VOLUME - 11, ISSUE - 11, NOVEMBER - 2022 • PRINT ISSN No. 2277 - 8160 • DOI : 10.36106/gjra

Original Research Paper

Neurology

ATYPICAL MANIFESTATIONS OF SUBACUTE SCLEROSING PANENCEPHALITIS: CASE SERIES

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ABSTRACT Subacute Sclerosing Panencephalitis (SSPE) is a rare, progressive degenerative disease of the brain caused by reactivation of aberrant measles virus. Classical symptoms of SSPE are described as behavioral changes, declining scholastic performance, myoclonic jerks etc. However, at times it presents with varied atypical manifestations, which makes diagnosis difficult. Hereby we report two patients with unusual manifestations of SSPE. In first case, 14-year-old boy presented with recurrent falls of 9 months duration. Initially, it was 1-2 times per week, progressing over 9 months to frequency of 40-50 falls per day. In second case, 21-year-old gentleman presented with behavioral changes and memory disturbance of 1-year duration. However, his family members noticed worsening of symptoms 1 month before presentation as he started developing involuntary jerky movements involving the right upper and lower limb. In both of them electroencephalogram (EEG) was showing long interval periodic discharges and cerebrospinal fluid was showing elevated titers of anti-measles antibody overall suggesting SSPE. Hence it needs much attention for atypical presentations and also considering SSPE in differential diagnosis of unsolved atypical neurological presentations.

KEYWORDS : SSPE, measles, drop attacks, status hemi-myoclonus, electroencephalogram, periodic complexes.

INTRODUCTION:

Subacute sclerosing panencephalitis (SSPE) is a devastating brain disorder attributed to infection by the mutated measles virus. It is a rare complication of measles virus infection affecting children and young adults with an incidence of one case per million population.[1] Symptom and signs are frequently variable and are characterized by behavioral abnormalities, intellectual deterioration, and myoclonus to death.[2] There are few uncommon presentations of SSPE reported such as acute disseminating encephalomyelitis, hemiparesis, and visual disturbances often making an early diagnosis of this fatal disease difficult.[3] The mechanisms that facilitate measles to have varied neurological manifestations are not precisely known. However, it is very important to understand the atypical manifestations of SSPE for improving quality of life and for timely diagnosis. Hereby we present two uncommon cases of SSPE presenting with atypical manifestations.

Case 1:

A 14-year-old boy presented to us with recurrent falls of 9 months duration. Each time, it was associated with abrupt falls mainly observed while standing or walking, without any warning signs or any loss of consciousness, lasting for 2-3 seconds with complete recovery. Initially, the frequency of falls was 1-2 times per week, progressing over 9 months to frequency of 40-50 falls per day. He also sustained minor injuries to the forehead, back of the head during these episodes. There were no other involuntary movements or jerky movements of limbs, urinary incontinence, or tongue bite during these episodes. His parents also observed a decline in his scholastic performance in the past 1 year. Patient was not vaccinated with the measles vaccine and there was no clear history indicating measles during childhood.

On examination, patient was conscious and vital parameters were within normal limits. He was found to be less attentive and apathetic. Cranial nerve examination was normal. No Kayser Fleischer ring (KF ring) was detected. Motor and sensory examinations were found to be normal. There were frequent episodes of sudden falls with trunk extension and knee flexion with most of the time falling backward and sometimes forwards. The propensity to fall was more while standing and walking. These episodes were not associated with any loss of consciousness and with complete recovery in posture within 10- 15 seconds of fall **(Video-1).** In between the attacks, there were no other neurological signs.

Video 1: Video of case 1 showing Generalized bursts of high amplitude slow-wave discharges in EEG (average reference montage, 10 sec/page) with the concurrent clinical event of myoclonus.

Routine hematological parameters were within normal limits. Liver and Kidney function tests were normal. Cerebrospinal fluid (CSF) was acellular with protein of 48mg/dl and glucose of 64mg/dl. Electroencephalogram (EEG) showed periodic, long interval generalized bursts of high amplitude slow-wave discharges with corresponding clinical episodes of myoclonic jerks of the axial structures, mainly paraspinal muscles. **(Figure 1)** Brain MRI was normal with non-specific subcortical white matter signal changes. CSF titer for IgG measles antibodies was 1:625 by ELISA, suggesting SSPE. The child was treated with Tab. Isoprinosine 100mg/kg/day and symptomatic treatment for myoclonus with Sodium Valproate 20mg/kg/day and Tab. Clobazam 10mg/day. Patient did not show response to treatment.

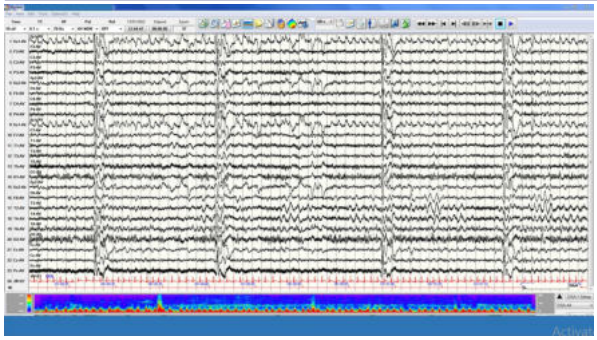

Figure 1: Scalp EEG (average reference montage, 60sec/page) of case 1 showing bilaterally symmetrical high amplitude long interval discharges suggestive of SSPE.

