Subacute Sclerosing Panencephalitis (SSPE) Following Measles Infection: A Case Report and Review of the Literature

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Abstract

Subacute Sclerosing Panencephalitis (SSPE) is a rare but fatal progressive neurodegenerative disorder caused by persistent infection with a mutant measles virus in the central nervous system. We present a pediatric case that developed SSPE years after a severe measles infection in early infancy. The diagnosis was confirmed by EEG, MRI findings, and detection of anti-measles IgG oligoclonal bands in cerebrospinal fluid. Despite immunomodulatory and antiviral therapies, the disease followed a progressive clinical course with poor prognosis. This report highlights the importance of early diagnosis and the critical role of measles vaccination in preventing SSPE.

***Keywords**: Subacute Sclerosing Panencephalitis, Measles, Neurodegeneration, Vaccination, Pediatrics*

Introduction

Subacute Sclerosing Panencephalitis (SSPE) is a progressive, chronic, and invariably fatal encephalopathy caused by the persistence of a mutant measles virus within the central nervous system (CNS). It typically manifests several years after primary measles infection, particularly in individuals who contracted measles at a very young age. SSPE remains an important reminder of the devastating consequences of inadequate immunization coverage. Although rare in countries with high vaccination rates, SSPE continues to pose a major public health burden in low resource settings where vaccine coverage is suboptimal.

A child who beats measles as a toddler, only to face a hidden enemy years later. That enemy is subacute sclerosing panencephalitis, or SSPE, a rare but brutal brain disorder. It creeps in quietly after the initial infection, turning a preventable illness into a lifelong nightmare. SSPE shows why measles shots matter so much. It hits the central nervous system hard, causing steady decline in movement, thought, and basic functions. This article shares a real case of SSPE seven years later after measles infection.

Measles Epidemiology in the Context of Vaccine-Preventable Diseases: Before the introduction of the measles vaccine, the United States reported millions of infections annually, with thousands of hospitalizations and deaths. Vaccination dramatically changed this epidemiologic landscape. Today, the CDC reports fewer than 1,000 measles cases per year in the U.S.

Globally, measles remains far more dangerous. The WHO estimated over 140,000 measles-related deaths in 2018—mostly in young children. Declining vaccination rates in several regions have contributed to measles resurgence. Low vaccine uptake increases the risk of severe primary infection and subsequent complications such as SSPE. Skipping vaccination invites preventable morbidity and the rare but catastrophic development of SSPE. (1,2,3)

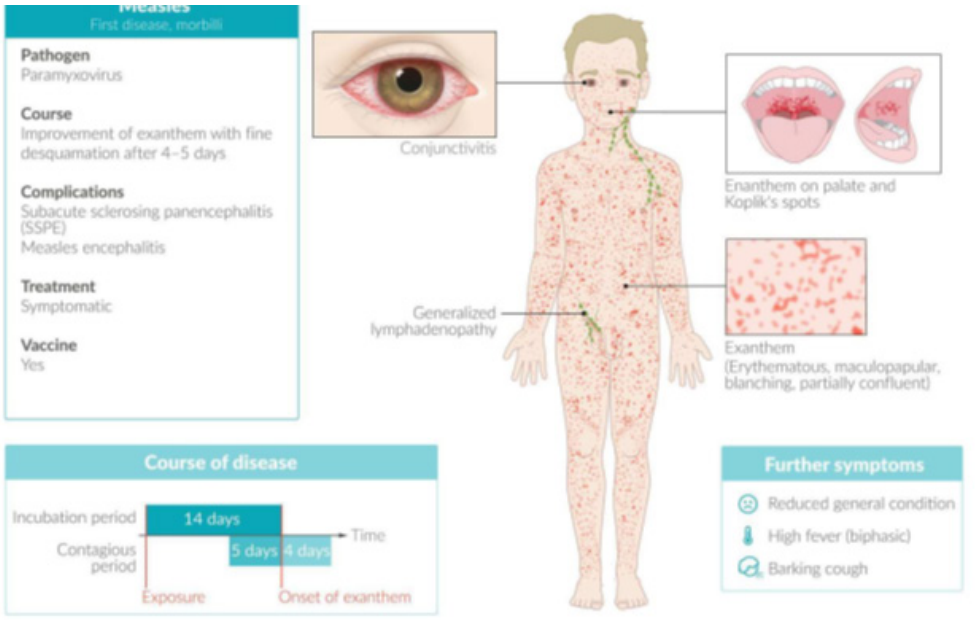
The Defective Measles Virus and SSPE Pathogenesis

The virus responsible for SSPE is a persistent, mutated form of the wild-type measles virus. After primary infection, the virus remains dormant in neurons and glial cells. Mutations—particularly in the M (matrix) gene—allow the virus to replicate intracellularly without lysing host cells. Over time, this leads to chronic inflammation and progressive demyelination, disrupting neuronal signaling and causing widespread neurodegeneration.

