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Case report

Early-Onset of Subacute Sclerosing Panencephalitis in a 3-Year-Old Girl Following Measles Infection: A Case Report

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Abstract

Subacute sclerosing panencephalitis (SSPE) is a rare, invariably fatal neurodegenerative disorder caused by persistent central nervous system infection with mutant measles virus. Early-onset SSPE in toddlers poses significant diagnostic challenges owing to low clinical suspicion and frequently absent documented measles history. A 3-year-old girl presented with a 15-day history of abnormal behavior, myoclonic jerks, progressive gait disturbance, and excessive somnolence. She had not received the measles-rubella vaccine, as her mother reported a presumed, laboratory-unconfirmed measles illness at two months of age. Empirical treatment with intravenous immunoglobulin and high-dose methylprednisolone for suspected autoimmune encephalitis yielded no improvement. Cerebrospinal fluid analysis revealed markedly elevated measles-specific IgG antibodies, and electroencephalography demonstrated high-amplitude quasi-periodic slow-wave complexes time-locked with myoclonic jerks. Diagnosis of SSPE was established using Dyken's modified criteria, and the patient was commenced on isoprinosine and lamivudine, though her condition continued to deteriorate. This case illustrates the diagnostic primacy of CSF anti-measles serology and EEG in early-onset SSPE, the frequent initial misdiagnosis as autoimmune encephalitis, and the heightened SSPE risk associated with primary measles infection in infancy due to relative immune immaturity and shortened incubation intervals. Withholding measles vaccination based on unconfirmed natural infection contributed to a preventable outcome, reinforcing the critical importance of documented immunization irrespective of clinical history.

Key word: SSPE, measles, early-onset, myoclonus, vaccination

Introduction

Subacute sclerosing panencephalitis (SSPE) is a rare, progressive, and invariably fatal neurodegenerative disorder caused by persistent central nervous system infection with a mutant measles virus (MeV).[1] It is classified as a delayed complication of primary measles, typically presenting months to years after the initial infection. Four overlapping stages characterize the clinical course: an early phase of subtle behavioral and cognitive change; a second phase of myoclonic jerks, seizures, and progressive motor deterioration; a third phase of extrapyramidal and pyramidal involvement with increasing disorientation; and a terminal vegetative state.[2,3]

The incubation interval between measles infection and SSPE onset is variable, typically ranging from 6 to 8 years, though shorter intervals—including early-onset disease in young children are increasingly reported, particularly in regions with ongoing measles transmission or incomplete vaccine coverage.[4] Importantly, SSPE risk is attributable to wild-type measles virus; epidemiological evidence consistently demonstrates that immunization reduces SSPE incidence by preventing primary measles infection.[5,6]

Diagnosis is established using Dyken's modified criteria,

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which require the presence of at least two major and one minor criterion. Major criteria comprise progressive subacute neurological deterioration—characteristically including myoclonus—and elevated anti-measles antibody titers in serum and/or CSF. Minor criteria include periodic stereotyped high-voltage EEG complexes, elevated CSF gamma globulin or oligoclonal banding, histopathological evidence on brain biopsy, and molecular detection of measles virus.[7]

The non-specific early symptoms of SSPE frequently lead to diagnostic delay, with initial misdiagnosis as autoimmune encephalitis, metabolic disorders, or other progressive myoclonic epilepsies.[3,4] Early-onset cases, particularly in toddlers, pose an additional diagnostic challenge owing to a low index of suspicion and the frequent absence of a clearly documented antecedent measles infection. We report a 3-year-old girl diagnosed with SSPE following an atypical measles illness in infancy, to raise clinical awareness of early-onset SSPE and highlight the diagnostic value of CSF measles serology and EEG.

Case Presentation

In October 2023, A 3-year-old girl presented with abnormal behavior and jerk movements persisting for 15 days, followed by progressive neurological symptoms. These included difficulty walking, inability to sit without support, and worsening imbalance. Excessive somnolence had developed over the preceding 10 days. There was no preceding history of head trauma, suspected toxin ingestion, or known structural renal disease. The child was developmentally normal before this presentation (milestones, speech, motor). Considering the possibility of autoimmune encephalitis, the child had received one 10 g dose of immunoglobulin and 3 high doses of methylprednisolone between October 2023 and November 2021. However, the child's condition was not significantly improved.

The child's mother reported that the child had received all age-appropriate EPI vaccines except the measles-rubella (MR) vaccine. She stated that the child had previously experienced a febrile illness clinically suggestive of measles at her 2 months of age, and therefore, the MR vaccine was not administered thereafter. However, no laboratory confirmation was obtained at that time. The absence of a prominent cutaneous rash raises the possibility that the illness may have represented an atypical or subclinical measles infection.

