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CASE REPORT

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A Rare Case Report of Doose Syndrome with Attention Deficit Hyperactivity Disorder

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Abstract

Doose syndrome also known as myoclonic-astatic epilepsy of early childhood (MAE), a rarely diagnosed entity, a kind of cryptogenic epilepsy with myoclonic astatic seizures as per the International League against Epilepsy (ILAE). A 9 year-old male child was bought by his mother with complaints of hyperactivity, excessive talking, and difficulty in paying attention noticed since 4 years of age. At 4 years of age, child had multiple episodes of unprovoked seizures on different occasions for which he was evaluated. EEG showed Myoclonic jerks in sleep with generalized poly spikes. The MRI Brain was normal. Genetic tests done showed no reportable genomic sequencing. The child was started on multiple anticonvulsants and advised for Ketogenic diet. At our hospital, a detailed psychological evaluation was done. The child was evaluated using Vanderbilt ADHD diagnostic parent rating scale and diagnosed with ADHD. Cognitive Behaviour Therapy was initiated. There was significant reduction in the scores following intervention indicating clinical improvement and quality of life of the child. Conclusion: This case report concludes that a holistic approach is needed in treating such cases to improve the quality of life. Most of the time behavioural problems remain unaddressed in search of primary ethology.

Keywords: Doose; Epilepsy; Syndrome

1 Introduction

Doose syndrome also known as myoclonic-astatic epilepsy of early childhood (MAE), a rarely diagnosed entity, a kind of cryptogenic epilepsy with myoclonic astatic seizures as per the International League against Epilepsy (ILAE).[1] MAE, which accounts for 1%-2.2% of childhood-onset epilepsy cases, is characterized by normal development before seizure onset, which generally occurs between 7 months to 6 years of age.[2] The defining characteristics include normal development prior to seizure onset, no identifiable cause of seizures, onset of myoclonic-atonic seizures between the ages of 7 months and 6 years, a 2:1 male: female ratio,

multiple generalized seizure types including recurrent status epilepticus, and initially normal electroencephalogram (EEG; or centroparietal theta) followed by generalized polyspike-and-wave epileptiform activity.[3] Here by we are reporting a rare case of Doose syndrome with Attention Deficit Hyperactivity Disorder (ADHD).

2 Case Report

A 9 year-old male child was bought by his mother with complaints of hyperactivity, excessive talking, and difficulty in paying attention noticed since 4 years of age. He also has difficulty in waiting for his turn and is always disturbing his peer group. His teachers noticed that he is slow in copying notes and is forgetful in his daily activities. At 4 years of age, child had multiple episodes of unprovoked seizures on different occasions for which he was evaluated. EEG showed Myoclonic jerks in sleep with generalized poly spikes. The MRI Brain was normal. Inborn errors of metabolism screening were normal. Genetic tests done showed no reportable genomic sequencing. The child was started on multiple anticonvulsants and advised for Ketogenic diet. His parents noticed increasing hyperactivity, inattention and impulsivity behaviour. There were regular complaints from school regarding his inattention.

At our hospital, a detailed psychological evaluation was done. The child was evaluated using Vanderbilt ADHD diagnostic parent rating scale and diagnosed with ADHD. The report was as follows:

Table 1. Scores Pre-Intervention

Pre intervention or Baseline Scores	
Inattention	9
Impulsivity/Hyperactivity	9
Total Score	18

Cognitive Behaviour Therapy was initiated. It was given for 45 minutes once a week. Twelve such sessions were conducted. These strategies helped him to increase his attention span, self-control and problem-solving abilities.

Table 2. The attention enhancement techniques used

Activities	Report
1. Grain Sorting 2. Coloring 3. Painting 4. Letter Cancellation 5. Digit Cancellation	His attention span was only 02 minutes 32 seconds pre intervention Post therapy ie by 12th session his attention span was 14 minutes15 seconds.

- **Parental activities that helped the child to modify the behaviors were such as**

1. Maintaining a daily Schedule
2. Keeping distractions to a minimum
3. Setting small reachable goals
4. Rewarding positive behaviour
5. Using charts and checklists to help the child stay on task
6. Finding activities in which the child can be successful (sports/games)
7. Using calm discipline (time out being avoided, distraction, removing the child from the situation)

Table 3. To improve his self-control self-instruction and covert modelling technique was used

Academic Activities	Intervention	Effect seen	Result
Delay in copying Notes from the Black Board.	The Therapist modelled him on how to talk to himself i.e. give instructions to self without uttering but only in thought "I will complete copying my work from the black board today".	The child would talk to self in thought that he should complete copying the notes from black board without spelling mistakes and would try to do it as much as Possible.	The child do this task to an extent of 65% by the end of 12th session of therapy.

Table 4. Role Playing

Behavior	Therapist	Result
Blurt out answers to questions before having finished asking questions.	The Therapist demonstrates "blurting out answers with impulsivity" while doing role playing with other people and also the consequences were projected.	Child showed improvement by the end of 12th session of therapy.

Table 5. Scores of Mast L Post Intervention

Post Intervention Scores	
Inattention	4
Impulsivity/Hyperactivity	3
Total Score	7

There was significant reduction in the scores following intervention indicating clinical improvement and quality of life of the child.

3 Discussion

Doose syndrome, also known as epilepsy with myoclonic atonic seizure (EMAS), was previously called myoclonic

astatic epilepsy (MAE), a rare childhood EE. First reported by Doose in 1970, the International League Against Epilepsy (ILAE) in 2010 changed its name to epilepsy with myoclonus-atonic seizures based on the characteristics of epileptic seizures.[4] A similar case was reported by Singh A et al.,in a 2 year old child with isolated speech delay and seizures.[1] Hinokuma N et al.[2] reported 29 cases of Doose syndrome with their genetic analysis. They reported that these patients had genetic heterogeneity and Febrile seizures prior to epileptic seizures and myoclonic-atonic seizure at onset indicated a genetic predisposition.[2] Magsi R et al.[5] reported Doose Syndrome in an 8-year-old female who presented with multiple types of seizures with Genetic testing showing SUOX gene mutation.[5]

About 80–90% of children with Doose syndrome may exhibit normal cognition or only minimal cognitive impairment.[1] These children can have behavioural disorders also and such problems may not be addressed adequately by parents or treating physicians. As in the present case, the child was initially treated only for seizures without psychological intervention. This did not make difference in quality of child's life until the ADHD was treated. Currently child is under follow-up with clinical improvement.

4 Conclusion

This case report of a 9-year-old male with Doose Syndrome who has seizures and ADHD without remarkable genetic sequencing concludes that, a holistic approach is needed in treating such cases to improve the quality of life. Most of the time behavioural problems remain unaddressed in search of primary ethology.

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