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A Report of Long QT Syndrome that Mimics Epilepsy: A Case Report

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Abstract

Introduction: Long QT syndrome is a rare hereditary disorder that could be a potentially fatal condition. One of the symptoms of long QT caused by ventricular arrhythmia is seizure. The diagnosis of this syndrome might be delayed when an initial diagnosis of epilepsy is made.

Case presentation: The patient to be studied in this research was a 24-year-old right handed female. She had the spells since she was 14; which were characterized by uncomfortable anxiety, nausea, pallor, and palpitation followed by generalized weakness and occasionally generalized clonic jerks with obvious impairment of consciousness. She was treated with Depakine and Carbamazepine. During the video-EEG monitoring, she had one habitual attack accompanied with ventricular tachycardia and cardiac arrest for which cardiorespiratory resuscitation was immediately started, and fortunately the patient returned to normal condition. Cardiac evaluation was requested and diagnosis of long QT syndrome was confirmed. Implantable cardiac defibrillator was placed for her.

Conclusion: Long QT syndrome possesses considerable mortality decreased with proper therapy. Long QT syndrome imitates seizure disorders. Hence, taking electrocardiography is required for individuals with vague causes of seizure and uncommon semiology.

Keywords: Long QT Syndrome, Seizure, Syncope

Introduction

Long QT syndrome (LQTS) is an inherited cardiac ailment induced by defects in the channels of cardiac ion, clinically specified by syncope, palpitations, and abrupt cardiac death, with different QT prolongation rates and T-wave morphological defects on the ECG surface. LQTS can be initially recognized as syncopal attacks, mainly in young adults and children. Current estimates of the prevalence of LQTS vary from 1 in 2000 to 1 in 7000. The mentioned rates may still underestimate the problem, considering that LQTS is not mostly diagnosed and possesses changing penetrance (1). LQTS can raise the death risk of the affected individuals and members of their family. Epileptologists have to know that patients with seizure and abrupt loss of consciousness may have LQTS. For those misdiagnosed with epilepsy, the diagnostic delay was in the range of 9.5 to 23 years, the median time of which was 11.8 years. This was a more critical delay compared with those receiving other types of diagnoses (1). The LQTS diagnosis has to be assumed when the patient has ventricular tachycardia episodes. Standard 12 lead ECG can approve a lengthy QT interval. Prolonged QT interval is described as QTc> 0.47 to 0.65 (2). Eleven genes related to LQTS are known. There exist more than 600 mutations so far within these genes, pointing to the notable genetic heterogeneity (3, 4). The various genetic subtypes of LQTS cause clinical defects, with their own presentation patterns, prognosis, ECG disorders, and selective management (5, 6). The tendency for LQTS to mimic other circumstances has been 1 in secondary hypoxia and convulsions. LQTS misdiagnosis as epilepsy was introduced in 1983; since then

other cases have been reported (7, 8). However, the LQTS diagnosis is still unknown. Rapid LQTS diagnosis is necessary since the death rate can be decreased with developed interventions. Beta-Blocker treatment has been proven to be efficient in treating LQT2 and LQT1 (9). Left-sided cardiac sympathetic denervation is beneficial in patients with higher risk (10). Overdrive cardiac pacing is also helpful for some patients. With regard to high-risk patients, the presence of implantable cardioverter defibrillators (ICD) with LQTS has decreased the death rate (11). The results demonstrate that delayed diagnosis of LQTS is still popular; 39% of the patients experience the delay between initial diagnosis and disease presentation.

