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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 Intensivits/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 609[1] 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; Spo_2- 86% on room air; ECG- Normal rate, rhythm; Temperature-100^0 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
- H_2- receptor blockers
- Intravenous fluids

All investigations sent and chest X-ray done

ABG analysis showed

- pH- 7.2
- Po_2- 88
- pCo_2-55
- HCo_3- 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.

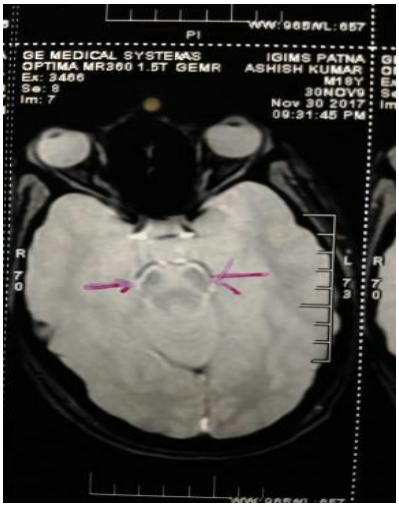
Considering the above history- CSF examinations were sent and were found normal.GCS was deteriorating hence after improvement of general condition like control of respiratory infection; fever and laboratory investigations- patient was transported to MRI suite for scan and EEG to be done.

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?

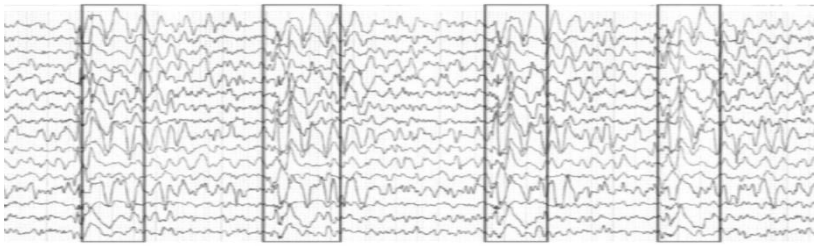
A subsequent thorough history from parents revealed the history of measles at the age of 5 years; upon this CSF for measles Antibody titer was sent and was found to be 212.33U/ml (Normal- 8 U/ml).

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[2].

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[3] 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
- Multilpe Sclerosis

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