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ORIGINAL ARTICLE

Subacute Sclerosing Pan-Encephalitis (SSPE): A ten year experience in Dhaka Shishu (Children) Hospital

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Abstract

Introduction: *Number of Subacute Sclerosing Panencephalitis (SSPE) in hospital patients remain unchanged since 2003 although measles immunization coverage has increased to about 88%.*

Objective: *To see the clinical and laboratory findings of children presenting with SSPE and to correlate with measles and its immunization.*

Methods : *SSPE patients in Neuroscience Department of Dhaka Shishu Hospital were categorized clinically in differrent stages. Diagnosis was based upon history of deteriorating neurological functions, typical clinical signs of SSPE, EEG findings and positive measles antibody titers in CSF. Socio-economic status, history of measles and immunization were recorded.*

Results : *Since January 2002 till August 2011, 82 cases were diagnosed. All children had normal development prior to the appearance of neurological signs. Sixty two (75.6%) cases had been immunized with measles vaccine of whom 56 (68%) gave no history of measles. Most of the cases came to hospital at stage 3 (42.7%) and 2 (33%). Fifty two (63.4%) came from rural areas, and the rest were mainly from Dhaka city. EEG feature was suggestive of SSPE in 70(85%) cases.*

Conclusion : *Recent cases of SSPE among children admitted in Dhaka Shishu Hospital has raised concerns about its public health importance. SSPE is still existing at an alarming state in our country despite good measles immunization coverage.*

Keywords: *SSPE, children, measles vaccine*

Introduction

Subacute sclerosing panencephalitis (SSPE) is by far the most common of the chronic encephalitides.Risk of SSPE is 2/100000 after natural infection of measles and only 1/1000000 after vaccination against measles.[1] There is no reported data of SSPE incidence in Bangladesh. SSPE is a progressive neurological disorder characterized by inflammation of the brain

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i.e., encephalitis. Measles virus infection normally cause an acute self limiting disease in which a virus-specific immune response leads to the establishment of lifelong immunity. Complications associated with acute measles can, on rare occasions, involve the central nervous system (CNS). These are post-infectious measles encephalitis which develop soon after infection; and, months to years after the acute disease, results in measles inclusion body encephalitis (MIBE) and SSPE which are based on a persistent measles virus infection of brain cells.[2] The disease may develop due to reactivation of the measles virus or an inappropriate immune response to the measles virus. SSPE usually develops 2 to 10 years after the original viral attack. Children and young adults are primarily affected with SSPE. Males are more affected than females, with a male-to-female ratio of 4:1. There is a geographical component to the infection, with those in rural areas being much more susceptible[1] and approximately 85% of cases arise in rural environments.[3] Specific risk factors include large family, overcrowding in the home, older age of the mother, higher birth order of the parents and rural place of birth.[4] The initial symptoms may include memory loss, irritability, seizures, involuntary muscle movements, and/or behavioral changes, leading to neurological deterioration.

Though measles immunization coverage has increased, number of admitted SSPE cases in Dhaka Shishu Hospital is still alarming. This study is conducted to see the clinical, laboratory and immunization profile of SSPE cases.

Methods

This prospective study was done at Paediatric Neurosciences Department of Dhaka Shishu (Children) Hospital which is a tertiary care paediatric hospital.

Among the admitted patients of Neuroscience department from January 2002 to May 2011, SSPE cases were identified and categorized by stages clinically. Socio-economic status and immunization status against measle were determined. Whether the child suffered from measles or not identified from clinical history.Then antibodies (IgG, IgM) for measles in the CSF were done by ELISA method with help of Armed Forces Institute of Pathology (AFIP), Dhaka.

EEG and other relevant investigations were done. Children who were positive for antibody (IgG, IgM or both) for measles in the CSF were taken as SSPE cases. Total 82 cases were diagnosed during this period.

Data were collected and analyzed by SPSS 11. Chi-square test was done to see co-relation between immunization status and history of measles.

Results

Total 82 cases of SSPE were diagnosed since 2003 (Fig 1). Sixty two (75.6%) cases had been immunized with the measles vaccine. Most of them (68%) presented between 5-12 years of age (Table I). Most

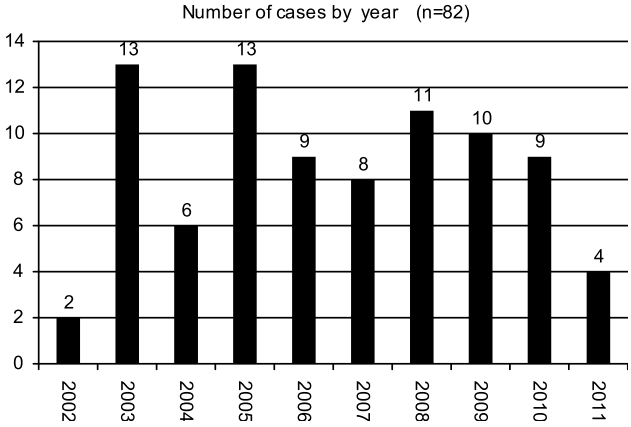

Fig 1 *Number of SSPE cases (2002-2011 yrs)*

of them came to hospital at stage 3 (42.7%) and 2 (33%) (Table III). Fifty two (63.4%) came from rural areas, and the rest were mainly from the capital Dhaka (Table I).

