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Case Report

Marked improvement in febrile infection-related epilepsy syndrome after lidocaine plus $MgSO_4$ treatment in a 12-year-old girl

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ABSTRACT

Purpose: This report sheds light on a successful treatment in febrile infection-related epilepsy syndrome (FIRES) with the combined use of lidocaine and $MgSO_4$.
Methods: We report a 12-year-old previously healthy girl who experienced an upper respiratory infection with fever and headache for 2 days, then suddenly went into a coma followed by repetitive status epilepticus. All tests for CNS infection, metabolic and toxic diseases, and autoimmune encephalitis were negative. Hence, the diagnosis of FIRES was made. During 5 weeks of hospital treatment, various antiepileptic drugs were administered at different times without success. To achieve seizure control, we then attempted the use of lidocaine first, then followed by $MgSO_4$.
Results: The SE was successfully controlled when lidocaine plus $MgSO_4$ was introduced. At follow-up, almost no neurological sequelae remained.
Conclusion: This is the first report describing the combined use of lidocaine and $MgSO_4$ with successful treatment outcomes. This experience has indicated that even FIRES can be controlled if treated promptly with certain agents. However, more studies are needed to explore the mechanisms and effects of lidocaine and $MgSO_4$ in FIRES.

1. Introduction

Febrile infection-related epilepsy syndrome (FIRES), also known as devastating epileptic encephalopathy in school-aged children or acute encephalitis with refractory repetitive partial seizures, is an acute and explosive-onset epileptic encephalopathy with a fatal outcome, which occurs in previously healthy children following a nonspecific febrile illness [1,2]. The precise etiology of this syndrome and the effective treatment options remain unclear. Herein, we present our experience in treating a 12-year-old girl with FIRES, who dramatically recovered from this devastating encephalitis after treatment with a combination of lidocaine and $MgSO_4$.

2. Case report

A 12-year-old girl was admitted to our emergency department with complaints of severe headache, dizziness, and fever (38 °C). Prior to her admission, she was healthy without any epileptic disorders and had normal development. The patient did not have a familial history of any epileptic or genetic metabolic disease. A week prior to admission, she experienced fever, sore throat, and fatigue and was treated for acute pharyngitis. On admission, she had a Glasgow Coma Scale (GCS) score of 12. On Day 2 of admission, her condition worsened rapidly, and she fell into a state of coma (GCS score of 3). She then manifested intractable status epilepticus (SE) with generalized tonic–clonic seizures, accompanied by tachycardia, respiratory compromise, and hypotension alternating with myoclonic jerks of both lower limbs. Although SE was temporarily stabilized by administration of BZD agents, the focal myoclonic seizures, involving both lower limbs alternatively, did not stop. Interictal electroencephalography (EEG) revealed diffuse slow delta activity (Fig. 1), distributed with frequent bouts of ictal fast activity. Initially, central nervous system infection was highly suspected, and so we initiated antibiotic and antiviral treatments; however, these were

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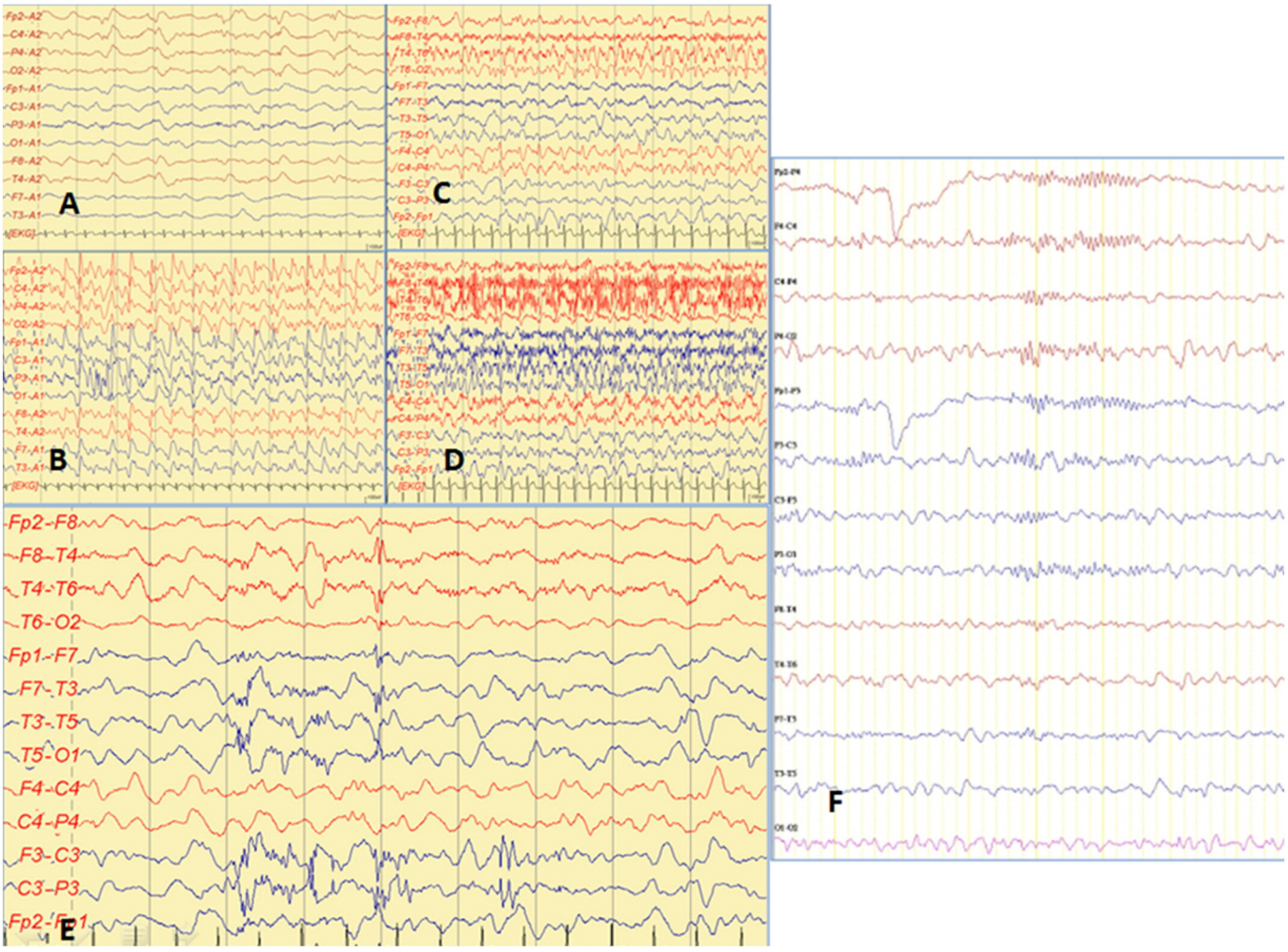

