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Dr. Hashma Mohammed P
Junior Resident, Department of Paediatrics, Sree Balaji Medical College and Hospital, Chromepet, Chennai, Tamil Nadu, India

Nasin Usman
Assistant Professor, Department of Radiology, Sree Balaji Medical College and Hospital, Chromepet, Chennai, Tamil Nadu, India

Shanthi Ramesh
Professor, Department of Paediatrics, Sree Balaji Medical College and Hospital, Chromepet, Chennai, Tamil Nadu, India

Sundari S
HOD & Professor, Department of Paediatrics, Sree Balaji Medical College and Hospital, Chromepet, Chennai, Tamil Nadu, India

Corresponding Author:
Dr. Hashma Mohammed P
Junior Resident, Department of Paediatrics, Sree Balaji Medical College and Hospital, Chromepet, Chennai, Tamil Nadu, India

An interesting case of Anti-N-Methyl-D-Aspartate receptor encephalitis

Dr. Hashma Mohammed P, Nasin Usman, Shanthi Ramesh and Sundari S

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Abstract
Anti-N-Methyl-D-Aspartate Receptor Encephalitis (Anti NMDARE) is an immune mediated encephalitis. It is one of the most common cause of acute and sub-acute encephalopathy. Clinical features include behavioral disturbances, psychiatric manifestations, seizures, movement disorders [1]. Here we report a case of 3 year 1 month old girl who presented with difficulty in walking and involuntary movements. There were no other complaints, necessary investigations were sent which pointed towards anti NMDARE, awake EEG showed characteristic delta brush and CSF analysis showed NMDA receptor antibody was positive.

Keywords: Anti-NMDA receptor encephalitis, ataxia, choreiform movements, dystonia, delta brushes

Introduction
Anti-NMDA receptor encephalitis is an autoimmune neurological condition more preponderant in females [2]. It usually follows an upper respiratory tract infection.
It is characterised by production of antibodies against NMDA receptor targeting synaptic proteins [2]. It usually presents as a diagnostic dilemma as it presents with both neurological and psychiatric features [3]. When child presents with complaints of behavioral disturbance, abnormal movements, seizures with or without fever, viral encephalitis should be considered as the most common cause; When viral etiology cannot be confirmed, immune mediated encephalitis like autoimmune encephalitis and acute disseminated encephalitis are the next common cause [4]. The diagnostic test used is CSF analysis; EEG abnormality detected is generalised beta and delta slowing [4].
Ovarian teratoma is a common association therefore transvaginal ultrasonography must be performed [5]. Early and prompt diagnosis helps in reducing sequelae. Treatment is with immunosuppressive and Immunomodulatory therapies with steroids, IVIG (immunoglobulin) and plasmapheresis [5].

Case Report
We report a case of 3 year 1 month old girl child brought with complaints of walking difficulty and involuntary movements since 10 days. She also had choreiform movements of bilateral upper and lower limbs, swaying to sides while made to sit. No history of fever, rash, seizures, cough, breathing difficulty, joint pain, vomiting. No history of trauma. No history of similar complaints in the past. No history of past hospital admissions. She was born via normal vaginal delivery, her birth weight was 3.2 kg, uneventful post natal history. The child was immunized for age according to NIS. Developmentally normal child. On examination child was irritable, vitals were stable, CNS examination revealed hypotonia of bilateral upper and lower limbs, but deep tendon reflexes were normal. Higher mental functions and cranial were normal. On local examination there was no swelling or deformity, other systems were within normal limits. Possibility of Sydenham's chorea, acute disseminated encephalomyelitis (ADEM) and autoimmune encephalitis were considered. Neurometabolic diseases like organic academia, Wilson's disease, mitochondrial cytopathy and ataxia telangiectasia were also kept as rare possibilities as the baby was very young. Necessary investigations sent, hemogram, ESR, CRP, ASO titre, TFT, CPK came normal. MRI brain was also normal. ECHO was normal which ruled out rheumatic chorea. EEG showed slowing with 1-2 Hz delta waves with superimposed beta activity on both sides (delta brush).

Lumbar puncture was done and CSF sent for analysis including NMDAR antibody testing. Keeping autoimmune encephalitis as a provisional diagnosis, child was given pulse dose methyl prednisolone. CSF analysis showed normal picture, but the cell based immunofluorescence of CSF demonstrated antibodies against NMDA receptor. IV immunoglobulins (at 1 g/kg) was also given. USG abdomen was done to rule out teratoma of ovary. Following treatment child got symptomatically better in due course of time, started walking with support, involuntary movements reduced and was discharged on oral steroids. On subsequent reviews the child was clinically better; walking normally and with no involuntary movements.

