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A Study of Subacute Sclerosing Panencephalitis (SSPE): Clinical and Investigation Profile in a Tertiary Care Hospital

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Abstract **Original Research Article**

Subacute Sclerosing Panencephalitis (SSPE) is a rare and slowly progressive neurodegenerative disorder caused by persistence of mutant measles virus in the central nervous system. Myoclonic jerk is the predominant feature. ***Objective:*** To identify common clinical, demographic and investigation findings of diagnosed cases of Subacute Sclerosing Panencephalitis. ***Material and Methods:*** This is a Cross sectional observational study was done in Department of Paediatric Neuroscience, Bangladesh Shishu Hospital and Institute during the period from September 2024 to February 2025. Total 10 cases were included after diagnosis of SSPE. The diagnosis was based on a combination of clinical features and investigation findings. When clinical and investigation findings supported the diagnosis of SSPE then the case was included in this study. All data were recorded on a previously prepared standard data collection form ***Results:*** 90% cases were in 5-15 years age group. 80% cases were male and 20% were female. Among 10 patients, 60% patients had past history of measles infection and 80% patients were vaccinated against measles. Presenting symptoms were behavioral change (40%), cognitive decline (70%), decreased school performance (50%) and motor regression (100%), myoclonic jerks (80%), fall to ground (20%), altered speech (60%), vision loss (20%), spasticity (80%) and clonus (30%). Measles specific IgG in CSF was positive in (80%) cases and positive serum measles antibody was in (100%) cases. Patients had an abnormal EEG finding which includes periodic complexes (70%), generalized spike (20%) or slow wave (10%). Total 80% patients had abnormal findings on neuroimaging. ***Conclusion:*** Children presenting with developmental regression, myoclonic jerks, characteristics EEG changes, measles specific antibody in CSF and serum may help in the diagnosis of SSPE.

Keywords: SSPE, Myoclonic jerks, Measles antibody, EEG.

INTRODUCTION

Subacute Sclerosing Panencephalitis (SSPE) is a rare and slowly progressive neurodegenerative disorder caused by measles virus. It is a subacute inflammation of brain which is caused by persistence of mutant measles virus in the central nervous system [1, 2]. It leads to degenerative changes in the brain. As a result, progressive regression of acquired milestone of development, behavioral changes, myoclonic jerks, visual loss, alteration of speech, and death may occur [3]. Its prognosis is very poor. Patients may die within 1-3 years of disease manifestation [4].

The incidence of SSPE in worldwide is 1 per million [2,3]. SSPE is rare in developed country due to widespread immunization against measles [1-3]. But it is high in developing country due to poor immunity to measles and poor vaccination.[2] According to WHO global incidence of SSPE is 4-11 per 100000 measles cases [4]. It is a disease of childhood and early adolescence [3].

The diagnosis of SSPE is usually based on clinical features, characteristics EEG findings, MRI findings, anti-measles antibody in CSF and serum [5]. Diagnosis of SPE is also done by dykan criteria including- clinical features of decreasing cognition and myoclonus, periodic discharge on EEG, raised CSF Ig level, raised titers of anti-measles antibody and brain biopsy. Confirmed diagnosis of SSPE is done with 3 or more criteria based on dycan criteria [4]. EEG findings in SSPE include periodic discharge which is characterized by synchronized bursts of sharp-slow wave

[5]. MRI may normal initially but it become abnormal at later stages. Advanced stages of SSPE shows cortical atropy in MRI [5].

Many treatment protocols like Isoprinosine, Interferons have been tried but no effective treatment is available till now for this disease [6]. This study was done to observe the clinical findings and the investigation findings of SSPE patients.

MATERIAL AND METHODS

This study was carried out in the Department of Paediatric Neuroscience, Bangladesh Shishu Hospital and Institute during the period of September 2024 to February 2025. During this period all patients compatible with the clinical diagnosis of SSPE were included. Total 10 patients were included. The diagnosis was based on history that included age at presentation, past history of measles, history of measles vaccination, developmental regression, behavioral change, cognitive decline, decreased school performance, myoclonic jerks, falls to ground, alteration of speech, vision loss. Thorough clinical examinations were done. The diagnosis was based on a combination of clinical features and investigations like measles specific antibody titers in CSF and serum, EEG and neuroimaging findings. When clinical and investigation findings supported the diagnosis of SSPE then the case was included in this study. All findings were recorded on a previously prepared standard data collection form.

