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Case Report

Unique Presentation of Subacute Sclerosing Panencephalitis as Post OPV Vaccine Falls in a Four Year Old Indian Girl: A Case Report

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Abstract

Background: Subacute sclerosing panencephalitis (SSPE) is a chronic encephalitis occurring after infection with measles virus. We present a child with SSPE whose initial presentation was like post-polio syndrome.

Case Report: This Four year old girl presented with complaint of multiple episodes of falls for 1 week after OPV vaccination. Central nervous system examination was normal. The initial diagnosis of post-polio syndrome was considered. Her MRI brain showed cerebral cortical T2 hyperintensities, EEG showed periodic generalised delta slow waves intermixed with spikes. SSPE was considered and the diagnosis was confirmed with detection of measles antibodies in cerebrospinal fluid.

Conclusion: Presentation of our case was like a post-polio syndrome, so whether there is any association between live virus vaccination and reactivation of latent measles virus needs to be confirmed by future larger studies.

Keywords: *Measles; SSPE; Falls; Isoprinosine*

Introduction

Subacute sclerosis panencephalitis (SSPE) is a persistent and chronic encephalitis secondary to measles virus infection that causes widespread demyelination of the central nervous system [1]. SSPE has 4 clinical stages. Falling episodes due to myoclonus are troublesome for patient and these falls restrict patient's daily life. Clinical features are poor school performance, progressive intellectual deterioration, personality changes, behaviour abnormalities; followed by steady motor decline, myoclonus, focal paralysis, seizures, autonomic failure, rigidity, finally leading to death with akinetic mutism [5-9]. Isoprinosine and Interferon are therapeutic agents for SSPE [2]. Here, we report a unique case of SSPE as Post OPV vaccine falls.

Case Report

Four years girl was brought with complaints of multiple episodes of forward falls every day. 2 weeks back, she was given oral polio vaccine, after 1 week from OPV vaccine administration she had low grade fever, cough, cold, managed with symptomatic measures, after another 1 week parents noticed episodes of multiple falls per day while walking, forward, unprotected falls, without loss of consciousness, she used to get up on her own after every fall, she was brought to our health facility. Her vital parameters were normal, anthropometric parameters were within normal range, detail central nervous system examination was normal, other systemic examinations were normal, possibilities considered were post-polio syndrome, epileptic axial myoclonus. Complete blood counts, serum electrolytes were within normal range, AFP authorities were informed and 2 stool samples were sent to referral laboratory which were reported as negative, nerve conduction study was normal, CSF routine analysis including cells, protein, sugar, gram stain, AFB stain, bacterial culture, AFB culture were normal, contrast MRI brain showed nonspecific cerebral cortical T2 hyperintensities, Contrast MRI Spine was normal, EEG showed periodic generalised delta slow waves intermixed with spikes. After seeing EEG pattern family was asked in detail about past history of

any exanthematous fever and they admitted at 1 year of age child had fever with rashes, probably must had been measles. Possibility of subacute sclerosing pan encephalitis was considered so CSF and serum measles antibodies were sent, Serum IgG measles antibodies 14268 U/ml, CSF IgG measles 14824 U/ml, Serum total IgG 1650 mg/dl (normal range 700 - 1600 mg/dl), CSF total IgG 14.2 mg/dl (normal range 0 - 3.4 mg/dl), CSF measles IgM 0.12 (Negative < 0.9, equivocal 0.9 - 1.09, positive > 1.1), CSF to serum measles specific IgG quotient reference-positive: 5.23 (normal range < 1.3, equivocal 1.3 - 1.5, positive > 1.5). Child was initiated on oral Isoprinosine, oral sodium valproate, clobazam, family was explained in detail about natural course and prognosis of illness. Our case is unique as presentation was like a post-polio syndrome, whether there is any association between live virus vaccination and reactivation of latent measles virus needs to be confirmed by future larger studies.

