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Case Report

Subacute sclerosing panencephalitis (SSPE): Late onset of measles complication - Case of IVIG treatment failure

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ABSTRACT

Subacute sclerosing panencephalitis (SSPE) is characterized by gradual deterioration of cognitive motor and cognitive functions. SSPE more often occurs in individuals who had a history of measles viral infection. We present a case of 8 year old male child who was presented with generalized progressive weakness, recurrent falls to one side, multiple episodes of seizure, urinary incontinence. On liaison it was understood that he had a history of measles at the age of 3 years. Examination showed elevated serum measles antibody titer, electro-encephalogram (EEG) analysis revealed stereotyped polymorphic discharge seen periodically, suggestive of SSPE. He was treated with IVIG and other symptomatic treatment. However he had recurrent episodes of vomiting after IVIG administration, Ribavirin administration caused multiple oral lesions. Upon discharge he was given Syr. Valproate, Oral Steroid (Prednisolone) tapering therapy, THP 2 mg BD, PPI and Laxatives. Parents were advised to maintain RT feed of 150 mL, once in 3 hours. After 1 week, he succumbed to his illness at his residence.

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1. Introduction

Subacute sclerosing panencephalitis (SSPE) is characterized by gradual deterioration of cognitive motor and cognitive functions. SSPE more often occurs in individuals who had a history of measles viral infection. The annual incidence of SSPE is reported to be 10-20 per million populations. The virus can remain inactive or dormant for years, after which an inflammatory response against infected cell occurs. The inflammatory response then give rise to proliferation of astrocytes, lymphocytic and plasma cell infiltration, neuronal degeneration. Electroencephalogram, CSF analysis for anti-measles IgG remains as a strong diagnostic parameter.[1–4]

2. Case Report

A 8 year old male child was bought to the care center with generalized weakness and had recurrent falls to one side or forward since 2 months. He was the third child born to NCM parents. The antenatal period was found to be uneventful as reported by the parents. His mother reports that he had developmental delay, but remained asymptomatic prior to 2 months of presentation. Fall towards one side or front occurred daily lasting 30 minutes after waking up. Mother says the frequency of falls has reduced over past 1 week. He had history of 6 episodes of focal seizure (L). Also, the child had reduced attention, educational regression +, altered sleep pattern, occasional twisting movements of upper limb and intermittent tonic posturing. Mother says there were tightening or stiffening of UL & LL along with urinary and bowel incontinence, since 3 – 4 days.

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Upon liaison, it was understood that the child had a past history of measles infection at the age of 3 years. Neurological examination revealed severe cognitive decline, positive SNOUT reflex and positive PALMOMENTAL reflex. All hematological and biochemical examination were normal.

The electro-encephalogram (EEG) analysis revealed stereotyped polymorphic discharge seen periodically, suggestive of SSPE.

The CSF study was carried out, which also revealed a highly positive CSF measles IgM antibody titer.

Table 1: Cranial nerves analysis

Cranial nerves	Characteristics
I	Nil special
II	Bilateral PEARL, No visual defects
III, IV, VI	No ptosis, No restriction of EOM
V	No jaw deviation
VII	No facial palsy
VIII	Nil special
IX, X	GAG reflex +, palate: central
XI	Nil special
XII	No tongue deviation

Table 2: Motor Reflexes Analysis

Motor reflex	Characteristics
Tone	Cogwheel rigidity + in all 4 limbs
Power	Moving all 4 limbs, > 3/5 in all 4 limbs
Reflex	Choreoathetoid movements +

In view of recurrent jerky movements, falls and postural defects, he was initially treated with Sodium Valproate, Benzodiazepine and Antispasmodics. Then the child was put on IVIG for 2 day at a dose of 2 g/kg. However, the child could not tolerate the drug and exhibited Cyclical Vomiting. In view of poor financial status, the parents owed for DAMA (Discharge Against Medical Advise). He was discharged with Cap. Ribavirin 200 mg TDS for 2 weeks, Tab. Sodium Valproate 200 mg 2-0-2 for 2 weeks, Tab. Risperidone 0.5 mg BD for 2 weeks, Laxatives and Proton pump inhibitors. The child was followed up in OPD basis and had 4-5 OPD visits. However, the diseases progressed and exhibited worsening akinesis.

After 6 months of first episode, he was readmitted. This time the child was totally bed ridden, not interacting with his mother, mutism +, persistent upward gaze +, choreiform movements +, perioral dyskinesias +, bilateral tremors + and had cog wheel rigidity in limbs. Now the child was given IVIG 1 g/kg over 4 days. Child had 1 episode of seizure during hospitalization for which oral Valproate was changed to IV form. Since his oral intake was poor, Ryles tube feed was initiated. His parents were counseled regarding the progressive nature of illness. In view of multiple numbers of oral lesions, Ribavirin was stopped and treated with Inj. Betamethasone for 2 days. Upon discharge he was given Syr. Valproate, Oral Steroid (Prednisolone) tapering therapy, THP 2 mg BD, PPI and Laxatives. Parents were advised to maintain RT feed of 150 mL, once in 3 hours. After 1 week, he succumbed to his illness at his residence.

3. Discussion

SSPE is a long standing/ chronic progressive neurodegenerative disease caused by latent measles virus. This might occur 2 – 10 years following primary infection. Acute measles encephalitis, measles inclusion body encephalitis are some of the neurological complications following a measles infection.[5] Myoclonus seizure (generalized/ focal), cognitive impairment, EPS, visual defects are common manifestations of SSPE. Early rare symptoms of SSPE includes transcortical sensory aphasia, hemiparesis, cortical blindness, optic neuritis, aphasia, choreoathetoid movements.[6]

There are some anecdotal case reports which describes a relation between Pseudotumour Cerebri and SSPE. Pseudotumour Cerebri is a condition characterized by elevation in ICP without an underlying lesion or any other cause. However, the exact mechanism for the elevation in ICP in SSPE is unclear; it might be due to acute inflammatory reaction. A case study reported that most patients with SSPE who had an elevated ICP remained asymptomatic.[7–9] In the present case, the child had a history of fatigueness, mild head ache, aggressiveness/ anger, myoclonus jerks, cognitive and intellectual defects. Also, on examination it was understood that the child also had partial rise in ICP, with marked papilloedema without any intracranial mass/ lesion. Here, the child had a presence of highly positive measles titer in the CSF, Electro-encephalogram (EEG) analysis revealed stereotyped polymorphic discharge seen periodically, suggestive of SSPE.[10] The presenting symptoms of SSPE are often confusing. SSPE might mimic acute encephalitis.[11]

4. Conclusion

Our case paves a path to uplift the need coverage of measles vaccination. The vaccination to measles remains at base and 50% of death occurs, associated with deaths in India. Our case study highlights that SSPE can be presented as acute encephalitis at the primary sight. Hence it is important to rule out the differential diagnosis of SSPE by taking into consideration about neuroimging, CSF study, anti- measles antibody.

5. Conflicts of Interest

None.

6. Source of Funding

None.

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