Table I
Socio-demographic characteristics of SSPE patients

Parameter	Number	Percentage
Distribution by Residence		
Urban	30	36.6
Rural	52	63.4
Total	82	100
Distribution by Sex		
Male	72	88.2
Female	10	11.8
Total	82	100
Age at presentation in years		
0-<5	9	11
5-12	56	68
>12	17	21

Table II
H/O measles, measles vaccination and measles antibody titre in CSF

	Number	Percentage
History of measles		
Yes	26	31.7%
No	56	68.3%
Total	82	100%
History of measles vaccine		
Immunized	62	75.6
Not immunized	16	19.5
No information	4	4.9
Total	82	100
Measles antibody titre in CSF		
IgG positive	67	81.8
IgM positive	01	1.2
IgG and IgM positive	14	17.0

EEG feature was suggestive of SSPE in 70 (85%) cases. As most of the children came from long distances and due to lack of logistic support, follow up could be possible only for 7 cases. Three had died, other 4 were severely handicapped, showing features of terminal stages of SSPE.

Chi-square value of co-relation between immunization status and H/O measles is 0.92 which is statistically not significant.

Table III
Clinical stages of SSPE at presentation

	Number	Percentage
Stage1	2	2.4
Stage 2	33	42.4
Stage 3	35	42.7
Stage-terminal	12	14.6
Total	82	100

Table IV
Co-relation between immunization status and H/O measles

	Number	Chi-square value
Immunization	62	.92
Measles	26	

Discussion

Though number of SSPE cases falls significantly worldwide after the introduction of measles vaccine but it is still persisting at alarming state in Bangladesh. Since January 2002 till May, 2011 we have diagnosed 82 cases.

An important point for consideration is that 62 (75.6%) have been immunized with the measles vaccine of whom 56(68%) gave no history of measles.

One study in Israel showed a statistically significant positive correlation between risk of SSPE and early measles infection, large family, overcrowding in the home, older age of the mother, higher birth order, fewer years of schooling of the parents, and rural place of birth.[4]

EEG of 70 patients out of 82 were studied and most of them showed (85%) characteristic finding of SSPE. A study in Turkey EEG studies in SSPE revealed periodic high amplitude complexes in all except one case.[5] It is comparable with our EEG findings.

As most of the children came from long distances and due to lack of logistic support, follow up information on mortality and morbidity could not be presented for the majority of cases. We were able to, however, follow up 7 children within Dhaka city. Three had died, other 4 were severely handicapped, showing features of the terminal stages of SSPE. One patient died within 3-6 months and was reported at advanced state. Sometimes very rapid progression occurs and one report from Turkey showed a patient died approximately 2 months from onset of disease.[6]

Cases with SSPE have no satisfactory treatment. Recent studies suggest that treatment with oral Isoprinosine plus alpha-interferon is effective for SSPE. [7] One study in Poland analyzed treatment with Antineoplaston AS2-1 plus isoprinosine are comparable with those observed during isoprinosine alone treatment but significantly worse than those after administration of Propionibacterium granulosum with isoprinosine. This suggests that Antineoplaston AS2-1 fails to modify importantly the natural course of SSPE.[8]

Bangladesh has achieved considerable success in its measles immunization with a coverage of 88% in 2007.[9] Recently a campaign advocating a second dose

of the vaccine has been launched.[10] While the benefits of the measles vaccine in preventing child mortality and morbidity in developing countries is irrefutable,[11] we found majority of SSPE patients were immunized with measles vaccine. A few reports showed measles vaccine strains causing encephalitis.[12,13]

Conclusion

There is no cure of SSPE. As considerable number of SSPE cases are confirmed every year in a tertiary hospital and most of them are immunized with measles vaccine. So we can assume that SSPE existing in an alarming state in our country despite good measles immunization coverage. Health authority in Bangladesh should take comprehensive measures to ensure the standard practice for preservation and transport of vaccine. Cold chain should be properly maintained as there is unstable power supply in the country.

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