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Research Article

Differences of Socio-Demographic Characteristics, Clinical Profiles and Investigational Findings between Pediatric and Adult Subacute Sclerosing Panencephalitis Patients Attended at a Referral Neurology Hospital in Bangladesh

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ABSTRACT

Background: Subacute Sclerosing Panencephalitis (SSPE) though more common in the pediatric age group, can also occur in adult patients. There may be a variation of socio-demographic, clinical, and investigation profiles in these two groups.

Objective: The purpose of the present study was to find out the differences in socio-demographic, clinical, and investigation profiles between the two groups.

Methodology: This cross-sectional study was conducted at the national institute of neurosciences and Hospital, Dhaka, Bangladesh from September 2017 to August 2020. Patients of SSPE diagnosed by Dyken's Criteria were included in the study. As per the hospital admission protocol, patients up to the age of 14 years were admitted to the pediatric neurology department while those above 14years were admitted to the adult neurology department. Details of socio-demographic characteristics, clinical profiles and investigational findings were recorded; then comparison of these parameters among the patients admitted in the pediatric and adult neurology department was performed.

Result: A total of 34 SSPE patients were recruited of which 24 cases were from the pediatric neurology department (group A) and the rest 10 patients were from the adult neurology department (group B). Male patients outnumbered females in both groups which was 18 (75.0%) cases and 7 (70.0%) cases respectively. Most of the patients were from rural areas and low socio-economic background. History of definite Measles infection was present in 41.0% in Group A and 30.0% in group B patients. The frequency of vaccination against measles was 100.0% in Group B whereas 91.7% in Group 1 patients. Fall was the commonest presenting symptom in both groups. Visual disturbance was more common among Group B patients ($p<0.05$). Antimeasles antibody was positive in all cases and EEG revealed periodic bursts in all patients in both groups.

Conclusion: The profile of SSPE patients admitted in the pediatric Neurology department is not varied from those admitted in the adult neurology department except for visual disturbance which has found to be more common in the late-onset group.

Keywords: Measles; Visual disturbance; Anti-measles antibody

INTRODUCTION

Subacute Sclerosing Panencephalitis (SSPE) though more common in the pediatric age group, can also occur in adult patients. There may be a variation of socio-demographic, clinical, and investigation profiles in these two groups. The purpose of the present study was to find out the differences in socio-demographic, clinical, and investigation profiles between the two groups. This cross-sectional study was conducted at the national institute of neurosciences and hospital, Dhaka, Bangladesh from

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September 2017 to August 2020. Patients of SSPE diagnosed by Dyken's Criteria were included in the study. As per the hospital admission protocol, patients up to the age of 14 years were admitted to the Pediatric Neurology Department while those above 14years were admitted to the adult neurology department. Details of socio-demographic characteristics, clinical profiles and investigational findings were recorded; then comparison of these parameters among the patients admitted in the pediatric and adult neurology department was performed.

Subacute Sclerosing Panencephalitis (SSPE) is a relentlessly progressive neurodegenerative disorder caused by the persistence of defective measles virus in the brain [1]. It is characterized by progressive intellectual and motor decline along with characteristic periodic myoclonic jerks ultimately progressing to a mute, bed-ridden, and incontinent state finally leading to death [2]. It is invariably fatal within 1 to 3 years of symptom onset [3] although spontaneous improvement or stabilization has been reported in a small number of patients [4].

SSPE has been reported from all parts of the world; however, it is considered a rare disease in the West and in the United States this is fewer than 10 cases per year. The trend of the incidence is also measured and it has been found that the reported frequency of SSPE in the United States is approximately one per million childhood population from 1960 to 1970. The incidence declined substantially after introduction of an effective measles vaccine though this does not decrease the rat of SSPE. The annual incidence of SSPE is still quite high but variable among developing countries. In India have reported annual incidence of 21 per million populations are suffering from SSPE which is in comparison with 2.4 per million populations in the Middle East [5].

The onset of the disease is mostly in childhood between 5 to 15 years of age, although a good number of cases have been reported to have an onset in late childhood or even adults. There may be a difference in sociodemographic, clinical, and investigation profiles between the early and late-onset groups. Although wide vaccination has led to a substantial decline in the incidence, the disease still remains to be higher in developing countries. In India the annual incidence is 21 (per million populations) while in Middle East it is 2.4 (per million populations). The clinical features are also varied in different socio-demographic group of people [6].