Case 2:

A 21-year-old gentleman from the eastern part of India presented with behavioral changes and memory disturbance of 1-year duration**.** It was initially noticed by his mother that he developed reduced interest in interacting with others and was searching for belongings in the wrong place. However, he was able to carry out his daily activities without assistance. Initially, he was diagnosed as functional neurological disorder and was advised behavior therapy and supportive care in nearby place. However, his family members noticed worsening of symptoms 1 month before presentation as he started developing involuntary jerky movements involving the right upper and lower limb. It was present continuously and used to worsen with anxiety. With these symptoms, he was finding difficulty in carrying out his daily activities requiring assistance of an attendee. He did not receive measles vaccination during childhood.

At the time of presentation, the patient was conscious and alert. His vital parameters were within normal limits. On neurological examination, higher mental function showed impairment of attention with relative preservation of visuospatial, language, and other cognitive domains. Cranial nerves examination was normal. Right-sided hypertonia was present in both upper limbs and lower limbs with occasional swaying towards the right while walking. Romberg's sign was negative. There were continuous slow myoclonic jerks involving the right upper and lower limbs, mainly involving proximal joints in the upper limb **(Video 2).** Jerks did not resolve with attempted distraction. The rest of the systemic examination was unremarkable.

Video 2: Video of case 2 showing continuous slow myoclonic jerks involving left upper and lower limb, almost at the rate of once in two seconds present in all body postures.

His routine hematological parameters were normal. Liver and renal function tests were normal. MRI brain was showing diffuse cerebral atrophy with normal brain parenchyma **(Figure 2).** CSF analysis was acellular with protein of 101.6 mg/dl and glucose of 69.18 mg/dl with high titer of IgG antibodies to measles virus (1:625). EEG revealed long interval frontally dominant generalized high amplitude periodic discharges time-locked with myoclonic jerks recorded in surface electromyogram (EMG) placed over the right forearm. Overall features were suggestive of subacute sclerosing panencephalitis. Patient was treated with Tab. Isoprinosine 100mg/kg/day and symptomatic treatment for myoclonus with Sodium Valproate 20mg/kg/day and Tab. Clobazam 10mg/day. There was no response to treatment at the time of discharge 1 week later.

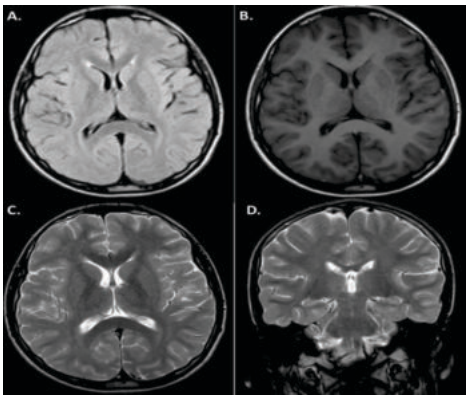

Figure 2: Magnetic resonance imaging (MRI) brain of case 2 with FLAIR axial (A.) T1 axial (B.) T2 axial (C.) and coronal (D.) sections showing diffuse cerebral atrophy

DISCUSSION:

Hereby we describe our experience with atypical manifestations of SSPE and recurrent falls in young and status hemi myoclonus. SSPE is a rare, potentially preventable devastating complication of the measles virus.[4] The disorder is more common in the pediatric population and can be misdiagnosed as sole epilepsy in the initial stages of the disease.[5] Patients can develop atypical manifestations of the disease in 10% of the cases like spastic hemiparesis, cerebellar ataxia, acute disseminated encephalomyelitis (ADEM), and unilateral myoclonus and refractory epilepsy.[6]

Falls in children can occur as a result of neurogenic syncope, transient ischemic attack, posterior circulation stroke, and most notable epilepsy syndromes.[7] With regards to epilepsy syndromes, falls are more common in patients with atonic seizures resulting from complete loss of tone. In our first case report, loss of balance and falls is probably due to myoclonic jerks involving the paraspinal muscles. On a few occasions it was not associated with falls, though the patient had myoclonus, as evidenced by an initial segment of the video, this could be due to intact protective reflexes.

SSPE may mimic drop attacks and can be the presenting feature of SSPE.[7] Our first patient did not manifest with any jerky movement of limbs at the time of presentation or during evaluation. Recurrent falls in our patient may be explained due to slow axial myoclonus leading to loss of postural tone and fall as seen in the EEG.

There are only a handful of cases that have been described to present with hemi-myoclonus. Initially, it was described in a 13-year-old girl with strictly unilateral SSPE complexes in EEG and strictly contralateral myoclonus, which remained the same even after 3 months of follow-up.[8]

One of the hypotheses postulated to be due to involvement of one hemisphere leading to side locked symptoms, though SSPE is considered due to diffuse involvement.[9] Similar presentation has been described in a 35-year-old man who was previously misdiagnosed with herpes encephalitis.[10] Hence it is very important to have a familiarity with these atypical presentations of SSPE for timely diagnosis. SSPE may be considered as a differential diagnosis in children with unexplained recurrent falls, Hemi-myoclonus without any other possible explanation.

CONCLUSION:

Though SSPE presents with cognitive decline and myoclonic

jerks, atypical presentations have to be kept in mind to make a swift diagnosis of this potentially fatal disease. SSPE is associated with a bad prognosis; however, early diagnosis and institution of appropriate treatment may help improve the quality of life.

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