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Wolff-Parkinson-White Syndrome in a Patient with Mitochondrial Encephalopathy, Lactic Acidosis, and Stroke-like Episodes Syndrome Mimicking Juvenile Myoclonic Epilepsy

Joo Hye Sung
Jung Hoon Han
Hayom Kim
Jung Bin Kim

Department of Neurology,
Korea University College of Medicine,
Seoul, Korea

Dear Editor,

Cardiac involvement in mitochondrial encephalopathy, lactic acidosis, and stroke-like episodes (MELAS) syndrome reportedly presents mainly as cardiomyopathy, but there is a paucity of reports of cardiac conduction disturbance in MELAS syndrome. Although a few studies have described Wolff-Parkinson-White (WPW) syndrome in patients with MELAS syndrome,[1-3] the electroencephalography (EEG) findings in patients with concurrent MELAS and WPW syndromes were not described in detail. We present a case of MELAS with WPW syndrome mimicking juvenile myoclonic epilepsy (JME).

A 19-year-old man presented with recurrent episodes of loss of consciousness for the previous 3 years. His past medical history, general physical examination, and brain MRI findings were unremarkable. A laboratory examination revealed no abnormal findings except for slightly elevated lactic acid [3.2 mmol/L (reference range: 0.7−2.5 mmol/L)]. Electrocardiography revealed a short PR interval and a delta wave with QRS-complex widening, suggesting WPW syndrome (Fig. 1A). He was therefore initially referred to the Department of Cardiology. Since no evidence was found of atrioventricular reentrant tachycardia preceded by loss of consciousness, he was referred to the Department of Neurology having not undergone any interventional procedure. The findings of a neurologic examination were unremarkable. His height was 161 cm, which is lower than the third percentile. History-taking revealed that he often experienced myoclonus, which had developed when he was 14 years old. His myoclonus occurred irrespective of exposure to light; however, all of the six generalized tonic-clonic seizures that he had experienced occurred when he was exposed to bright sunlight. He had not experienced an absence seizures. Abundant 2−3 Hz generalized spikes/polyspikes and wave discharges were found in an EEG examination (Fig. 1B). The electroclinical features led to an initial consideration of JME as a diagnosis. A detailed family history-taking revealed that his mother and maternal grandmother had been diagnosed with epilepsy, hearing defect, and diabetes. His maternal great-grandmother also had a hearing defect, and his aunt was diagnosed with epilepsy (Fig. 1C).

Considering the patient's short stature and family history, genetic studies for identifying hereditary causes of epilepsy were performed. The mitochondrial 3243A>G mutation in the *MT-TL1* gene was confirmed, and so he was diagnosed as MELAS syndrome. Further evaluations for identifying the involvement of other organs were performed. Electromyography, brainstem auditory evoked potentials, and an ophthalmologic examination showed no abnormalities. Although he did not complain of any deterioration of his hearing, pure-tone audiometry revealed moderate sensorineural hearing loss with preserved speech discrimina-

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Correspondence
Jung Bin Kim, MD
Department of Neurology,
Korea University College of Medicine,
73 Inchon-ro, Seongbuk-gu,
Seoul 02841, Korea
Tel +82-2-920-6277
Fax +82-2-925-2472
E-mail brainbin80@gmail.com

