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New Onset Refractory Status Epilepticus Case And Their Management.

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Abstract
32 year old male laborer by profession caught with seizure , in-spite of taking multiple medication in different health care the symptom didn't subside. Ultimately, he landed to emergency where he started on multiple medication for seizure as the symptom was refractory and later diagnosed with New Onset Refractory Status Epilepticus (NORSE).

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I. Introduction

NORSE is a specific clinical presentation (but not a specific diagnosis) without a clear acute or active structural ,toxic ,infectious or metabolic cause occurring in patient without earlier epilepsy or neurological disorder which is known to precipitate seizure. It is important to Diagnose and Manage the NORSE as it is a very complicated and resistant to anti-seizure medication diseases of unclear etiology. It seen in < 10 % of patient presenting with status epilepticus but large proportion of patient with refractory non convulsive statues epilepticus. A cause of NORSE is ultimately found in approximately one and half of the patient mostly paraneoplastic or autoimmune encephalitis. When no underlying cause is found the symptom is considered to be cryptogenic in nature. Systematic review and metanalysis study conducted in 2023 which included 109 studies with 395 adult cases of NORSE, the etiology (identified after a negative initial evaluation) was cryptogenic in approximately 50 percent of cases, autoimmune in 36 percent, and infectious in 8 percent.[1] Mortality and morbidity are high patients with NORSE fatality rates of 20 to 30 percent or more, and under 25 percent return to their prior baseline health with long term treatment follow up. [2]

II. Case Scenario

32 year old male , Laborer by profession, Hindu by religion, Residence of Puducherry, Admitted to Clinic with a chief complaint of Fever 14 days, Seizures 14 days. Patient was apparently normal and later develop fever that was low grade. Fever not associated with chills and rigors. No history of rash/ Joint pain. No history of nausea, vomiting constipation and diarrhea. He took self-medication for symptom. Symptoms subside, patient went for one day trip and later got one episodes of GTCS presenting with Up-rolling of eye, Tongue fits, drooling of saliva, facial deviation, which lasted 5 minutes involved spontaneous regained of consciousness past seizure activity. Before admitting to higher center he went to multiple hospital and started on multiple antiseizure medication.

On examination in Higher Centre Clinic he was confirmed a case of NORSE (New Onset Refractory Status Epilepticus). The status was found to be infective, and the causative organisms was Acinetobacter baumanii.

III. Methods

On GCS examination $E3VTM_4$, Papilledema was found to be negative , pus discharge from nose sign of pansinusitis on MRI . On repeated EEG Examination Frequent rush of bilateral ictal rhythm (Frequent multifocal sharp waves) in posterior region of brain seen which was a sign of Electroencephalographic seizure . On further CSF examination for autoantibody it was found to be positive for antibody. Later Pupil examination , not reacting to light, fever 101degree Fahrenheit. Patient was started on Antiseizure medication and sedatives, For infectious condition it was started on first line Antibiotic treatment and Immunotherapy is given simultaneously.

IV. Results

Despite of multiple medication and lots of efforts patient was not able to cope up the Symptoms.

V. Discussion-

NORSE is a severe condition that occurs suddenly without any known prior neurological condition, it can lead to super refractory status epilepticus, where seizure persist despite aggressive treatment, including

multiple anticonvulsant, anesthetic,and immunotherapy. Patient with NORSE despite being on multiple antiseizure medication the seizure persists at 24-72 hours despite appropriate treatment, clear etiology is difficult to evaluate on initial evaluation. Overall etiologic evaluation of Brain MRI with gadolinium contrast, Serum, and CSF monitoring for infection evaluation and relevant to region, season, and individual comorbid condition. Comprehensive rheumatologic ,autoimmune and paraneoplastic antibody, evaluation CSF cytokine needs to be considered as well. Malignancy screening for CT chest, abdomen and pelvis required if its common etiology is not able to make the diagnosis of disease . If the etiology is unknown than genetic testing needs to be done .Multidisciplinary expertise in epilepsy , immunology, critical care ,rheumatology, infectious diseases is needed to consider treating the diseases. Begin immunotherapy within 48-72 hrs. of status epilepticus onset as soon as infectious etiology ruled out. Immune therapy I.V Methyl Prednisolone 20-30mg/kg per day (max 1g/day) for 3-5 days or IV IG 2g/day in divided doses for 2-5 days. For seizure sedative like benzodiazepine (doses -lorazepam 0.1mg/kg IV OR IO access) and antiseizure medication (Levetiracetam 60mg/kg IV/IO for 15-50 minutes) are given simultaneously via IV access line. If patient require Mechanical ventilation use vasopressor support (midazolam, propofol, or phenobarbital) as necessary. Phenobarbital is longer acting agent, proper cardiovascular monitoring needs to do Continuous Antiseizure medication with neurological follow-up need to be done. If the diseases is antibody mediated start with IV Rituximab even if the seizure continue than add IV Anakinra (IL-1-Receptor antagonist) and IV Tocilizumab (IL-6 -Receptor antagonist). Non -Pharmacologic intervention ketogenic diet and modified Atkin diet added within one week of status epilepticus condition or as soon as it got diagnosed[3]. Even after giving this drug if the seizure persists consider for therapeutic plasma exchange , intrathecal dexamethasone, high dose ketamine and magnesium to target the NMDA receptor, combination of previous therapy and thalamic neurotransmitter. If NORSE found to be cryptogenic than treatment needs to add rituximab and tocilizumab with antiseizure medication plus consider for therapeutic plasma exchange , intrathecal dexamethasone, high dose ketamine and magnesium to target the NMDA receptor, combination of previous therapy and thalamic neurotransmitter if the seizure does not subside .A retrospective study conducted in Columbia on 27 patient were 73% found to be cryptogenic,19% autoimmune, and 8%infectious .At discharge only 6 patient 23 percent were found to be good to fair outcome, while 14 had severe disabilities and 6 patient (23 percent) had died. The patient with anti NMDA receptor encephalitis survived with severe disabilities.[4]

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