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Original Research Paper

Neurology

SSPE (SUBACUTE SCLEROSING PANENCEPHALITIS) - THE GREAT MASQUERADER

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ABSTRACT

Background: SSPE is chronic progressive encephalitis affecting children and young adults which usually presents with cognitive decline and behavioural changes followed by periodic myoclonic jerks, seizures, vision loss and ataxia. High degree of suspicion is required as the presentation can be variable and can have many differentials.

Aim: We aim to study various presentations of SSPE

Methods and Material: Retrospective study was done to analyse various presentations in patients diagnosed with SSPE as per modified Dykens criteria from a tertairy care centre over a period of 2 years (1st January 2018-31st December 2020).

Results: 6 cases of SSPE were identified.

Case1 - 22 months old presented with subacute history of ataxia, multifical myoclonus and developmental regression. Gradually myoclonus worsened to involve trunk and developed drop attacks.

Case 2 - 17 years boy presented with single episode of seizure.

Case 3- 25 years female with 5 months gestation presented with subacute vision loss followed by progressive cognitive decline, behavioural changes, Parkinsonism, Dystonia and stimulus sensitive myoclonus.

Case4- 28 years female presented with rapidly progressive cognitive decline and behavioural changes.

Case 5- 32 years male presented with history of myoclonic jerks and dropping of objects. Gradually developed progressive behavioural changes and cognitive decline and became vegetative.

Case6 - 9 year old child presented with faciobrachial seizures (myoclonic jerks) and scholastic backwardness.

Conclusions: SSPE can manifest with varied presenting complaints. Also, Results of EEG, MRI and CSF examination can change during the disease course. Therefore, high degree of suspicion is required for early diagnosis of this challenging entity.

KEYWORDS : SSPE, Presentation, outcome.

INTRODUCTION-

SSPE is chronic progressive encephalitis affecting children and young adults which usually presents with cognitive decline and behavioural changes followed by periodic myoclonic jerks, seizures, vision loss and ataxia. High degree of suspicion is required as the presentation can be variable and can have many differentials.

METHODS-

We present 6 cases of proven SSPE diagnosed with Dyken's criteria over a study period of 1 year (October 2018 to September 2019) with varied initial presentations.

RESULTS-

Case 1 - 22 months old presented with two months progressive history of ataxia, multifocal myoclonus and developmental regression. During the ward stay, myoclonus worsened to involve trunk and developed drop attacks. MRI brain showed asymmetric white matter changes. CSF showed positive anti measles antibody titres confirming the diagnosis of SSPE. There was no history of measles and child was vaccinated with measles vaccine. On 6 months follow up, the condition of the child remained static without further regression on treatment with valproate and isoprinosine.

Case 2 - 17 years healthy boy presented with single episode of seizure and on evaluation MRI brain showed bilateral occipital white matter changes. EEG showed intermittent occipital slowing with sharp wave discharges. CSF examination confirmed SSPE. He is under observation for last 6 months and has worsened in scholastic performance and developed myoclonic jerks.

Case 3- 25 years female, 5 months gestation presented with vision loss for few weeks followed by progressive cognitive decline, behavioural changes, Parkinsonism, dystonia and stimulus sensitive myoclonus. EEG showed periodic discharges and MRI brain showed generalised cerebral atrophy with diffuse white matter changes. Relatives denied CSF examination. Fetus died in utero and patient s

Case4- 28 years female presented with progressive 2 year history of cognitive decline and behavioural changes. MRI brain showed white matter changes. EEG showed generalised slowing. Csf examination confirmed SSPE. Patient progessively worsened and became bed bound over next 6 months.

Case 5- 32 years male presented with history of myoclonic jerks and dropping of objects. EEG showed periodic discharge, CSF examination confirmed SSPE. Over next few months, developed progressive behavioural changes and cognitive decline and became vegetative.

Case6 - 9 years previously healthy male child presented with seizures and scholastic backwardness. Over next few months developed faciobrachial seizures and had worsening cognitive decline. EEG showed periodic discharges and MRI showed white matter hyperintensities. CSF examination confirmed SSPE. Child succumbed after 2years of symptom onset.

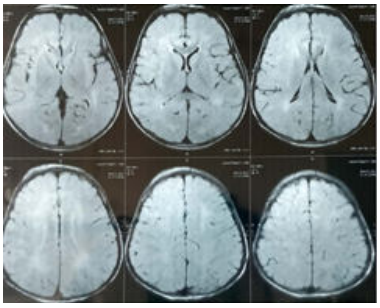

Figure no 1: MRI FLAIR showing multifocal white matter hyperintensities

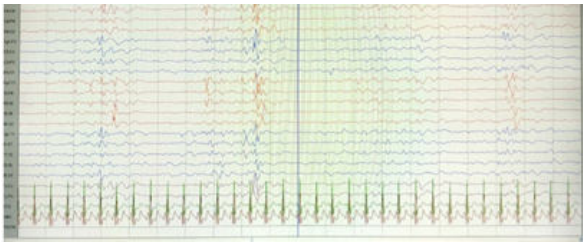

Figure no 2: EEG slowing slow sharp wave generalized quasiperiodic discharges

DISCUSSION-

SSPE is a late manifestation of measles infection occuring in childhood. Children affected in infancy carry 16 times the risk of SSPE than those affected after 5 years of age.[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] The earlier the age of measles infection, shorter will be the latent period for the development of SSPE.[1] history of antecedent measles infection is commonly seen. However, history of measles infection may not be there as the infection can be subclinical or mild in many of cases.[3] Only 3 out of 6 of our patients had antecedent history of measles infection. Atypical forms of SSPE occurs in about 10% of all patients in which unlike classical SSPE, there are no defined stages in clinical presentation due to rapid course. Atypical forms include varied initial presentations such as vision loss, seizure, focal symptoms, gait ataxia and ritualistic behaviors.[1,4] SSPE should be considered in the differential diagnosis of hemiparesis, cerebellar ataxia and acute encephalopathy.[5] SSPE presenting as headache with papillodema and multiple cranial neuropathies has also been described in the literature.[6] Atypical presentation with parkinsonism has also been described.[7] The initial presentation depends on the part of the neuraxis which is first to get involved. SSPE affects the occipital area initially followed by spread to the anterior portions of the cerebral hemispheres, subcortical structures, brain stem and spinal cord.[8] Atypical EEG patterns can be seen early in the disease course with focal slowing, focal epileptiform discharges at at times EEG can be normal.[9] CSF testing for measles antibody has 100% sensitivity and 100% positive predicitive value.[1]

Conclusion- SSPE can manifest with varied presenting complaints. Results of EEG, MRI and CSF examination change during the disease course. Also, Initial presenting complaints can be variable. Hence high degree of suspicion is required for early diagnosis of this challenging entity. Research needs to be done with regards to treatment response with stage of illness because early treatment with antivirals does seem to prolong/ stabilise the disease progression initially.

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