# Case Report On Subacute Sclerosing Panencephalitis


## Abstract

**Introduction:** Subacute sclerosing panencephalitis (SSPE), frequently referred 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 hyper mutated measles virus.

**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.

**Diagnostic Evaluation:** 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:** Tab. Valparin 200mg BD, Tab. Frisium 5mg BD, Tab. Baclofen 10mg BD, Tab. Samion D3 OD, Syp Q-Carni 5ml BD, Tab. Augment 375mg BD.

**Outcome:** After treatment, the child showed some improvement.

**Conclusion:** My 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 month.


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

**Introduction:**

Subacute sclerosing panencephalitis (SSPE) is indeed neurodegenerativeentropythat affects youngster 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).[1]

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. [3]

The process of SSPE evaluation is generally complex.Dyken criteria were first designed to aid in the diagnosis of SSPE. This series of criterion 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. [4] 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 feature regular, unspecified, two folded concurrent, and well-proportioned weak waves of great amplitude that reoccur every 5–15 s, Amount ofCerebrospinal 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, 6543individuals 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 antiepileptic drugs is divalproate sodium.For the treatment of SSPE, there are no other are no established therapy procedures.In the therapy of SSPE, antiviral medicines and immunosuppressive agents are employed. [3]

**Case Presentation**

**Patient Identification:** A male child of 12 year old from Yavatmal admitted to pediatric ward no- 22 in AVBRH on 25th 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 25th June 2021 by her parents with the complaint of swelling over the right jaw with carries tooth since 4 month. After the investigations he 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 5years for which he is on medication: Tab. Sodium valproate200mg 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.

**Etiology:**The measles virus infects the brain and promotes neuronal death.SSPE's pathophysiology is still a mystery, although wild strains, not vaccine strains, are to blame, according to genetic studies, which also corroborate this theory. [3]

**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 bleeds

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:** Tab. Valparin 200mg BD, Tab. Frisium 5mg BD, Tab. Baclofen 10mg BD, Tab. Samion D3 OD, 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 month 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.

A case report on a five year old child was hospitalized to the Pediatric Neurology Department with a background of fever, seizures that began at two months of age. Symptoms were associated withmyoclonic 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. His 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. On admission, 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 compose of two folded concurrent, well-proportioned weak waves of great amplitude that did not vanish after diazepam induction. Because the EEG image suggested subacutesclerosingpanencephalitisa sample of CSF was collected to be tested for anti-measles antibodies. [7]

A case study on a 10 year old boy, overall proper growth and development presented with just a 2-week background of automatic jerky motions on the left half side of body, as well as the head.His symptoms began 15 days prior, after a generalized seizure with mental decline. The patient was fully unvaccinated, and his parents had no idea he had measles as a youngster. The patient had left hemiplegia and was nonambulatory, according to physical examination. On theleft side, the Babinski's sign was positive, and the sensory test appeared to be normal. His condition deteriorated and he got utterly deaf. Routine blood readings were within normal limits, and the EEG demonstrated global slowing. There was a significant increase in anti-measles IgG antibody titer in both cerebrospinal fluid and serum. Magnetic resonance imaging of the brain revealed focal T2 hyperintensity in the right parasagittal cortex. The patient was given levetiracetam and clobazam. However, the patient's health deteriorated and he became akinetic and silent.The child died about a month after being discharged from the hospital. [8]

An 11 month old newborn came to the hospitalcomplaining 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-apt 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 thedelivery date.At eight months of age, he hada 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 IgGantimeasles 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 and clonazepam.However, after three months of constant monitoring, the child's cognitive abilities did not improve activities. [9]

**Conclusion:** Subacute sclerosing panencephalitis (SSPE) is still a frequent disease in India, characterized by progressive mental impairment, myoclonus, periodic encephalographic abnormalities, and an elevated anti-measles antibody titer in CSF fluid. The measles virus infects the brain and promotes neuronal death.My 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. www.ninds.nih.gov/Disorders/All-Disorders/Subacute-Sclerosing-Panencephalitis-Information-Page
2. Garg RK. Subacutesclerosingpanencephalitis. Postgraduate Medical Journal. 2002 Feb 1;78(916):63-70.
3. Jafri SK, Kumar R, Ibrahim SH. Subacutesclerosingpanencephalitis–current perspectives. Pediatric health, medicine and therapeutics. 2018;9:67.
4. Rocke Z, Belyayeva M. SubacuteSclerosingPanencephalitis. StatPearls [Internet]. 2020 Jul 15.
5. Paul L, Jain T, Agarwal M, Singh S. SubacuteSclerosingPanencephalitis manifesting as Bell’s palsy and bilateral macular necrotizing retinitis: an atypical presenting feature.

Journal of Ophthalmic Inflammation and Infection. 2021 Dec;11(1):1-6.
6. Saurabh K, Singh V, Pathak A, Chaurasia R. Subacutesclerosing pan encephalitis: An update. Journal of Clinical and Scientific Research. 2021 Jan 1;10(1):35
7. Kartal A, Çıtak Kurt AN, Hirfanoğlu T, Aydın K, Serdaroğlu A. Subacutesclerosingpanencephalitis in a child with recurrent febrile seizures. Case reports in pediatrics. 2015 Feb 24;2015.
8. Garg RK, Kumar N, Rizvi I, Jain A, Jaipuriar RS, Sharma PK, Malhotra HS, Khan DN, Uniyal R. Case report: subacutesclerosingpanencephalitis presenting as acute encephalitis. The American journal of tropical medicine and hygiene. 2019 Jul;101(1):260.
9. Saurabh K, Gupta R, Khare S, Sharma S. Atypical subacutesclerosingpanencephalitis with short onset latency. Indian pediatrics. 2013 Feb 1;50(2):244-5.