Case presentation

Our patient is a 24-year-old right handed woman admitted for video EEG monitoring in an attempt to better define her seizures. The patient's first seizure occurred at the age of eight without any provocation. The details of this spell are not obvious. Later on, she experienced these spells at the age of 14. Her seizures were characterized by (a) an uncomfortable aura of anxiety and nausea (b) followed by a short period and obvious impairment of consciousness. However, rarely, these seizures were followed by generalized clonic activity; postictally, she was alert without any postictal confusion. Seizure frequency was about 2 to 3 per year and often gets worse by fear, stress, and hunger. Since she was diagnosed with atypical seizure semiology and her serial EEGs were normal, the clinical history and the ancillary studies were not helpful. The patient was admitted for assessment of non-epileptic paroxysmal events. The patient was considered to be the product of a normal gestation. Her birth was spontaneous after the typical 9 month gestation period without any complication. The early development was normal. She walked at one year of age and could talk at the age of two. She had no history of febrile seizure, CNS infection, or CNS trauma with loss of consciousness. Her first seizure occurred at the age of eight; suddenly, she felt a shock like sensation in her head and then she fell down. It is certain that she did not have tonic clonic activities, but the details of this episode were not obvious. She mentioned the history of epileptic seizures in her brother who had experienced SUDEP six months ago. The patient was living with her parents, having no job, and no history of tobacco, alcohol, or illicit drug use.

Previous work up

(a) Brain MRI: The latest MRI (Haghighat Imaging Center) on November 13, 2016 was normal. (b) Interictal SPECT scans were not done. (c) Neuropsychological evaluation was not available.

Prior medication trials

Depakine, carbamazepine

Current medications

Depakine 500 mg/d, Carbamazepine 200 mg TDS. Routine labs, including FBS, Ca, P, alkaline phosphatase, SGPT, SGOT, and U/A were within normal limits. VDRL was negative. Her systemic exam was within normal limits. The patient was alert and oriented x3. Her mental status was within normal limits. Visual fields on confrontation were intact. Pupils were equal and reactive to light with extra ocular movement intact. There was no facial sensory or motor asymmetry. Her palate and uvula rose symmetrically, and her tongue protruded on midline. Force and tone assessment were normal. Reflexes were 2+ and symmetric. Cutaneous plantar reflexes were in flexion. Cerebellar functions including finger-to-nose and heel-to-sheen testing were normal. Gait and tandem walking were normal. A video EEG monitoring session was scheduled from 1/3/2017 to 8/3/2017, using modified international 10-20 system. Silverman’s true anterior temporal electrodes

(T1, T2) and T9 and T10 were also applied. Recorded EEGs were reviewed in bipolar and referential montages using reformatting. Computerized spike and seizure detection systems were employed. At first, Depakine was discontinued. Then Carbamazepine was tapered and discontinued. To provoke her habitual seizures, sleep deprivation was also performed from the third night of her admission.

Interictal findings

(A) Awake EEGs: The basic rhythm of her resting records consisted of medium amplitude well-organized 10 CPS alpha activity properly attenuated by eye opening. There were no lateralized, neither focal nor abnormal, paroxysmal discharges throughout her awake EEGs. (B) Sleep EEGs: The sleep pattern was proper in her sleep records. Neither lateralized nor focal or paroxysmal discharges were seen in her sleep EEGs. One paroxysmal event was recorded during this monitoring session. She was suddenly awakened from sleep, and immediately her cardiac rhythm became irregular, and ventricular fibrillation and then ventricular tachycardia appeared. Also, generalized jerky, clonic movements, and vocalization occurred. At this moment, CPR was started quickly, but she became asystolic for 30 seconds. While undergoing CPR, her heart rate returned. She was awake after a few minutes. Her awake and sleep EEGs were essentially normal. Her EEGs before this event were essentially normal, but during this event, her EEGs showed abnormal decreased amplitude without any epileptiform discharges. We also paid attention to her EKG lead during this period. Long QT interval was observed before starting this event that evolved to ventricular tachycardia and then ventricular fibrillation and asystole for 30 seconds. A cardiologic consult was immediately requested. Standard 12-lead EKG showed abnormal prolonged QT interval (QTc: approximately 550 msec). Chest x-ray and echocardiography of the patient were normal. Consultant cardiologist recommended emergency CCU admission. She was admitted to the CCU, and continuous holter monitoring was done for her, and she was scheduled for implantable cardioverter defibrillator (ICD). During this video-EEG monitoring, one convulsive cardiac syncopal attack was recorded (Fig.1: LTM record).