This silent and persistent viral activity gradually damages the brain, ultimately resulting in SSPE. (7,8,9)

Incubation Period and Typical Timeline from Infection to Onset

SSPE doesn't show right after measles. The wait averages seven to ten years. Some kids see signs as early as two years post-infection. Others wait into their teens or beyond. Case reports vary by age at first exposure. Younger kids, under two, face higher risk and quicker onset. A study in the Journal of Neurology found 80% of cases start before age 20. Factors like immune strength play a role. If measles hits early, the brain might not fight it off well. This long gap fools many into thinking the past infection is gone for good.



Clinically, SSPE progresses through four stages: (I) behavioral changes and cognitive decline, (II) myoclonus and dystonia, (III) extrapyramidal signs and spasticity, and (IV) akinetic mutism and vegetative state. EEG typically reveals periodic high-amplitude slow-wave discharges.

Subacute Sclerosing Panencephalitis (SSPE) is a rare yet devastating disorder that can follow a measles infection, particularly among children. This progressive neurodegenerative condition typically surfaces several years after an initial measles infection, posing significant challenges for diagnosis and treatment. In this article, we will delve into a remarkable pediatric case that underscores the complexities of SSPE, along with a comprehensive review of relevant literature to highlight the critical role of vaccination in preventing such outcomes. (10,11,12)

Case Presentation

A 7-year-old male presented with a one-year history of progressive neurological deterioration characterized by:

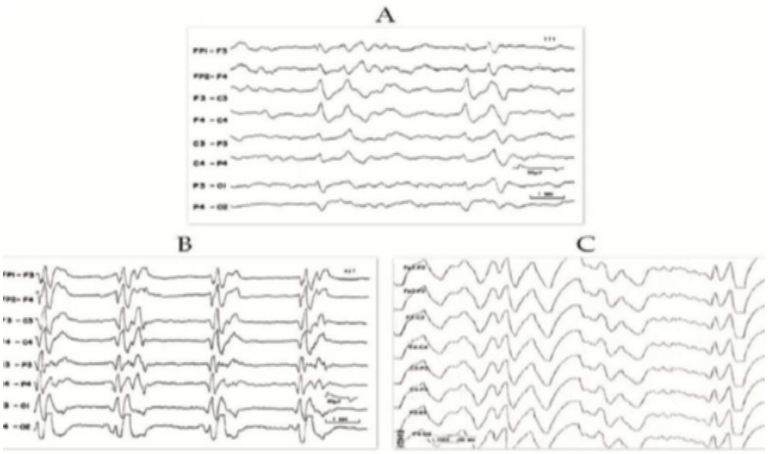
- recurrent seizures,
- myoclonic jerks,
- abnormal muscle tone,
- psychomotor regression.

The child had a documented history of severe measles at two months of age. Behavioral changes were first noted at school: declining academic performance,

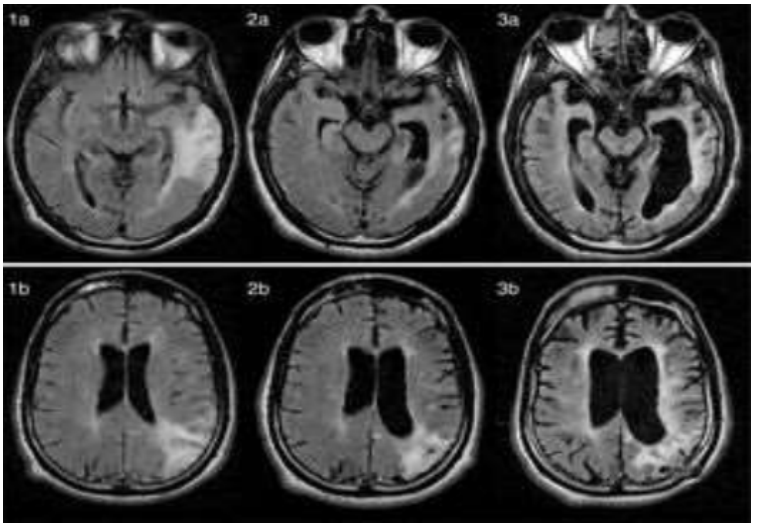
episodes of staring, and memory impairment. At home, the family observed sudden limb jerks consistent with myoclonus. These early manifestations may mimic ADHD, epilepsy, or even autoimmune encephalitis, often delaying SSPE diagnosis due to its rarity