On examination, she was drowsy, with a Glasgow Coma Scale score of 9/15. Myoclonic jerks were present in both the upper and lower limbs, accompanied by atonia and tremor. No meningeal signs were elicited. Higher mental functions were impaired, as evidenced by nonspecific behavioral disturbances and increased restlessness. Cranial nerve examination was unremarkable. Deep tendon reflexes were brisk, and bilateral extensor plantar responses were present. Frequent myoclonic drops were also noted.

Brain magnetic resonance imaging revealed small hyperintensities in the bilateral periventricular regions, with mild enlargement of the lateral ventricles, subtle hyperintensity in the putamen, and subcortical involvement. The CSF analysis revealed elevated levels of total IgG (9.4 g/L) and positive measles IgG antibodies (5.58 g/L), indicative of SSPE (Table 1). Other CSF parameters, such as cell count, neutrophil/lymphocyte ratio, protein, glucose, adenosine deaminase, and lactate levels, were within normal ranges. The virology panel results were negative.

Table 1: Study of CSF panel

Parameter	Result (g/L)	Normal range (g/L)
Serum total IgG	1,567	700 –1,600
CSF total IgG	9.4	0–3.4
CSF measles IgG antibodies (+ve)	5.58	<1.5
Measles virus IgG (+ve)	>300	<300

CSF = cerebrospinal fluid; IgG = immunoglobulin G

Additional investigations did not reveal any abnormalities (Table 2).

Table 2: Study of Blood investigations.

Parameter	Value
Hemoglobin	10.5 g/dL
TLC	11,100/µL
Ca^{2+}	9.3 mg/dL
CRP	5.12 mg/dL
Dengue	Negative
COVID -19 IgG	Negative
Platelets	3.22 lacs/mu L
N/L	52/35
PCV	41.8%
Na	139 mEq/L
K	3.55 mEq/L
Cl	95 mEq/L
Blood culture	No microbial growth

TLC = total leucocyte count; CRP = C-reactive protein; IgG = immunoglobulin G; N/L = neutrophil/lymphocyte; PCV = packed cell volume

Electroencephalography demonstrated High-amplitude quasiperiodic slow wave complexes time-locked with myoclonic jerks, a characteristic EEG pattern consistent with SSPE and fulfilling a minor criterion of Dyken's modified criteria.[7]

The differential diagnosis included autoimmune encephalitis (including anti-NMDAR and other antibody-mediated encephalitides), acute viral or bacterial encephalitis, progressive myoclonic epilepsies, and inherited metabolic or mitochondrial encephalopathies. However, the subacute and progressively deteriorating clinical course, the combination of cognitive decline with myoclonus and seizures, and the positive CSF anti-measles antibody result, together with the supportive EEG findings, were collectively consistent with SSPE using Dyken's modified criteria.[3,8] The absence of CSF pleocytosis and the lack of response to empirical antiviral or immunosuppressive therapy for alternative diagnoses further supported this conclusion.

The patient was managed with supportive care, nutritional support, and physiotherapy. Disease-modifying therapy with Isoprinosine, Lamivudin and/or intrathecal interferon-alpha was initiated for partial improvement in symptoms or to slow down disease progression. In keeping with current evidence that no definitive curative treatment exists for SSPE and that pharmacological interventions offer, at best, variable stabilisation.[4,9]

Despite optimal supportive management, the patient's condition continued to deteriorate. The family received counselling regarding the poor prognosis, the progressive nature of the disease, and the goals of long-term supportive care.

Discussion

SSPE is a rare, fatal neurodegenerative disorder caused by persistent mutated measles virus infection, progressing through personality changes, seizures, intellectual decline, and coma, predominantly affecting unvaccinated children in developing countries years after primary measles exposure.[8] The present case of a 3-year-old girl with a probable antecedent atypical measles illness at the age of 2 months, who developed progressive myoclonus, cognitive decline, and motor deterioration after 3 years.

