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Case Report

Fulminant Subacute Sclerosing Panencephalitis (SSPE) – An Ordeal for the Family and the Clinician!

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Abstract

Background: Subacute sclerosing panencephalitis (SSPE) is a rare condition notorious for causing a chronic, progressive encephalitis that affects children and young adults, resulting from persistent infection with an immune-resistant measles virus.

Methods: Descriptive study of a case of an eleven-year-old child who had a rapid and fulminant course of illness, suggesting a rare phenotype of "Fulminant SSPE". This was confirmed with an electroencephalogram (EEG) and measles antibodies in the cerebrospinal fluid (CSF).

Results: The clinical deterioration and electrophysiological features with imaging provided evidence of a grave entity. Our patient had a downhill course despite starting on intrathecal interferon therapy. The child succumbed to the illness very rapidly, and autopsy revealed predominant inflammation and minimal gliosis.

Conclusion: Awareness of this rare entity, fulminant SSPE, is of paramount importance to recognise it early (as the presentation may be atypical), prognosticate about the illness, and initiate available immunomodulatory therapy.

Keywords: *Subacute sclerosing panencephalitis (SSPE), Fulminant, Intrathecal interferon*

Introduction

Subacute sclerosing panencephalitis (SSPE) is a rare entity that is notorious for causing a progressive encephalitis due to persistent infection of immune-resistant measles virus.[1] Measles is a highly contagious infection, and it is one of the deadliest vaccine-preventable diseases of childhood. It is a WHO-notifiable disease.[2] The incubation period can vary from a few weeks to 27 years after acquiring the infection in childhood.[3] The Government of India gives a combined Measles-Rubella vaccine at nine and fifteen months of age.[4,5] Therapies involving oral administration of Inosine pranobex and intraventricular administration of interferon (IFN) have been attempted in SSPE and proven to be effective, though temporarily.[6] A rapidly progressive fulminant course has also been described, which can have a very misleading phenotype in the form of focal neurological signs and the absence of typical features of SSPE.[7] Fulminant SSPE is defined as death within 6 months of the onset of the disease.[7]

Case Presentation

An eleven-year-old boy with a normal perinatal and developmental history presented with a progressive decline in his scholastic performance, memory disturbances and recurrent falls in the last 3 months. The falls were characterised by a sudden jerk involving the head and trunk with the right arm, resulting in a loss of posture. Higher mental function testing revealed impaired recent memory, impaired calculation and psychomotor slowing. Neurological examination revealed bilateral incoordination and gait ataxia. He had frequent myoclonic jerks, suggestive of "slow myoclonus", a classical feature of SSPE. There was no history of measles infection in his childhood, and his vaccinations were appropriate according to the National Immunisation Schedule (NIS).

The electroencephalogram (EEG) (Figure 1) showed a slow background activity in the theta range with intermittent quasi-periodic "Radermecker complexes". Magnetic resonance imaging (MRI) of the brain (Figure 2) revealed nonspecific frontal white matter hyperintensities on fluid attenuation inversion recovery (FLAIR) sequence, with no significant abnormality on T1 and T1 post-contrast imaging. Cerebrospinal fluid (CSF) analysis showed an elevated IgG anti-measles antibody titer. SSPE was thus diagnosed based on Dyken's criteria.[1] He was started on intrathecal IFN and oral Isoprinosine as per the hospital protocol. After the first dose of intrathecal IFN, the boy developed a high-grade fever, a drop in sensorium and new-onset generalised seizures. He was shifted to our neurocritical care unit and intubated. The possibility of drug-induced aseptic meningoencephalitis (DIAM) was considered, and further doses of IFN were deferred. Repeating neuroimaging and repeat CSF analysis were normal. He gradually weaned off the ventilator and was discharged home after 3 weeks of hospital stay. He was readmitted to our hospital with a recurrence of seizures. Despite our best efforts, he succumbed to the illness. After obtaining consent from the parents, an autopsy was performed. Post-mortem microbiology reports were negative.

The coronal slice (Figure 3A) of the brain showed minimal greyish discolouration of the white matter with significant perivascular and parenchymal inflammation (Figure 3B). Cowdry inclusions in the neurons are highlighted by immunohistochemistry (Figure 3C). There was a significant inflammatory response with minimal demyelination and gliosis (Figure 3D), contrary to the typical description in "fulminant SSPE".

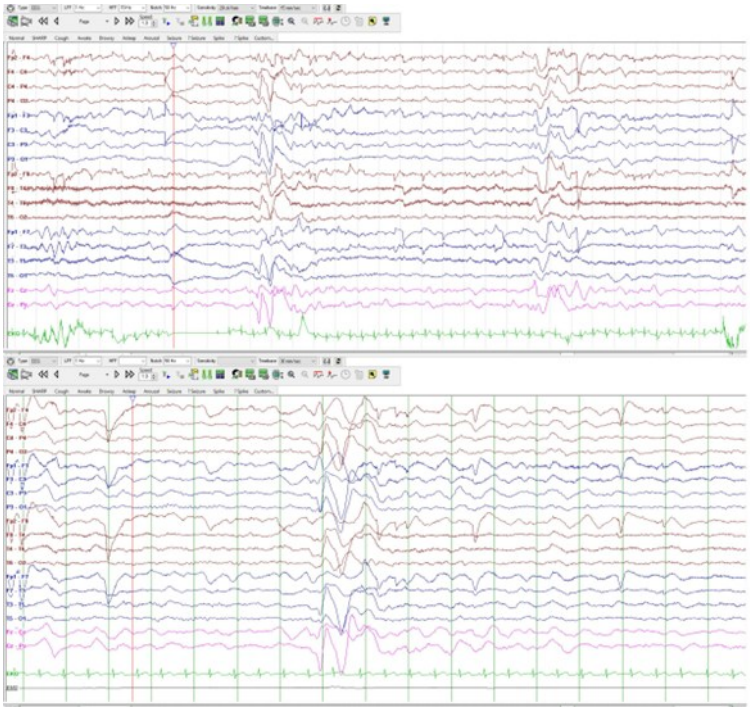

Figure 1. *Electroencephalogram (EEG) showing a slow background activity in the theta range with intermittent quasi-periodic "Radermecker complexes"*

