

Demonic Possession or Something else... Anti-NMDAR Encephalitis: A Case Report

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Abstract

Acute confusional state is a challenging condition especially when present in young patients. I am presenting a challenging case of acute confusion in a young patient which was not diagnosed in a timely manner.

A 33-year-old pleasant lady was initially admitted with fits and managed with sodium valproate. There were no past medical problems. No cause was found and she was discharged with outpatient neurologist follow up. One week later she re-admitted with confusion, bizarre behaviour and personality change. Her confusion got worse and her personality was changed to that extent that her family and other ward members including nursing staff felt about demonic possession.

This presentation was considered secondary to sodium valproate which was changed to phenytoin. Afterward, she went through extensive investigations including CT head, MRI head and Spine, Lumbar puncture, Septic & autoimmune screen. Her pregnancy test was negative. LP results confirmed leucocytosis and EEG showed diffuse abnormality. MRI head showed encephalitis changes, MRV was normal. CSF culture did not grow any organism. She was treated with 14 days course of IV Aciclovir for? encephalitis which did not help and her condition deteriorated further and she was transferred to ITU.

Neurologist subsequent review advised for VGKC, NMDA, GAD & Paraneoplastic antibodies. NMDA receptor antibodies result later came back positive. She had a CT TAP & TV ultrasound which ruled out any ovarian malignancy.

She was treated with immunoglobins & IV steroids & her condition improved slightly however later she aspirated during fitting and was intubated and transferred to tertiary hospital where she received plasma exchange and got almost full recovery.

Keywords: *Demonic possession; Autoimmune encephalitis; NMDAR Encephalitis*

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Background

Demonic possession is believed by some, to be the process by which individuals are possessed by malevolent preternatural beings, commonly referred to as demons or devils.

Various forms of mental illness are commonly attributed to magic spells or demonic possession. These illnesses are usually manifested by overt motor behavioural disturbances. Top of the list of these disorders is epilepsy and schizophrenia. Recently anti-NMDA receptor encephalitis has been widely recognized a cause for such presentations.

I am presenting an interesting case of a young patient with anti-NMDAR encephalitis which was not diagnosed in a timely manner.

Case Presentation

A 33-year-old Caucasian lady was admitted to AMU with 2 brief witnessed episodes of tonic-clonic fits. Associated history of tongue biting and urinary incontinence. There was no past medical problem. She was not on any medications including OTC medications. Her LMP was 2 weeks ago and she had 3 healthy children. There was no recent travel history. She was non-smoker and drinks alcohol socially.

Systemic examination was entirely normal. Initial investigations e.g. FBC, U&Es, LFTs, CRP, Clotting were all in the normal range. Chest X-Ray was normal. A urine Pregnancy test was negative. CT head was normal as well. She was treated with Sodium Valproate, driving advice was given and discharged with outpatient Neurologist follow up.

One week later she re-admitted with strange bizarre behaviour including personality change. Her language was very much abusive and noticed on many occasions about rolling of eyeballs with flickering eye movements along with opisthotonos body postures. There were also spells of Bipolar-like presentations.

Her Systemic examination revealed GCS 14/15, rest was all normal. All bloods were normal as well. Initially this presentation was considered secondary to Sodium Valproate which was stopped and changed to Phenytoin.

Later she had Lumbar Puncture under sedation and results showed CSF: Clear, Protein: 0.50, Glucose: 3.7, WCC: 26 (100% Lymphocytes). She was treated for Viral Encephalitis with IV aciclovir however her condition did not improve and deteriorated further.

At this stage, the family was deeply concerned and father kept asking to discharge her and preferring to be seen by Priest? Demonic possession. Nursing Staffs were also terrified by her strange behaviour. They were trying to avoid going near to her & having the same comments? Demonic Possession.

She was transferred to side room because of the frightening nature of acts and complaints from other patients & relatives. She was kept on IV antibiotics, IV Aciclovir and IV fluids with no help and her condition deteriorated further.

Meanwhile, Viral PCR came back as negative. She had an MRI Head with DWI which showed diffuse signal abnormality at right parietal and temporal lobe grey matter. A focus of increased signal in the right frontal lobe and these changes suggestive possibility of encephalitis. MRV was normal. Based on MRI result IV aciclovir was continued.

Later she developed status epilepticus and was transferred urgently to ITU for further management i.e. IV phenytoin.

In ITU she was seen by Neurologist who advised for IV methylprednisolone trial as her condition did not improve despite IV aciclovir. Further blood tests including a complete autoimmune screen were sent as per Neurologist advice.

She was also reviewed by Psychiatrist who put diagnosis? Functional/Conversion Disorder. While waiting for additional blood results patient condition deteriorated further and she aspirated during fitting and comatose and intubated. Later serum NMDA receptor antibodies result came back positive. She was treated immediately with Immunoglobins. Urgent CT TAP and TV Ultrasound ruled out malignancy. She was later transferred to the tertiary centre where she received plasmapheresis and got almost full recovery within a few months.

