Case Report

# Atypical subacute sclerosing panencephalitis presenting in a toddler with a short latency period: Evolving epidemiological trend

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## ABSTRACT

Subacute sclerosing panencephalitis (SSPE) is a slowly progressive degeneration of the central nervous system (CNS) caused by a persistent defective measles virus infection. We report the case of a 2.5-year-old girl who presented with complaints of sudden onset frequent falls while walking, myoclonic jerks, and loss of speech. The electroencephalogram showed generalized slowing with irregular activity and cerebral magnetic imaging revealed T2 hyperintensities in the periventricular region in the parietal and occipital lobes. SSPE was considered and the diagnosis was confirmed with the identification of measles antibodies in cerebrospinal fluid. Our findings show that SSPE should be in mind in the differential diagnosis of meningoencephalitis even when a short latency period with measles infection.

**Key words:** Electroencephalogram, Measles, Short latency, Subacute sclerosing panencephalitis

Measles is a highly contagious disease caused by a virus belonging to the paramyxovirus family. Measles-related neurological syndromes encompass primary measles encephalitis, acute post-measles encephalitis, measles inclusion-body encephalitis, and subacute sclerosing panencephalitis (SSPE). SSPE is a slowly progressive degeneration of the CNS caused by a persistent defective measles virus infection. It is the most common cause of myoclonus in a young child. The disease has a gradually progressive course leading to death in many cases within 1–3 years [1]. The latent period between measles infection and SSPE is commonly 6–8 years but may range between 3 months and 18 years [2].

Children infected with measles under the age of 1 year carry a 16 times greater risk of SSPE than those infected at age of 5 years or later [3]. The estimated risk of SSPE is 4–11/100,000 cases worldwide, unlike developing countries like India where the current reported incidence rate is 21 cases per million population [4]. We report a 2.5-year-old with a very short latency which can help pediatricians all over the world to differentiate this grave diagnosis from other causes of acute encephalopathy.

## CASE REPORT

A 2.5-year-old developmentally normal child was admitted with complaints of sudden onset frequent falls for 1 month while walking and myoclonic jerks associated with loss of speech for the past 2 weeks. Frequent falls progressed and the patient was unable to stand or sit without support by the end of 1 month. Myoclonic jerks started 2 weeks after regression in motor milestones. Initially, the child had 1–2 episodes per day which have increased to 8–10 episodes per minute. Myoclonic jerks were always succeeded by head drop which would disappear in sleep and were not associated with loss of consciousness. Regression in the speech domain was also present. Initially, she could tell her name and 8–10 words but after 1 month, she developed mutism.

A history was suggestive of fever with measles-like maculopapular rashes starting over the trunk followed by the involvement of the whole trunk at the age of 10 months. These rashes occurred 1 month after vaccination and subsided after 14 days for which the patient did not take any treatment for the same. The child was immunized for measles at the end of 9 months and 18 months. There was no history of irritability before the onset of symptoms, loose stools, febrile seizures, or trauma. A family history of seizure was also absent. The child was a product of non-consanguineous marriage and was born by normal vaginal delivery to a 26-year-old female. The antenatal and postnatal periods were uneventful and there was no history of measles in the mother either during pregnancy or at the time of delivery.

On examination, vitals were stable. On CNS examination, there was no cranial nerve palsy, bulk, the tone was normal, deep tendon reflexes in both upper and lower limbs were brisk

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and bilateral plantar upgoing. The sensory system examination was normal. Higher mental function and power could not be assessed. Eye examination and hearing assessment were normal. Other system examination was within normal limits (Fig. 1). A provisional diagnosis of acute encephalopathy was kept. The patient was started on sodium valproate at 20 mg/kg/day, and gradually, lamotrigine at 0.2 mg/kg/day and levetiracetam at 40 mg/kg/day were also added.

Investigations revealed hemoglobin of 11.2 g/dl, total leukocyte count of 6500 with a lymphocytic predominance of 63%, and neutrophils of 28% with platelets of 215,000/mm$^3$. Electrolytes, liver, and kidney function tests were within normal limits. On cerebrospinal fluid (CSF) examination, total cells were 5, the protein was 63 mg/dl, and glucose was 60 mg/dl with concomitant blood sugar of 90 mg/dl, IgG measles antibodies titer ratio in CSF to blood was 1:625, respectively, and confirmed the diagnosis. Electroencephalogram (EEG) was suggestive of abnormal slow-wave spikes (Fig. 2). Magnetic resonance imaging (MRI) was suggestive of T2/T2 FLAIR hyperintensities in the periventricular region in the parietal and occipital lobes which is a non-specific finding for SSPE. The fundus was normal (Fig. 3).

The prognosis was explained to the parents and the patient was planned to be started on interferon therapy, isoprinosine, and ribavirin. However, the parents refused the treatment due to financial constraints. At present, she is in Stage 3 of SSPE classification by Jabbour [5]. On follow-up after 3 months, she developed spasticity and dystonia with no improvement in the cognitive functions (Fig. 4).

