

### Case Report on Subacute Sclerosing Panencephalitis

#### **ABSTRACT**

**Introduction:** Subacute sclerosing panencephalitis (SSPE), frequently referred to as Dawson disease, is a kind of sclerosing panencephalitis, and is an uncommon long-term, continuous cerebral inflammatory condition triggered by a gradual infection with particular faulty types of hypermutated measles virus. **Main Symptoms and/or Important Clinical Findings:** Patient came with the complaint of gait abnormalities, sudden fall while walking, loss of speech, staring look, abnormal smile, and loss of bladder control, hemiplegia and contracture of all four limbs, seizures, Grade III Malnutrition. **The Main Diagnoses, Therapeutic Interventions, and Outcomes:** Patient undergone investigation i.e Blood Test: Hb- 9.8 gm%, TLC- 14,700/mm, Platelet- 2.24 lacs/cu.mm, Total Protein- 8.4 g/dl, HCT- 29.8, Serum Bilirubin- 1.0mg/dl, ALP- 99IU/L, Serum Urea- 32mg/dl, Serum Creatinine- 0.5mg/dl, Serum Sodium- 153meq/L, Serum Potassium- 4.2mmol/L. CSF IgG Measles test was done EEG which revealed burst of periodic complexes with well-preserved background activity. Doctor manages with Tab. Valparin 200mg BD, Tab. Frisium 5mg BD, Tab. Baclofen 10mg BD, Tab. Samion D3 OD, Syp Q-Carni 5ml BD, Tab. Augmentin 375mg BD. **Outcome:** After treatment, the child showed some improvement. The patient was admitted to Pediatric Ward No- 22, AVBRH with known case of Subacute sclerosing panencephalitis (SSPE) and he had complaint of swelling over right jaw with caries tooth since 4 months.

**Keywords:** Subacute sclerosing panencephalitis, Dawson disease, hyper mutated, measles virus, cerebral inflammatory condition, seizures, infection.

## Introduction:

Subacute sclerosing panencephalitis (SSPE) is indeed neurodegenerative entity that affects younger and adolescent central nervous systems. It's a gradual and sustained infectious illness produced by a faulty measles virus.[1] It is also known as Dawson's Disease, Dawson's Encephalitis. Dawson described for the first instance in 1933 a kid having gradual mental decline with spontaneous motions whom, at necropsy, was discovered to also have a prominent engagement of grey matter with numerous neuronal inclusion bodies.[2]

In emerging nations such as India and Eastern Europe, the prevalence of SSPE tends to be high. Males have a greater prevalence than females (male/female: 3:1).[3] This was triggered by the measles virus's interaction in the brain, which destroys neurotransmitters. SSPE's pathophysiology is still a mystery, although wild strains, not vaccine strains, are to blame, according to genetic investigations, which further corroborate this conclusion. [4]

The process of SSPE evaluation is generally complex. Dyken criteria were first designed to aid in the diagnosis of SSPE. This series of criteria was revised in 2010 due to the fact that SSPE presentations might vary widely. Two main and one minor criterion are required for diagnosis when using this new recommended diagnostic criteria, which is part of the SSPE. Histomorphological and genetic investigations can be used if clinical findings or accompanying requirements are lacking but the diagnosis is still likely. High anti-Measles levels of antibodies in CSF and standard or unusual patient findings are two major criteria. Radermecker complexes are typical EEG results typically featuring regular, unspecified, twofolded concurrent, and well-proportioned weak waves of great amplitude that reoccur every 5–15s, Amount of Cerebrospinal fluid globulin containing more than 20% CSF protein, Brain sample histopathological features, Test to identify the altered genome of the wild-type measles virus using molecular diagnostics. In most cases, two major criteria and one minor requirement are necessary. [5]

The commonest typical age of onset is 5–15 years, usually 5–8 years after infection with measles. The most prevalent symptoms include myoclonus, cognitive deterioration, poor academic performance, and behavioral problems. The route is a steady downhill that may be divided into four distinct segments (Jabbour staging). Stage 1 is characterized by behavioral shifts and cognitive deterioration, both of which impair academic achievement. Stage 2 begins with the appearance of myoclonus. In the third stage, 6543 individuals acquire a variety of pyramidal and extrapyramidal symptoms such as stiffness, dystonia, trembling, spasms, and hemiparesis. Akinetic-mute condition with periods of profuse sweats, blood pressure fluctuations, and irregular breathing rates constitute Stage 4. [6]

