ORIGINAL RESEARCH PAPER

Paediatrics

A CASE REPORT OF PAEDIATRIC SUBACUTE SCLEROSING PANENCEPHALITIS

KEY WORDS:

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ABSTRACT

Subacute sclerosing panencephalitis (SSPE) is a devastating disease of the central nervous system (CNS) caused by persistent measles virus infection. The diagnosis of SSPE is based on characteristic clinical and EEG findings and demonstration of elevated antibody titres against measles in cerebrospinal fluid. Subacute sclerosing panencephalitis can have atypical clinical features at the onset.We report a 10 year old boy presented with convulsions and visual disturbances. The disease progressed with behavioural and cognitive disturbances and periodic high amplitude generalised complexes on EEG, and elevated titers of measles antibodies in cerebrospinal fluid leading to the final diagnosis of subacute sclerosing panencephalitis.

INTRODUCTION:

Measles is still a common communicable disease, particularly in Africa and Asia. As per the recent WHO report, 7 million people were found to be affected with measles in 2016.(1) Approximately, 38% of Indian children fail to receive the basic immunisation in the first year of life.(2) The Government of India introduced a second dose of measles vaccination drive in 14 high-risk Indian states with approximately 134 million children to prevent around 60,000 to 100,000 child deaths annually.(3)

Subacute sclerosing panencephalitis (SSPE) is a progressive neurological disorder caused by persistent measles virus infection.(4) SSPE is characterised by progressive mental decline, myoclonus, and raised anti-measles antibody titer in the cerebrospinal fluid. A periodic EEG record, typically, consists of generalised and synchronous bursts of sharp–slow wave complexes. Pathologically, there are diffuse perivascular lymphocytic infiltration and intranuclear eosinophilic inclusion bodies.

CASE REPORT:

A 10 year old boy , with normal growth and development ,came to tertiary care hospital with complaints of convulsion since last 10 months. It was generalised tonic clonic convulsions, while awake , while afebrile lasting for 1-2 mins, associated with clenching of teeth and uprolling of eyeball ,not associated with bowel-bladder incontinence. Initially convulsions were 1-2 episodes per week,later increased to 2-3 episodes per day since last 2 months . It is associated with behavioural abnormalities in the form of irrelevant and abusive speech. Patient had multiple staring episodes lasting for 10-20 seconds without loss of consciousness since last 4 months.Then the patient also had brief ,jerky,shock-like movements of both upper limbs not associated with loss of consciousness.Patient had difficulty in vision since the last 2 months.MRI done at that time suggestive of hyperintense areas of preferential subcortical white matter abnormality seen in posterior occipito parietal lobes, subinsular region and left frontal region.Patient was treated with IV immunoglobulin for 2 days, Inj rituximab and Inj MPSS for 5 days followed by oral steroids with tapering doses. Patient had a past history of measles at one year of age. Patient is unvaccinated ,with uneventful birth history and family history, development wise appropriate for age.

On examination, patient was conscious, confused and disoriented to time ,place and person ,hemodynamically stable, with memory disturbances of short and long term, and irrelevant speech and language with normal tone, power and reflexes,gait, pupils reacting to light, with no signs of cerebellar abnormality. During Course of hospitalisation, MRI brain done on D2OA suggestive of asymmetrical abnormal area of T2 and FLAIR hyperintensity in cortical and subcortical white matter of bilateral parieto-occipital lobes(right>left) and head of left caudate nucleus- suggestive of -subacute sclerosing demyelinating encephalitis.Also there is white matter volume loss(right> left) and generalised dilatation of both lateral ventricle suggestive of cerebral atrophy.CSF Tapping was done on day two of admission and CSF anti measles antibody was found to be positive. EEG done on day two of admission suggestive of generalised periodic complexes occurring at an interval of 2-7 secs. Periodic complexes consisted of predominantly high amplitude slow waves with background slowing s/o SSPE.The patient was treated with tab clobazam, tab quetiapine, tab levetiracetam on dayone of admission. On day three of admission, tab isoprinosine was added.

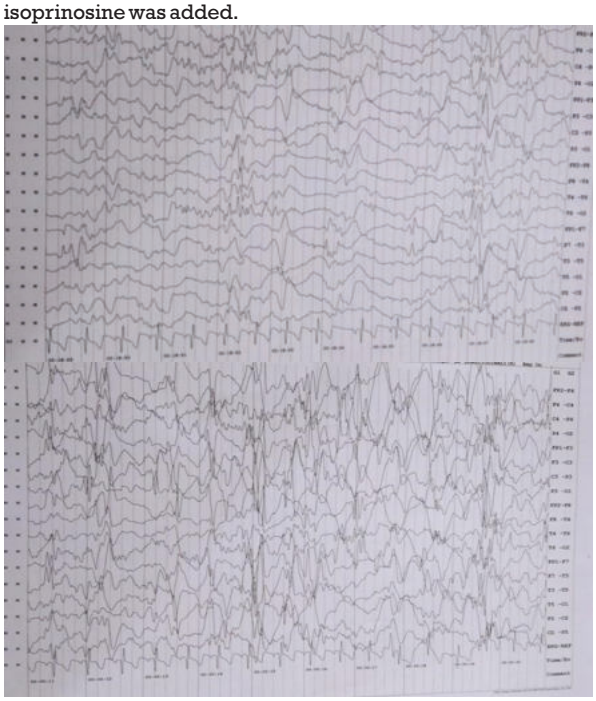

Figure 1: EEG Showing Generalised Periodic Complexes

Consisted Of Predominantly High Amplitude Slow Waves With Background Slowing

DISCUSSION:

SSPE is one of the most frequent causes of progressive cognitive decline in developing countries. The disease may present with varying symptoms. Uncommonly, subacute sclerosing panencephalitis may manifest with different types of seizures, such as generalized tonic-clonic seizure and myoclonic atonic seizures (5) .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.(6) 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 (7). 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.(6)In this child, a latent period of 7-8 years was noted after which symptoms started in form of seizures initially generalised tonic-clonic followed by jerky movements associated with behavioural abnormalities..There was difficulty in vision(8) .There was no associated fever , any other systemic complaints .There is a positive history of measles infection with unvaccinated patient.

In light of these findings, subacute sclerosing panencephalitis was suspected and the diagnosis of subacute sclerosing panencephalitis was confirmed by the detection of cerebrospinal fluid measles antibodies(9).EEG suggestive of generalised periodic complexes predominantly high amplitude slow waves s/o SSPE.In conclusion, subacute sclerosing panencephalitis is a rare complication of measles infection. We strongly recommend screening for SSPE in children with new onset cognitive deterioration and myoclonic jerks. Increasing awareness of SSPE in the paediatrician and neurologists can help in the early diagnosis of the patients and prevent unnecessary investigations.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 which were seen in this child.(10)

CONCLUSION:

SSPE is a slow virus infection caused by an aberrant measles virus. This disease is still common in developing and underdeveloped countries. One of the most important limitations in treatment of SSPE is difficulty in recognising early manifestations of disease, when the inflammatory changes are, possibly, still reversible.Treatments available are very costly and are available only at a few centres in the world. Moreover, these treatments are not curative and only help in buying time for these patients. (11)The families of patients with SSPE have a lot of physical, psychological, and economical stresses to endure.At present effective measles vaccination seems to be the only solution to the problem of this dreaded neurological disorder .

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