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Case Report

West Syndrome – Infantile Spasms: A Rare Paediatric Case Report

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Abstract

West syndrome (WS) is a rare, severe form of epilepsy with onset in infancy and early childhood. It involves clustered epileptic spasm episodes, the aberrant interictal electroencephalogram pattern known as hypsarrhythmia, and neuropsychomotor delay. In this view, we present a case of a 6-year-old female child a known case of west syndrome, global developmental delay, hypothyroidism, and post-operative laparoscopic gastrotomy performed under general anaesthesia on March 31/2022, in view of recurrent aspiration and came with the chief complaints of seizure 6-7 episodes per day, decreased urine output, constipation for two days, straining to pass urine, and three episodes of vomiting. The history included that the baby cried immediately after birth, seizure on day 5 of life, and was shifted to the Neonatal intensive care unit (NICU), admitted there for 5 days because of respiratory distress syndrome and hyperalbuminemia. During their stay in the hospital, the child has been prescribed Lamotrigine 50mg, Levocarnitine 500mg, Levetiracetam 250mg, Clobazam 5mg, Phenobarbital 60mg, Sodium Valproate 100mg, Thyronorm 50mcg. Now the patient is symptomatically better. This case report intends to sensitize physicians in this region toward this rare neurological syndrome.

Keywords: Hypsarrhythmia, Infantile, Seizure, West syndrome.

Introduction

West Syndrome (WS) and Infantile Spasms (IS) are often used as synonyms.[1] WS is a severe and infrequent type of childhood epilepsy. Even if part of the symptoms is absent, WS is defined by a triad of symptoms, including epileptic spasms, stop or regression of psychomotor development, and hypsarrhythmia on interictal electroencephalography (EEG).[2,3] William West, an English doctor, first characterized the disease in 1841. Dr. Gastaut, Poirier, and Pampiglione coined the term "West Syndrome" in the early 1960s. [4] WS affects between 2 to 3.5 out of every 10,000 live births, with males being affected 60:40 more frequently than females.[5-7] It is divided into three primary categories based on the linked etiological factors symptomatic, idiopathic, and cryptogenic.[8] The etiological cause has an impact on the long-term prognosis. Compared to children with symptomatic infantile spasms, those with idiopathic WS have a better prognosis. [9] Mutations in the Aristaless Related Homeobox (ARX) gene and the cyclin-dependent kinase-like 5 (CDKL5) gene on the X chromosome's short arm have been related to an increase in infantile spasms.[9] According to the most recent recommendations from the American Academy of Neurology and the Child Neurology Society for the medicinal treatment of WS, which examined the available evidence as of 2004, adrenocorticotropic hormone (ACTH) is most likely effective and Vigabatrin may be effective in the cessation of spasms of hypsarrhythmia.[10] The following anti-epileptic drugs may also seem beneficial for WS: Valproate, Topiramate, Pyridoxine, Zonisamide, Lobazam, or Clonazepam. Sometimes multiple therapies are administered simultaneously, such as ACTH a, Vigabatrin, hydrocortisone, and valproate. In some studies, a high-fat, sufficient-protein, low-carbohydrate diet (also known as a ketogenic diet) has produced positive results; however, its effectiveness in treating infantile spasms has not yet been confirmed.[8] This case report intends to sensitize physicians in this region toward this rare neurological syndrome.

Patient information

A 6-year-old female child, a product of non-consanguineous marriage, who is a known case of WS, global developmental delay, hypothyroidism, and post-operative laparoscopic gastrotomy performed under general anaesthesia on March 31/2022, in view of recurrent aspiration, presented in the medical outpatient department of Sri Ramachandra medical college and hospital with the chief complaints of seizure 6-7 episodes per day, decreased urine output, constipation for two days, straining to pass urine, and three episodes of vomiting. The history was narrated by the mother of the child. A 2kg female child born to a primigravida mother at term gestational age via lower segment caesarean section (LSCS). The history included that the baby cried immediately after birth, a history of 1st episode seizure on day five of life, and was shifted to the Neonatal intensive care unit (NICU), admitted there for 5 days

because of respiratory distress syndrome and hyperalbuminemia. No history of seizures or developmental delay in the family.

Clinical findings

Following her admission, the child is drowsy and dull, a general examination was performed and respiratory rate (RR) - 24 breaths/min, blood pressure (BP) - 90/60 mmHg, and pulse rate of 100 beats/min. On systemic examination, the cardiovascular system both heart sounds normal, central nervous system exhibited microcephaly positive and hypertonia in all 4 limbs, respiratory system was discovered to be normal. On laboratory examination, everything is found to be normal except the thyroid function test revealed elevated TSH of 8.580 suggestive of hypothyroidism.

Diagnostic assessment

EEG: Abnormal EEG with multifocal and generalized discharges as described may suggest an atypical variant of Hypsarrythmia. (Supplementary file 1)

MRI: Shows periventricular white matter hyperintensities. Evidence of gliosis in the right posterior parietal and occipital region with mild dilation of the occipital horn of right lateral ventricle.

