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LETTER TO THE EDITOR

Periodic Jaw-Opening Myoclonus in Subacute Sclerosing Panencephalitis

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Dear Editor,

Subacute sclerosing panencephalitis (SSPE) is an inexorable neurological disorder caused by the persistence of mutant measles virus in the brain. SSPE is progressive and fatal in 95%–96% of patients and has no curative treatment to date.[1] Primary measles infection usually occurs before the age of 2 years, followed by a latent period of 7–10 years, after which progression sets in.[1] The clinical features are dominated by a constellation of rapid cognitive decline, myoclonic jerks, seizures, and typical electroencephalographic (EEG) findings. SSPE usually occurs in childhood, although adult-onset cases have also been reported. Movement disorders are also reported.[2] We report the occurrence of a highly unusual jaw-opening myoclonus in a child with SSPE, thereby expanding the phenotypic repertoire of this condition.

CASE PRESENTATION

A 6-year-old girl presented to us with a history of frequent falls, recurrent mouth opening, and behavioral disturbances for the past three months. Her parents reported that she had become increasingly inattentive and irritable and had an increase in temper tantrums. For the past two months, she had developed sudden jerks involving her trunk and limbs, leading to the dropping of objects from her hands. These jerks did not persist in sleep. She often required help to dress herself and eat, although she had been previously able to perform these activities independently. She denied fever, visual or hearing disturbance, or speech or swallowing difficulties. She was born at term by vaginal delivery, with no perinatal complications. Her developmental milestones had been normally achieved without any history of febrile exanthem or seizures. However, she had not been vaccinated before.

On examination, the child was cooperative, although inattentive. She was able to follow commands. She exhibited frequent and periodic jaw-opening myoclonus, accompanied by neck extension and myoclonic jerks involving the flexor muscles of the limbs (upper limbs more than lower limbs and trunk). These myoclonic jerks showed a slow relaxation phase (Supplementary Video 1 in the online-only Data Supplement). She had brisk deep tendon reflexes. The findings from examinations of extraocular movements, the fundus, coordination, and the extrapyramidal system were normal. SSPE continued to be clinically suspected.

The results of routine blood investigations, including a complete hemogram, hepatic and renal function tests, thyroid function test, serum lactate test, tests for viral markers for human immunodeficiency virus, and erythrocyte sedimentation rate and C-reactive protein tests, were normal. EEG showed high-amplitude periodic discharges, occurring every 8–10 seconds, consistent with Radermecker complexes (Figure 1). Cerebrospinal fluid (CSF) analysis showed acellular CSF, with protein 89 mg/dL (elevated) and normal sugar 78 mg/dL (concomitant blood sugar 92 mg/dL). CSF analysis also showed elevated anti-measles antibodies (CSF/Serum Quotient reference 7.75 (normal < 1.3), confirming the presence of SSPE, as per the Dyken criteria.[3] Brain magnetic resonance imaging findings were normal.

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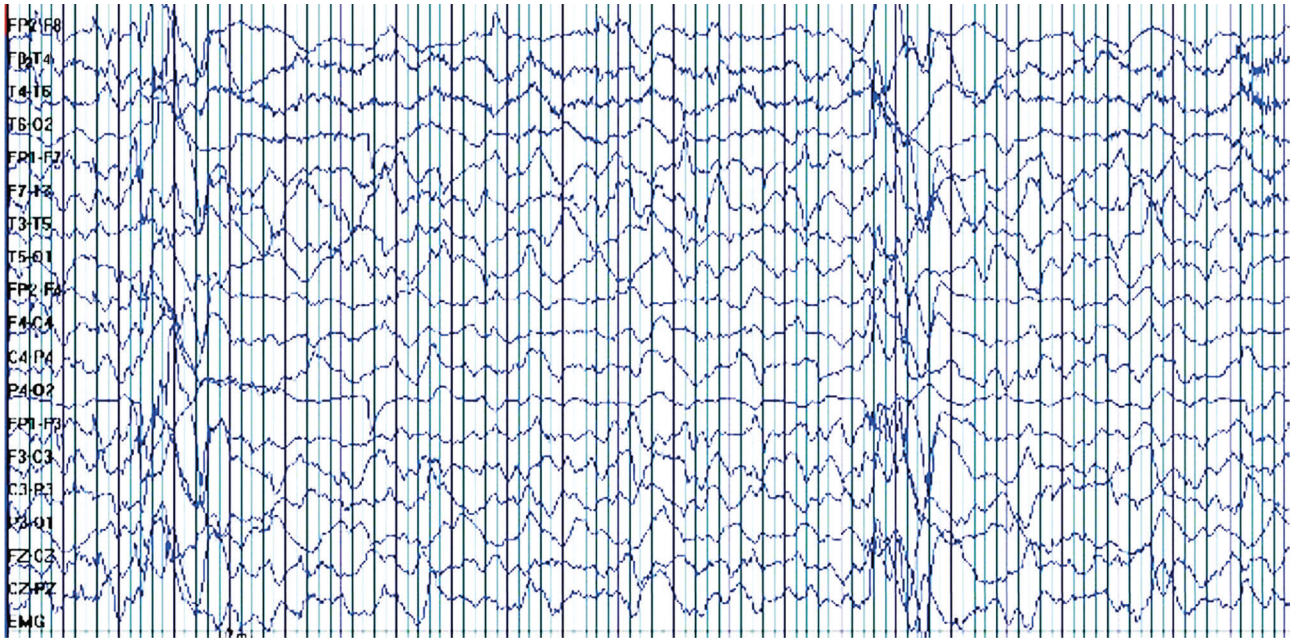