Fig. 1. (A) On Day 1 of admission, the patient was in a state of deep coma; electroencephalography (EEG) background consisted of slow delta waves (3–4 Hz, 25–50 μV). (B) On Day 2, the EEG pattern shifted to a slow background mixed with numerous spikes or sharp-wave activity from both hemispheres. (C and D) On Day 3, 8–9 s before the generalized seizure occurred, the EEG pattern in the right frontal–temporal region transformed into fast low-voltage activities (C) and quickly entered a state of generalized fast low-voltage activities. In the clinical setting, generalized tonic–clonic seizure appeared (D). (E) On Day 31 of admission (3 days of lidocaine use prior to the introduction of $MgSO_4$), the EEG background consisted of a slow theta wave (4–5 Hz, 25–50 μV) interspersed with paroxysmal spike activities. (F) On Day 35, after the introduction of lidocaine for 7 days and $MgSO_4$ for 4 days, neither clinical seizures nor epileptiform activities on EEG were noted.

ineffective. Simultaneously, multiplex polymerase chain reaction, microarray, high-throughput sequencing, and molecular biological detection to detect encephalitis with unknown pathogens based on Taiwan Pathogenic Microorganism Genome Database were performed, but all test results were negative.

Unrevealing laboratory investigations included arterial blood gas analysis; complete blood count; blood glucose level; electrolyte panel; serum concentrations of blood urea nitrogen, creatinine, calcium, magnesium, phosphate, and liver enzymes (aspartate aminotransferase, alanine transaminase, coagulation studies, and serum concentrations of bilirubin and ammonia); and cultures of blood and cerebrospinal fluid. Specialized metabolic tests were also conducted including quantitative plasma amino acids, evaluation of plasma lactate and pyruvate levels, acylcarnitine profile, and qualitative urine organic acids. Drug testing using appropriate panels was performed on samples of blood and urine to determine the presence of drugs that can cause altered levels of consciousness, such as sedatives, salicylates, hallucinogenic agents, anticholinergic agents, opioids, monoamine oxidase inhibitors, acetaminophen, selective serotonin reuptake inhibitors, and tricyclic antidepressants and was negative. In addition, we examined for the presence of environmental toxins, such as organophosphates, lead or other heavy metals, and hydrocarbons in the aforementioned biological samples — all were unremarkable. Screening for autoimmune and paraneoplastic panel in blood or cerebrospinal fluid [i.e., antibodies of anti-Hu, anti-Yo, anti-Ri (ANNA2), anti-MA2 (Ma2/Ta), anti-CV2/CRMP5, antiamphiphysin, anti-NMDAR, anti-Caspr2, anti-AMPAR, anti-LGI1] revealed negative results. Based on these test results, a diagnosis of FIRES was made.

Status epilepticus (SE) was treated using various drugs at different time points, specifically using phenobarbital (PHB; 15 mg/kg for loading, 3 mg/kg/day for maintenance), phenytoin (15 mg/kg for loading, 5 mg/kg/day for maintenance), valproic acid (15 mg/kg/day gradually titrated to 60 mg/kg/day), levetiracetam (LEV, 1500 mg q12hr), intravenous immunoglobulin (1 g/kg; up to 3 courses), dexamethasone (7 days), midazolam (20 mg/kg/min continuous intravenous infusion), pyridoxine, and a ketogenic diet (stopped by gastrointestinal bleeding). The levels of all these drugs were closely monitored to ensure their effectiveness. However, SE occurred up to dozens of times a day (>30 times overall; Fig. 2).

Drug-induced comas were induced four times (two times with pentobarbital, one time with propofol, and one time with thiopental). Throughout the course of hospital admission, frequent EEG monitoring was performed. Electroencephalography showed burst-suppression features with suppression periods lasting 4–5 s and without electroclinical seizures. However, every time the dose of