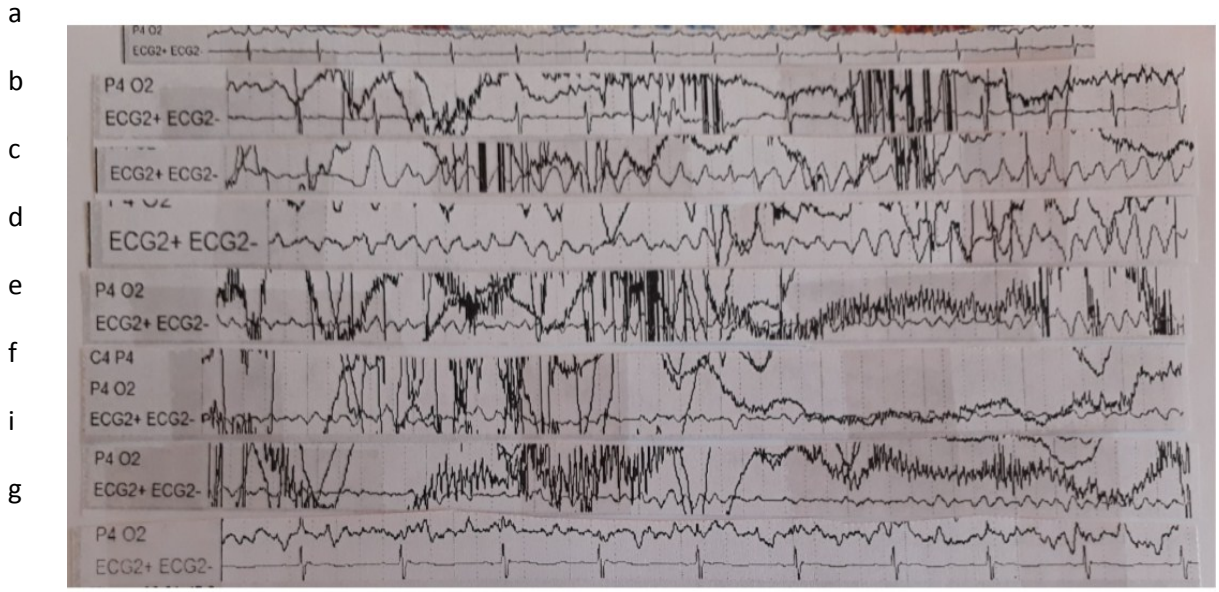

Figure 1. a) the patient's ECG had normal rhythm. b) QT interval was prolonged. c) At this moment, ventricular tachycardia occurred. d) Ventricular fibrillation occurred immediately after ventricular tachycardia. e), f), i) ventricular asystol was appeared and lasted for 20 seconds. g) Normal cardiac rhythm returned.

