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Anti-NMDA receptor encephalitis presenting as an acute psychotic episode

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Abstract

Anti-NMDA receptor (NMDAR) encephalitis is a autoimmune disorder with prominent psychiatric symptoms. Patients usually present with acute behavioural change, psychosis, catatonic symptoms, memory deficits, seizures, dyskinesia, and autonomic instability.

We describe a 28 yrs old female patient who had previous 2 brief episodes of psychosis and has now again presented with psychosis but then clinical condition gradually worsened and she developed dyskinesia, catatonic symptoms and seizures. Though her routine blood work, brain imaging was normal, a confirmative diagnosis of NMDAR encephalitis was made by positive CSF studies. Patient was treated with methylprednisolone and plasmapheresis to which she showed response. This case illustrates the importance of considering NMDA R encephalitis as a differential diagnosis especially in young female with neuropsychiatric symptoms and progressive disturbance of consciousness, and associated involuntary movements, since prognosis depends on early recognition and treatment.

Introduction

Anti N Methyl D Aspartate (NMDA) receptor encephalitis is an autoimmune encephalitic syndrome with a specific pattern of presentation, course, and outcome. The syndromepredominantly presents in young women, with 60% of them having a neoplasm (usually ovarian teratoma).(1)

The classic presentation of anti-NMDA-R encephalitis involves a confluence of psychiatric, neurologic and autonomic symptoms, often with a viral prodrome.

Psychosis, hallucinations, memory loss and personality changes are the earliest symptoms due to which psychiatrist is usually consulted first. Dyskinesias(especially orofacial), chorea, ataxia, seizures and decreased level of consciousness may follow, prompting referral to a neurologist. Days to weeks later, autonomic instability may occur, manifesting as cardiac arrhythmia, hypotension and hypoventilation, requiring supportive care in the intensive care unit (ICU).(2-3)

Results of conventional investigations including examination of cerebrospinal fluid (CSF), brain imaging and electroencephalogram (EEG) are non-specific. Lymphocytic pleocytosis, oligoclonal banding, increased CSF protein, hyperintensity on T2 magnetic resonance imaging (MRI), decreased uptake in hippocampal structures on functional MRI and epileptiformactivity on EEG have been described.

Diagnosis is confirmed by presence of anti NMDA antibodies in serum and CSF. Concomitant tumours were confirmed in 60% cases with the most common being teratoma of ovaries and testis.

Treatment consists of methylprednisolone, immunoglobulin, plasmapheresis, or immunosuppressive drugs (cyclophosphamide, azathioprine, rituximab) as well as operation to remove the tumour.

Prognosis depend on early recognition and treatment of illness with symptoms disappearing within 1-12 months.

Case Presentation

Mrs L a 28-year-old married female with past history of 2 brief psychotic episodes was referred to our hospital from a ER of a multispeciality hospital with a current 2 weeks'history of being fearful, suspicious, withdrawn and aloof with gradual detoriation and 1 week history of poor oral intake, sleeplessness, and talking to self.

Family reported that she had the above symptoms for 2 weeks after which she was taken to psychiatrist where she was diagnosed with acute psychosis and prescribed olanzapine 5mg. then when patient went to hometown was seen by another psychiatrist where she was again diagnosed with psychosis and advised admission. Family took her to neurologist who advised CT Brain which had a finding of arachnoid cyst with calcified granuloma. Patient was then referred to neurosurgeon in view CT findings who opined that its incidental finding and no active intervention required. In next 1 week, there was gradual deteriotion in her earlier clinical state and new symptoms like patient started talking to self, poor oral intake and sleeplessness. Pt was seen in ER of a multispeciality hospital where ER physician / neurologist referred to our psychiatric institute. When patient was brought she was in a wheel chair due to generalisedweakness. On examination she had psychomotor retardation, no ETEC, rapport was not established. But during the course of interview suddenly she became agitated and started being fearful, screaming, rolling on floor, tearing her clothes. She had to be shifted to ward and sedated for behavioural control. Next day in the ward she continued to have poor oral intake, had incomprehensible speech, had oro facial dyskinesia, lip

biting leading to swollen lips, and had abnormal upper limb movements, down going planters, altered sensorium, and a seizure episode.

Due to changing clinical state Patient was shifted to multispecialty hospital. There routine investigation, EEG, MRI Brain was normal. Finally, CSF studies was positive for NMDA receptor antibodies confirming the diagnosis of NMDA R encephalitis.

Patient was treated with methylprednisolone, plasmapheresis after which she gradually responded too treatment.

Discussion

This report illustrates a patient, with past history of 2 brief episodes of psychosis, now presented again with psychotic episode. Eventually the bizarre limb movements, orofacial dyskinesia, lip smacking, with status like picture led to clinical suspicion of organicity and referral to multispeciality hospital where diagnosis of NMDA R encephalitis was made based on definitive confirmation on CSF studies.

Anti NMDA R encephalitis is an auto immune disorder with a complex presentation including psychiatric symptoms, cognitive symptomsand autonomic instability. About 4% of patients have only isolated psychiatric episodes.[(4)]

80% are females with majority presenting during early adulthood. In nearly 60% of females' patients an underlying neoplasm, most commonly ovarian teratoma is identified. There was an absence of clinically apparent tumour in this case. Based on immune pathogenesis, early removal of teratoma andimmunotherapy is mainstay of treatment. Prognosis depends on early recognition and treatment of illness.

Since patient presents with combination of symptoms in which psychiatric symptoms are usually seen initially it is common to miss this diagnosis unless NMDA encephalitis is considered as differential diagnosis.

In our case, it was further more likely to miss this, given the patient already had 2 definite prior episodes of psychosis and also that it was evaluated by 2 neurologists/ neurosurgeon before being referred to us.

This case illustrates the importance of considering NMDA R encephalitis as a differential diagnosis especially in young female with neuropsychiatric symptoms and progressive disturbance of consciousness, and associated involuntary movements. This case draws attention to the characteristic course of disease and typical progressive clinical signs which allows for early diagnosis before its confirmation by anti NMDA R antibodies in serum/CSF.

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