Recurrent Ventricular Tachycardia Due to Long QT Syndrome

Rambabu Singh, Kshitiz Nath, Archana

Abstract

Long QT syndrome (LQTS) is a rare inherited heart condition in which delayed repolarization of the heart following a heartbeat, increases the risk of episodes of Torsades de pointes (TdP, a form of irregular heartbeat that originates from the ventricles). These episodes may lead to palpitations, fainting, and sudden death due to ventricular fibrillation. Episodes may be provoked by various stimuli, depending on the subtype of the condition. We are reporting a case of 37 years old male whom we diagnosed to have long QT syndrome on the basis of clinical and ECG findings.

Introduction

The long QT syndrome (LQTS) is a primary electrical disease which is characterized by prolongation of the disordered ventricular repolarization of the corrected QT on the surface ECG. This disease is caused by various mutations in at least seven genes coding for cardiac ion channels. There are seven known types of inherited LQTS. The most common ones are LQTS 1, 2, and 3. In LQTS 1, emotional stress or exercise (especially swimming) can trigger arrhythmias. In LQTS 2, extreme emotions, such as surprise, can trigger arrhythmias. In LQTS 3, a slow heart rate during sleep can trigger arrhythmias.

Long QT syndromes can be associated with

The congenital long QT syndrome (LQTS) is characterized by abnormally prolonged ventricular repolarization due to inherited defects in cardiac sodium and potassium channels, which predispose the patients to syncope, seizure like activity, ventricular arrhythmias, and sudden cardiac death. We describe one case that fulfilled the electrocardiographic, historical diagnostic criteria and epinephrine stress test suggestive of LQT syndrome.

Case Report

We report a case of 37 years old male resident of Jhansi, who suffered from sudden onset unconsciousness for 4-5 minutes. This episode was not associated with any abnormal body movements, frothing from mouth, any urinary or fecal incontinence, or limb weakness. Patient came to our hospital on 15th June at 3:15 am. Patient also had a history of 4-5 similar episodes in past. These episodes, as described by patient himself, were precipitated on doing some heavy work. His BP was 100/70 mm Hg, PR -98/min, chest had bilateral crepitation. He was admitted for evaluation. On 15 June at 8 am patient again become unconscious, an urgent ECG was done which showed ventricular tachycardia with a rate around 360/min. (Figure 1).

Urgent DC shock of 150 J was given and the VT reverted. Patient again developed VT at 11 am, 11:45 am, 1:30 pm and 3:00 pm; every time he was reverted by giving DC shock of 150 to 250 J and he was started on injection Amiodarone 150 mg IV stat over 10 min and then 360 mg over 6 hours and was further maintained on oral treatment.

ECG of our patient showed prolonged QT interval with corrected QT, of about 500-510 msec and notched T waves (Figures 2 and 3). Hemogram, cardiac enzymes, liver profile, renal profile and electrolytes were normal.

Patient was discharged on propranolol and was advised for ICD implantation. On subsequent follow-up, patient did not undergo ICD insertion and remained symptom free for 2 years, but in Sept 2015, he again developed similar kind of episode (Figure 4) which was again reverted by DC shock. Patient was again advised for ICD implantation and was referred to higher center.
mortality to <5 %.2  

Both, which has reduced long-term or surgical antiadrenergic therapy or poor prognosis has been significantly varies among different families. This affected. 3 While cardiac events are more LQTS is influenced largely by the gene value of 9 points.

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The new diagnostic criteria are listed in Table 1, with relative points assigned to various ECG, clinical and familial findings. The score ranges from a minimum value of 0 to a maximum value of 9 points.

The clinical course of the congenital LQTS is influenced largely by the gene affected. 3 While cardiac events are more frequent and occur at a younger age in patients with LQT1 and LQT2, they are potentially more fatal in patients with genotype LQT3. Patients with LQT1 and LQT2 genotype typically benefit from high dose beta-blocker therapy. 4 However, patients with LQT3 are at higher risk at lower heart rates and potentially may benefit from pacemaker therapy. In addition, they shorten their QT-interval more with sodium channel blockers.

Provocative tests using catecholamine or exercise testing have long been used to unveil concealed LQT1 syndrome 5. An epinephrine test may be applied not only for unmasking silent mutation carriers with LQT1 syndrome but also for predicting genotypes.

Facilities for genetic analysis are not easily available. However, in view of the growing importance of genotype specific treatment of this potentially fatal syndrome, one can utilize the ECG criteria and epinephrine QT stress test as a reliable indicator of the underlying genotype and accordingly tailor the management.

Discussion

The idiopathic prolongation of the QT interval and by the occurrence of life-threatening tachyarrhythmias, particularly long QT syndrome (LQTS) is a congenital disease with frequent familial transmission, characterized primarily by an association with emotional or physical stress. 1 Among untreated symptomatic patients, lethality is high, with 20% mortality in the first year after the initial syncope and approximately 50% within 10 years; however, the risk of death varies among different families. This poor prognosis has been significantly improved by the use of pharmacological or surgical antiadrenergic therapy or both, which has reduced long-term mortality to <5 %. 2

In its most characteristic presentation, with obvious QT prolongation and stress-induced syncope, the diagnosis of LQTS is quite straightforward for physicians aware of the disease. In cases of borderline QT prolongation and/or absence of symptoms, however, a correct diagnosis may be more difficult.

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Provocative tests using catecholamine or exercise testing have long been considered to unmask some forms of congenital LQTS. Recent preliminary data by Ackerman et al. have suggested the usefulness of an epinephrine test to unveil concealed LQT1 syndrome 5. An epinephrine provocative test should only be done by cardiologists, under enough preparation of intravenous beta-blockers and direct cardioverter for unintentionally induced ventricular fibrillation.

Both experimental and clinical studies have suggested a differential response of action potential duration (APD) and QT interval to sympathetic stimulation among LQT1, LQT2, and LQT3. 12 Persistent and paradoxical prolongation of APD and QT interval at steady state conditions of catecholamines is reported in LQT1 syndrome. 6 In LQT2 syndrome, catecholamines are reported to initially prolong but then abbreviate APD and QT interval, probably because of an initial augmentation of INa-Ca and a subsequent stimulation of IKs. In contrast to the LQT1 and LQT2 syndromes, catecholamines are reported to constantly abbreviate APD and QT interval as a result of a stimulation of IKs in the LQT3 syndrome, because an inward late sodium current (INa) was augmented in this genotype. The epinephrine test may be applied not only for unmasking silent mutation carriers with LQT1 syndrome but also for predicting genotypes.

Facilities for genetic analysis are not easily available. However, in view of the growing importance of genotype specific treatment of this potentially fatal syndrome, one can utilize the ECG criteria and epinephrine QT stress test as a reliable indicator of the underlying genotype and accordingly tailor the management.

Conclusion

The availability of effective therapy for this often lethal disease emphasizes the importance of early and accurate diagnosis. Unfortunately, there is often delay in the diagnosis of LQTS, and patients with syncope are often misdiagnosed, most commonly as affected by a seizure disorder.

References