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Original Research Article

Initial presentation and clinico-radiological profile of subacute sclerosing panencephalitis in an Eastern Indian tertiary care hospital

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Abstract

Introduction: Subacute sclerosing panencephalitis (SSPE) is a rare but fatal progressive neurodegenerative disorder caused by persistent measles virus infection in the brain. Although its incidence has declined in countries with widespread measles vaccination, SSPE continues to pose a significant challenge in developing regions. Increasing reports of atypical clinical presentations further complicate early diagnosis, emphasizing the need for heightened clinical awareness.

Aim and Objectives: To find out Initial presentation and clinical and neuro-imaging findings of patients with Subacute sclerosing panencephalitis.

Materials and Methods: This cross-sectional study was conducted at Neurology ward of Calcutta National Medical College and Hospital, a tertiary care hospital in West Bengal. Between the period June 2022 to May 2024, total 23 patients of SSPE were evaluated in neurology ward. Diagnosis was based on Modified Dyken's Criteria. MRI Brain and Ophthalmological evaluation were done for all patients. Special emphasis was given to initial presenting symptoms.

Results: Total 23 patients were studied. Mean age of our patients were 11.13 years and male female ratio was 1.875:1. More than half (69.5%) patients were from poor socio-economic status. Among 23 patients of SSPE 56.5% were vaccinated and 60 % had history of measles infection. On a thorough history it was noted that initial presenting features were myoclonus (43.5%), cognitive decline /poor school performance (52.2%), visual disturbance (17.4%), seizure (17.4%), behavioural abnormality (8.7%), Unilateral tremor (4%), and gait disturbance (8.7%). While on first examination after admission clinical signs were myoclonus (82.6%), cognitive decline (82.6%), visual disturbance (26%), dysarthria (21.7%), seizure (17.4%), behavioural abnormality (52.2%), urinary and faecal incontinence (21.7%), Unilateral tremor (4%) and gait disturbance (8.7%). Ocular findings like optic neuritis, chorioretinitis, papilledema were present in 34.8%. Neuroimaging abnormalities in 69.6% cases included parieto-occipital white matter hyperintensity, cortical atrophy and ischaemic changes. Abnormal EEG in 91.3% cases. Measles specific IgG antibody in CSF were present in 100% of cases.

Conclusions: In developing countries like India where measles is still prevalent, high index of suspicion regarding uncommon presentations of SSPE like vision loss, seizures, behavioural disturbances, unilateral tremor and gait disturbance will help in early diagnosis of the disease.

Keywords: Subacute sclerosing panencephalitis, Measles virus, Atypical clinical presentation, Neurodegenerative disease, Myoclonus, Cognitive decline.

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1. Introduction

Subacute sclerosing panencephalitis (SSPE) is a progressive, fatal brain disease that primarily affects children and early adolescent.[1,2] Persistent infection of the brain due to inability of the patients' immune system to eliminate the measles virus leads to the disease. As a result, inflammation, cell death and gliosis of the CNS takes place.

The incidence of SSPE is 4-11 cases per 100000 cases of measles approximately.[3] If measles occurs under 1 year of age it has a prevalence of 20-per 100,000. But for cases where measles occurred after 5 years of age prevalence is 1 per 100,000 for infections.[4] In the developed countries cases of SSPE substantially decreased following measles vaccination programme. But SSPE still exists in the developing nations, with high incidence.[5] A higher incidence of SSPE has noted in boys, although primary measles infection shows no such sex disparity.[2] It is also more common in children with a lower birth order and in children living in overcrowded environments.[6]

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The clinical course of SSPE varies significantly from case to case. The symptoms, duration, and intensity vary with stages of the disease. Typically, four stages of disease are observed in SSPE. Stage I is characterized by irritable mood, forgetfulness, social withdrawal, fatigue, deterioration in school performance and speech regression. The second stage is characterized by different movement disorders like dystonia, dyskinesia, myoclonus, seizures and dementia. In the third stage extra pyramidal symptoms, rigidity, and progressive unresponsiveness ensue. The fourth stage is characterized by loss of cortical functions with signs of vegetative state. Eventually autonomic failure, and akinetic mutism also occurs. The mean survival period after onset of symptoms is usually between 1 to 3 years.[7-9]

Over the years, it has been observed that pattern of clinical presentation of SSPE has change to some extent, and there are also some changes in the laboratory features of SSPE.[1,2] Many unusual presentations have been described. These includes isolated extrapyramidal symptoms such as chorea, hemi-parkinsonism, dystonia; isolated psychiatric manifestations, and seizures etc. Occasionally a stroke-like onset has also been described. In some patients' transient plateau period or slight improvement has been observed. But classically it has a relentless progression of disease associated with high mortality.[10]

The uncommon modes of presentation of SSPE pose diagnostic difficulties. These are highlighted in this study.

