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LETTER TO THE EDITOR

An early diagnostic tool for subacute sclerosing panencephalitis: Sleep-deprived electroencephalography

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Dear Editor,

Subacute sclerosing panencephalitis (SSPE), caused by the measles virus, is associated with characteristic neurological symptoms, electroencephalography (EEG) findings, and brain imaging results. Diagnostic criteria for various presentations of the disease were established in 2010 (1). However, in some cases, EEG and imaging findings remain normal for months.

The typical presentation of SSPE includes either an acute, rapid, subacute, or chronic progressive course, or a chronic relapsing-remitting course, often accompanied by hallmark signs such as myoclonus. Following an average asymptomatic period of seven years, progressive neurological deterioration occurs. Common symptoms include behavioral changes, intellectual problems, myoclonic seizures, blindness, and ataxia. Currently, there is no cure for SSPE (2).

Electroencephalography findings in SSPE often feature periodic complexes described as stereotyped, bilaterally synchronous, and symmetrical waves with amplitudes ranging from 100 to 1000 μV and frequencies 1 to 3 Hz. While these typical EEG patterns are more commonly observed in SSPE, atypical patterns—including asymmetric periodic complexes, spike-and-slow wave patterns, paroxysmal rhythmic delta activity between periodic complexes, and normal EEG patterns—can also be present (3).

Neuroimaging rarely helps in diagnosing SSPE. In the early stages, magnetic resonance imaging (MRI) findings may appear normal, and the observed changes are not pathognomonic for SSPE. However, MRI can be utilized to monitor disease progression and exclude other pathologies. In later stages, asymmetrical hyperintense lesions in the cortical and subcortical regions, including the thalamus, corpus callosum, and basal ganglia, may be observed on MRI T2 sequences (4, 5).

A 22-year-old female patient presented to our hospital with complaints of continuous involuntary jerky movements of the left side of her body, persisting for one month, along with speech and gait disturbances. She exhibited a grimacing appearance resembling risus sardonicus. Neurological examination findings were normal. Brain computed tomography and MRI, including diffusion-weighted and T2 sequences, revealed no pathology. Electroencephalography results were also normal. Routine blood tests and cerebrospinal fluid (CSF) analysis showed no abnormalities, except for significantly elevated anti-measles immunoglobulin G (IgG) antibody titers in both the CSF and serum. The elevated CSF measles IgG level was >230 IU/mL (normal range: <25 IU/mL). The CSF total IgG level was measured at 9.31 IU/mL (normal range: 0.33–6.1), with a CSF-to-serum IgG ratio of 0.09 (normal range: 0.03–0.06), and an IgG index of 2.20.

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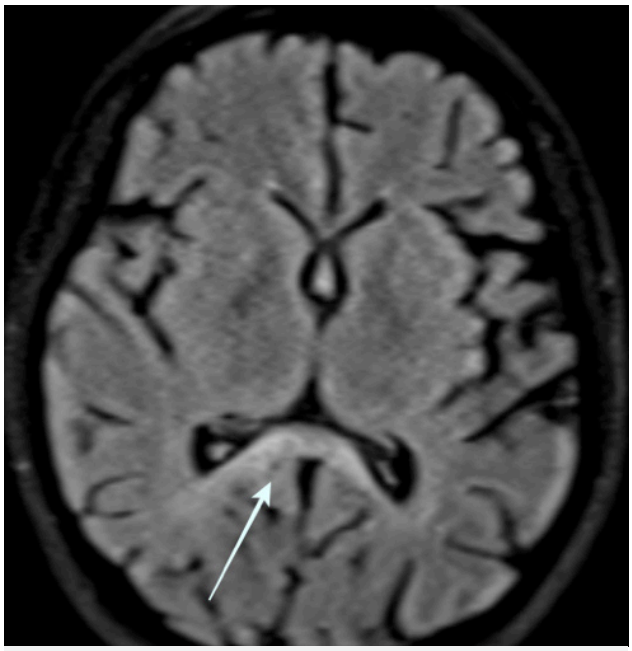

Figure 1. Hyperintensity in the splenium of the corpus callosum on fluid-attenuated inversion recovery (FLAIR) sequence.

The patient's history revealed that she had been completely unvaccinated and had contracted measles during early childhood.

The patient was treated with carbamazepine and intravenous immunoglobulin for five days. She was started on oral isoprinosine (100 mg/kg per day). Despite these treatments, her myoclonic jerks persisted. The patient's condition continued to deteriorate, and she became akinetic and mute. The illness was explained to her family, who opted to care for her at home. MRI performed in the fourth month revealed hyperintensity in the splenium of the corpus callosum on T2-weighted and fluid-attenuated inversion recovery (FLAIR) sequences, as well as brain atrophy (Fig. 1).

Cerebrospinal fluid analysis confirmed the diagnosis of SSPE, but the MRI and routine EEG findings did not support the diagnosis. We proceeded with a sleep-deprived EEG, which showed bilateral generalized pseudoperiodic slow-wave paroxysms, particularly during the transition between sleep and wakefulness in the superficial stage of non-rapid eye movement (NREM) sleep (Fig. 2). The sleep-deprived EEG findings ultimately supported the diagnosis.

