ORIGINAL RESEARCH PAPER
Volume - 13 | Issue - 12 | December - 2024 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

A RARE CASE REPORT OF ANTI-NMDAR ENCEPHALITIS

Neurology

Indhuja Karunakaran* Post Graduate, DM Neurology, Department Of Neurology, Government Chengalpattu Medical College And Hospital, Tamilnadu, India – 603001. *Corresponding Author

Thamil Pavai Arulnambi Professor And HOD, Department Of Neurology, Government Chengalpattu Medical College And Hospital, Tamilnadu, India - 603001

Balaji G Assistant Professor, Department Of Neurology, Government Chengalpattu Medical College And Hospital, Tamilnadu, India – 603001

Hariharan S Assistant Professor, Department Of Neurology, Government Chengalpattu Medical College And Hospital, Tamilnadu, India – 60300

ABSTRACT

Anti-N methyl D aspartate receptor encephalitis (anti-NMDAR encephalitis) is one of the most common forms of autoimmune encephalitis, that is frequently underreported. It can be triggered by a variety of causes like ovarian teratoma, and viral encephalitis like herpes simplex encephalitis, etc., Neuropsychiatric features, cognitive impairment, seizures, and movement disorders are some of the various clinical manifestations of this autoimmune encephalitis. Females are affected more often than males. A high index of suspicion based on clinical features is needed to diagnose this autoimmune condition. If identified early and treated appropriately, good outcomes can be expected in a majority of cases. This case report details anti-NMDAR autoimmune encephalitis provoked by a preceding mumps infection in a thirteen-year-old boy. His predominant manifestations were a combination of movement disorders including myoclonus and chorea. He was treated with immunotherapy and improved clinically over a span of one month.

KEYWORDS

Anti-NMDAR encephalitis, mumps virus, movement disorder , myoclonus, chorea

INTRODUCTION

Anti-NMDAR encephalitis, one of the most common autoimmune encephalitis[1] in India, has a predilection for children and young adults [2]. Movement disorders are one of the common manifestations in this age group [3]. There are many triggers for anti-NMDAR encephalitis. Mumps viral infection is one of the rarer triggers. Here we report a case of a young boy who presented with a unique combination of movement disorders, subsequently diagnosed with anti-NMDAR encephalitis.

Case Report

A 13-year-old boy, with normal birth and developmental history, without any co-morbidities, presented with imbalance while walking with frequent falls, which progressively worsened over a period of ten days. He was unable to stand with frequent buckling of his knees when he presented to us. All these symptoms were more during standing and walking and disappeared in sleep. His mother complained that he often spilled food and water while eating. He gradually became withdrawn and ultimately became bedbound due to the above complaints.

On examination, it was observed that he was apathetic. His mini-mental state examination and lobar function tests were normal. He had frequent myoclonic jerks, that were generalized. He also had chorea of both hands and legs. This was hampering his ability to walk and causing frequent falls. Bilateral spasticity with pyramidal type of weakness only confined to his lower limbs, with exaggerated deep tendon reflexes and extensor plantar was observed. Two days after admission, on his second week of illness, he developed perioral dyskinesias and one episode of focal seizures with impaired awareness.

His basic blood tests, echocardiogram, ultrasound of abdomen & pelvis, CT BRAIN, MRI BRAIN, and electroencephalogram were normal. Cerebrospinal fluid (CSF) analysis showed an elevated total cell count of 163 cells/cu mm. Other CSF parameters were normal. However, the serum antibody panel turned out to be positive for anti-NMDAR encephalitis.

On probing history, it was further revealed that he had a fever with bilateral parotid swelling suggestive of mumps viral infection, three months before current admission. He was treated conservatively and his illness subsided spontaneously. With this history, a clinical diagnosis of anti-NMDAR encephalitis precipitated by mumps infection was made. To confirm this, serum IgG antibodies for mumps were sent for testing in this clinical context. They came out to be positive in high titers of more than 300 AU/ml. This confirmed the possibility of a recent mumps viral infection that could have triggered the autoimmune encephalitis.

The patient was treated symptomatically. Pulse intravenous methylprednisolone and intravenous immunoglobulin were given for five days according to his weight. All the movements subsided with treatment. He improved and returned to his premorbid state over a period of one month.

DISCUSSION

Anti NMDAR encephalitis is often underdiagnosed due to its variety of presenting symptoms. The pathophysiology is due to the binding of anti-NMDA antibodies to NMDA receptors, causing neuronal dysfunction and the disruption of fronto-striatal connections. This disease preferentially affects children (from 2 months) and young adults (less than 5% of patients are over 45 years) with a male/female ratio of 1/4.[4]

Patients Often Present With Six Main Symptoms: including neuropsychiatric symptoms or cognitive problems, seizures, speech dysfunction, movement disorders, decreased level of consciousness, and autonomic dysfunction. A prodromal phase with features like headache, fever, and nausea was seen in a majority of patients.[5] There is a difference in presenting features between children and adults. Typically children present predominantly with non-psychiatric features like movement disorders. Myoclonus and chorea are some of the lesser common manifestations of anti-NMDAR encephalitis in this age group [6]. This patient had a combination of both.

Tumors and viral infections are the two most common triggers of anti-NMDAR encephalitis. This case illustrates that mumps is one of the rare triggers for anti-NMDAR encephalitis [7]. Mumps is a highly infectious RNA virus spreading by droplet and contact infection. Mumps, now uncommon in developed countries, is still seen frequently in India despite widespread vaccination [8]. The central nervous system (CNS) is the next most common site of involvement in children after salivary glands. Meningitis and encephalitis are the most commonly reported CNS complications of mumps. Anti-NMDAR encephalitis triggered by preceding mumps viral infection is rarely reported.

Treatment consists of immunotherapy including high-dose corticosteroids, intravenous immunoglobulin, and plasma exchange. If patients do not respond to this first-line immunotherapy, then

second-line immunotherapy like rituximab and cyclophosphamide can be tried. Younger age and milder symptoms have a better prognostic outcome than older age of onset and severe disease.

CONCLUSION

Timely identification and treatment of movement disorders as one of the presenting features of anti-NMDAR encephalitis helps in preventing complications and reducing morbidity. This case is one such example of the same.

Ethical Compliance Statement

1) The authors confirm that the approval of an institutional review board was not required for this work.
2) Informed written consent was obtained from the patient for publication.

We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this work is consistent with those guidelines.

Financial Support And Sponsorship

Nil.

Conflicts Of Interest

There are no conflicts of interest

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