MATHEWS JOURNAL OF CYTOLOGY & HISTOLOGY

MATHEWS Open Access Journals

Case Report

ISSN: 2577-4158

Vol No: 3, Issue: 2

Received Date: Nov 21, 2019

Published Date: Dec 6, 2019

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Anti-N-Methyl-d-Aspartate Receptor Encephalitis: A Single Case Report

ABSTRACT

Anti-N-methyl-d-aspartate receptor (anti-NMDAR) encephalitis usually presents with behavioral manifestations and subsequently emerge by an acute or subacute movement disorders. Although in adults the movement disorders seem to appear after a prodromal period with psychiatric features, in children diagnosed with anti-NMDAR encephalitis they may be the initial manifestation. We report a case of a 7 years old male with history of choreiform movements and two isolated seizures episodes.

Keywords: Encephalitis; Anti-N-Methyl-d-Aspartate Receptor; Seizures; Choreiform Movements.

INTRODUCTION

Anti-NMDAR encephalitis is autoimmune limbic encephalitis related with antibodies against NR1 or NR2 subunits of the NMDA receptor [1-3]. There is a suggestive correlation between underlying malignancy and also predominance in women [3]. The initial manifestations are often psychiatric (auditory and visual hallucinations, delusions, and agitation), as the syndrome go on other features as autonomic dysfunction, seizures, motor disturbance (fluctuating from dyskinesia to catatonia) and compromised consciousness, leading to coma may also present. We describe a case of anti-NMDAR encephalitis presenting with choreiform movements and punctuated seizures episodes.

CASE REPORT

A 7-year-old Hispanic male presented for evaluation of 4-5 days worsening right-sided unilateral arm and leg, non-rhythmic involuntary movements and progressive weakness. On examination we observed a purposeless, non-rhythmic, athetoic dyskinesia on the right upper and lower extremities. Speech, attention, verbal expression and comprehension, cranial nerves, muscle strength and tone, reflexes and sensory pathway were intact. Finger to nose and heel to shin was abnormal. Right arm with pronator drift. The patient was admitted to the pediatric floor at this institution, where ancillary tests such as CBC, CMP, ASO, DNAse titers, TSH, C reactive protein, Lyme antibody, Mycoplasma pneumonia antibody, strep culture, ESR, EKG and MRI were ordered and came back normal. Neurology and Infection disease (ID) teams were consulted. He was started on clonidine as recommended by neurology and had some improvement. The ID team

DOI: https://doi.org/10.30654/MJCH.10012

considered Sydenham's chorea less likely due to normal MRI, inflammatory markers and patient history, therefore he was discharged with a follow up in two weeks at neurology clinic.

A week later, patient had worsening choreoathetoid right extremity movements. He also began to complain of left foot and hand numbness, and intermittent, dull headaches 2/10. Those symptoms brought him to the ER, where he experienced 2 seizures episodes characterized by rapid blinking, smacking of the lips, stiffening of his arms and legs. His eyes were deviated to the right, without loss of conscience. They lasted 2 minutes and 30 seconds, respectively and self resolved. On examination we observed a purposeless, non-rhythmic, athetoic dyskinesia on the right upper and bilateral lower extremities that persisted during sleep, corresponding to choreia and 3-4 beats of clonus appreciated in LE bilaterally. The patient was admitted to the pediatric intensive care unit (PICU). Cerebral spinal fluid (CSF) studies, paraneoplastic panel, NMDA antibodies, Anti-Basal Ganglia antibodies, ANNA-1 (Antineural Nuclear Antibodies type 1), CRMP-5, IgG-ANA, Anti-ds-DNA, IL-2 and IL-6 were ordered. Neurology recommended starting levotiracetam for the seizures, and carbamazepine associated with a 3 day course of intravenous methylprednisolone, followed by solumedrol dose pack for the chorea. A continuous vEEG, MRA, MRV was also ordered and came back normal.

He had a mildly improvement at the beginning and after two days of treatment he could sleep restfully. Carbamazepine was discontinued due to nauseas and lorazepam was initiated. NMDA antibodies came back positive.

DISCUSSION

Anti-NMDA receptor encephalitis is a rare neuroautoimmune disorder with a wide range of symptomatic presentation, a high mortality rate and a potential for treatment [4]. It is easily diagnosed by screening for antibodies to the NMDA receptor subunit in the serum or CSF sample. Early recognition and treatment may have significant prognostic implications.

Our patient particularly illustrated hyperkinetic movement disorder associated with anti-NMDAR encephalitis. Usually, chorea is dominant in younger patients and therefore anti-NMDAR chorea should be on a higher consideration on any child presenting choreiform symptoms. Symptoms may include oral facial dyskinesias, choreathetosis, myoclonus, dystonia, and ataxia [2,4,5]. Generally there is a non-specific prodrome phase, followed by psychiatric prominent symptoms, cognitive, motor and then autonomic dysfuntion [2].

Beside the positive NMDAR antibody in the serum or/and in CSF, there are some other tests that could present with some abnormality. CSF could describe a lymphocytic pleocytosis and CSF-specific oligoclonal bands [2,6]. Electroencephalogram (EEG) regularly shows diffuse background slowing or focal slow waves frequently in the fronto-temporal regions [1,2]. Brain magnetic resonance imaging (MRI) although are more commonly reported as normal, may show hypersensitivities in multiple regions including hippocampi, brainstem, basal ganglia, cerebellar and cerebral cortex) [7].

The treatment is most often obtained with immunotherapy, first or second-line. The first-line immunotherapy includes corticosteroids, intravenous immunoglobulin or plasma exchange. Second-line immunotherapy, rituximab or cyclophosphamide, may be used in refractory cases, individuals who received a delay diagnosis or in those with no tumor [4].

CONCLUSION

The phenomenology of movement disorders in children affected with anti-NMDAR encephalitis may be difficult to characterize, although chorea usually is more frequent in younger children. These hyperkinetic movements tend to improve with first-line immunotherapy.

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