2. Materials and Methods

This cross-sectional study was conducted at Neurology ward of Calcutta National Medical College and Hospital, a tertiary care hospital in West Bengal. Between the period June 2022 to May 2024, total 23 patients of SSPE were evaluated in neurology ward. Diagnosis was based on Dyken's criteria[7] stated as:

2.1. Major criteria

1. Clinical history
2. Elevated CSF measles antibody titer

2.2. Minor criteria

1. Generalized long-interval periodic complexes in the electroencephalography (EEG).
2. Elevated cerebrospinal fluid (CSF) globulin levels.
3. Histological findings in brain biopsy or Molecular diagnostic test for measles genetics detection 2 major and 1 minor criterion were required for diagnosis.

After the patients fulfilled the above criteria, they were selected and enrolled for the study. Their data has been collected. Following details were taken:

1. Preliminary data: name, age, sex of the patient. Presenting symptoms and duration of symptoms; history of measles infection if present and history of measles vaccination.
2. Presence of symptoms: if cognitive decline, behavioral change, loss of vision, seizures, and myoclonic jerks, sphincter dysfunction or any other symptoms present.

Clinical evaluations of these patients were done. Abnormalities of higher mental functions, presence or absence of long tract signs and motor or sensory deficit, visual acuity, fundus examination was looked for.

Clinical examination was followed by investigations. These included MRI Brain, EEG, cerebrospinal fluid (CSF) and serum examination for measles antibody titers.

Data were collected in structured data collection sheets.

3. Results

A total of 23 cases were included in the study as per inclusion criteria. Out of total 23 cases 15 (65%) were male and 8 (15%) females with a male: female ratio of 1.857. Age at presentation varied from 4 years to 23 years with a mean age of 13.1 years. Out of these 16 patients (69.5%) were from rural area while rest 7 patients (30.5%) were from urban area. Majority of the patients (18, 78%) were from poor socio-economic background.

In our study 8 (34.8 %) patients were vaccinated and 15 (65.2%) unvaccinated. Among vaccinated 6 (75%) were from urban and 2 (25%) from rural area. In unvaccinated 5(33%) from urban and 12(67%) from rural area. The mean age of disease onset was 14.3 years in vaccinated and 9.5 years in unvaccinated group of patients. History of measles infection were present in 14 (61%) of patients. The mean latency period in these patients were 9.7 years for vaccinated and 4.6 years for unvaccinated patients. The unvaccinated group had a rapid worsening of symptoms than the vaccinated patients. Using neurology disability index scale 12 unvaccinated and 3 vaccinated were found to have acute progression. The rest of the patients had subacute progression. The average duration of illness from the onset of first symptom to seeking medical attention was 4.8 months in the vaccinated group while it was 2.3 months in the non-vaccinated group.

In our study after taking a thorough history from the parents and relatives it was found that while some patients presented with typical clinical features while a number of patients had atypical symptoms at presentation. Myoclonus (43.5%), cognitive decline /poor school performance (52.2%), visual disturbance (17.4%), seizure (17.4%), behavioural abnormality (8.7%), Unilateral tremor (4%), and gait disturbance (8.7%) were the initial presenting symptoms.(**Table 1**)

After hospital admission on examination following findings were present.(**Table 2**)

Table 1: Initial presenting symptoms of the patient

Initial presenting symptoms	No of patients
Myoclonus	10 (43.5%)
Cognitive decline /poor school performance	12 (52.2%)
Visual disturbance	4 (17.4 %)
Seizure	4 (17.4 %)
Behavioural abnormality	2 (8.7%)
Unilateral tremor	1 (4.3 %)
Gait disturbance	2 (8.7%)