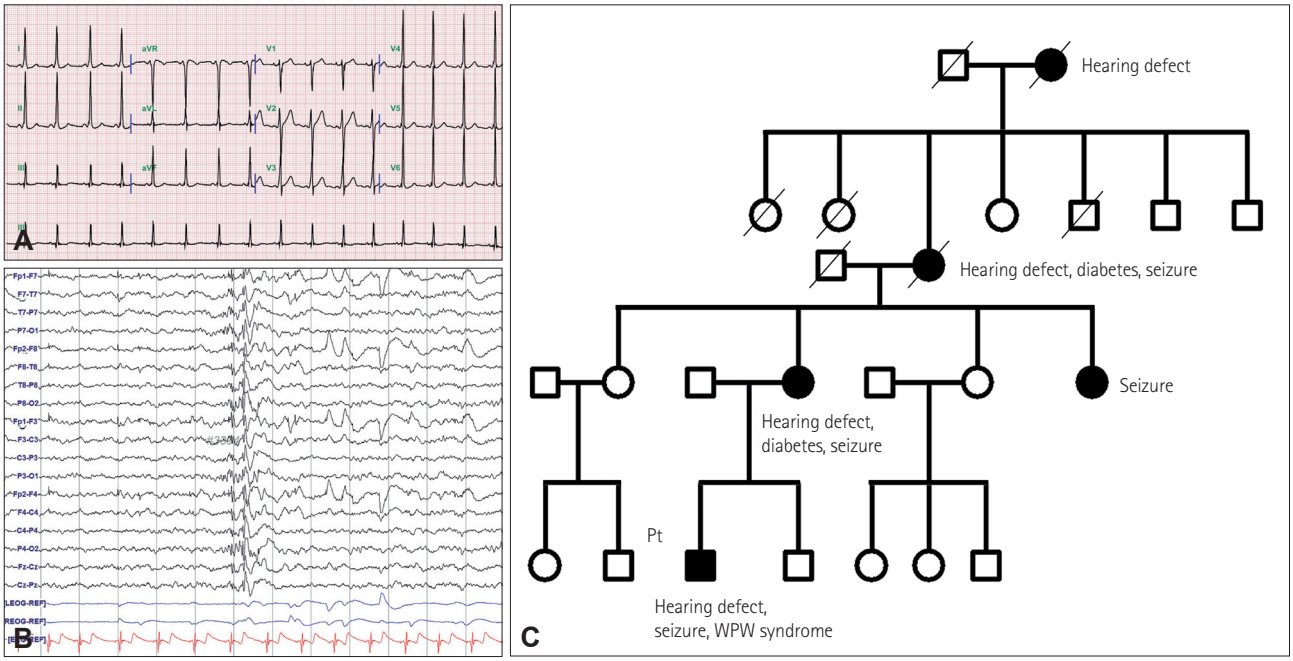

Fig. 1. A: Electrocardiography shows a short PR interval (96 msec) and QRS-complex widening (114 msec) with a delta wave. B: Electroencephalography shows 2–3 Hz frontal-dominant generalized polyspikes and wave discharges. C: Pedigree of the patient shows maternal inheritance traits. WPW: Wolff-Parkinson-White.

tion. He was receiving ubidecarenone and an antiepileptic drug (valproate). He experienced no seizure attack during the 4-month follow-up after discharge. Due to the possibility of mitochondrial toxicity of valproate, we are planning to replace this with other antiepileptic drugs.

While there are a few reports of WPW syndrome in patients with MELAS syndrome,[1-3] to our best knowledge EEG findings mimicking JME in patients with concurrent MELAS and WPW syndromes have not been described. Various patterns of epileptiform discharges can be found in patients with MELAS syndrome, although none of the findings are pathognomonic.[4,5] Focal and multifocal spikes in association with cerebral lesions have been reported more frequently than generalized spikes in patients with MELAS syndrome.[4,5] The present case implies that an EEG examination is essential in WPW syndrome with loss of consciousness without evidence of preceding atrioventricular reentrant tachycardia. In addition, it has been unclear whether WPW syndrome might be an epiphenomenon in MELAS syndrome that mainly involves tissues with high energy demand. However, a genetic study should be considered when an epileptogenic condition has been confirmed in patients with WPW syndrome. Given the electroclinical features of the present MELAS case mimicking JME, a genetic study should be considered if WPW syndrome or other symptoms suggesting MELAS are comorbid in patients who are diagnosed electroclinically as JME.

Conflicts of Interest

The authors have no financial conflicts of interest.

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