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Case Report

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Adult Onset Subacute Sclerosing Panencephalitis: A case report

Mohammad Akter Hossain[1], Romal Chowdhury[2], Nazmul islam[3], Md. Azharul Hoque[4], Mohammad. Enayet Hussain[5]

[1]Assistant Professor, Department of Neurology, National Institute of Neurosciences and Hospital, Dhaka, Bangladesh; [2]Phase-A Resident, Department of Pulmonology , National Institute of Chest Diseases and Hospital, Dhaka, Bangladesh; [3]Phase-A Resident, Department of Cardiology, National Institute of Cardiovascular Diseases and Hospital, Dhaka, Bangladesh; [4]Professor, Department of Neurology, National Institute of Neurosciences and Hospital, Dhaka, Bangladesh; [5]Assistant Professor, Department of Neurology, National Institute of Neurosciences and Hospital, Dhaka, Bangladesh

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Abstract

Subacute sclerosing panencephalitis (SSPE) is a chronic encephalitis of childhood and young adolescence due to persistent measles virus infection of the central nervous system (CNS). In majority of cases, onset occurs between 5-10 years of age. SSPE generally occurs 5-10 years after measles virus infection1. The diagnosis of SSPE is based on characteristic clinical and electroencephalogram (EEG) findings, increase measles antibody titer in cerebrospinal fluid (CSF) and serum. As onset of SSPE in adults is rare and may have atypical feature it requires high index of suspicion for early and accurate diagnosis. Herein, we report a case of SSPE in a male of 26 years with recurrent episodes of myoclonic jerks. [Journal of National Institute of Neurosciences Bangladesh, 2016;2(1): 40-42]

Keywords: Subacute sclerosing panencephalitis; measles; myoclonic jerks; electroencephalogram; antimeasles antibodies

Correspondence: Dr. Mohammad Akter Hossain, Assistant Professor, Department of Neurology, National Institute of Neurosciences and Hospital, Dhaka, Bangladesh; Email : dr.akter1972@gmail.com; Cell no.: +880172084804

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Introduction

Subacute sclerosing panencephalitis, a progressive fatal neurodegenerative disease caused by an aberrant measles virus in the CNS[2] is largely considered to be a disorder of childhood and adolescence and may not readily be recognized when presenting later in life. The typical clinical presentation of SSPE includes behavioral and intellectual impairment followed by myoclonic jerk and complete neurological deterioration depending on the degree of neuroanatomical structure involvement[3]. However, the initial characteristics and clinical course of the disease can be highly variable and prior reports have suggested that adult-onset SSPE may have atypical features[4].

Case report

A 26-Year-old normotensive, non-diabetic male was admitted to the neurology department of National Institute of Neurosciences and Hospital with the complaints of recurrent, sudden, brief jerks involving the head, limbs and trunk for last one and a half months, accompanied by frequent fall and decreased attention span. His antenatal period was uneventful and was born at term by normal vaginal delivery. Milestones of development were achieved normally and was vaccinated according to national immunization schedule. On query his mother did not give any definite history of measles in his childhood but he had history of viral illness with skin rash eight years back. His family history was negative for psychiatric or neurologic

illness. On neurologic examination, he was conscious and communicable. There was myoclonic jerks almost regularly at an interval of 8-15 seconds involving the arms, legs, head and shoulder. Cranial nerves and fundoscopic examination were normal. The examination of motor system, tone, power and reflexes was unremarkable. General examination did not reveal any abnormality. Laboratory investigation showed normal values of blood count, chemistry and electrolytes. Urine routine microscopic examination, CSF analysis and magnetic resonance imaging (MRI) were all normal. EEG revealed a slow background with periodic generalized complexes consisting of bilaterally symmetrical, high-voltage slow wave complexes for 2-3 seconds at an interval of 8-15 sec which were time locked with the myoclonic jerk (figure-1). The anti measles antibody was found to have a high titer in CSF (IgG- 774.0 concentration/AI). A diagnosis of SSPE was made in view of myoclonus, deterioration of cognitive function, elevated antimeasles antibody level in CSF and periodic discharge in the EEG.

