Research Paper Nursing

A Case Study on West Syndrome

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KEYWORDS

Present history with Chief Complaints

A baby boy aged 11 months was brought to a hospital, with complaints of spasms and convulsions with deviation of eye to left side fever and cough diagnosed with symptomatic west syndrome.

The past history reveals that the baby from the initial 2 months of life was irritable but consolable and had exaggerated startle reflex.From 3 month of life mother complained that child had convulsions which was deviation of eye to left side, tonic clonic movement of both upper limbs lasting for 1 minute and subside as own with frequency of 25 times per day initially and now with medicine it has decreased to 8-10 times per day. The child was on: syp. Gardinal 3.5ml oral Od and Tab. Bexel 1\4 Tds.

The antenatal history of mother was normal, it was a full term normal delivery but baby did not cry soon after birth and needed stimulation and suctioning to breathe out and on 3rd day of life baby developed severe respiratory distress and shifted to NICU where he was kept for 2 weeks and then discharged. He belongs to a low class nuclear family and was on exclusive breast feeding till 6 months of age and thereafter weaning was started. There is no significant family history and the baby is immunized to age according to the National Immunization schedule.

The disease started its course in child from 2 month of age and had a symptomatic west syndrome with the possible cause being the asphyxia at time of birth which required stimulation and suctioning. The baby has all the three types of spasms appearing simultaneously and also had irritability but was consolable and regression of development was also present.

The investigations done were; EEG- Showed the typical hypsarrythmia, blood investigations included; WBC=13.8x 10^3\cum, RBC= 4.30x 10^6\cum, HGB=10.8gm\dl, HCT=35.2%, MCV= 82fl, MCH= 25.1pg, MCHC= 30.6%, PLT= 2.13x 10^5\cum, ESR= 19mm\hr and BERA test= no abnormality seen.

The treatment measure opted was with: Tab. Topiramate 25mg oral Od, Syp. Gardinal 3.5ml oral Od, Inj. ACTH 150µ\ m^2 IM Od, Tab. Bexal 1\4 oral Tds, Syp. Paracetamol 3.5ml oral Bd, Syp. Emeset 3.5ml oral Tds and Ketogenic diet [60ml milk 1 tsp. sugar+1ml MCT oil] every second hourly.

Introduction

West syndrome or West's syndrome is an uncommon to rare epileptic disorders in infants. It is named after the English doctor and surgeon William James West (1793-1848) who lived in Tonbridge. In 1841, he observed this type of epilepsy in his own son, James Edwin West (1840-1860) who was approximately, four months old at the time. He published his observations from a scientific perspective in an article in "The Lancet". He named the seizure "Salaam Tics" at the time.

It is also known by other names like:

- Generalized Flexion Epilepsy
- Infantile Epileptic Encephalopathy
- Infantile Myoclonic Encephalopathy
- Jackknife Convulsions
- Massive Myoclonia
- Salaam Spams, also as Infantile Spasms.

Definition

- West syndrome is a triad of infantile spasm a pathognomonic EEG pattern (called hypsarrythmia) and developmental regression. Presence of any two out of three elements is required.

Incidence

Incidence is around 1:3200 to 1:3500of live births. Statistically boys are more likely to be affected than girls at a ratio of around 1.3:1. In 9 out of every 10 children affected, the spasms appear for the first time between third and twelfth month of age. In rare cases, spasms may occur in first two month or during second to fourth year of age.

Causes

- If a case presents itself, the syndrome is referred to as symptomatic west syndrome, as the attacks manifest as symptom of another problem.
- One third of children, there is evidence of profound organic disorder of brain which includes: microcephaly, cortical dysplasia, cerebral atrophy, bacterial meningitis, tuberous sclerosis, cephal hematoma, vascular malformation, neurometabolic diseases, congenital infections (CMV), hypoglycemia, brain damage due to asphyxia or hypoxia during birth.
- First time after vaccination against measles, mumps and rubella or tetanus, pertussis, diphtheria, polio, hepatitis B, and Hib.
- Cryptogenic west syndrome: when a direct cause cannot be determined bu child has other neurological disorder.
- In cases where multiple children within the same family develop it, where it appears in successive generation in boys, X- chromosomal heredity.
- Idiopathic west syndrome: when the cause cannot be determined.

Clinical features

- The epileptic seizures which can be observed in infants with west syndrome fall into three categories, collectively known as infantile spasms. Typically following triad of attack, types appear while the 3 types usually appear simultaneously, they also can occur independently of each other.
- Lighting attacks: sudden, severe myoclonic convulsions of entire body or several parts of body in split seconds and legs in particular are bent.

- Nodding attacks: convulsions is of throat and neck flexor muscles during which the chin is fitfully jerked towards breast or head is drawn inward.
- Salaam or Jackknife attacks: a flexor spasm with rapid bending of head and torso forward and simultaneous raising and bending of arms while partially drawing the hands together in front of chest and or flailing (looks similar to oriental ceremonial greeting Salaam from which it derived its name)

♠ Irritability in child
♠ Regression of development till it is treated.

Management

Compared to other forms of epilepsy, it is difficult to treat. Success of treatment depends upon early diagnosis and prompt treatment, yet there is no guarantee. The treatment depends on

- Etiological classification
- State of brain development at time of damage.

♠ **The pharmacological management consist of the medications like:**

- Prednisolone
- ACTH
- Vigabatrin (Sabril)
- Topiramate
- Lamotrigine
- Levitiracetam

♠ Other treatment mode- The Ketogenic Diet which has shown to be effective in treating infantile spasms up to 70% of children having a 50% or more reduction in seizures.

Nurses Role

- Observe for the order of the events(before, during and after)
- Duration, onset- time, precipitating events, behaviors, movements with change of positions if any.
- Postictal period- observe for state of consciousness, arousibility, motor ability, sensations and recollection of pre seizure sensations.
- Educate the parents and family members regarding the disease, medications and side effects with a stress given on care of the child with the same.
- Parental attention to be brought on the precautions to be taken for the child with west syndrome.

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