CASE REPORT

Subacute Sclerosing Panencephalitis (SSPE)
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Introduction

SSPE is a rare chronic, progressive demyelinating disease of the CNS associated with a chronic non permissive infection of brain tissue with measles virus. The frequency has been estimated at 1 in 1,00,000 to 5,00,000 measles cases.

Most patients give a history of primary measles infection at an early age (2 years), which is followed by a latent interval of 6-8 years by the development of a progressive neurologic disorder. Some 85% of patients are between 5 and 15 years old at diagnosis.

Key Words: Subacute Sclerosing Panencephalitis, neurologic disorder.

Case report

A 14-years-old male presented with 4 months history of

1) Involuntary jerky movements of the whole body involving all four limbs.

2) History of poor school performance, inappropriate laughing, and altered behaviour was noticed by parents.

3) No history of fever, headache, seizures

4) He was immunised but details not known.

5) Not a known diabetic and seizure disorder. No history of head injury.

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6) Not a known diabetic and seizure disorder. No history of head injury.
   No family history of movement disorders.

7) History of measles infection in his one year of age.

   On examination: He was conscious, not obeying commands, no neurocutaneous markers.

**CNS Examination:**

Higher functions:
Right handedness, memory impaired, attention decreased. There is no obvious cranial nerve involvement.

Ophthalmic examination was normal.
Sensory system could not be examined.
Motor system examination: Increased muscle tone in all four limbs, increased deep tendon reflexes with plantar extensor response.

   Involuntary movements:
Generalised myoclonus seen.

Other systemic examination - Normal.
**Investigations:**

Complete blood counts, renal function and Liver function tests were within normal limits.

CT Brain- Normal

MRI brain- Normal study except pansinusitis.

EEG- Semi periodic pattern (36, 44, 58, 64 sec) with background of slow waves.

CSF analysis shows normal sugar and protein, no significant abnormality in cytology and cell count.

CSF IgG measles antibody- 11592.0, Serum IgG measles antibody- 2291.5, CSF Total IgG- 7.5 gm/dl (0-3.4), serum Total IgG=1460 mg/dl (700-1600),

CSF/Serum measles antibody quotient =4.8 (CSF/Serum quotient- Normal =< 1.3, Positive = >1.5). Patient was diagnosed to have Subacute Sclerosing Panencephalitis. Patient was treated with T.Sodium valproate and T.Clobazam to control myoclonus.

**Discussion**

SSPE initially manifested with poor school performance and mood and personality changes.

As the disease progresses, progressive intellectual deterioration, focal and/or generalized seizures, myoclonus, ataxia and visual disturbances develops. In the late stage of illness, patients are unresponsive, quadriparietic, and spastic, with hyperactive tendon reflexes and extensor plantar responses.

MRI is often normal. EEG may initially shows nonspecific slowing but with disease progression pts develop a characteristic periodic pattern with bursts of high voltage, sharp, slow waves every 3-8 secs followed by a periods of attenuated background. CSF Antimeasles antibody levels are invariably elevated.

No definite therapy is available.

Treatment with isoprinosine (100mg/kg per day), alone or with intrathecal or intraventricular alpha interferon has been reported to prolong the survival, but has never been subjected to a controlled clinical trial.
## Reference

1) William J. Moss, Measles (Rubeola), Harrison’s principles of Internal Medicine, 18th edition.