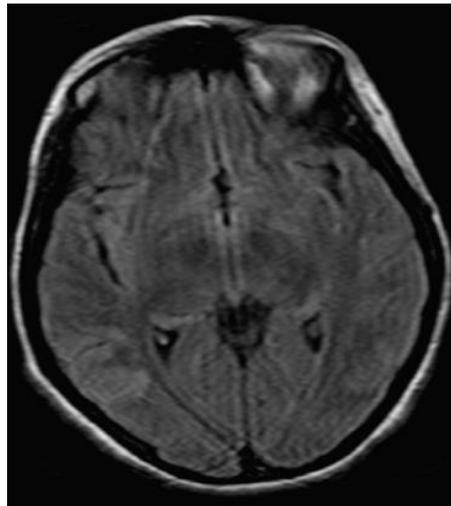


Figure 1: MRI head showing swollen grey matter of the right temporal and temporo-occipital areas.

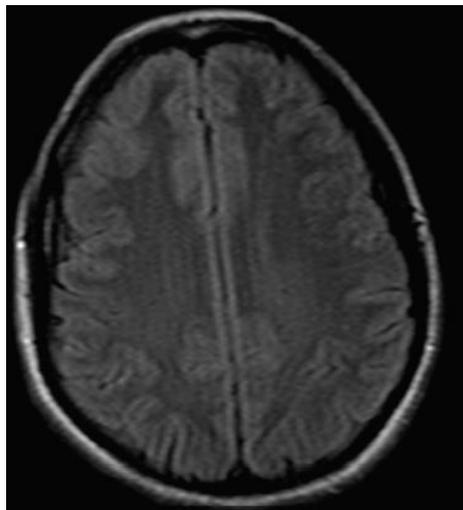


Figure 2: MRI Head showing swollen grey matter of the right frontal lobe.

Epidemiology

Epidemiological studies suggest that anti-NMDA receptor encephalitis may be the most common cause of autoimmune encephalitis after acute demyelinating encephalitis [1]. The number of new cases a year is unknown [2]. According to the

California Encephalitis Project, the disease has a higher incidence than its individual viral counterparts in patients younger than 30. This study provides the best approximation of disease distribution. It found that women are disproportionately affected, with 81% of cases reported in female patients. Disease onset is skewed toward children, with a median age of diagnosis of 21 years. Over a third of cases were children, while only 5% of cases were patients over the age of 45 [3].

While in Northern Europe 40 % of cases are infectious and 40 % are due to unknown causes, at least 20 % are immune-mediated, with the largest groups being anti-NMDA-receptor encephalitis (4 %) and VGKC-complex antibody positive encephalitis (3%). Wright et al have found an incidence of 0.85 per million children per year in the UK [4].

The largest series of over 400 patients found that at least 80% of sufferers are female [5], with the preceding paper finding 20% of sufferers under the age of 19 [6], and a smaller series finding the mean age of presentation at 18.5 years [5]. Anti-NMDAR encephalitis is more common in Asian and Pacific Islanders [5] and associated ovarian teratoma is more common in Black females [6].

Discussion

Anti-NMDA receptor encephalitis was first diagnosed in 2005 however was not widely recognized till 2013. It is a serious condition however if diagnose & treat in a timely manner has a very good prognosis [7-10].

It is caused by an autoimmune reaction primarily against the NR1 subunit of the NMDA receptor [11].

It is common in young patients with a female predominance. Herpes simplex encephalitis as a potential cause of anti-N-methyl-D-aspartate receptor antibody encephalitis [12].

There is a strong association between ovarian teratoma and anti-NMDA receptor encephalitis. It can also present with total insomnia as presenting complaint [13].

It mainly presents with psychiatric manifestations e.g. anxiety, agitation, bizarre behavior, hallucinations, delusions, disorganized thinking. Memory problems, autonomic instability, language dysfunction, and dyskinesias have also been reported [14].

There are cases where it can mimic HaNDL syndrome. (Headache, Neurological Deficit, CSF Lymphocytosis) & neuroleptic malignant syndrome [15].

Patients with anti-NMDAR encephalitis may develop concurrent or separate episodes of demyelinating disorders, and conversely, patients demyelinating disorders with atypical symptoms (eg, dyskinesias, psychosis) may have anti-NMDAR encephalitis [16].

Diagnosis is mainly detection of anti-NMDA receptor antibodies in serum and CSF. (CSF has 100 % sensitivity and 100% specificity) [17].

Treatment is mainly by steroids, immunoglobins and plasmapheresis as first line. While Rituzimab and cyclophosphamide are used as the second line. These patients may benefit from long-term rehabilitation and multi-disciplinary approach.

Learning Points/Take Home Messages

1. Anti-NMDAR Encephalitis should be suspected in young patients with prominent psychiatric symptoms accompanied by seizures, autonomic instability, hypoventilation and dyskinesias.
2. Anti-NMDAR encephalitis is less severe in patients ≥ 45 years old than in young adults, but the outcome is poorer in older patients due to late diagnosis.
3. Extreme delta brush EEG pattern can be present in one third patients and can lead towards early diagnosis.
4. Long term integrated and multi-disciplinary input by a variety of therapies and health disciplines is required in order to improve the long term outcome and quality of life for NMDAR patients and their families, and ultimately leads to improved positive outcomes [18].

Conflict of Interest

No conflicts of interests.

Patient Consent

Obtained

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