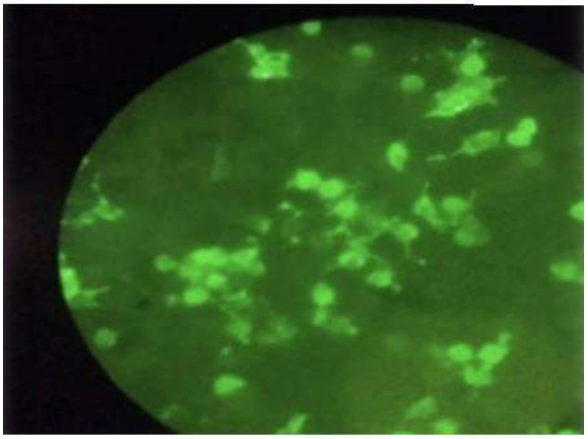

Fig 1: Cell based immunofluorescence showing positive anti-NMDA receptor antibody.

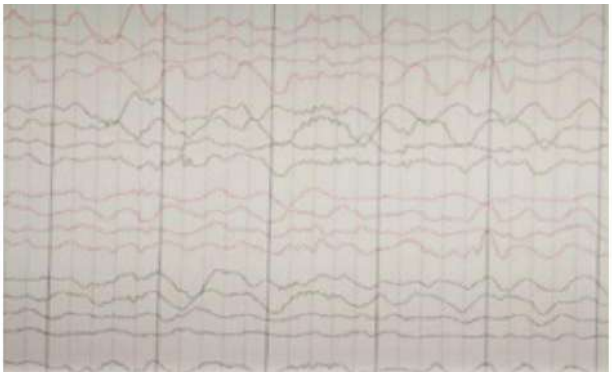

Fig 2: EEG Showing delta brush

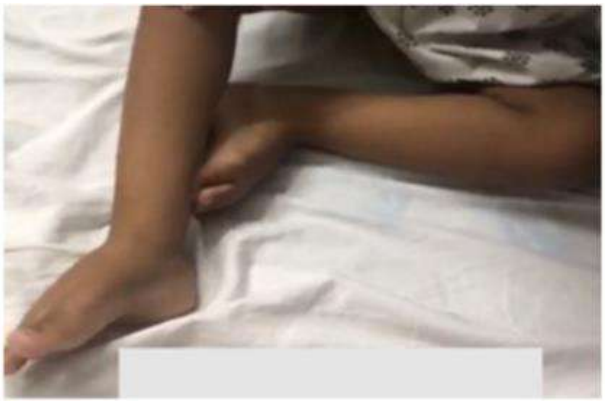

Fig 3: Dystonia of left foot

Discussion

Anti NMDARE was first described in 2007 by Dalmau [1]. In this condition IgG antibodies are targeted against GluN1 subunit of NMDA receptor. Following ADEM, it is the most common cause of auto immune encephalitis [6]. Anti NMDARE can be classified as paraneoplastic encephalitis, autoimmune encephalitis, immune mediated encephalitis but immune mechanism not studied. It has female preponderance. A common association seen is the teratoma of ovary [7].

Anti NMDARE may be seen in association with infections such as Herpes simplex virus, Mycoplasma pneumoniae, Influenza virus etc [7]. Clinical features can vary from flu like illness to behavioral disturbances, speech defects, psychiatric manifestations, autonomic instability.

MRI brain might be abnormal showing transient cortical and meningeal enhancement in a T2 FLAIR image. CSF may show a lymphocytic pleocytosis picture. Signature EEG finding is the presence of Delta brush- moderate to high amplitude 1-2 Hz delta waves superimposed with beta waves. Gold standard is demonstrating the anti NMDARE antibody in CSF using cell based immunofluorescent assay [8].

The case reported here is not typical as the child had only complaints of walking difficulty and behavioral disturbances, no seizures, fever, oro-facial dyskinesias. The behavioral disturbances will be severe usually but not in this case, the child was irritable but her higher mental functions were all intact[8].

Treatment includes methyl prednisolone and IVIG. Diagnosis early and initiation of treatment is the key. Plasma exchange can made use of in advanced situations.

Next line of treatment includes Rituximab and cyclophosphamide. For at least one year steroids or azathioprine or mycophenolate mofetil should be continued as it prevents recurrences [9].

It has a mortality rate of 7%. Early diagnosis and treatment initiation is the key to better prognosis [10].

Conclusion

The most common causes of immune mediated encephalitis is anti NMDA receptor encephalitis. Definitive diagnosis is by demonstration of anti NMDA receptor antibody in CSF [9]. Other diagnostic tools include cell based immunofluorescent assay of blood. Widespread availability of the above tests have increase the detection rates. Early diagnosis and aggressive immunotherapy has improved the outcome and reduced the relapses in this eminently treatable disease [10]. From this case we can identify an atypical presentation of anti NMDA receptor encephalitis.

Conflict of Interest

Not available

Financial Support

Not available

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