**Figure 1: Initial image of the child**

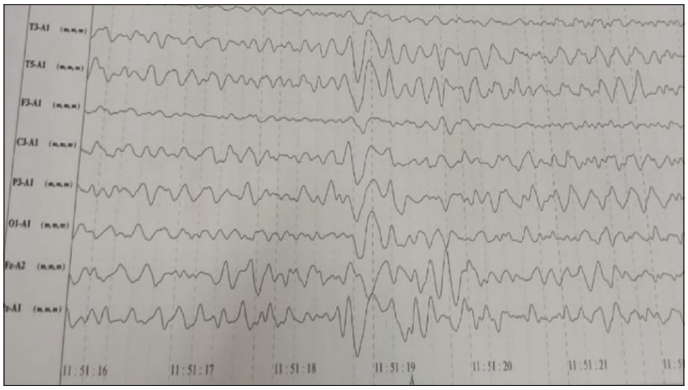

**Figure 2: Electroencephalogram showing decrease in amplitude background in Stage 3**

## DISCUSSION

Extensive vaccination programs against measles have led to a decrease in the incidence of measles in developed countries but it is still an important cause of morbidity in our country due to various risk factors. Various risk factors for measles include poverty, low socioeconomic status, rural area, uneducated parents, high birth order, and immature immune system [6]. Even though a number of elimination strategies and preventive methods have been taken to prevent measles, as per the recent studies, India ranks fourth among 194 countries in the number of measles cases registered between July 2018 and June 2019 [7].

Subacute sclerosing encephalitis is a late complication of measles. It is generally characterized by progressive slow myoclonus, cognitive decline, and extrapyramidal symptoms. As per Jabbour's classification [5], SSPE can be divided into four stages. Stage 1 comprises subtle changes in behavior such as irritability, reduced attention span, and temper outbursts.

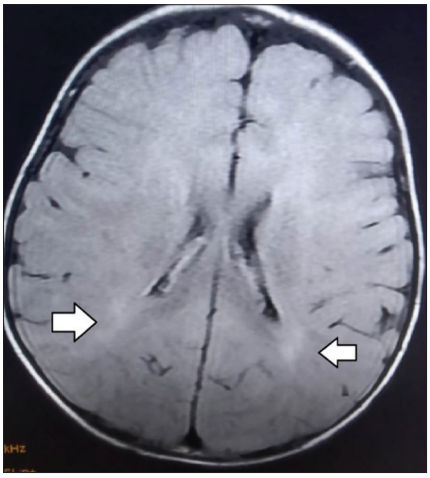

**Figure 3: T2 hyperintensities in periventricular region of the parietal lobes**

**Figure 4: Follow-up after 3 months showing generalized hypertonia**

Slow myoclonic jerks involving both the axial and appendicular muscles are the characteristic finding of Stage 2. Here, consciousness remains intact. In Stage 3, the patient develops choreoathetosis, immobility, spasticity, dementia, stupor, and gradually coma. There is a loss of critical centers that support breathing, heart rate, and blood pressure which finally leads to death in Stage 4.

An atypical form of SSPE occurs in about 10% of all patients. Unlike classical SSPE, in atypical form, there are no defined stages in clinical presentation due to rapid course. Atypical features also include unusual age of onset, visual loss, seizures, and other focal symptoms as initial presentations, a lack of SSPE-specific EEG pattern, and atypical fast progression of the disease. A patient could have more than 1 of these atypical features [3,7]. Our patient had an atypical presentation of SSPE as the latency period was short, the age of onset was young, and atypical EEG pattern of slow-wave spikes with absent burst suppression.

The latency period between onset of SSPE and measles infection is usually 6–8 years. Early-onset SSPE with short onset latency is generally associated with congenital and neonatal measles infection. Zwiauer *et al*. diagnosed a case of SSPE as early as 4 months of age after perinatally acquired measles infection [8]. It is seen that children with early age of onset of measles infection have a shorter latency period as seen with our case. At present, there are only a few case reports of SSPE seen in toddlers.

The protective efficacy of measles vaccination is very high and the role of measles causing SSPE is controversial. Our child developing measles after a month of immunization points out that the patient would have contracted a wild virus secondary to post-vaccination cannot be ruled out. The child had normal immunoglobulin levels and did not have any risk factors for measles. Recently, an association with programmed cell death protein 1 and children has been reported. The wild-type measles D3 and D6 are the most prevalent genotypes identified [9].

At present, no curative treatment is available for SSPE but therapy with immunomodulators such as isoprinosine, interferons, and antiviral drugs like ribavirin may help in halting the progression of the disease [1,10,11].

## CONCLUSION

SSPE is a significant clinical issue in developing nations. EEG and MRI of these patients must be assessed. Despite these, serum and CSF measles antibodies should be examined for confirmation. The possibility of SSPE developing after post-vaccination measles vaccination can also not be excluded. Due to the atypical presentation of SSPE, a high record of doubt must be kept to determine SSPE even with a short latency period.

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