Table 2: Clinical signs on admission

Clinical signs on admission	No of patients (%)
Myoclonus	19 (82%)
Cognitive decline	19 (82%)
Visual disturbance	6 (26%)
Dysarthria	5 (21.7%)
Seizure	4 (17.4%)
Behavioural abnormality	12 (52.2%)
Urinary and faecal incontinence	5 (21.7%)
Unilateral tremor	1 (4.3%)
Gait disturbance	6 (26%)

Different ophthalmological abnormalities were also found like optic atrophy, papilledema, chorioretinitis. It was found 15 (60%) children. Rest of 8 (40%) children were normal in eye evaluation.(**Table 3**)

Table 3: Ophthalmological findings of the patients

Features	No of patients
Normal	15 **(**65.2)
Optic atrophy	7 **(**30.4)
Papilledema	2 **(**8.6)
Chorioretinitis	1 **(**4.3)

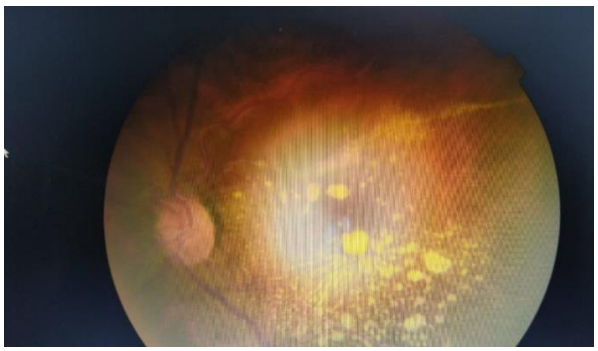

Figure 1: Chorioretinitis found in a case of SSPE

MRI of Brain was done in all patients. While 7 patients had normal MRI of Brain 16 had different abnormalities. White matter changes in form of diffuse and focal areas of hyperintensity were the most common finding. Deep grey matter hyperintensity were also present in few patients. DWI and ADC sequences were normal in all patients.(**Table 4**, and **Figure 1-3**)

Table 4: MRI Brain findings of the patients

MRI Brain findings	No of patients
Normal MRI	07(30.4 %)
White matter changes (diffuse/focal)	
Temporo-parieto-occipital	9 (39.1 %)
Patchy, periventricular	5 (21.7 %)
Thalamic/cerebellar/basal ganglia involvement	05 (21.7 %)
Cortical atrophy	04 (17.4 %)

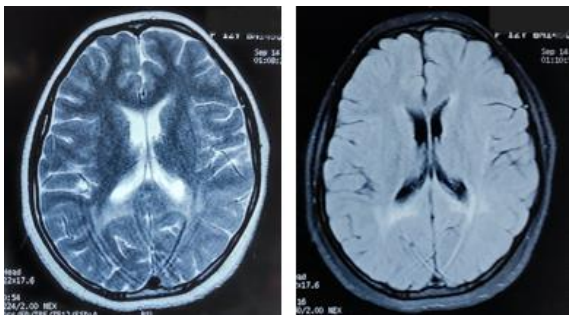

Figure 2: T2 and FLAIR image of a patient showing hyperintensity in parieto- occipital region

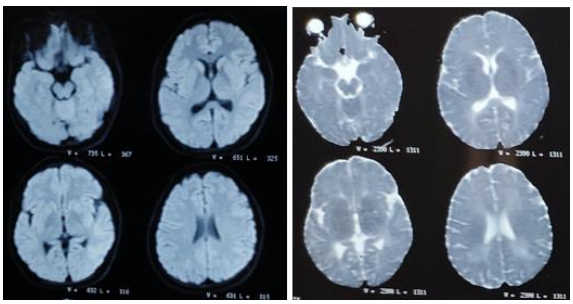

Figure 3: DWI and ADC image of a SSPE patient without any apparent changes

EEG was done in all patients. In 2 (8.7) patients EEG was normal. Rest 21 (91.3 %) patients showed abnormal periodic complexes in the EEG.(**Figure 4**)