METHODOLOGY

This single centre, analytical cross-sectional study was conducted in the Department of Neurology and Pediatric Neurology at the national institute of neurosciences and hospital, Dhaka, Bangladesh from September 2017 to August 2020. All the referral patients were attended in this only neuroscience institute in Bangladesh. Written informed consent was taken from the parents/guardians before inclusion in the study. Patients fulfilling 3 out of the 5 criteria were included as the study population. These were progressive cognitive decline with stereotyped myoclonic jerks, generalized long-interval periodic complexes in the Electroencephalography (EEG), elevated cerebrospinal fluid globulin levels, elevated cerebrospinal fluid measles antibody titers, and typical histological findings in brain biopsy or autopsy. As per the hospital admission criteria, patients up to 14 years of age are admitted under the department of pediatric neurology and those above 14 years of age are admitted to the adult neurology department. Patients were initially evaluated clinically either by pediatric neurologists or adult neurologists which was followed by investigations including EEG, cerebrospinal fluid, and serum examination to look for measles antibody titers. Neuroimaging, wherever possible, was also carried out. Details of socio-demographic, clinical profiles, and investigations were recorded. Data were analyzed using SPSS version 16. Continuous data were presented as means and standard deviations whereas categorical data were presented as proportions. Chi-square tests were done to test differences in proportions. For continuous variables, the Student-test or Mann-Whitney U test was used to compare values between groups. $P<0.05$ was considered to be significant for all statistical tests. The study was approved by the local ethics review committee of the institute before starting the data collection [7].

RESULTS

A total of 34 SSPE patients were recruited for this study of which 24 (70.59%) cases were from the Pediatric Neurology department (Group A) and the rest 10 (29.41%) patients were from the Adult Neurology department (Group B). The number of male patients surpassed that of females in both groups, (Group A-75%, Group B-70%) (Table 1). Most of the patients were from rural areas (Group A-70.8% and Group B-70%) and low socio-economic groups (Group A-50%, Group B-60%).

Table 1: Socio-dmographic profile of studied population (n=34).

Variables	Group A (n=24)	Group B (n=10)	P value
Age on admission mean SD (years)	12.34+1.91	19.05+4.51	<0.001
Age range (years)	6-14	14.5-26	-
Sex			
Male	18 (75%)	7 (70%)	0.76
Female	6 (25%)	3 (30%)	
Habitat			
Rural	17 (70.83%)	7 (70%)	0.96
Urban	4 (16.67%)	2 (20%)	
Semi urban	3 (12.50%)	1 (10%)	
Socio-economic condition			

Lower	12 (50%)	6 (60%)	0.61
Middle	11 (45.83%)	3 (30%)	
Higher	1 (4.17%)	1 (10%)	

The commonest presenting symptom was recurrent fall in both groups. Visual disturbance turned out to be significantly more common in group B (P-value<0.05). Anti-measles antibody in CSF was present in both groups. All patients in both groups had periodic bursts in EEG. History of definite Measles infection was found in 10 (42.7%) and 3 (30.0%) of cases respectively. The frequency of vaccination against measles was 22 (91.7%) in Group A and 10 (100%) in Group B (Table 2).

Table 2: Clinical characteristics of studied population (n=34).

Variables	Group A (n=24)	Group B (n=10)	P-value
Age of onset of disease (mean ± SD) (years)	11.69+2.01	18.30+4.83	0.001
Age range (years)	5.80-13.75	13-25.90	-
Measles vaccinated	22 (91.67%)	10 (100%)	0.34
History of measles infection	10 (41.67%)	3 (30%)	0.65
Symptoms			
Intellectual deterioration	20 (83.33%)	10 (100%)	0.16
Change of personality	20 (83.33%)	8(80%)	0.81
Myoclonic jerks	23 (95.83%)	8 (80%)	0.13
Generalized tonic-clonic seizure	13 (54.17%)	3 (30%)	0.19
Focal seizure	5 (20.83%)	2 (20%)	0.57
Limb weakness	2 (8.33%)	0	0.34
Speech difficulty	16 (66.67%)	6 (60%)	0.17
Swallowing difficulty	2 (8.33%)	0	0.34
Ataxia	12 (50%)	2 (20%)	0.10
Visual disturbance	4 (16.67%)	5 (50%)	0.04
Apathy	14 (58.34%)	7 (70%)	0.52
Unresponsiveness	6 (25%)	1 (10%)	0.32
First symptoms noticed			
Fall	11 (45.84%)	5 (50%)	0.82
Myoclonic jerks/seizure	6 (25%)	3 (30%)	0.76

DISCUSSION

SSPE usually has an early onset, although some cases are reported to have a late-onset. The socio demographic, clinical, and investigation profiles might differ in the early and late-onset groups. This study was intended to find out any such difference.

