



SUBACUTE SCLEROSING PANENCEPHALITIS IN A YOUNG FEMALE:CASE REPORT AND LITERATURE REVIEW

Neurology

Prashant Kumar Thakur*

Senior Resident, Department of Neurology, AIIMS Patna. *Corresponding Author

Anand Kumar Rai Assistant Professor, Department of Neurology, AIIMS Patna.

ABSTRACT

Subacute Sclerosing Panencephalitis(SSPE) is a degenerative disease resulting from persistent infection of brain by a mutated variant of measles virus. We are reporting case of a 12 year old girl who had history of myoclonic jerks and gait abnormalities since last three years followed by language impairment since last six months. Based on clinical presentation, raised measles antibody in serum and classic Electroencephalography (EEG) findings, diagnosis of SSPE was made. She was kept on antiepileptics, steroids and antivirals. Her symptoms improved partially. She had history of measles at four years of age. As SSPE ends in debility, coma and death, clinicians must be aware of this entity, especially in developing countries and awareness about vaccination programmes should be created among general population.

KEYWORDS

Subacute Sclerosing Panencephalitis, SSPE, Measles, Vaccination

INTRODUCTION

Subacute Sclerosing Panencephalitis is a slow viral infection, still frequently found in developing nations like Pakistan and India. Better immunization has resulted in its decline in developed countries. (1,2) Incidence of SSPE varies globally, from around 0.2 to 40 cases per million population per year, considering geographical variation, with a decrease of 82–96% in nations that have attained better vaccination coverage, especially from 2000 to 2016. (3) Rural background, overcrowding, higher birth order and measles infection at a younger age (less than 5 years) are important risk factors for SSPE.

Case Report

A 12 year old girl presented with history of jerky movement of limbs and trunks since last 3 years. She had history of progressively worsening gait impairment since last 3 years and language impairment since last 6 months. She had history of measles at 4 years of age and was never vaccinated properly. Clinical examination revealed myoclonic jerks involving limbs and trunk associated with some choreiform movements in left lower limbs.

Speech output was reduced. Routine blood investigations and serum Thyroid Stimulating Hormone (TSH) were normal. Cerebrospinal fluid (CSF) was acellular with normal sugar and marginally raised protein. Magnetic Resonance Imaging (MRI) of brain too came normal. EEG (Figure.3) revealed quasiperiodic sharp and slow wave discharges. Previous EEG (Figure.1) had background slowing with sharp and slow wave discharges. We tested her for serum measles IgG antibody. Antibody level was significantly high. As per Dyken's criteria, she was diagnosed with SSPE and kept on valproic acid, levetiracetam, steroids, amantadine.

Patients family could not afford expensive drugs. There was partial improvement in the frequency of jerks and general condition over next one month. We discharged her in a stable condition after explaining the prognosis of SSPE.

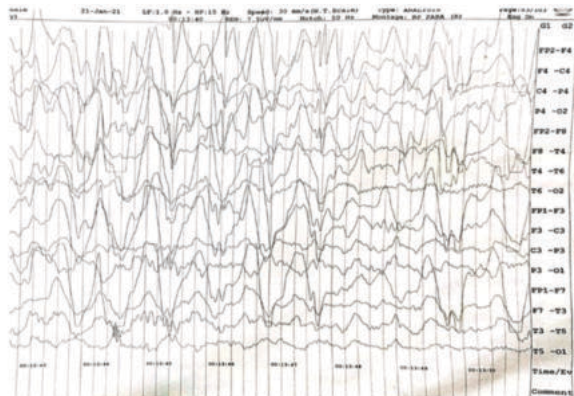


Figure 1. EEG Of year 2021

Test Name	Results	Units
MEASLES (RUBEOLA) ANTIBODY, IgG, SERUM (CLIA)	>300	AU/mL

Interpretation	
RESULT IN AU/mL	REMARKS
<13.5	Negative
≥13.5-<16.5	Equivocal
≥16.5	Positive

Figure 2. Positive serum measles IgG antibody



Figure 3. EEG Of September 2023

DISCUSSION

Latent period of SSPE is 6-8 years conventionally. (4,5). Our patient too had a latent period of 8 years. Evidences reveal that mutations which change viral envelope glycoproteins, especially the fusion (F) protein, are causing neuro-virulence. (6) Studies substantiate that SSPE patients have poor cellular immune response and high humoral immune response, which would act as a hurdle in viral elimination. (7) Amantadine, an anti RNA (Ribonucleic Acid) agent, which inhibits viral replication and hence maturation, has been tried in past successfully. (8) Another study showed that amantadine had satisfactory effect on SSPE cases, though less than interferon alpha and Isoprinosine. (9) Partial improvement in our case too substantiates the effect of amantadine on natural course of SSPE. It can be helpful for patients of poor socioeconomic strata.

CONCLUSION

With this case report we want to emphasize the need of effective implementation of vaccination programmes in developing nations, so that incurable diseases like SSPE can be eradicated. Public awareness about vaccination programmes and timely intervention in case of SSPE can help reduce morbidity and mortality.

REFERENCES

1. Garg RK, Mahadevan A, Malhotra HS, Rizvi I, Kumar N, Uniyal R. Subacute sclerosing panencephalitis. Rev Med Virol 2019;29:e2058.

2. World Health Organization. Progress towards regional measles elimination – Worldwide, 2000–2018. Reform of the International Coordinating Group for vaccine provision: A new framework for coordination, accountability and transparency. *Wkly Epidemiol Rec* 2019;94:581-600. □
3. Alexander, J.P.; Patel, M.; Goodson, J.; Alexander, J. Progress Toward Regional Measles Elimination—Worldwide, 2000–2019. *MMWR Morb. Mortal. Wkly. Rep.* 2020, 69, 1700–1705. [CrossRef].
4. Saurabk K, Singh VK, Pathak A, et al. Subacute sclerosing pan encephalities: An update. *J Clin Sci Res* 2021;10:35-42.
5. Samia P, Oyieke K, Tunje D, et al. Options in the treatment of subacute sclerosing panencephalitis: Implications for low resource areas. *Curr Treat Options Neurol* 2022;24:99-110.
6. Sato Y, et al. Cell-to-cell measles virus spread between human neurons is dependent on hemagglutinin and hyperfusogenic fusion protein. *J Virol.* 2018;92(6).
7. Gutierrez J, Issacson RS, Koppel BS. Subacute sclerosing panencephalitis: an update. *Dev Med Child Neurol.* 2010;52(10):901–7.
8. Robertson WC Jr, Clark DB, Markesbery WR. Review of 38 cases of subacute sclerosing panencephalitis: Effect of amantadine on the natural course of the disease. *Ann Neurol* 1980;8:422-5.
9. Nasirian A, Ashraf MR, Nasrabady SE. Use of A-Interferon, Amantadin and Isoprinosine in Subacute Sclerosing Panencephalitis (Sspe): Comparing the Effectiveness. *Iran J Child Neurol.* 2008;2:27–32.