

Case Scenario of Sub Acute Sclerosing Pan Encephalitis In Tertiary Care Hospital Karnataka , India



Medical Science

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Raghavendra M	Post graduate, Department of Paediatrics Cheluvamba Hospital , MMC& RI, mysore, Karnataka , India
Rajendra kumar	Assistant Profosor Department of Paediatrics Cheluvamba Hospital , MMC& RI, mysore, Karnataka , India
Sudha Rudrappa	Profesor & Head , Department of Paediatrics Cheluvamba Hospital , MMC& RI, mysore, Karnataka , India
Keerthan N	Post graduate Department of Paediatrics Cheluvamba Hospital , MMC& RI, mysore, Karnataka , India
Vinayaka Hegade p	Post graduate Department of Paediatrics Cheluvamba Hospital , MMC& RI, mysore, Karnataka , India

ABSTRACT

Subacute sclerosing panencephalitis (SSPE) is a slowly progressive degeneration of central nervous system caused by persistent defective measles virus infection. The latency period between measles infection and the onset of symptoms of subacute sclerosing pan encephalitis is commonly 6-8 years. To see clinical and laboratory findings of children presenting with SSPE and to correlate with measles and its immunisation. A differential diagnosis of SSPE should be considered in all cases of acute encephalopathy in younger age group for early diagnosis and further management.

INTRODUCTION:

Measles virus infection normally cause an acute self limiting disease in which a virus specific immune response leads to the establishment of lifelong immunity. Complications associated with acute measles can, on rare occasions, involve the central nervous system (CNS). These are postinfectious measles encephalitis which develop soon after infection; and months to years after the acute disease, results in measles inclusion body encephalitis (MIBE) and SSPE which are based on a persistent measles virus infection of brain cells(1). The disease may develop due to reactivation of the measles virus or an inappropriate immune response to the measles virus. SSPE usually develops 2 to 10 years after the original viral attack. Children and young adults are primarily affected with SSPE. Subacute Sclerosing Panencephalitis (SSPE) is a subacute inflammation of brain with a predominant childhood onset, caused by persistence of mutant measles virus in the central system (CNS)(2,3). The persistence of infection leads to degenerative changes in the brain resulting in progressive regression of acquired milestone of development, intelligence, alterations in behavior and myoclonic seizures and finally to death(4,5). SSPE risk is at least 10 times lower (5-50 times) in individuals after vaccination compared to the risk in individuals who had measles . Though many treatment protocols have been tried , no effective treatment is available till now. Here we present 2 cases of SSPE presented to us in the age group of 5-8 years

case reports:

case report 1:

A 7 year 6 month old male 2nd child born to non-consanguineous married couple with normal birth and developmental history with a past history suggestive of measles at the age of 2 years presented to cheluvamba hospital attached to MMC&RI with progressively increasing myoclonic jerks, ataxia, generalized tonic clonic convulsions and cognitive decline over a period of past 6 months. There was no history of measles in mother during antenatal period. On examination he was conscious and oriented. His vital parameters and anthropometric measurements being within normal limits.

On CNS examination, he had myoclonic jerks, spasticity, normal

deep tendon reflexes and extensor plantars with no focal neurologic deficit or cranial nerve palsy. Rest of the systemic examination was normal. Complete blood counts, ESR, serum electrolytes, liver and renal function tests were normal. His chest X ray was normal and Montoux test was negative. CSF examination showed normal cell count, protein and sugar.

An EEG was done which showed diffuse high amplitude bursts of periodic slow wave complexes suggestive of SSPE. Based on EEG picture, CSF was sent for IgG anti-measles antibodies which were positive with titre of 1:625. MRI of brain was non enhancing signal alteration in bilateral subcortical cerebral white matter. During the hospital stay his condition was progressive with increasing myoclonic jerks, generalized tonic clonic type of convulsions and gradually declining cognition and he was started on sodium valproate(20mg/kg/day) and discharged .

Case report 2:

5 year old boy born of non consanguineous married couple through full term normal vaginal delivery with normal perinatal history presented with loss of attained milestones since 4 months and myoclonic seizures since 2 months ,dystonic posturing of limbs,cognitive decline.past history of measles at 7 month of age. . On examination he was conscious and oriented. His vital parameters and anthropometric measurements were within normal limits.

On CNS examination, he had myoclonic jerks, spasticity, normal deep tendon reflexes and extensor plantars. There was no focal neurologic deficit or cranial nerve palsy. Rest of the systemic examination was normal. Complete blood counts, ESR, serum electrolytes, liver and renal function tests were normal.