There has been presently none treatment for SSPE and elimination through an efficient vaccination programme is thought to be much highly beneficial along with expense than other distinguished modes of control. The cornerstone is supportive care, which includes seizure stabilization and associated problems. One of the most commonly prescribed anti-epileptic drugs is divalproate sodium. For the treatment of SSPE, there are no other established therapy procedures. In the therapy of SSPE, antiviral medicines and immunosuppressive agents are employed.[7]

**Patient Identification:** A male child of 12-year-old from Yavatmal admitted to pediatric ward no- 22 in AVBRH on 25<sup>th</sup> June 2021 with a known case of Subacute sclerosing panencephalitis (SSPE). He is 22kg and height 125cm.

**Present Medical History:** A male child of 12 years was brought to AVBRH on 25<sup>th</sup> June 2021 by her parents with the complaint of swelling over the right jaw with caries tooth since 4 months. After the investigation she was diagnosed as extra-oral abscess (cutaneous fistula).

**Past Medical History:** Patient was diagnosed to have Subacute Sclerosing of Panencephalitis (SSPE) since 7 years of age.

**Birth History:** Full term normal vaginal delivery, place of delivery was at home. He did not cry immediately after birth. Cried after 5 minutes of birth. Birth weight was 3.5kg.

**Immunization History:** Patient did not receive all vaccines according to IAP (Indian Academy Pediatrics) schedules. Measles vaccine not received.

**Developmental History:** Achieved all milestones till 7 years of age.

**Family History:** Not significant.

**Past Interventions and Outcomes:** Patient was diagnosed with Subacute Sclerosing of Panencephalitis (SSPE) since 7 year of age. As the condition of the patient deteriorating they went to Nain hospital, Mumbai at the age of 7 years for 1 and half month. Since 5 years for which he is on medication: Tab. Sodium valproate 200mg BD, Tab. Frisium 5mg BD, Tab. Baclofen 10mg BD, Tab. Samion D3 OD, Syp Q-Carni 5ml BD.

**Clinical Findings:** Gait abnormalities, sudden fall while walking, loss of speech, staring look, abnormal smile, and loss of bladder control, hemiplegia and contracture of all four limbs, seizures, Grade III Malnutrition.

**Physical Examination:** There is not much abnormality found in head to toe examination, only found asymmetry of face due to swelling present on right side of face. Face was oval in shape, black in color, tenderness was present. Generalized gingival growth over teeth. Gingiva bleed on manipulation. Patient is in vegetative state. AEBE present and rhonchi positive.

**Diagnostic assessment: Blood Test:** Hb- 9.8 gm%, TLC- 14,700/mm, Platelet- 2.24lacs/cu.mm, Total Protein- 8.4 g/dl, HCT- 29.8, Serum Bilirubin- 1.0mg/dl, ALP- 99IU/L, Serum Urea-32mg/dl, Serum Creatinine- 0.5mg/dl, Serum Sodium- 153mEq/L, Serum Potassium-4.2mmol/L. CSF IgG Measles test was done. **EEG:** The EEG revealed burst of periodic complexes with well-preserved background activity.

**Therapeutic Intervention:** Doctor

prescribed Tab. Valparin 200mg BD, Tab. Frisium 5mg BD, Tab. Baclofen 10mg BD, Tab. Samion D3OD, Syp Q-Carni 5ml BD, Tab. Augment 375mg BD.

**Discussion:** A male child of 12 years was brought to AVBRH on 25th June 2021 by her parents with the complaint of swelling over the right jaw with carries tooth since 4 months with a known case of Subacute sclerosing panencephalitis (SSPE) associated with Gait abnormalities, sudden fall while walking, loss of speech, staring look, abnormal smile, and loss of bladder control, hemiplegia and contracture of all four limbs, seizures.

This study supported to the study which clear the idea that background of fever, seizures that began at two months of age. Symptoms were associated with myoclonic twitches in the axial direction, head dips, along with diminished span of concentration for the last two months. Benchmarks were met properly, although he had not had any of his vaccines, with the exception of a dose of diphtheria, tetanus, and pertussis vaccine. The family background was devoid of mental or neurological disorders. This resulted in epileptic seizures that prolonged 30 minutes. Due to a one-hour febrile generalized status epilepticus, his treatment was supplemented with valproate when he was three years old. The patient's physical assessments for physiological parameters as well as laboratory investigations were also normal. EEG indicated a sluggish backdrop with generalized cyclic complexes composed of two folded concurrent, well-proportioned weak waves of great amplitude that did not vanish after diazepam induction. Because the EEG images suggested sub-acute sclerosing panencephalitis sample of CSF was collected to be tested for anti-measles antibodies. [8]

