

West Syndrome in a Nepalese Boy: A Case Report

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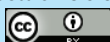
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ABSTRACT

West syndrome is characterized by the triad of epileptic spasms, arrest of psychomotor development, and hypsarrhythmia. It usually begins during the first year of life. We present a case of West syndrome in a child of 2 years.

INTRODUCTION

West syndrome (WS) is a unique epilepsy disorder characterized by a triad of infantile spasms, hypsarrhythmia, and arrest of psychomotor development.¹ 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.² It was first described by Dr. Williams James West (1841) in his own son.³ Its incidence is approximately 0.249 cases per 1000 live births.³ It comprises epileptic spasms, arrest of psychomotor development, and hypsarrhythmia, although the second feature may be absent.⁴ Most characteristic pattern in electroencephalography (EEG) is hypsarrhythmia, or one of its variants.³ We report a case of West Syndrome that presented to the Pediatrics outpatient department (OPD) in Nepalgunj Medical College, Kohalpur.

CASE REPORT

Mr. JB, 2-years male patient reported to the Pediatric OPD with irritability, bowing, and relaxing of the head and trunk with stretching of limbs several times a day since 9 months of age. The child appeared frightened and screamed. Each attack lasted for about one to two minutes and was used to get resolved on its own. There were 10-15 attacks each day. Consciousness was preserved and no other associated symptoms were seen. The child is full-term normal delivery with an uneventful prenatal, natal, and postnatal period. No history of head injury and there were no similar episodes in any family members.

On clinical examination, the patient was conscious, afebrile with a normal respiratory rate. His pulse rate and blood pressure were below the 90th percentile for age. No pallor/icterus/cyanosis/pedal edema was seen. However, he had dental caries and tooth decay. The cardiovascular sounds S1 and S2 were normal and P/A was soft, with

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no organomegaly. The tone was normal in all four limbs with bilateral extensor plantar. Gross developmental delay was present. Complete blood picture, serum electrolytes, calcium, and blood sugar tests were reported to be normal. CT head was normal. EEG showed high amplitude multi-focal spikes or hypsarrhythmia (Figure 1). He was diagnosed with West Syndrome.

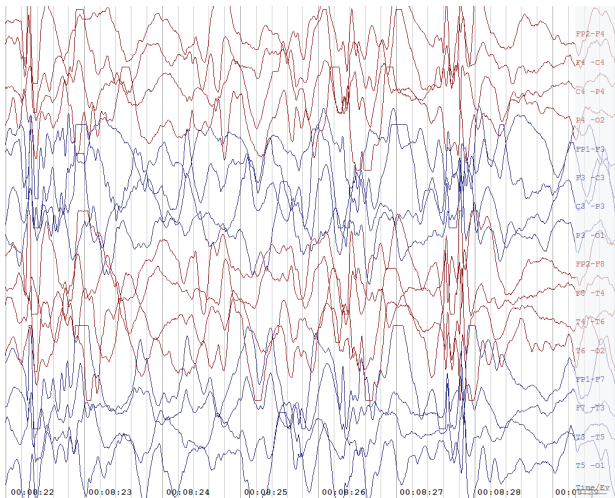


Figure 1: EEG of the child displayed at displayed at 7.5 μ V/mm, LFF= 1Hz, HFF=70 Hz, timebase = 30 mm/sec.

Sodium valproate was started at 30 mg/kg daily. Significant response was appreciated on a one-week follow-up. Episodes of attacks decreased in frequency. Frightening and screaming improved after starting therapy. However, bowing and relaxing of the head and trunk, stretching of limbs persisted. During a three-week follow-up the child recovered significantly.

DISCUSSION

WS is a rare and severe childhood epilepsy syndrome. WS and Infantile Spasms (IS) are often used as synonyms.⁵ An infantile spasm is a distinct epileptic disorder that occurs in infancy and early childhood, and its clinical features are well documented.⁶ This syndrome is also referred to in the literature as massive spasms, salaam tics, infantile myoclonic seizures, or Blitz-Nick-Salaam Krämpfe.⁷ At least 76 other different synonyms have also been used to describe it such as massive spasms, infantile massive spasms, flexion spasms, jack-knife convulsions, nodding convulsions, and lightning fits.⁸

WS can be classified into two categories based on the etiology of IS; symptomatic spasms (etiology can be identified in more than 70% of cases, such as hypoxic-ischemic encephalopathy), and cryptogenic (etiological diagnosis cannot be identified and normal development precedes the spasms).³ Among patients with cryptogenic etiology, there is a subgroup that shows normal psychomotor development and a complete cessation of epilepsy. This group can be called idiopathic West syndrome.⁴ Its incidence is approximately 0.249 cases per 1000 live births, most cases begin during the first year of life (peak incidence at 6 months), and familial occurrence is rare. It is more frequent in males; it is responsible for 2-10% of the causes of childhood epilepsies.³

The most typical manifestations are spasms. Spasms consist of brief muscle extension, flexion, or mixed flexion-extension jerks involving the neck, trunk, or extremities. Lateralizing or localizing clinical features may be present during the spasms.² Spasms often appear in clusters, their number may vary and can also during sleep or awakening. Irritability or crying is frequent during or after the spasms.⁷ Marked abnormalities of the EEG are usually present. In idiopathic cases the initial EEG, if the onset of symptoms is very recent, can be normal or borderline. The most typical finding is called hypsarrhythmia. It consists of a mixed pattern of high voltage slow waves mixed with spikes or polyspikes. These abnormalities are continuous or almost continuous.¹ In our case also, this characteristic feature on EEG was demonstrated.

There are two drugs with evidence of efficacy in the treatment of WS: ACTH hormone and Vigabatrin. Pyridoxine, Valproic Acid, Lamotrigine, Topiramate, Prednisone, Phenobarbitone, Carbamazepine, or Phenytoin are also used.⁶ There is growing evidence that some of the newer drugs, high-dose intravenous immunoglobulin, and the ketogenic diet may be effective.⁸ WS has unpredictable outcome.⁹ The disease may recover spontaneously, may remain static or progress as intractable seizures or evolve into other epileptic syndromes like Lennox Gaustat or Davert syndrome.¹⁰ However, early diagnosis with careful evaluation and proper therapy can lead to normal development or a much-improved situation in some cases, especially in cases with unknown etiology.^{11,12}

Our case had above-described spasms since 9 months of age but presented to our pediatrics

OPD only at the age of 2 years. There was no history of previous visits to a pediatrician or neurologist for the problem. This shows a general lack of awareness in the parents about epilepsy and the availability of various management options. As in our case, the utilization of EEG plays a central role in the diagnosis and management of patients with epilepsy, especially in children.^{13,14}

CONCLUSIONS

Diagnosing West Syndrome in a child can sometimes be challenging as developmental parameters may be difficult to access. Hence a strong suspicion, utilization of EEG and regular follow-up are the keys to correct diagnosis and management.

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