Subacute Sclerosing Panencephalitis (SSPE)- A Case Report

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I. Introduction

Anaesthesiologists come across a wide spectrum of cases which are critical and have a diagnostic dilemma. Rare cases are referred to ICU management by Intensivists/Anaesthesiologist for symptoms and features for life support makes the treatment and outcome of such rare cases a challenge for the ICU team. Here also we are reporting a case of Subacute Sclerosing Panencephalitis (SSPE) which was a diagnostic challenge with a challengable management. SSPE is a chronic form of progressive brain inflammation caused by mutated measles virus. In developing countries where immunization is still poorly practised health-care. The incidence of SSPE is as high as 1 in 609¹ cases and often has a fatal outcome. Incidence is high in Asia and Middle East.

II. Pathogenesis

A large number of Nucleocapsids are produced in Neurons and glial cells. The viral genes that encode “Envelope Proteins” have “Restricted Expressions”; so infectious particles like M-protein are not produced and virus survives for a longer period without evoking the immune response. Eventually later in life leading to Subacute Sclerosing Panencephalitis (SSPE).

III. Case- Report

18 year old boy was referred to ICU with signs of respiratory failure; myoclonic jerks from emergency department seen by Neurologist.

On evaluation- Patient was in respiratory failure; very mild gap reflex and GCS < 8 and responding only on painful stimuli.

Vitals- BP-110/70mmHg; PR-120/min; RR-30/min; Spo₂- 86% on room air; ECG- Normal rate, rhythm; Temperature-100° F.

Considering the respiratory failure; poor GCS and suspected aspiration, patient was put on mechanical ventilation after securing the airway.

Treatment:-
- Intravenous Levicetram with Midazolam for convulsion
- Antibiotics
- H₂ receptor blockers
- Intravenous fluids

All investigations sent and chest X-ray done

ABG analysis showed
- pH- 7.2
- P O₂- 88
- P CO₂- 55
- HCO₃⁻- 24

Patient was treated symptomatically for two days and history revealed- Generalised frequent convulsive attacks for last six months and was being treated with sodium valporate since then but for last one week had slurring of speech; difficulty in swallowing and deterioration in the level of consciousness.

Considering the above history- CSF examinations were sent and were found normal. GCS was deteriorating hence after improvement of general condition like control of respiratory infection; fever and laboratory investigations- patient was transported to MRI suite for scan and EEG to be done.

Once again IgE:IgM was also sent and was found normal.

On evaluation of EEG and MRI findings and discussion with Neurologist we suspected viral infection?

A subsequent thorough history from parents revealed the history of measles at the age of 5 years; upon this CSF for measles Antibody titer was sent and was found to be 212.33U/ml (Normal- 8 U/ml).
**MRI findings-**

- Hyperintensities in Ventral Pons
- Bilateral Cerebellar Peduncles

**EEG findings-**

Generalised, periodic, stereotyped high amplitude sharp and slow wave discharges lasting 1-2 seconds occurring every 5-7 seconds

**IV. Discussion**

SSPE is a diagnosis of high clinical suspicion; clinical assessment; EEG findings and high Anti-measles Antibody titre in CSF and serum.

MRI helps in raising a doubt but not in diagnosis nor outcome.

In SSPE normal changes seen in MRI are the changes seen in cerebral cortex; subcortical and periventricular white matter.

Changes in corpus callosum; thalamus and brainstem are very rare. Progressive atrophy starts in occipital region which may be missed during early scan and may not correlate with the clinical stages. In our patient slow progressive nature of onset; convulsion; EEG pattern and presence of high Anti-measles Antibody titer confirmed the diagnosis. Yilmaz et al. have also reported two patients with brainstem involvement and only one patient with pontine involvement on MRI scan which is a very rare presentation and was similar to our patients’ MRI findings.

**V. Conclusion**

Proper clinical history taking is a must and plays pivotal role in management of such rare cases. High Index of suspicion among Anaesthesiologists in Intensive Care Unit (ICU); confirmed with measles Antibody titer and MRI scan helps in clinching the diagnosis.

SSPE is a rare entity and commonly involves cerebral cortex but in our case report involvement of ventral pons and cerebellar peduncles are rarest MRI findings of this rare disease - Hence this case report.

**Differential diagnosis**

- Progressive Myoclonic Epilepsy
- Juvenile Myoclonic Epilepsy
- Multiple Sclerosis

**References**