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Letter to The Editor

An Unusual Cause for the Sudden Death: Short QT Syndrome

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The short QT syndrome (SQTS) is a cardiac channelopathy associated with a propensity for cardiac arrhythmias in persons with a structurally normal heart [1-3]. Although victims of sudden cardiac death (SCD) tend to fit in certain categories, the cause of SCD may be sometimes completely unpredictable. Herein, we present the case of a young man with a history of succesfully resuscitated SCD and recurrent syncopal attacks with unknown etiology.

A 20-year-old man presented to our cardiology out-patient clinic due to several episodes of transient loss of conciousness not associated with emotional stress or exercise during the last 6 months. The patient was afebrile with blood pressure of 110/70 mmHg, heart rate of 60 bpm, and oxygen saturation of 99% by pulse oximetry. Cardiac examination showed a regular heart rhythm with no murmurs. The rest of the physical examination was unremarkable. Electrocardiography (ECG) demonstrated a sinus rhythm, 55 bpm; PR interval: 132 ms; QRS duration: 85 ms; QTc: 313 ms (Bazzet formula); and J-point to T-wave-peak interval: 140 ms (Figure 1).

Biochemical tests were within normal limits. Cardiac biomarkers were negative. Transthoracic echocardiography displayed a normal heart with ejection fraction of 69% and no evidence of structural disease. The father reported one episode that took place approximately 1 year ago in which the patient was taken to the nearest hospital, a 3-5 minute drive. He was unconscious and unresponsive to stimuli and cardiopulmonary arrest in asystole was confirmed by the physician at the hospital. Cardiopulmonary resuscitation (CPR) had been performed with total CPR time of 30 minutes. He had been monitored and treated in the intensive care unit with undetermined etiology and discharged after 25 days without any neurological deficit. At that time the patient underwent a coronary angiography, which was found to be normal. He had no other known disease or history of sudden death in the family. He denied smoking, the use of illicit drugs or medications. Twenty four-hour Holter monitoring was carried out in our clinic and showed infrequent premature ventricular contractions with no repetitive patterns. A likely diagnosis of SQTS is made after other precipitating factors and structural heart disease have been excluded, and ECGs have showed a shortened QT interval. Based on diagnostic criteria by Gollob et al, the patient is categorized as high-probability SQTS with a score of 5 or more. Malignant arrhythmias due to SQTS have been considered as a primary cause of the syncopal attacks and sudden cardiac death. According to the current guidelines, an implantable cardioverter-defibrillator is indicated but the patient refused, and therefore, he was treated with quinidine, with uneventful follow-up of 6 months. Genetic testing for both the patient and his family could not be performed due to financial and logistic reasons.

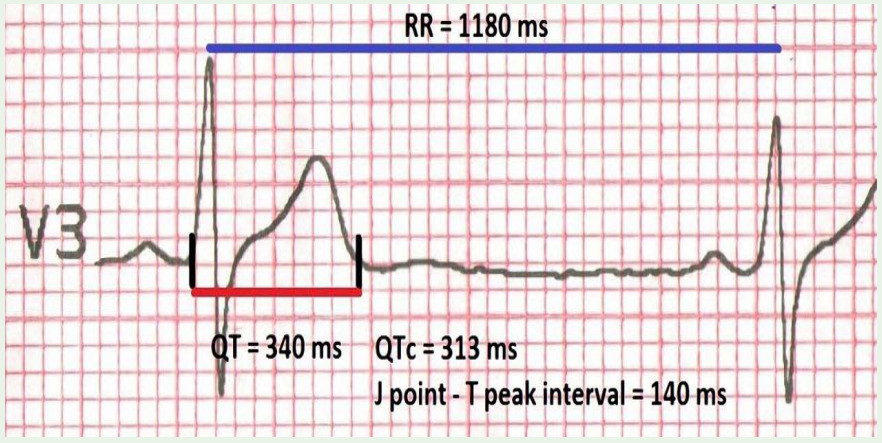

Figure 1: Electrocardiography shows short QT interval.

SQTS is a recently defined entity included in the group of inherited primary electric disorders [1-3]. Since its first report in 2000, six different genes encoding various cardiac ion channels have been identified in the pathogenesis of SQTS [2]. It involves the relation of a significantly shortened QTc interval on the surface ECG with atrial fibrillation and sudden cardiac death [1-3]. However, the presence of a short QT interval alone is not always predictive of an elevated cardiac arrhythmic risk [3]. Therefore, diagnostic criteria for SQTS have been developed based on a point system, which includes QTc, Jpoint-Tpeak interval, clinical history, family history, and genotype. In this scoring system, patients are deemed high-probability (> or equal to 4 points), intermediate probability (3 points) or low probability (2 or less points) [3].

In a young person who has no alternative diagnosis for the life-threatening events, SQTS should be kept in mind by virtue of its possible lethal arrhythmogenic effect. In suspected cases of SQTS, diagnostic criteria can be used to facilitate diagnostic assessment.

Disclosure

This case has been presented as a poster presentation at the 31th Turkish Cardiology Congress with International Participation, which was held in Antalya, Turkey, on October 22-25, 2015.

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