CASE REPORT

Mexiletine Suppressed Recurrent Ventricular Tachycardia Triggered by Hemodialysis in an Old Patient with LQT2

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Summary
The first onset of cardiac event of long QT syndrome (LQTS) was at young age and caused by emotional or physical triggers. We presented a 64-year-old woman who experienced recurrent ventricular arrhythmia after hemodialysis initiation because of end-stage renal disease. Persistent prolonged QTc interval and diagnosis of inherited LQT2 were missed at her first 3 years of hemodialysis. The patient was beta-blocker nonresponder for ventricular arrhythmias suppression and experienced multiple ICD discharge. We reported an inherited LQT2 case with uncommon clinical manifestations and the successful experience of mexiletine use in such a patient.

Key words: Long QT syndrome, Ventricular arrhythmia

Inherited long QT syndrome (LQTS) is a potential lethal cardiac channelopathy and is associated with life-threatening ventricular tachycardia (VT) or torsades de pointes (TdP) usually triggered by physical or emotional stress. Most patients experience first onset of cardiac event before 40 years old. It is one of causes of autopsy-negative sudden cardiac death (SCD) in young patients.1-6 Beta-blocker is the first-line drug recommended for all LQTS patients regardless of genotype. Mexiletine, one of the Na+ channel blockers, is recommended as add-on therapy limited for LQT3.2 Implantable cardiac defibrillator (ICD) is recommended for patients with high SCD risk, but it sometimes can trigger norepinephrine release, subsequently resulting in electrical storms. Left cardiac sympathetic denervation (LCSD) is a promising mini-invasive procedure to reduce cardiac events demonstrated by small sample size studies.6 Here, we reported an LQT2 patient, who experienced first onset of cardiac event after hemodialysis initiation at the age of more than 60 years, and recurrent syncope and ICD discharge occurred even under compliant bisoprolol treatment but were finally suppressed by mexiletine.

Case Report
A 64-year-old woman was admitted to our department due to convulsive syncope 1 hour after hemodialysis in 2014. She experienced palpitation for seconds and quickly progressed to loss of consciousness which was resuscitated by minutes through cardiopulmonary resuscitation performed by her son. By history taking, her first onset of syncope was during her first hospitalization in the nephrology department for oliguria and elevated creatinine (668 umol/L) in 2011 at which she was diagnosed as having end-stage renal disease and initiated hemodialysis. Her medical records in 2011 revealed that VT-associated syncope occurred twice during that hospitalization and was thought to have resulted from prolonged QT interval secondary to hemodialysis-induced hypokalemia (K+ = 2.9 mmol/L). Potassium supplement terminated VT, but further ECG monitor was overlooked. From 2011 to 2014, she sometimes felt palpitation and presyncope symptoms. Additionally, she had hypertension for more than 20 years, treated by amlopidine and irbesartan. She denied any family history of cardiac disease or sudden death. On admission, her vital signs were stable, ECG revealed a short run of VT (Figure 1), and laboratory tests revealed serum potassium was 3.1 mmol/L, magnesium was 0.85 mmol/L, brain natriuretic peptide was 6200 pg/mL, and cardiac troponin I was 39.2 ng/L. Transthoracic echocardiography (TTE) showed normal cardiac structure and function with the measurement of left ventricle end-diastolic diameter (LVEDD) of 46 mm and left ventricular ejection fraction (LVEF) of 66%. After bisoprolol (10 mg orally once a day) administration and potassium and magnesium supplied to a high level normal (K+ > 4.5 mmol/L, Mg2+ > 0.8 mmol/L), episodes of nonsustained VT (NSVT) decreased, but QTc interval was still prolonged (QTc = 583 ms, Figure 2), and syncope recurred. The patient was highly likely to have inherited LQTS with the high risk of sudden cardiac death, ICD was implanted, and atrial pacing rate of 80 bpm was programmed to expect to minimize cardiac events. The patient was discharged with a prescription for bisoprolol and potassium supplement, and frequent electrolyte monitoring was planned especially during hemodialysis.
The patient was admitted again after 4 months with complaints of multiple recurrent palpitation, syncope, and ICD discharge, most of them occurred during hemodialysis, sometimes at rest. Medical records revealed that blood potassium fluctuated around 4.5 mmol/L. She gradually developed exercise intolerance and paroxysmal nocturnal dyspnea. After admission, coronary angiography was performed and excluded the diagnosis of coronary artery disease. TTE showed LVEDD enlarged to 56 mm and LVEF reduced to 49%. Bedside electrocardiograph monitor revealed intermittent episodes of VT and frequent premature ventricular beats (PVB). After empirical intravenous bolus administration of lidocaine 100 mg, the frequency of PVB and VT were decreased. By literature review, we tried to prescribe oral mexiletine (150 mg three times a day) on her, and episodes of VT were further suppressed even during hemodialysis. The patient was discharged after having stabled for days. Oral mexiletine and bisoprolol were taken as long-term regimen. Days after discharge, the gene test results came back and showed two mutations (c.656T>A; c.1661delA and c.1660delT) located in the KCNH2 gene, indicating the diagnosis of LQTS2.

TTE revealed LVEDD recovered to normal size of 50 mm and LVEF increased to 71% 6 months after discharge, and 24-hour Holter monitoring revealed no VT and only one PVB per 24 hours, and QTc interval was 425 ms. Atrial pacing rate was reduced to 60 bpm. She was doing well without syncope recurrence and better tolerated regular hemodialysis during the subsequent 4-year follow-up.

Figure 1. ECG showed ventricular tachycardia (black arrow) after heart arrest during the first admission to cardiology department in 2014. ECG, electrocardiogram.

Figure 2. ECG showed prolonged QT interval of 560 ms (two-way arrow) and HR of 65 bpm, and then the calculated QTc interval was 583 ms. The ECG was obtained after potassium supplementation to a level of > 4.5 mmol/L. ECG, electrocardiogram; HR, heart rate; bpm, beat per minute.
Inherited LQTS is a rare disease with heterogeneous phenotypes. The first onset of cardiac event at age of more than 60 years and triggered by secondary causes are uncommon. Clinicians should enhance the awareness and ability to differentiate inherited from acquired LQTS in clinical practice. Mexiletine might also be an add-on therapy in LQT2 patients when beta-blocker was insufficient to suppress VT-associated events supported by weak evidence.

References