In this study of 34 cases, a clear male preponderance has been noted in both (M: F 3:1) which is consistent with other studies although primary measles infection doesn't show such sex disparity. The cause of this disparity remains inconclusive although some have speculated hormonal influence as a contributor while others have implicated social circumstances to this disparity. Most of the patients in this study were from a rural area and a lower socio-economic group. investigated the risk factors of SSPE and had a similar observation. They concluded that the incidence of SSPE is higher among children from lower- socioeconomic levels, larger family sizes, and rural areas as the measles virus is transmitted by direct contact with infectious droplets or by airborne spread from an infected person. Other studies also supported the findings [8].

Surprisingly in both groups, quite a good number of patients developed SSPE despite vaccination against measles. This has been explained in several reports that the occurrence of SSPE in measles vaccinated patients may be due to subclinical measles infection before vaccination, high prevalence of malnutrition in developing countries, improper vaccine coverage, poor quality, improper storage and transport of vaccine, poor seroconversion or vaccine failure, or circulation of atypical/wild measles virus strain. Recurrent fall was the commonest presenting symptom of the patients in both groups. The initial subtle clinical changes like mild intellectual deterioration, behavioral changes, progressive deterioration in scholastic performance might initially remain unnoticed by parents and teachers. Hence most of the patients were brought when the parents or guardians noted recurrent falls. The visual disturbance was more common in the late-onset group and statistically significant (P-value<0.05) although other presentations did not differ significantly. Other studies also found a higher frequency of ophthalmic manifestations in the late-onset group. EEG had been done in all patients which revealed the classical periodic burst in all cases in both groups. Actually, periodic bursts are found in around 63-85% of the SSPE patient. In this study, the patients with

classical periodic bursts were recruited for the study. So the burst was present in all patients. The classic clinical findings along with a history of measles infection in early life and a supportive EEG warrants testing of CSF anti-measles antibody to arrive at a definitive diagnosis. Anti-measles antibody in CSF was found to be positive in all cases in both groups. In this comparative study of SSPE between the early and late-onset groups, visual disturbance was found to be significantly more common in the adult-onset groups whereas swallowing difficulty was found to be 2.2 times more frequent in early onset group.

In a study the author has mentioned that most of the patients with SSPE have a history of primary measles infection at an early age which is followed by the onset of progressive neurological disorder after a latent period of 6 to 8 years; furthermore, children who have infected with measles under the age of 1 year carry a risk of 16 times greater than those infected at age 5 years or later. Since the incubation period is typically less than a decade, SSPE is commonly a disease of childhood. A higher incidence (male/female ratio 3:1) has been noted in boys, although primary measles infection shows no such sex disparity. The incidence is higher among rural children, children with two or more siblings, and children with mental retardation which is consistent with the other study. It is also more common in children with a lower birth order and in children living in overcrowded environments. In another study it has been suggested that these features like age of exposure, sex, and geography are indicative of intensive measles exposure as a risk factor. Other factors are also identified as risk factors for SSPE which may modify the course of acute measles infection. These are close temporal relationship of measles with another viral infection such as Epstein-Barr virus or parainfluenza type-1 virus.

This study has few limitations. The number of adult patients is fewer in the study. The inclusion of a larger number of patients would have given a clearer picture.

CONCLUSION

SSPE might have some atypical presentation when it is manifested in adolescents and young adults have been reported previously in several case series and research articles. In this study, the socio-demographic, clinical, and investigation profile of pediatric and adult patients with SSPE did not differ significantly except for visual disturbance which was found to be more common in adult patients whereas swallowing difficulty was more frequent in children. A large-scale study involving more patients is warranted to confirm the findings.

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