RESULTS

Table I shows, 90% of patients was in 5-15 years age group and 80% were male and 20% were female among all the cases. Among 10 patients, 60% patients had history of measles infection and 80% patients were vaccinated against measles (Table-II). The presenting symptoms were behavioral change (40%), cognitive decline (70%), decreased school performance (50%) and motor regression (100%), myoclonic jerks (80%), fall to ground (20%), altered speech (60%), vision loss (20%), spasticity (80%), clonus (30%) (Table-II). Table III shows, measles specific IgG in CSF was positive in (80%) cases and positive serum measles antibody was in (100%) cases. Table III also shows, all patients had an abnormal EEG finding which includes periodic discharge (70%), generalized spike (20%) or slow wave (10%) and this table also shows, 80% patients had abnormal findings in neuroimaging.

Table-I: Age and Gender distribution among the study population (n=10)

Variable	Number of Patient	Percentage (%)
Age in years		
Below 5 years of age	1	10.0
5-15 years of age	9	90.0
Gender		
Male	8	80.0
Female	2	20.0

Table-II: History and Clinical presentation of study population (N=10)

History and Clinical Presentation	Number of Patient	Percentage (%)
History of Measles	6	60.0
History of Measles vaccination	8	80.0
Behavioral change	4	40.0
Cognitive decline	7	70.0
Decreased school performance	5	50.0
Motor regression	10	100.0
Myoclonic jerks	8	80.0
Falls to ground	2	20.0
Altered Speech	6	60.0
Vision loss	2	20.0
Spasticity	8	80.0
Clonus	3	30.0

Table-III: Measles antibody in CSF and serum, EEG and Neuroimaging findings

Investigation Findings	Number of Patient	Percentage (%)
EEG		
Periodic discharge	7	70.0
Generalized spike	2	20.0
Slow wave	1	10.0
Measles antibody		
IgG positive in CSF	8	80.0

IgG positive in Serum	10	100.0
Neuroimaging		
Cortical atrophy	5	50.0
White matter hyperintense signal	3	30.0

DISCUSSION

This study reported the common clinical and investigation findings of SSPE in a tertiary care hospital. Usually the age of presentation worldwide is between 5 and 15 years [7]. In this study, 90% cases were within age group of 5-15 years, which is similar to Akram *et al* and Saha *et al* [7,8]. SSPE is more common in boys than girls [3]. In this study 80% cases were male which is similar to the report of Garg and Cruzeiro *et al* [3,9].

In this study 60% cases had past history of measles infection. SSPE may also develop in vaccinated children. Several reports showed that it occurs due to malnutrition, improper vaccine coverage, poor quality, subclinical measles infection prior to vaccination, or vaccine failure [10].

In our study 80% of the patients were vaccinated who developed SSPE. Akram *et al* also reported that 86% cases had history of measles vaccination who developed SSPE later [7]. Their study is similar to our study.

Developmental milestones were normal in all patients before illness which deteriorated during the illness. In this study, cognitive decline was present in 70% cases and 100% cases presented with motor regression. Akram *et al* reported the cognitive decline in 86% cases and motor regression in 100%, which is similar to this study [7]. Myoclonic jerks were present in 80% cases of our study. Akram *et al* showed that 76% cases had myoclonic jerks [7]. Other types of seizures also reported by Garg *et al* in 35% cases besides myoclonic seizure [11]. Our study also reported other types of seizure in 30% cases and behavioral change in 40% cases. Visual loss of variable type and severity has also been reported by different observers [3,7]. This study also showed visual loss in 20% cases. The clinical characteristics of this study is also similar to Alam *et al* and Saha *et al* [12,8].

In this study measles specific IgG antibody in CSF was positive in 80% cases whereas the antibody in serum was positive in 100% cases. Confirmed diagnosis of SSPE was done by raised titers of measles specific antibodies in CSF and serum [8]. Akram *et al* and Alam *et al* reported anti measles antibody in CSF was positive in 100% cases [7,12].

All cases showed abnormal EEG. Periodic discharge was observed in 70% cases and other EEG changes documented generalized spikes or slow waves. Ekmekci *et al* and Praveen kumar *et al* also showed similar findings like our study [13,14].

The most striking findings of the neuroimaging were cortical atrophy and white matter hyper intense signals. Other study also reported same findings similar to our study [8,15].

CONCLUSION

Children presenting with regression of acquired milestones of development along with myoclonic jerks, characteristics EEG changes, CSF and serum antibody to measles virus may help in the diagnosis of SSPE.

Limitation: Small sample size, single centre study and short duration of study period were the limitation of this study.

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