The risk of developing SSPE is markedly higher when primary measles infection occurs at a young age, rising to approximately 18 per 100,000 measles cases in children under five years old.[4] This elevated risk is attributable to the relative immaturity of the infant's immune system, which is thought to favor viral persistence in neural tissue and hasten the development of disease.[10] In a systematic review of SSPE cases in children aged three years or younger, most patients had not received measles vaccination, and measles infection frequently preceded SSPE by as little as one to five years, with a reported prognosis that was dismal in this age group, with the majority progressing rapidly to a severe neurological outcome.[10]

According to a Chinese study, the measles antibodies in the vast majority of mothers in China are induced by vaccination rather than by the natural infection. Therefore, it is speculated that infants may acquire fewer antibodies from their mothers, resulting in the reduced duration of protection against measles and leading to the onset of measles before the age of receiving the vaccine. [11]

If the mother had neither natural measles infection nor measles vaccination, she would not be expected to pass protective measles-specific IgG to her infant. Since maternal antibodies are transferred through the placenta, a mother without measles immunity would provide little or no passive protection. As a result, the infant could become susceptible to measles in early life.

The decision to withhold the measles-rubella vaccine on the basis of a presumed prior natural infection, without serological confirmation, left the child unprotected against a pathogen whose neurotropic consequences can be catastrophic. Epidemiological evidence consistently demonstrates that SSPE incidence has an inverse relationship with measles vaccination coverage, and vaccination prevents SSPE by preventing the primary measles infection that underlies it.[4] This case therefore exemplifies a preventable tragedy arising from an unverified assumption of natural immunity.

The presenting features—progressive behavioral change, myoclonic jerks of the upper and lower limbs, atonia, tremor, increasing somnolence, and evolving motor deficits—are consistent with the overlapping clinical stages described in SSPE.[12] The initial consideration of autoimmune encephalitis, for which the child received immunoglobulin and high-dose methylprednisolone without meaningful improvement, is common in clinical practice. Differential diagnoses for SSPE include epileptic encephalopathies such as Dravet syndrome and Lennox-Gastaut syndrome, autoimmune encephalitides (including anti-NMDA receptor and other antibody-mediated conditions), progressive myoclonic epilepsies, and inherited metabolic or mitochondrial encephalopathies.[13] In this patient, the subacute and relentlessly progressive clinical course, the combination of cognitive regression with prominent myoclonus, the absence of CSF pleocytosis, and the lack of response to immunosuppressive or empirical antiviral therapy for alternative diagnoses collectively argued against these entities. Ultimately, the combination of a compatible clinical history with measles exposure and the serological findings secured the diagnosis.

The diagnostic work-up in this case illustrates the central role of CSF anti-measles antibody testing and EEG in confirming SSPE. Among tested paired samples, CSF measles-specific IgG levels at a threshold of 0.5 IU/mL or greater, together with a CSF-to-serum ratio of at least 0.05,

effectively distinguished SSPE from non-SSPE encephalitis.[13]

The EEG demonstrated high-amplitude quasi-periodic slow-wave complexes, time-locked with the observed myoclonic jerks—characteristically described in SSPE. These EEG periodic complexes are identified in approximately 65% to 83% of SSPE cases, and their presence in the appropriate clinical context constitutes a minor criterion under Dyken's modified diagnostic criteria.[4,14]

The diagnosis in this patient was established by fulfilling two major criteria and one minor criterion as specified by Dyken's modified diagnostic framework.[7] The major criteria met were: a progressive subacute neurological deterioration characterized by myoclonus, and markedly elevated serum and CSF anti-measles antibody titers. The minor criterion was the characteristic quasi-periodic EEG pattern. Compared to other techniques, measles antibody detection in CSF using ELISA has demonstrated superior diagnostic performance in SSPE, and its role in establishing the diagnosis in this young child, where clinical suspicion may otherwise be delayed, cannot be overstated.

No curative therapy currently exists for SSPE. Treatment is directed at disease modification, symptom control, and palliative support. Oral isoprinosine stabilizes or improves symptoms in approximately 30% of patients, while intrathecal interferon-alpha is used with variable results.[15]

A randomized study comparing oral isoprinosine alone with combined oral isoprinosine and intraventricular interferon-alpha found no significant difference in survival or neurological disability between the groups, though treatment was superior to no treatment, with satisfactory outcomes observed in approximately 34-35% of patients.[16] SSPE is almost always lethal, typically within one to three years of onset, with approximately 60% of patients surviving less than two years.[17]

A recent surge in SSPE cases in developed countries has been attributed to reduced vaccination coverage, aggravated by misinformation and a decline in immunization following the COVID-19 pandemic.[18]

Conclusion

SSPE is preventable by vaccinating children against measles before they become infected; the strains of measles virus contained in measles vaccines do not cause SSPE. The present case, in which the MR vaccine was withheld based on a presumed and unconfirmed measles illness, illustrates the critical need for clinicians and public health authorities to reinforce the importance of documented immunization, irrespective of clinical history.

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