Based on medical history, clinical examination, and laboratory findings; the confirmatory diagnosis was "WEST SYNDROME"

Therapeutic intervention

Day 1 – She had complaints of seizures 6-7 episodes per day, decreased urine output, constipation for two days, straining to pass urine, and three episodes of vomiting. Upon admission, the child has prescribed the following medication: Tablet Sodium valproate 100mg TDS, Clobazam 5mg BD, Levocarnitine 500mg BD, Lamotrigine 50mg BD, Levetiracetam 250mg BD, Junior lanzol 15mg OD, Phenobarbitone 60mg HS, Pyridoxine 100mg OD, Zonisamide 25mg-0-50mg, Thyronorm 75mcg on Monday, Thursday and 50mcg OD on Friday, Saturday, & Sunday.

Day 2 – The child reviewed. No complaints of vomiting and seizure after admission. The patient was advised to continue the same drugs as per the physician's order.

Day 3 - No new complaints were noted. Thyroid Function Test(TFT) and ammonia were repeated. Paediatric endocrinology review was sent. The patient was advised to continue the same drugs as per the physician's order.

Day 4 - The child reviewed one episode of vomiting and 4 episodes of seizures. Follow for TFT. Levocarnitine 500mg BD is switched to 250mg of Levocarnitine. Thyronorm 50mg is stopped. The patient was advised to continue the same drugs as per the physician's order.

Day 5 – The child reviewed, 3 – 4 episodes of seizure, continue anti-epileptic drugs as prescribed by the physician. A review of paediatric endocrinology found that she had congenital hypothyroidism and increase the 75mcg OD dose of Thyronorm.

Day 6 – The patient is symptomatically better and discharge with adequate counselling. To continue the discharge medication as per the physician's advice. Drugs: Tablet Sodium valproate 100mg, Clobazam 5mg, Levocarnitine 250mg, Lamotrigine 50mg Levetiracetam 250mg, Junior lanzol 15mg, Phenobarbitone 60mg, Zonisamide 25mg-0-50mg, Thyronorm 75mcg, Pyridoxine 100mg, and syrup cremaffin 10ml.

Follow-up and outcomes

At the time of admission, she had complaints of seizures, decreased urine output, constipation, and vomiting. Now the child's symptoms improved, and she is symptomatically better. Her mother was instructed to use the medication as prescribed, and the patient-caregiver was given the necessary counseling.

Discussion

Although WS was first described over 160 years ago, diagnosing, assessing, and managing the condition continue to provide numerous difficulties to healthcare professionals and families that are impacted.[11] The onset of infantile spasms varies from the first week of life to 3 years of age, with a peak at 6 months, and usually ceases before 5 years of age.[9] contrarily the child in our case was diagnosed with WS at the age of 6 years old. The various etiological causes of West syndrome trigger a stress response, which increases the synthesis of corticotrophin-releasing hormone (CRH) from the immature brain, culminating in epileptic spasms. Through a negative feedback mechanism, ACTH inhibits the effect of CRH, which is an established therapy for the syndrome.[12] In our case, the child was treated with Sodium valproate, Clobazam, Levocarnitine, Lamotrigine, Levetiracetam, Junior lanzol, Phenobarbitone, Zonisamide, Thyronorm, Pyridoxine, and syrup cremaffin. Similar orofacial features are found in a wide range of illnesses, including Rett's syndrome, Angelman's syndrome, and Lennox-Gastaut syndrome. A thorough knowledge of the specific pattern of infantile spasms and associated neuropsychomotor deficits in such children can help differentiate this condition from others. Previous studies using Valproate as a monotherapy in children with WS showed promising in hypsarrhythmia or epileptic spasm management.[13,14] In our case the child is treated with valproate along with clobazam, lamotrigine, phenobarbital, and Zonisamide. In addition to Valproate, Topiramate and Nitrazepam (single or combined therapy) have shown positive outcomes in WS.[15] In 60 to 80% of newborns with WS, short-term hormonal therapy with ACTH has been shown to be effective.[16] Nelson GR[17] concluded that first-line drugs for the treatment are hormonal therapies, including ACTH and corticosteroids, and vigabatrin has the most evidence to support their use in infantile spasms. Since first-line treatment is not supported for our child, the cost and side effects of medications vary substantially and may limit choices in some regions and patient populations. It is important to note, however, there is a second-line treatment that includes topiramate, Zonisamide, pyridoxine, and a ketogenic diet in the ratio of 3:1-4:1 also proven effective for the child with WS. This way of treatment intends to sensitize physicians in this region toward this rare neurological syndrome.

Conclusion

The burden of the diseased state is itself crippling for the parents leading to a neglect of the health of the patient. Children with WS often require an evaluation for early intervention programs for developmental impairment. Early detection and referral to a pediatric neurologist for clinical evaluation and prompt effective treatment are strongly recommended as they may improve prognosis.

Patient perspective

Prior to admission, the patient's mother complained of a wide range of problems, but the doctor and other medical experts were able to correctly identify and treat my child, allowing her to recover rapidly and alleviate her ailments. I'm happy with the treatment she's received.

Informed Consent

The authors certify that they have obtained appropriate patient consent forms. In the form, the patient mother has given consent for her clinical information to be reported. She understood that her name would not be published, and outstanding efforts will be made to conceal her identity.

The mother granted her explicit agreement for this case report to be published. We have avoided disclosing information that could identify the patient and all data have been released anonymously.

Authors' Contribution

Sabitha Panchagiri contributed to the idea and design of the study. Akila Murugan gathered the data. Sabitha Panchagiri and Akila Murugan drafted the manuscript, and all authors critically revised it for relevant intellectual content and approved the final version.

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Conflicts of interest

There are no conflicts of interest.

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