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WEST SYNDROME: A RARE EPILEPTIC SYNDROME OF INFANCY

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ABSTRACT West syndrome (WS) is one of the causes of refractory epilepsy with infantile onset and characterized by epileptic spasms and neuro-developmental problems. We report a case of 6 month old boy with history of severe asphyxia at birth who presented with complaint of irritability, jerky head nodding and developmental delay. Neuro imaging had features suggestive of severe acute hypoxic injury in a term child and EEG showed hypsarrythmia. Later diagnosed as a case of West syndrome and managed with ACTH and prednisolone therapy.

KEYWORDS : West Syndrome, irritability, jerky head nodding, hypsarrythmia,ACTH

INTRODUCTION

West syndrome is a rare form of epilepsy usually affecting children in early infancy. The peak age of onset is between 3 and 7 months; onset after 18 months is rare, though onset up to 4 years of age has been reported [1].

It is one of the common causes of refractory epilepsy with infantile onset and characterized by epileptic spasms, neuro-developmental problems due to cessation and regression of development and common association with electroencephalogram finding of hypsarrythmia [1,2]. It was first described by Dr. Williams James West 75 years ago in his own son but still its diagnosis, evaluation and management remains to be a challenge for medical professionals [3]. Incidence of West syndrome in children ranges from 2 to 3.5 per 1000 thousand live births [4]. West syndrome comprises of characteristic triad of infantile spasms, developmental delay and hypsarrythmia on EEG [5]. Prognosis of children is usually poor despite proper treatment [6].

Perinatal asphyxia (61.4%), neonatal sepsis/meningitis (10.6%), and postnatal meningitis (11.4%) were the predominant causes. The etiology could not be ascertained in 16.6% of children.[7]

CASE STUDY

We present a case of six month old male who is the first issue of non consanguineous parents with history of severe birth asphyxia (APGAR 1 at birth and 3 at one minute). Patient was intubated just after birth and was kept on positive pressure ventilation for almost 1 hour. Patient had an episode of seizure within first hour of birth and was given intravenous Levetiracetam .Seizures were subsided but patient was neurologically dull and reflexes were weak which improved gradually over a period of time. At around third week of life exclusive breastfeeding was established. MRI Brain was done before discharge (at 28th day of life) which showed increased T1W signal within the lentiform nucleus, thalami and the crura of midbrain. Cortical highlighting was seen in high frontal and medial temporal lobes and loss of signal in posterior limb of internal capsule. These features corresponded with those of acute severe hypoxic injury in a term child. EEG was also done at the same time but it was inconclusive. Patient was hence discharged on maintenance oral dose of Levera and was closely monitored on follow up. Developmemtal delay was noticed as child did not achieve neck holding even at 5 months. At around 6 months of life the mother reported that the child is having jerk like movements of limbs with head nodding and irritability. Serum levels of levetiracetam were checked which came out to be in normal range. Another anti epileptic drug (sodium valproate) was added and patient was called after 7 days.

Child showed no improvement and was referred to Pediatric neurologist for opinion. Repeat EEG was done which showed hypsarrythmia pattern and with supporting clinical evidence of jerk like movements (infantile spasm) diagnosis of West syndrome was made. Oral prednisolone was started at 2 mg/kg/day and was given for 2 weeks with gradual tapering. Even after oral steroid therapy patient showed no improvement and hence Inj. ACTH at dose of 40 IU/day was started and was given intramuscularly for 4 weeks. On completion of ACTH therapy there was significant decrease in frequency of head nodding and spasm but EEG continue to show intermittent generalized rapid spike bursts. There was no improvement in developmental quotient. Patient is now on Syrup Sodium Valproate (at 30 mg/kg/day), Tablet Clobazam (2.5 mg BD) and other supplements and is monitored closely by pediatric neurologist team for growth , development and neurological issues.

DISCUSSION

We described a case of 6 month old male infant with West syndrome. Infantile spasm, developmental delay and hypsarrythmia in EEG are the characteristic triad of West syndrome which was seen in our case. Infantile spasms are resistant to routine anti epileptic drugs like Phenobarbitone, phenytoin, Levetiracetam and sodium valproate but responded to ACTH therapy [8]. Although EEG and neuro developmental behavior did not show much changes even after completion of ACTH therapy.

These findings were similar to the results of Richard et al who studied west syndrome outcome in 214 children. On the contrary, Riikonen in 1996 and Kimura et al in 1999 when managed there cases with low dose ACTH therapy showed significant improvement in controlling of seizures and developmental delay [9,10]. Later Sharma et al in 2008 suggested that if ACTH therapy is given in first month of occurrence of infantile spasm outcome can be not as bad as it has been known [11].

CONCLUSIONS

West syndrome is a rare form of epilepsy with poor outcome. Birth asphyxia is one of the most common cause and hence any follow up case of birth asphyxia if presents with abnormal or jerk like movements; then West syndrome should be ruled out with high index of suspicion. Infantile spasm is resistant to phenytoin, levetiracetam and sodium valproate. ACTH therapy is effective means to control spasms but has little role in improvement of neuro developmental outcome. There is an absolute need for further studies which can discover the early diagnostic measures and more effective treatment.

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