Anti-NMDA receptor encephalitis in children – tale of ten years at a tertiary care center

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INTRODUCTION

- Anti-NMDAR encephalitis is a common childhood autoimmune encephalitis
- Varied presentations and mimics many infectious/post-infectious etiologies
- Not many studies are available describing the anti-NMDA encephalitis in pediatric age group, from south Asian region

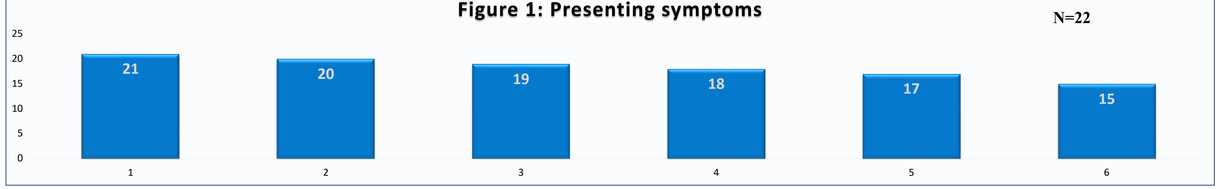
OBJECTIVES

- To study the clinical profile of patients with confirmed NMDA encephalitis in a tertiary hospital in northern India
- To assess the therapeutic response of immunotherapy in these children

MATERIALS AND METHODS

- **Type of study:** Retrospective observational study
- **Place:** Tertiary hospital in North India
- **Population:** Children (1 month to 18 years of age) with diagnosed NMDA encephalitis
- **Study period:** Jan-2012 to Feb-2022
- **Procedure:** Demographic details were collected. EEG was done for those presenting with seizures. CSF characteristics were documented along with MRI findings. The treatment received, the response along with follow-up data was collected

Figure 1: Presenting symptoms



1- Seizures 2- Behavioral changes 3- Altered sleep 4- Emotional lability 5- Oromotor dyskinesia 6- Choeroathetosis

RESULTS

- N = **22 patients** (Males:**9**; Females:**13;** median age:**10 years (IQR: 5.5-12year))**
- Most **common symptom – seizure - 21 (95%**), [12 (57%) had refractory seizures], followed by **behavioral changes – 20 (91%),** sleep disturbances and **altered sleep wake cycle – 19 (86.4%), emotional lability-18 (82%), oromotor dyskinesias – 17 (77.2%**) and **choreoathetosis-15 (68.2%),** focal motor deficits (hemiparesis and facial palsy) were seen in 6 of them
- **CSF** studies - **normal 12 (55%) patients** and **abnormal 7 (32%)[elevated protein-4, hypoglycorrhachia-1, lymphocytic pleocytosis-3**]
- **MRI Brain: normal - 15**, temporal involvement - **6**, changes in the thalamus and substantia nigra - **1**
- **EEG was done in 19 children** (**Normal: 4 (18%);** Diffuse cerebral dysfunction: 8 (42.1%); Focal/Multifocal discharges:9 (41%)); Delta-brush pattern - None
- **All patients** received **immunotherapy** (pulse steroids and intravenous immunoglobulin(IVIG)). 2nd line therapies (**Plasma exchange:4 patients**; **Rituximab-3 patients**) was given to those who either did not respond or responded partially to the 1st line agents
- Fourteen patients **(64%) showed complete response, 5 (23%)** responded **partially, 2 (9%) didn't respond** and **one (4%) died of sepsis**

CONCLUSIONS

- Anti-NMDAR encephalitis in children presents as fever, seizures and significant movement disorders and encephalopathy.
- If diagnosed promptly and timely treatment initiated, outcomes are favourable.

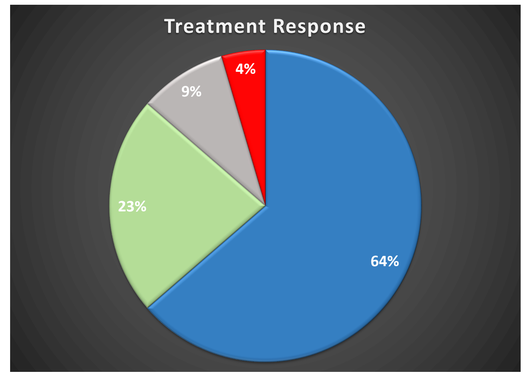


Figure 2: Pie chart depicting the clinical outcome in children with anti-NMDA encephalitis after receiving immunotherapy – with 64% of them showing complete clinical response

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