In conclusion, SSPE is a progressive slow-virus infection. Clinical features often raise suspicion of the illness. However, in the early stages, MRI and routine EEG findings may appear normal, only becoming pathological in later months. MRI abnormalities, such as those observed in advanced stages of the disease, are not pathognomonic for SSPE. Sleep EEG is superior to awake EEG for

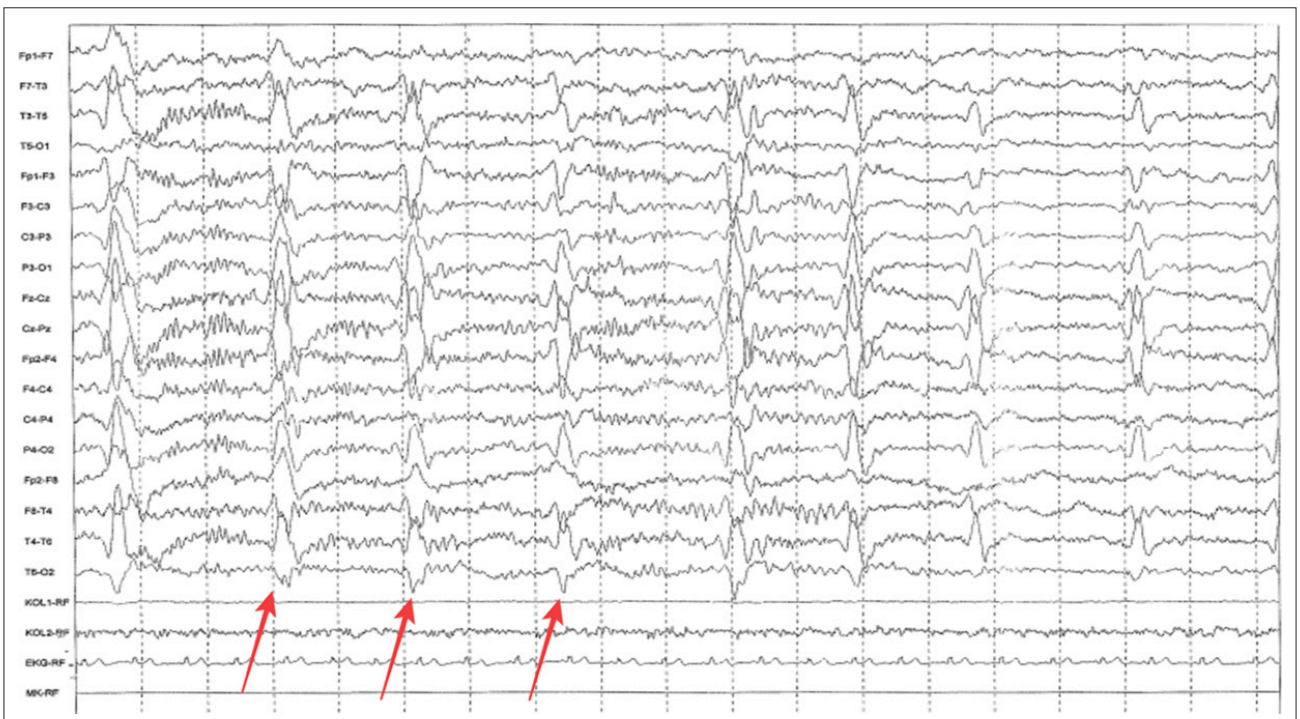

Figure 2. Bilateral generalized pseudoperiodic slow-wave paroxysms.

detecting epileptiform anomalies, as epileptiform discharges are modulated by sleep and occur with higher frequency during NREM sleep compared to the awake state. Most clinical studies suggest that sleep EEG has added diagnostic value compared to standard EEG (6). Therefore, when SSPE is suspected, a sleep-deprived EEG can serve as a valuable tool for early diagnosis. Its use can shorten the time required for diagnosis and help prevent misdiagnosis.

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REFERENCES

1. Jafri SK, Kumar R, Ibrahim SH. Subacute sclerosing panencephalitis - current perspectives. Pediatric Health Med Ther 2018; 9:67-71. [CrossRef]
2. Gutierrez J, Issacson RS, Koppel BS. Subacute sclerosing panencephalitis: An update. Dev Med Child Neurol 2010; 52:901-907. [CrossRef]
3. Kurata T, Matsubara E, Yokoyama M, Nagano I, Shoji M, Abe K. Improvement of SSPE by intrathecal infusion of alpha-IFN. Neurology 2004; 63:398-399. [CrossRef]
4. Tuncay R, Akman-Demir G, Gokyigit A, Eraksoy M, Barlas M, Tolun R, et al. MRI in subacute sclerosing panencephalitis. Neuroradiology 1996; 38:636-640. [CrossRef]
5. Oguz KK, Celebi A, Anlar B. MR imaging, diffusion-weighted imaging and MR spectroscopy findings in acute rapidly progressive subacute sclerosing panencephalitis. Brain Dev 2007; 29:306-311. [CrossRef]
6. Ng M, Pavlova M. Why are seizures rare in rapid eye movement sleep? Review of the frequency of seizures in different sleep stages. Epilepsy Res Treat 2013; 2013:932790. [CrossRef]

EDITOR'S NOTE

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