Investigations performed

- Electroencephalogram (EEG)
- Lumbar puncture for CSF analysis
- MRI of the brain
- PCR for measles RNA in CSF and blood



EEG: Classic bursts: sharp waves every few seconds,
mixed with slow ones. This pattern screams SSPE to experts.



MRI scans painted a grim view—shrunk brain tissue and patchy white matter spots.

Next came a spinal tap for CSF. His fluid had sky-high measles antibodies and special bands just for the virus. (This test for measles analysis in LCS was performed in Italy Bambino Gesu hospital)

The child was in stage 1. After 6 months he went in stage 2. Treatement with immunoglobuline every month stopped the progress of illness,

Actually he became spastic, with tetraparesis, dystonic movements and multiform seizures with trucal spasms. Extrapyramidal and pyramidal signs. (8,9.10)

Diagnosis and Differential Diagnosis of SSPE

Essential Laboratory Markers. Key diagnostic indicators include:

- Serum measles IgG titers extremely elevated (up to 1,000× normal).
- CSF measles antibody index showing intrathecal synthesis.
- Oligoclonal bands specific to measles virus.
- PCR for measles RNA, with ~90% sensitivity in active stages.

Differential Diagnosis. SSPE must be distinguished from:

- Creutzfeldt–Jakob Disease (CJD): more rapid course, no measles link, different EEG patterns.
- Viral encephalitides: lack long latency and periodic myoclonus.
- Leukodystrophies: early onset but without characteristic EEG pattern.

A history of prior measles infection strongly supports SSPE. (13,14)

Prognostic Indicators

SSPE progression is divided into four stages:

Stage I – behavioral changes, cognitive decline
Stage II – myoclonus, dystonia, visual disturbances
Stage III – severe neurological impairment, increased spasticity
Stage IV – akinetic mutism, vegetative state

Early detection improves short-term stabilization rates. Younger age at onset and poor immune status worsen prognosis. (13,14)

Management and Emerging Therapies

Standard of Care
There is no cure. Treatment aims to slow disease progression through combined therapy:

- IVIG to reduce inflammation,
- Inosiplex (Isoprinosine) to enhance immune response,
- Ribavirin (systemic or intrathecal) to reduce viral replication.

Trials show stabilization in 30–50% of early-stage patients.

Experimental Therapies

Ongoing research includes:
RNA-silencing technologies targeting mutant viral genes, anti-inflammatory glial modulators, viral gene therapy vectors (early trials in India), stem-cell–based approaches for remyelination. Due to the rarity of SSPE, clinical trials are limited but progressing.

Supportive Care Includes:

- antiepileptics (valproate, clonazepam),
- nutritional support (NG tube or PEG),
- physiotherapy for contracture prevention,
- psychosocial and palliative support for families (15,16,17)

Discussion

The pathogenesis of SSPE involves a persistent "wild-type" measles virus that undergoes mutations, particularly in the M gene, allowing viral replication without host cell lysis. The disease usually manifests 7–10 years after measles infection. Epidemiologically, the incidence has declined dramatically with universal measles vaccination, from approximately 4–11 cases per 100,000 measles infections globally, to almost none in vaccinated populations.

Clinically, SSPE progresses through four stages: (I) behavioral changes and cognitive decline, (II) myoclonus and dystonia, (III) extrapyramidal signs and spasticity, and (IV) akinetic mutism and vegetative state. EEG typically reveals periodic high-amplitude slow-wave discharges. (18,19)

Conclusion

Subacute Sclerosing Panencephalitis serves as a stark reminder of the potential consequences of measles infections in childhood. Our case report illustrates the profound impact of this neurodegenerative disorder and its tragic progression, emphasizing the need for vigilant monitoring and swift diagnosis in patients with a history of severe measles. As we reflect on this case and the broader implications of SSPE, it becomes increasingly clear that vaccination plays an essential role in safeguarding children against preventable diseases. (18,19)

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