDA type of glutamate receptor antibody. EEG monitoring showed moderate degree of focal nonspecific disturbance of electrical function maximum over right hemisphere. MRI brain showed normal study. USG scan revealed no ovarian teratoma.

Under definitive treatment we have tumour resection (if present) and immunotherapy. Corticosteroid plus Intravenous immunoglobulin or plasmapheresis is used as first line agents. Rituximab or cyclophosphamide is used as second line agent if the patient does not improve with first line agents alone. 3. The patient was treated with Inj IV Methylprednisolone 900 mg and IVIg. Since the patient did not respond to first line treatment, she was started on second line agent as per protocol.

This case illustrates the importance of early diagnosis of Anti NMDA Encephalitis as its symptoms mimic psychiatric syndrome. Prompt intervention will improve patient outcome

CONCLUSION

Physicians should have adequate knowledge about the disease and its differential diagnosis. Early diagnosis is essential to curb the disease progression. Clinical pharmacists should be aware of the appropriate selection of drugs and dose which helps in preventing major adverse drug events as the side effect profile of these immunotherapy drugs are high. This will ultimately improve the therapeutic outcome.

REFERENCES


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