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CASE REPORT OF WEST SYNDROME: A RARE AND EARLY ONSET EPILEPTIC SYNDROME

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West Syndrome, Seizures, Hypsarrhythmia, Developmental delay

Abstract
This is the case report of a 2 years old female child who presented with the chief complaints of seizures like activity multiple times since 6 months and developmental delay. The Magnetic resonance imaging of brain revealed no abnormalities, but the Electro Encephalogram showed abnormal record with Hypsarrhythmia and generalized polyspike wave complexes. The final diagnosis was West Syndrome and the patient is treated with Sodium Valproate and Prednisolone

Introduction

West syndrome is a severe epileptic syndrome which is characterized by the triad of infantile spasms, hypsarrhythmia, and mental retardation. The International League Against Epilepsy's (ILAE) revised the classification and terminology of seizures and epilepsies which were published in 2010, terms West syndrome as an electroclinical syndrome with onset in infancy, and classified it as a form of seizure.[1] Dr W J West was the first to give the comprehensive picture of infantile spasms that happened in his own child.[2] The pathophysiological mechanism defines that the effect of various stressors in the immature brain produces an atypical and increased production of corticotropin - releasing hormone (CRH)which results in infantile spasms.[3] The incidence of West syndrome in early stages of life is about 2 to 3.5 per 1000 live births. [4] The prognosis for the West Syndrome patients is poor and takes time, which is directly linked with the etiology of the disease. [5, 6] The commonly used first – line drugs are ACTH, Vigabatrin, Prednisolone and Sodium Valproate.[7]

Case Report

A two year old female child of weight 8.4 kgs, the first born of a non – consanguineous parentage who is immunized completely according to the immunization schedule was admitted into the pediatrics department our hospital with the chief complaints - seizures like activity multiple times since 6 months She was a full term normal delivery baby.

The general examination revealed that her vitals were normal, but she looks pale, there are no significant abnormalities were noticed other than that. There was a delay in the development of the patient with the developmental quotient of 54%. The laboratory investigations revealed Hemoglobin was 10 g/dl, and the remaining lab parameters like complete blood count, Liver functions tests, Renal function tests were normal. EEG revealed abnormal record with hypsarrhythmia and generalized polyspike wave complexes. MRI of the Brain showed no abnormalities. The final diagnosis was West Syndrome.

Initially patient was started on Inj. Midazolam 2ml slow IV SOS. Even after two days there was no improvement in the condition of the patient. Then, on third day the patient was prescribed with Syrup Sodium Valproate 2.5ml PO BD and Tab Prednisolone 2mg/kg/day for two weeks. After two weeks there was a significant improvement in the patient condition, but EEG revealed intermittent generalized polyspike wave complex. There was no improvement in her developmental quotient from the previous value. The patient was also prescribed with Tab B – complex PO OD, Tab Ranitidine 150mg 1/3rd tab PO OD, Cap A & D PO OD, Syp Iron 10 drops PO OD and Syp Calcium 3ml PO BD for two weeks.

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Discussion

This is a case report is on West Syndrome, seen in a female child of age two years. West Syndrome is Characterized by three features that are Salaam seizures, Hypsarrhythmia and developmental delay as seen in this case.

The normal Anti – epileptic drugs does not work in the case of Salaam seizures or infantile spasms. The commonly used first – line choice of drugs are ACTH, Vigabatrin, Prednisolone and Sodium Valproate.[7] In this case the patient was prescribed with Sodium Valproate and Prednisolone. As there is no availability of ACTH in the hospital the patient was prescribed with Sodium Valproate instead of ACTH which is the best choice. ACTH therapy has been reported to succeed in 60 – 80% of the patients.[8] Sodium Valproate was proved to be effective in improving hypsarrhythmia or salaam seizures.[9, 10] A study on the current treatment of West Syndrome by the Japanese Epilepsy Society disclosed, vitamin B6 was the best choice. The second line choice was vitamin B6 in combination with Sodium Valproate/ Sodium Valproate monotherapy and the third choice was ACTH.[11] According to Elterman et al, vigabatrin is the best choice for treating West Syndrome which is due to tuberous sclerosis, the recovery rate from West Syndrome rages from 91% to 100%.[12] Even though there is an availability of drugs for treating West Syndrome, the prognosis is slow.

Conclusion

The West Syndrome is a rare and severe epileptic form seen in infants. Even though there are many drugs which are effective in treating this disease, there are still challenges in the diagnosis, assessment and management which require further studies. Early detection and referral to the neurologist may increase the prognosis of the disease and helps in the effective treatment.

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List of Abbreviations:
ACTH – Adenocorticotrophic hormone
BD – Twice a day
EEG – Electroencephalogram
IV – Intravenous
MRI – Magnetic resonance imaging
OD – Once a day
PO – Orally
SOS - If necessary

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