Figure 1. Electroencephalography of the patient. Electroencephalography showing periodic, high-amplitude discharges occurring every 8–10 seconds, accompanied by a slow background, consistent with Radermecker complexes.

The child was initiated on oral isoprinosine (100 mg/kg per day), valproic acid (500 mg thrice daily) and clobazam (10 mg once daily at night). Mild improvement in myoclonic jerks was observed at three months. However, cognitive and behavioral issues continued to worsen.

DISCUSSION

Movement disorders are an underrecognized but frequent feature of SSPE.[2] Myoclonus in SSPE is described to be characteristically periodic and stereotyped, usually involving the head, trunk, and limbs. A slow relaxation phase, defying the otherwise rapid shock-like nature of myoclonus, is sine qua non. Initial involvement of the head followed by the trunk may lead to head drops and falls. Muscle contraction is followed by a relaxation phase for 1–2 seconds. A comprehensive description of movement disorders in SSPE has been rarely reported. We previously assessed 50 patients with SSPE in a cross-sectional study from Lady Hardinge Medical College, New Delhi, India.[2] The most frequent movement disorders were observed to be myoclonus, ataxia, chorea-athetosis, dystonia, tremor, repetitive behaviors and parkinsonism. Notably, we did not observe the occurrence of jaw myoclonus in this series.

Unlike the jaw myoclonus reported with other conditions, the jaw myoclonus observed in our patient had a slower phenomenology. Furthermore, the jaw myoclonus in our patient occurred in conjunction with limb and neck myoclonus, which was followed by slow relaxation. This was synchronized with the characteristic EEG findings, Radermecker complexes. Radermecker complexes are characterized by quasiperiodic, high voltage (300–1,500 microvolts), sharp and slow wave complexes occurring every 0.5–2 seconds, with a slowing of background activity every 4–12 seconds. These complexes are logged with myoclonic activity.

Jaw myoclonus has not been reported in association with SSPE thus far. There is controversy on whether the precise mechanism of the periodic dystonic myoclonus observed in SSPE has cortical, subcortical or cortical-subcortical origins.[4] One electrophysiological study demonstrated that the origin of myoclonus varies with the stage of disease progression, with cortical and cortical-subcortical origins in the earlier stages and progression to the caudal brainstem occurring in more advanced disease.[5] Brainstem involvement has been observed previously on radiology[6] and pathology.[7] In our patient, jaw myoclonus probably represents the extension of the disease from the cortical/subcortical origin to the brainstem, with consequent myoclonus affecting the trigeminal-innervated jaw muscles, occurring in association with axial myoclonus. Jaw-opening myoclonus has not been previously reported in association with SSPE; therefore, this case may serve as a diagnostic pointer.

Ethics Statement

The study was approved by the Ethics Committe for Human Research, Lady Hardinge Medical College, New Delhi, India (Approval number: F.LHMC/IEC/2022/02/31). Informed consent was obtained from the parents for publication in *Journal of Movement Disorders*.

Supplementary Video Legends

Video 1. Periodic jaw-opening myoclonus, accompanied by neck extension and prominent upper limb myoclonus involving the flexor muscles with slow relaxation, is observed. Myoclonus in the lower limbs and trunk is also observed to a lesser extent when the child is walking, leading to loss of postural tone.

Supplementary Materials

The online-only Data Supplement is available with this article at https://doi.org/10.14802/jmd.23015.

Conflicts of Interest

The authors have no financial conflicts of interest.

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Author Contributions

Conceptualization: Divyani Garg, Suvasini Sharma. Data curation: Ashna Kumar. Formal analysis: Divyani Garg. Investigation: Ashna Kumar, Suvasini Sharma. Methodology: Divyani Garg, Suvasini Sharma. Project administration: all authors. Resources: Suvasini Sharma. Software: Divyani Garg. Supervision: Suvasini Sharma. Validation: Suvasini Sharma. Visualization: Divyani Garg. Writing—original draft: Divyani Garg. Writing—review & editing: Ashna Kumar, Suvasini Sharma.

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