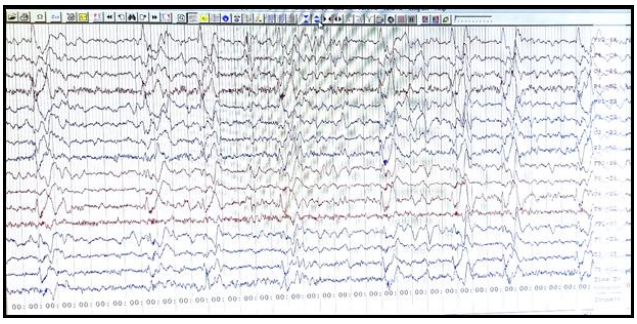

Figure 4: EEG of a patient showing both long and short interval periodic discharge

CSF measles IgG antibody titre of ≥0.5 IU/mL and a CSF/serum ratio ≥0.05 were present in all 23 patients.[17]

4. Discussion

Subacute Sclerosing Panencephalitis (SSPE) is a progressive neurodegenerative disease that attacks the central nervous

system, due to a persistent infection of measles virus in childhood or early adolescence.

With a very successful Extended Programme on Immunisation (EPI) in India prevalence of measles has decreased by almost 62 % in last few years. The efficacy of measles vaccine is usually between 90-95 %. Bust still there is an unmet need of larger vaccination coverage and also poor seroconversion in some cases leading to measles infection in many children. In some cases, these viruses persist in CNS, become mutated and later manifested as SSPE.

Worldwide average age of presentation of SSPE is between 5 to 15 years and mean age is around 9-10 years.[1,2] In our study it was 13.1 years. This may be due to increasing average of SSPE globally because of better vaccination coverage.[11,12]

In our study, unvaccinated patients had more rapid course of disease, from the onset till the diagnosis, being 2.3 months as compared to 4.8 months in the vaccinated group. This showed that the rapidity of initial progression somewhat less in vaccinated group which is also supported by other studies.[9,10]

Prevalence of measles infection is equally common in both sexes but higher incidence of SSPE (male/female ratio) found in boys in our study similar to Prashant et al.[3] Which may be due to increased risk of being intensively exposed within the home for boys as compared to girls as per Aaby et al.[4] Several other studies also suggested higher incidence of SSPE in male Saha et al (2.4 :1),[7] Bhat et al (3:1).[8]

As found in earlier studies children from lower socioeconomic status, large family size, and rural area have higher incidence, as measles virus is transmitted by respiratory secretions.[7,8] Our study also shows similar clinical picture.

Unusual symptoms like visual disturbance (4), seizures (4), unilateral tremor (1), gait disturbance (2) and behavioural changes (2) were initial presenting symptoms in total 13 patients. Though several studies[15,16] reported loss of vision as presenting symptom, age ranging was higher 20-35 than our study with age range of 13-19 years with a mean age of 16.3 years. These patients had diffuse white matter lesions in the parieto-occipital lobes. Fundus abnormality was present in total 8 patients with Optic atrophy (7), chorioretinitis (1), papilledema (2) but visual difficulty was not their initial manifestations. So cortical vision loss was an important feature in the early phases of SSPE.

Seizure was initial presentation in 4 patients. Out of 4 patients 3 were presented with generalized tonic clonic seizure (GTCS) and one with focal seizure. Most of these patients continued to experience seizures in spite of anti-seizure medications. Eventually they developed other symptoms which finally led to diagnosis of SSPE. Similar incidence of seizure also noted in other studies.[14]

Two patients presented with gait difficulties. One patient at age 14 years who had periventricular hyperintensities in T2 imaging and other at age 17 years who had cerebellar changes in imaging.

One 15 years old female presented with right sided postural tremor followed by cognitive symptoms and myoclonus. Imaging showed changes in contralateral basal ganglia region.

Characteristic EEG changes with abnormal periodic complex was present in 21 out of 23 patients. In 5 EEG both long and short interval periodic discharges found as shown in the picture above.

5. Conclusion

SSPE is a fatal neuroinflammatory disorder. In developing countries like India where measles is still prevalent, high index of suspicion regarding uncommon presentations of SSPE like vision loss, seizures, behavioural disturbances, unilateral tremor and gait disturbance will help in early diagnosis of the disease and prevent unnecessary investigations.

6. Ethical Clearance

Ethical clearance has been taken from institutional ethical committee.

7. Source of Funding

None.

8. Conflict of Interest

None.

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