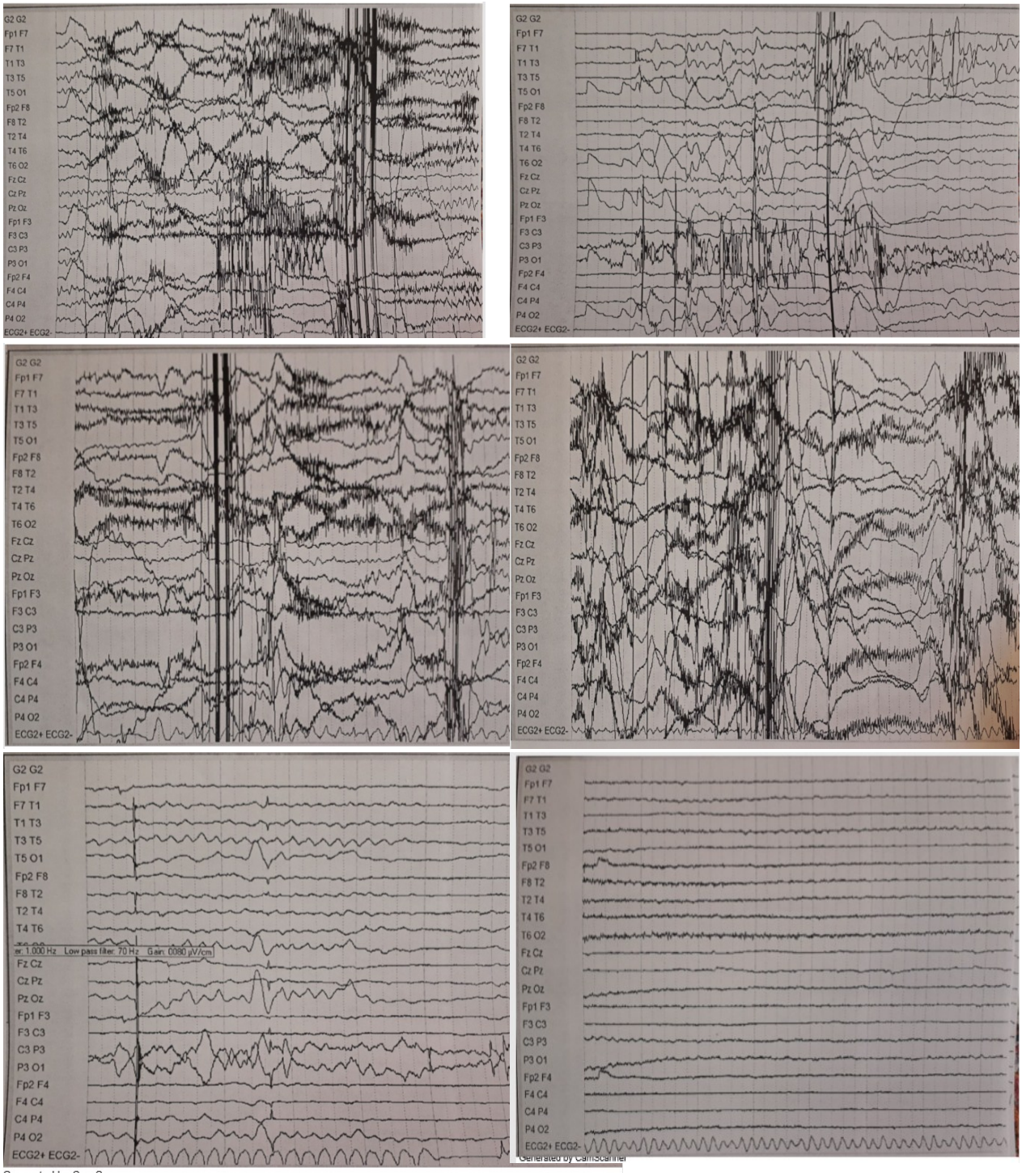

Figure 2. Long-term video- EEG monitoring was recorded one attack of ventricular tachycardia that progressed to ventricular fibrillation then cardiac asystol. Simultaneous EEG recording was shown generalized theta and delta slowing that turned to electrical silence during cardiac asystol.

After cardiologic consultation, long QT syndrome was diagnosed, and ICD was done for her. Her antiepileptic drugs were discontinued permanently.

Discussion

LQTS may be manifested as epilepsy convulsion or seizure. However, abrupt mortality can be prevented through early diagnosis. This syndrome can simply be

recognized and appropriate management can be done if an EKG is taken. The LQTS history shows death rate more than 20% one year after the first syncopal incidence, with about 50% mortality in 5 years. Proper intervention can considerably decrease the morbidity and death rate, making rapid diagnosis necessary (1).

Conclusion

LQTS delayed diagnosis is seen frequently. Symptoms are assigned to alternative diagnoses, the most common of which is seizure. Epileptic patients experience a lengthy diagnostic delay. Although ECGs are requested frequently, wrong interpretations delay the correct diagnosis (12). Hence, emergency physicians who study seizure and syncope are required to possess a great index of suspicion, considering the potentially preventable mortality of LQTS (1).

Ethical issues

Not applicable.

Authors' contributions

All authors equally contributed to the writing and revision of this paper.

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