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Anti-NMDA Receptor Refractory Status Epilepticus: Case Report

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ABSTRACT:

Anti-NMDA receptor (anti NMDA-R)encephalitis is an immune-mediated form of encephalitis, with paraneoplastic and nonparaneoplastic forms.

We will report the case of a 39year-old patient in our ICU ward that presented a refractory status epilepticus related to anti NMDA-R secondary to an ovarian teratoma that didn't match neither EEG, SCF nor MRI criteria.

The purpose of this case report is to sensibilize to this misdiagnosed etiology of encephalitis.

Key-words: Teratoma, anti-N-methyl-d-aspartate receptor encephalitis; encephalitis; intensive care, antibody, paraneoplastic , refractory status epilepticus, autoimmune, extreme delta brush,

Introduction:

Anti-N-Methyl-D-Aspartate receptor encephalitis is a rare and easily misdiagnosed syndrome with a

Myriad of complications (psychosis ,seizure, death …) (1). It is an autoimmune neurological disorder caused by antibody anti- NMDA receptors due mostly due to a teratoma (2)

Clinically, it could manifest by refractory status epilepticus, in over 75% of patients(3)

35% of patients pass away and 35% return to their neurological baseline, and the rest have an intermediate outcome (4).

25 % of patients have severe sequelae or die, while the rest recover with only minor deficits (5)

Status epilepticus doesn't have a specific protocol due to the very specialized nature of the disease.

The frequency of it remains uncertain. So, our purpose is to raise awareness in the emergency department.

Case report:

We report the case of a 39-year-old female with a medical history of autoimmune hypothyroidism and depression under.

Originally presented to the emergency department with anxiety, abnormal movement , psychosis , and confusion. A few days later, she came back to the emergency department with partial seizures (left leg and right side of the face) and hypersalivation.

Despite multiple anti-epileptic drug (AED) regimens, seizures persisted. Attempted measures included thiopental , keppam, phenobarbital, midazolam , clonazepam, valium the case of the patient got worse and evolved to status epilepticus that led to her intubation.

CSF (gly : 0.45 Alb : 0.5 cellularity 5element) was normal

A CT-Scan and MRI were performed and came back normal

Toxicological tests :were only positive to the treatment of her depression

EEG: came back normal.

The biological tests :

Serology :HCV (-) HBV (-) VIH (-) SYPHILIS (-) HSV (-)

After excluding all reasons of encephalitis (infectious ,tumoral metabolic…) the diagnostic of an anti NMDA related encephalitis was pointed . (tab 1)

	13/8/23	16/8/23	21/8/23
Hb	13	11.5	12
Plat (10*3)	358	244	375
Wc(10*3)	7.4	13	12.28
Uree	0.12	0.25	0.19
Creat	0.7	6	4
Na+	138	140	136
K+	4.53.8	3.7	3.7
Ca+	89	110	88
TSH	1.02		
Ac anti tpo	600 (N<4)		
Gly	0.8	0.89	0.87

Tab 1 : biological results

The anti – NMDA results came back highly positive . She was treated with steroids ,clonazepam, olanzapine and sodium valproate and she showed an improvement in her clinical status.

However , the patient died from a ventilator associated pneumonia. The post-mortem examination found out an ovarian teratoma

Discussion:

The clinical manifestations of patients with anti NMDA receptor are complex and vary in severity

In our case, the patient presented psychosis, seizures , abnormal movements as described in literature (6). Patients evolve progressively from psychosis, memory deficits, seizures, and language disintegration into a state of unresponsiveness with catatonic features often associated with abnormal movements, and breathing instability (6).

Spinal fluid was normal in our case . it's abnormal in up to 90% of cases : include mild-to moderate lymphocytic pleocytosis (up to 90%, with a median WBC count of 23/mm3), mildly elevated protein (up to 30%, with a median protein level of 24mg/dl) (6)

Electroencephalography was normal in our patient .according to literature it shows nonspecific focal or generalized slowing with predominant rhythmic activity in the delta range, especially in those manifesting catatonia "extreme delta brush" is present in up to 30% of patients (7.10)

The MRI was not concluding in our case .30–50% of patients present at MRI : mild and nonspecific T2 , fluid attenuation inversion ,recovery signal hyperintensity in cortical/subcortical areas of the hemispheres, cerebellum, brain stem, or spinal cord (8)

In the case of our patient, she seemed to be receptive to steroids alone that are describe as the cornerstone of the initial therapy(9). However , the patient died from a ventilator associated pneumonia which is the most common cause of death in Anti-NMDA encephalitis (11).

Steroids (methylprednisolone 1g/d for 5 d) are typically used concomitantly with immunotherapy (IVIG 0.4g/kg/d for 5 d) or plasmapheresis. It is followed by second-line immunotherapy (rituximab and/or cyclophosphamide) after failure (lack of improvement in 10–14 days of the former). (1,9)

Conclusion:

Anti NMDA receptor encephalitis remain misdiagnosed illness.

Just few cases have been reported in literature

Nowadays, the most effective therapeutic strategy remains undefined, particularly in the ICU.

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