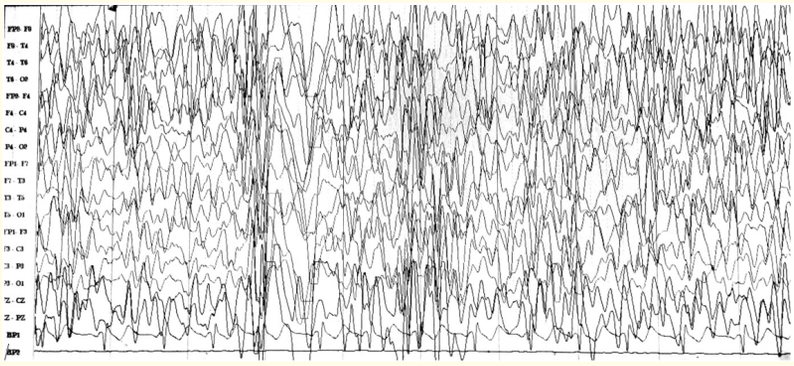

Figure 1: *EEG epoch showing periodic generalised delta slow waves intermixed with spikes.*

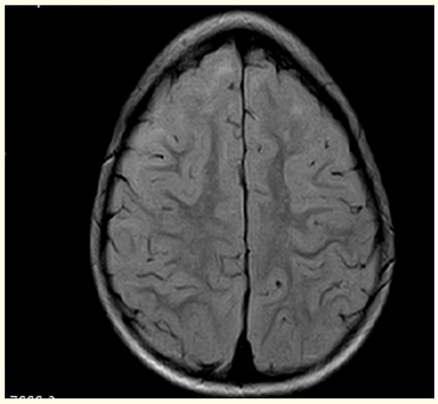

Figure 2: *MRI FLAIR sequence showing cerebral cortical hyperintensities.*

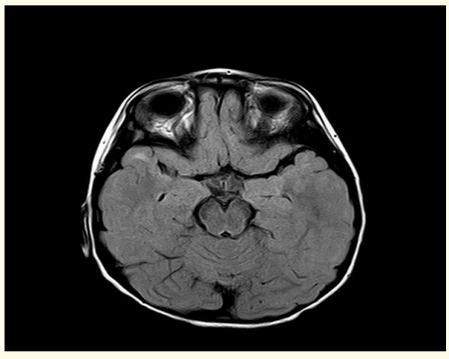

Figure 3: *MRI FLAIR sequence showing cerebral cortical hyperintensities.*

Discussion and Conclusion

Subacute sclerosis panencephalitis (SSPE) is a persistent and chronic encephalitis secondary to measles virus infection that causes widespread demyelination of the central nervous system [1]. SSPE is caused by persistant infection of the brain by a mutated form of measles virus, earlier the age of exposure to measles virus, greater the likelihood of developing SSPE because of immune system immaturity [3]. The average period between exposure and onset of SSPE ranges between 3 and 12 years [4]. SSPE has four stages, Myoclonus involves the extremities, trunk, or head, generalized or focal seizures may begin simultaneously. As the disease progresses, speech and intellectual function deteriorate, myoclonus increases and other neurologic manifestations, such as choreoathetosis, bradykinesia, or rigidity, appear. Approximately half of patients have chorioretinitis and visual impairment. Our case had falls after 1 week from Oral polio vaccine administration so post-polio syndrome was considered as a differential, but MRI Brain and EEG led us to diagnosis of SSPE so CSF measles antibodies were sent, which confirmed diagnosis of SSPE. Patients with SSPE have elevated CSF immunoglobulin levels, oligoclonal bands, and increased IgG synthesis rates. High CSF titers of measles-specific IgG establish the diagnosis of SSPE, and measles virus RNA can be detected in CSF or plasma by reverse transcription PCR. MRI changes in SSPE shows initial cortical and subsequent subcortical or periventricular white matter involvement, our case had T2 FLAIR cortical hyperintense areas. These changes occur initially in parietal or occipital lobes [10] and eventually cortical atrophy. The EEG shows diffuse slowing, focal or generalized epileptiform discharges, bilaterally synchronous spike-wave or slow-wave bursts with regular periodicity which eventually assume a suppression-burst pattern. There is no cure for SSPE, is a relentlessly progressive disease, oral Isoprinosine, intrathecal INF-a, ribavirin, immunoglobulin, H2 receptor blockers, have been tried, alone and in combination, with little or no effect. Antiepileptic drugs for myoclonic jerks have been suggested, divalproex sodium, clonazepam, or levetiracetam may provide some relief. SSPE ends in debility, coma and death. In conclusion, interaction between host's immune system and measles virus is poorly understood so whether there is any association between live virus vaccine administration and reactivation of latent measles virus needs to be confirmed with future larger studies. Currently high vaccination coverage rate is most crucial factor in preventing SSPE.

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