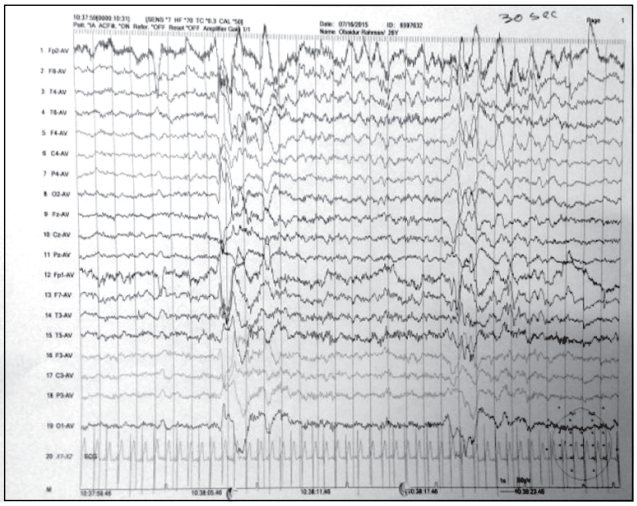

Fig- 1: Periodic burst of generalized discharges on a slow background

Discussion

SSPE is a progressive neurological disorder caused by persistent measles infection in the brain and failure to clear up primary infection due to defective measles virus replication cycle. The mutated measles genome produces abnormal matrix protein (M-protein) which is important for viral budding and establishing extracellular infection. The preserved hemagglutin and fusion protein allow spreading of measles virus from cell to cell[5]. SSPE patients develop a hyperimmune response to measles virus, which fails to control persistent infection[6]. The patients go through several stages. In stage I, behavioral and intellectual changes are noted, rarely SSPE is diagnosed at this stage[7,8]. In stage II, continued intellectual deterioration and onset of typical pattern of myoclonic jerk, both are hallmarks of SSPE. Visual impairment and dressing apraxia may be apparent indicating parieto-occipital involvement[9]. In stage III, prominent hyperkinetic movements, choreoathetosis are noted indicating basal ganglia involvement. Ambulation is clearly affected at this stage. Autonomic dysfunction is a prominent feature[10]. In stage iv, the patients become completely bedridden, the myoclonic jerks and seizures may disappear with the appearance of pathological laughter, persistence of autonomic dysfunction and startle reflex. Our case was in stage II. The progression of symptoms from stage I-IV takes about 1-3 years in 80% cases. Ten percent of patients, develop slower progressive course, some may survive up to 10 years but in another 10% , the disease has a more fulminant course[11]. EEG findings are quite characteristic of SSPE. The slow wave complexes occur in a periodic pattern with interburst interval of 4-12 seconds, the jerks are time locked to the complexes. With the progression of the disease, the interburst interval gets shorter with further background activity slowing[12,13]. Positive measles antibodies in serum and CSF are confirmatory tests, positive oligoclonal band in the CSF indicates intrathecal immunoglobulin synthesis. The level of both viral RNA and antigen in the brain may correlate with disease progression[14]. There is no curative treatment of the disease, virtually all patients are dead within 10 years[8]. In another series, 5% of patients went into remission for a variable duration but subsequently relapsed and died[7]. Several therapeutic trials were conducted. In this case, the patient had clear cognitive decline, as well as myoclonia. The EEG was diagnostic. The possible mechanism of SSPE in vaccinated patient is either due to the preceding primary measles infection or aborted vaccine.

Conclusions

SSPE is a rare complication of measles infection. We need to keep in mind during evaluation of any patient, even in young adult, presenting with cognitive dysfunction with myoclonic jerks to seek a history of previous measles infection as it may cause a latent infection. Although the treatment options are not promising but introduction of measles vaccination program reduce the prevalence of the disease.

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