Atypical Subacute Sclerosing Panencephalitis With Short Onset Latency

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An 11-month-old boy presented with focal seizures, myoclonic jerks and altered sensorium of one month duration, with a history of measles at eight months of age. A diagnosis of Subacute sclerosing panencephalitis (SSPE) was made on the basis of typical EEG changes and presence of anti-measles antibody in cerebrospinal fluid. A differential diagnosis of SSPE should be considered in all forms of acute encephalopathy in infants for early diagnosis and management.

Key words: *Diagnosis, Subacute sclerosing panencephalitis (SSPE).*

Subacute sclerosing panencephalitis (SSPE) is a slow progressive degeneration of the central nervous system caused by a persistent defective measles virus infection. The disease has a gradual progressive course leading to death in many cases within one to three years [1]. The latent period between measles infection and SSPE is commonly 6-8 years [2]. We report an infant with a very short latency.

CASE REPORT

An 11-month-old infant presented to us with complaints of right-sided focal seizures for 3 days followed by myoclonic jerks and altered sensorium for last one month. Prior to this illness, the infant was well and achieving age-appropriate milestones At presentation, the infant was unable to recognize his parents, unable to hold neck or sit, and not vocalizing bisyllables. Myoclonus of limbs was noted at the time of examination. Rest of the clinical examination was unremarkable.

Infant was a product of non-consanguinous marriage, and born by normal vaginal delivery. Antenatal and postnatal period was uneventful and there was no history of measles in mother either during pregnancy or at time of delivery. At eight months of age, he had history of fever, cough, coryza followed by maculopapular rash (first noticed at forehead then descended downward), which was diagnosed as measles by a pediatrician.

Complete blood count, serum electrolytes, liver and kidney function test, ESR, tandem mass screening, serum lactic acid, and ammonia were in normal ranges. Chest *X*-ray was normal and Mantoux test was non-reactive. Cerebrospinal fluid was clear with 4 cells (all lymphocytes), CSF protein 24 mg/dL glucose 55 mg/dL. EEG revealed periodic generalized complexes consisting of bilaterally symmetrical, high voltage (>200μV) bursts of sharp waves and delta waves which repeat at interval of 3 to 20 seconds interval with a slow background (***Fig.* 1**). Periodic burst was associated with each episode of myoclonus. As EEG picture was suggestive of SSPE, a sample of CSF and serum was obtained for anti-measles antibody. ELISA test using commercial kits for IgG antimeasles antibody was found positive both in CSF and serum (normal finding is CSF negative for IgG antimeasles antibody). While IgM anti-measles antibody was negative both in CSF and serum. Blood and CSF serology for *Herpes simplex*, Toxoplasma and Cytomegalovirus were all negative (both IgM and IgG).

MRI brain done at day five of admission revealed hyperintense signal in the cortex and subcortical white matter of frontal lobe. A repeat MRI done after one month revealed diffuse cerebral atrophy of brain. Child was treated with Isoprinosine (100mg/kg/day) but therapy with interferon was not affordable. Sodium valproate and clonazepam were added for control of myoclonus. However, after three months of continuous follow up, patient did not show any improvement in cognitive functions.

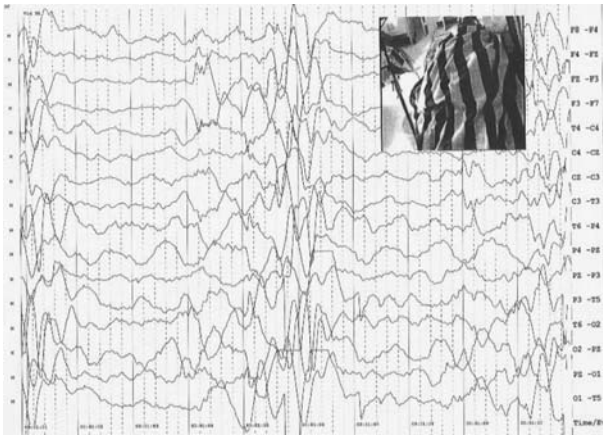

Fig.1 *Generalized periodic EEG pattern with a slow background.*

DISCUSSION

Most of the patients with SSPE have a history of primary measles infection at an early age. Children infected with measles under the age of one year carry a 16 times greater risk of SSPE than those infected at age five year or later. The diagnosis is based upon characteristic clinical manifestations, the presence of characteristic periodic EEG discharges, and demonstration of raised antibody titre against measles in the plasma and cerebrospinal fluid [1]. The latent period between measles infection and SSPE is around 6-8 years in most of the cases, but may range between 3 months to 18 years [2]. In this child, latent period of 2 months was noted which was much shorter.

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 [3]. 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 disease. A patient could have more than one of these atypical features [4]. This case is atypical as there is very early age of onset, a very short latent period of 2 months between measles infection and development of SSPE, and focal seizures as first symptom.

Early onset SSPE with short onset latency is generally associated with congenital and neonatal measles infection. Zwiauer, *et al*. [5] diagnosed a case of SSPE as early as 4 months of age after perinatally acquired measles infection. In four of the five cases described in the literature, onset of symptoms in the patients occurred under one year of age. However, the diagnosis of SSPE was made at 4 months, 13 months, 14 months, 18 months and 3 years of age in these series [5-7]. It appears that earlier the age of measles infection, shorter will be the latent period for development of SSPE.

The EEG pattern in our case was virtually diagnostic [1]. CSF IgG anti-measles antibody test in our patient was done with ELISA method, which has a sensitivity of 100% and a positive predictive value of 100% [8]. MRI commonly reveals focal abnormalities in the cortex and subcortical white matter early in the course of disease and diffuse cerebral atrophy at a later stage of disease [9].

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

A high index of suspicion is needed to detect SSPE with atypical presentation. As the disease can mimic acute encephalopathy, it is important to include SSPE on the list of differential diagnosis of acute encephalopathy, especially in infants.

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