CSF examination showed normal cell count, protein , sugar and 1:625 titre positivity for anti measles antibody. EEG showed periodic discharge suggestive of SSPE.child was on levetricitam(30 mg/kg/day) and sodium valproate(22 mg/kg/day) and discharged

Discussion:

Subacute Sclerosing Panencephalitis (SSPE) is a rare progressive neurological disorder that results as an indirect sequel to measles infection(3). SSPE incidence closely relates to measles incidence. This case reports shows the common clinical and investigation profile of SSPE cases admitted to cheluvamba hospital attached to MMCRI . Regarding age incidence, SSPE is a disease of childhood and early adolescence 3. The average age of presentation worldwide is between 5 and 15 years with the mean age being 9-10 years . In the present study both children were within age group of 5-10 year

The incidence is higher in males with a ratio of 2-4:1 female (3) , although primary measles infection shows no such sex disparity (6,7). In this case report both patients were males and came from low income group, which is in conformity with other studies that SSPE occurs mostly in lower socio-economic group (8,9).

SSPE is a post-measles complication , in this case report both patients had symptomatic primary measles infection. It may be due to subclinical measles infection or of ignorance of parents about identification of measles infection. As regards to vaccination, SSPE may develop in vaccinated children. Several reports suggested that this occurrence may be due to high prevalence of malnutrition in developing countries, improper vaccine coverage, poor quality, improper storage and transport of vaccine, subclinical measles infection prior to vaccination, poor seroconversion or vaccine failure, or circulation of atypical / wild measles virus strain (12).

In the study both patients were vaccinated, yet developed SSPE, which is not unlikely to happen in the presence of one or more factors mentioned above related to measles vaccination. There are no reported cases of vaccine associated SSPE because the DNA sequence of measles vaccine is different from those that of measles virus which causes SSPE(13,14).

Moreover ,epidemiological and virological data from meta-analysis suggests that measles vaccine does not cause SSPE

The milestones of development were normal in both cases before illness which deteriorated during clinical course of the disease. In this study, cognitive decline was present in both patients and one patient presented with h/o recurrent fall. The study by Akram et al(15) documented the cognitive decline in 86% patients and motor regression in 100%, which is consistent with the present study. In the clinical stage II, myoclonic seizure is the predominant feature of SSPE. Myoclonic seizures were present in both patient which was present in 74% of cases by Akram et al (15).

The most sensitive ELISA method was adopted for detecting measles antibodies in CSF . In the CSF, measles specific IgG antibody was positive in both patient. Diagnosis of SSPE was confirmed by elevated titers of measles antibodies in cerebrospinal fluid . In SSPE, measles specific antibodies are found in the

CSF due to intrathecal production of antibodies as specific immune response to virus in the central nervous system. The anti measles antibody in CSF was positive in 100% of patients with SSPE in another study by Akram et al(15) and comparing with few other studies.

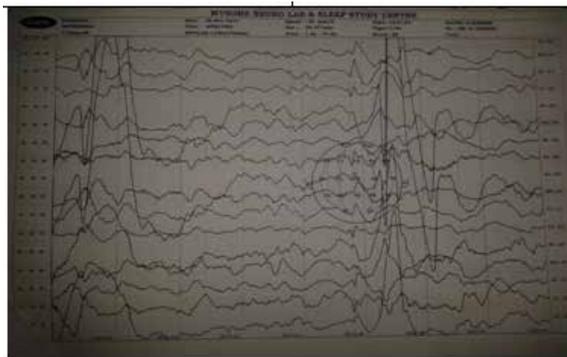
The classic EEG pattern in SSPE consists of periodic complexes (PC) with generalized, bilaterally symmetrical, high voltage bursts of polyphasic slow waves occurring synchronously throughout the recordings , which were also found in the both of the cases of the present study.

Conclusion:

SSPE is often not considered because of its rarity and the non-specific clinical manifestations at onset. Children presenting with deteriorating milestone of development along with myoclonic jerk should raise the suspicion for the diagnosis of SSPE . Investigations like CSF antibody to measles virus and characteristics EEG change may help further in the diagnosis.

Comparison of clinical and investigation profile of SSPE between two cases

	Case 1	Case 2
Age	5-10 year	5-10 year
Sex	Male	Male
Cognitive decline	Present	Present
Myoclonus	Present	Present
Altered speech	Present	Present
Vision	Normal	Normal
History of measles	Present	Present
Measles vaccine immunisation	Immunised	Immunised
Measles antibody Positive CSF IgG	Positive	Positive(1:625)
Clinical stage of SSPE at presentation	Stage 3	Stage 3
EEG Periodic complex	Present	Present



1: Electroencephalogram (EEG) at the time of presentation of case report 1. EEG reveals periodic bursts of high-amplitude, slow-wave complexes. The background rhythm is normal. This "burst-suppression" pattern is highly characteristic of sub-acute sclerosing panencephalitis

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