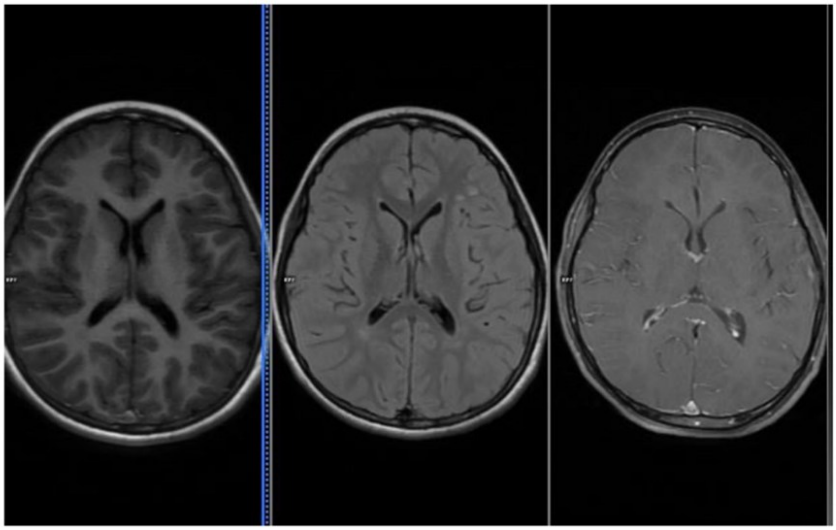

Figure 2. *Magnetic Resonance Imaging (MRI) of the brain revealed nonspecific frontal white matter hyperintensities on fluid attenuation inversion recovery (FLAIR) sequence, with no significant abnormality on T1 and T1 post-contrast imaging.*

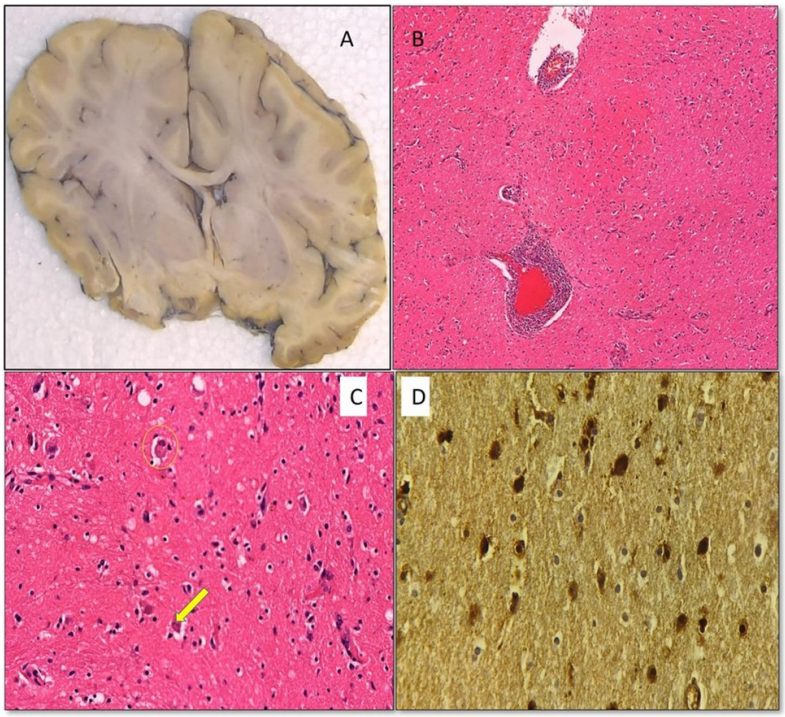

Figure 3. ***A.*** *The coronal slice shows minimal grey discolouration of the white matter.* ***B.*** *The cortex and white matter with significant perivascular and parenchymal inflammation.* ***C.*** *The presence of Cowdry inclusions.* ***D.*** *Inclusions highlighted by immunohistochemistry.*

Discussion

This case report highlights a grave presentation of a common pediatric illness. None of the available therapies have shown robust improvement in the neurological condition of the patient. Various IFN regimens have been designed by various institutes across the globe. The regimen we followed at our institute has been adapted from Garg et al. [4] Interferon alpha is given as a slow injection in the dose of 6 million units weekly for 6 weeks, which is followed by a monthly dose for 6 months or longer; usually combined with Isoprinosine.[4]

Our patient had a stormy course following the intrathecal INF therapy. Though intrathecal IFN has been shown to halt the progression of disability, many adverse events have been reported by various authors. [8,9,10]

The first hypothesis to explain the stormy course in our patient is the possibility of a “fulminant SSPE” due to more suppressed immunity or a rapid increase in the virus virulence.[7]

The second hypothesis is the possibility of immune dysregulation and worsening of clinical condition following the intrathecal IFN therapy with suspected "interferonopathies".[8] There is no pathological precedent to detect a possible interferonopathy, which could not be ruled out in our patient.

Conclusion

Awareness about fulminant SSPE is crucial for prognostication and early initiation of treatment. The most appropriate form of IFN and the most suitable dose of IFN are subject to variability and require standardisation.

Conflict of Interest

The authors declare no conflict of interest.

Acknowledgement

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