

Subacute sclerosing panencephalitis - A fatal complication of measles

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Abstract

Subacute Sclerosing panencephalitis (SSPE), also known as Dawson's disease or measles encephalitis, is an extremely rare and fatal subacute infection caused by a persistent defective measles virus (MV). It mostly develops 7-10 years after initial MV infection and is fatal within 3 years from onset of symptoms.¹

Measles virus infection before 2 years of age has 16 times higher risk of developing SSPE as compared to infection occurring after five years of age.²

SSPE is a progressive, degenerative disease of the central nervous system that occurs several years after measles infection.

Saha et al reported an annual incidence of 21 per million population in India³ as compared to 2.4 per million population in the middle east.^{4,5}

By this case report, we aim to stress on the certitude that the only definitive prevention against SSPE is timely measles vaccination.

Keywords: SSPE, Measles, Vaccination.

Case Report

Presenting the case of a 17-year-old female patient who was admitted to our intensive care unit (ICU) with disorientation, myoclonic jerks and rigidity. The Glasgow Coma Scale (GCS) of the patient was 6 at the time of admission. Mechanical ventilation and anticonvulsant drug treatment was initiated on arrival.

The history revealed that the patient had started having mental and behavioral changes, which included forgetfulness, irritability and lethargy along with myoclonic jerks since 2 years prior to the present episode. The patient gradually developed weakness in her lower limbs and subsequently lost the ability to walk.

On taking history of childhood illnesses, it was ascertained that the patient had suffered from a severe bout of measles at the age of 5 years, which had complicated into pneumonia leading to hospital admission for 10 days. Immunization for measles had not been administered during infancy.

The initial hematological examination revealed lymphocytosis and biochemical evaluation displayed mild hyponatremia with sodium levels of 130 mEq/L and serum creatinine of 1.8 mg/dl. Arterial blood gas analysis revealed respiratory acidosis with a Ph of 7.23, PCO₂ of 55mmHg and HCO₃ of 22 mEq/L.

Expert opinion by neurologist was sought and a Magnetic Resonance Imaging (MRI) and Electro Encephalogram (EEG) were advised.

The EEG showed periodic bilateral high voltage bursts every 5-10 seconds with background suppression

accompanied with stereotyped periodic complexes. Periodic complexes consisting of bilaterally symmetrical, synchronous bursts of polyphasic, stereotyped delta waves characterized the EEG picture.

MRI could not be performed as the patient remained in a highly critical state to be shifted from the ICU. Ophthalmoscopic examination showed peripheral chorioretinitis in both eyes. Cerebro Spinal Fluid (CSF) globulin levels were 23% of the total CSF protein. A provisional diagnosis of SSPE was made based on history, examination and investigations.

The treatment was initiated with intravenous methylprednisolone 1000mg/day. Other treatment modalities include Interferon alpha, Interferon beta, Isoprinosine and Ribavirin. However these were not available at our center.

The patient remained in the Intensive Care Unit (ICU) in a critical state for the next 6 days. Eventually decerebrate rigidity appeared, autonomic instability worsened and even after the best efforts of the clinicians, she succumbed to her downward spiraling illness.

Discussion

SSPE is progressive neurological disorder of childhood and early adolescence. Most patients of SSPE have early onset of measles (< 2 years of age) and then with a latent period of 7-10 years develop neurological disturbances.

Table 1: Diagnostic criteria of SSPE

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| Definitive: criteria 5 with three more criteria; probable: three of the five criteria. | |
| 1. Clinical | Progressive, subacute mental deterioration with typical signs like myoclonus |
| 2. Electroencephalogram (EEG) | Periodic, stereotyped, high voltage discharges |
| 3. Cerebrospinal fluid | Raised gammaglobulin or oligoclonal pattern |
| 4. Measles antibodies | Raised titre in serum ($\geq 1:256$) and/or cerebrospinal fluid ($\geq 1:4$) |
| 5. Brain biopsy | Suggestive of panencephalitis |

Three or more out of the five diagnostic criteria should be fulfilled prior to establishing a diagnosis of SSPE.⁶ Amongst the above-mentioned criteria, our patient exhibited 3 out of 5 points namely, clinical picture, EEG and positive CSF picture. On the basis of this, a diagnosis of SSPE was established. Post mortem examination was refused by the parents of the patient therefore brain biopsy could not be performed.

The initial changes of SSPE include mild behavior changes and intellectual deterioration which later progresses to motor weakness and myoclonic jerks.

In advanced stages of SSPE there is progressive decline in sensorium, decerebrate and decorticate rigidity appear, breathing becomes irregular. At this stage patients usually die of hyperpyrexia, cardiovascular collapse and hypothalamic instability.⁷ Our patient also experienced a similar pattern of disease progression.

“Prevention is better than cure”- this aptly summarizes the management of SSPE. As the role of timely measles vaccination overrules all other treatment strategies.

The treatment modalities for SSPE are not curative and are mainly used to sustain life. Relapse is frequent even after good initial results. Combination of intraventricular Interferon alpha and oral Isoprinosine is the best effective treatment available.⁸

Santoshkumar and Radhakrishnan⁹ reported a case of spontaneous recovery after SSPE in a woman after 17 years of progressive neurological deterioration. She experienced slow recovery over a period of 7 years to the extent of becoming ambulatory and performing her daily activities.

Many factors have been associated with spontaneous recovery, which include the age of onset of SSPE less than 12 years, disappearance of periodic complexes on EEG, rising measles antibody titers in the cerebrospinal fluid. Our patient did not experience any such remission symptoms and her neurological decline was steady and fulminant.

Spontaneous remission may occur at any stage in the course of the disease for a variable period of time before eventual relapse occurs.

Whitson J et al¹ in their case report also described a fatal case of SSPE of a 23 year old male patient who

presented with confusion, personality changes, myoclonic jerks and later progressed on fulminant encephalitis and flaccid paralysis.

This case is similar to ours with the regards to the rapid neurological deterioration and fulminant course of the disease. However, the onset of SSPE was 20 years after the first attack of measles contrary to our case where symptoms started 10 years following childhood measles.

Another rapidly progressing case of SSPE has been described by Lackmann et al¹⁰ in a 2 year old child who acquired primary measles virus infection at the age of 18 months following which she developed SSPE 5 months later and died within 6 months.

Overall, the present case of SSPE clearly highlights the fact although this fatal neurological disorder forms its place as a diagnosis of exclusion, it is still an entity encountered in the developing countries of the world as a result of inadequate coverage of measles vaccination.

Conclusion

SSPE is a slowly evolving disease caused by aberrant measles virus, which is still common in the developing countries of the world. The most important limiting factor in the treatment of the disease is the failure to recognize its early manifestations. Most present day clinicians and intensivists may not be aware of the clinical presentation of SSPE. It was therefore considered imperative to share this rare case presented in our ICU. Preventive strategy in the form of measles vaccination is a simple yet effective tool to prevent this disastrous complication.

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