Another study supported to the case of an 11-month-old newborn came to the hospital complaining of right-sided partial seizures for three days, followed by muscle spasm and impaired nervous system throughout the previous month. Previous to this sickness, the newborn was healthy and meeting age-appropriate developmental achievement. During the assessment, myoclonus of the limbs was observed. The antenatal and postnatal periods were uncomplicated, and the mother had no incidence of measles throughout gestation or at the delivery date. At eight months of age, he had background of temperature and coughing, and acute rhinitis, which was followed by a maculopapular rash (initially detected on the forehead, then descended lower), which was diagnosed as measles by a pediatrician. Other investigations were normal. Because the EEG image suggested SSPE, a sample of CSF and serum was taken to test for anti-measles antibody. An ELISA test for IgG anti-measles antibody utilizing commercially available kit revealed good outcomes in both CSF and serum. While anti-measles IgM antibodies were negative in both CSF and serum. The child was treated with isoprinosine (100mg/kg/day), but interferon therapy was not financially feasible. Myoclonus was controlled with sodium valproate. [9],[10]

**Conclusion:** Subacute sclerosing panencephalitis (SSPE) is still a frequent disease in India, characterized by progressive mental impairment, myoclonus, periodic cephalographic abnormalities, and an elevated anti-measles antibody titer in CSF fluid. The measles virus infects the brain and promotes neuronal death. The patient has made substantial progress since seeking treatment, and the treatment is still continuing as of my last visit.

#### **Consent**

While preparing case reports for publication guardian informed consent has been taken from parent of client

#### **Ethical approval**

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

#### **REFERENECE:**

1. Subacute Sclerosing Panencephalitis [Internet]. NORD (National Organization for Rare Disorders). [cited 2021 Dec 14]. Available from: <https://rarediseases.org/rare-diseases/subacute-sclerosing-panencephalitis/>
2. Jafri SK, Kumar R, Ibrahim SH. Subacute sclerosing panencephalitis – current perspectives. *Pediatr Health Med Ther.* 2018 Jun 26;9:67–71.
3. Subacute Sclerosing Panencephalitis Information Page | National Institute of Neurological Disorders and Stroke [Internet]. [cited 2021 Dec 14]. Available from: <https://www.ninds.nih.gov/Disorders/All-Disorders/Subacute-Sclerosing-Panencephalitis-Infor>

4. Measles infection and encephalitis [Internet]. The Encephalitis Society. [cited 2021 Dec 14]. Available from: <https://www.encephalitis.info/measles-infection-and-encephalitis>
5. Manning L, Laman M, Edoni H, Mueller I, Karunajeewa HA, Smith D, et al. Subacute Sclerosing Panencephalitis in Papua New Guinean Children: The Cost of Continuing Inadequate Measles Vaccine Coverage. *PLoS Negl Trop Dis*. 2011 Jan 4;5(1):e932.
6. Gutierrez J, Issacson RS, Koppel BS. Subacute sclerosing panencephalitis: an update. *Dev Med Child Neurol*. 2010;52(10):901–7.
7. Measles (for Parents) - Nemours KidsHealth [Internet]. [cited 2021 Dec 14]. Available from: <https://kidshealth.org/en/parents/measles.html>
8. Vaccine-safety-E-course-manual.pdf [Internet]. [cited 2021 Dec 14]. Available from: [https://www.who.int/vaccine\\_safety/initiative/tech\\_support/Vaccine-safety-E-course-manual.pdf](https://www.who.int/vaccine_safety/initiative/tech_support/Vaccine-safety-E-course-manual.pdf)
9. Epilepsy and Disorders of Consciousness | Adams and Victor's Principles of Neurology, 11e | AccessNeurology | McGraw Hill Medical [Internet]. [cited 2021 Dec 14]. Available from: <https://neurology.mhmedical.com/content.aspx?bookid=1477&sectionid=117184222>
10. Manning L, Laman M, Edoni H, Mueller I, Karunajeewa HA, Smith D, et al. Subacute Sclerosing Panencephalitis in Papua New Guinean Children: The Cost of Continuing Inadequate Measles Vaccine Coverage. *PLOS Neglected Tropical Diseases*. 2011 Jan 4;5(1):e932.