

Atypical Presentation of Sub Acutesclerosing Pan Encephalitis With Short Onset Latency

Wardhane S¹, Oswal J², Shrivastava K³

Abstract

The most severe sequel of measles virus infection is sub acutesclerosing pan encephalitis (SSPE), a fatal disease of the central nervous system that generally develops 7–10 years after infection. A 7 ½ year old boy presented with gradual difficulty in walking and speech along with jerky movements of right upper limb which progressed to become generalized and progressive loss of cognition was also observed within days of onset of symptoms. Diagnosis of SSPE was made on the basis of burst suppression pattern on EEG and anti measles antibody in CSF. A differential diagnosis of SSPE should be considered in all forms of acute encephalopathy for early diagnosis and treatment.

Introduction

Sub acutesclerosing pan encephalitis (SSPE) is a progressive fatal disease of the central nervous system that is caused by a persistent measles virus infection. Early clinical characteristics of SSPE may be variable, but they often include behavioural changes, cognitive deterioration, sporadic episodes of falling, and such optic disturbances as chorioretinitis^{1,2}. A 7 ½ year old male child presented with history of gradual difficulty in walking and speech along with jerky movements of right upper limb, which then became generalized. A diagnosis of SSPE was made on the basis of EEG findings and presence of anti- measles antibody in CSF.

The Case

A 7 ½ year old male child was brought with complaints of gradually increasing difficulty in walking along with associated speech abnormality. He also had sudden jerky episodes of right upper limb which gradually progressed to generalized myoclonic spasms. He was drowsy irritable on presentation and responding to mother's voice but had increased tone in all four limbs and brisk reflexes, clonus was also observed. Gradually his cognition deteriorated and he stopped responding to any stimuli. He continued to have multiple episodes of myoclonus.

There was no significant past history or history of suffering from measles during childhood. Until onset of symptoms, child had apparently normal behavior and cognition. He was also vaccinated for age with all primary vaccines.

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Initially other causes for sudden onset weakness with myoclonus were evaluated. CSF and fundus examination was normal. EEG done was suggestive of periodic epileptic discharges, bilaterally symmetrical and synchronous. MRI brain was also suggestive of discrete hyper intensities in sub cortical white matter of bilateral fronto-parietal regions, representing early changes of sub-acute sclerosing pan encephalitis. CSF for anti- measles antibody was reported positive. A diagnosis of atypical sub-acute sclerosing pan encephalitis was made as there is history of short onset latency.

Child was treated with Sodium valproate, Clonazepam and Amantidine, but continued to have episodes of myoclonus and there is no improvement in cognition observed.

Discussion

Sub-acute sclerosing pan encephalitis is a progressive inflammatory disease of central nervous system caused by a persistent aberrant measles virus infection³. In the classical form of SSPE, slow progression of the disease through four characteristic stages is observed:

1. Slowly evolving behavioural and intellectual deterioration
2. Various types of involuntary movements
3. Severe pyramidal and extra pyramidal hypertonus and disappearance of hyperkinesias
4. Chronic vegetative state and death^{4,5}.

Stage I may last from several weeks to years, stage II lasts from three months to one year. Stages III and IV last for a duration ranging from six months to one year^{4,5}. But in a series by Risk and Haddad approximately 10% of patients had a fulminant progressive form⁶. Similarly Bojinova et al in a 25 year epidemiology study found 10% of patients with a fulminant course⁷.

In early stages of SSPE, MRI shows lesions involving the parieto-occipital, cortico-subcortical regions asymmetrically. In time symmetric periventricular changes become more prominent, however MRI findings can be normal during early stages of disease⁸, as was observed in our patient. Diagnosis of SSPE is based on clinical findings, typical EEG changes (bilaterally symmetrical and synchronous periodic complexes) and elevated CSF titers of anti-measles antibody^{9,10}.

Treatment for SSPE is still undetermined. Antiviral agents such as Amantidine, Ribavirin or immunomodulators like Isoprinosine, interferons, immunoglobulins, and corticosteroids have been used. However the results of the treatment of SSPE are not totally satisfactory^{11,12}.

Conclusion

SSPE has recently been seen with more atypical symptoms and more acute, fulminant course. High index of suspicion should be present in patients with acute neurological signs like ataxia, dysarthria and

inability to walk. Early diagnosis is important for the effectiveness of treatment.

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