We present a case of a 23-year-old woman with no major cardiovascular risk factors, who presented with recurrent complete loss of consciousness during her driving-license examination. The patient was transferred to the emergency room, where she had the next episode of syncope with ventricular fibrillation (VF). Her ECG before VF revealed ST-segment elevation in leads II, III, aVF, and V3–V6 (Figure 1), although the patient did not mention any chest pain. On admission, her echocardiogram showed no abnormalities, her left ventricular ejection fraction (LVEF) was 70%, and myocardial necrosis markers were negative. Several days later, the patient was referred for coronary angiography, which was free of coronary artery stenosis; however, the second contrast pass and catheter positioning revealed long-lasting diffused spasm of left main, proximal part of left descending and circumflex arteries, which persisted despite intracoronary infusion of nitroglycerin. An ECG taken at this time demonstrated ST-segment elevation in the V2–V6 leads, which was followed by the occurrence of VF an hour later and recovered in 2 hours. Serially obtained blood samples showed a significant increase in cardiac troponin I (maximum, 101.6 ng/mL). On the next day, ECG showed hypokinesis of apical segments of anterior and interventricular wall, and LVEF was 40%. Those abnormalities resolved 2 weeks later, and a subsequent echocardiogram demonstrated LVEF of 60% and no wall motion abnormalities. Thereafter, an implantable cardioverter-defibrillator (Lexos DR, Biotronik) was implanted, and combination therapy with metoprolol (50 mg/d) and verapamil (60 mg/d) was initiated. At discharge, her LVEF was 40%. Those abnormalities resolved 2 weeks later, and a subsequent echocardiogram demonstrated LVEF of 60% and no wall motion abnormalities. Thereafter, an implantable cardioverter-defibrillator (Lexos DR, Biotronik) was implanted, and combination therapy with metoprolol (50 mg/d) and verapamil (60 mg/d) was initiated. At discharge, her ECG showed QT prolongation up to 480 ms (QTc, 570 ms), with negative T waves in the I, aVL, and V3–V5 leads. An ECG taken at 10-month follow-up showed positive T wave with a QT of 440 ms (QTc, 550 ms; Figure 2). At any time, the patient did not receive any drugs prolonging the QT interval.

The patient’s family history was positive for sudden cardiac death and sudden infant death syndrome. We performed genetic testing, and we found the R591H mutation in the KCNQ1 gene in the patient, her mother, and siblings. The clinical evaluation, particularly QTc prolongation during exercise tests, confirmed clinical diagnosis of long QT syndrome (LQTS) in all genetically affected family members.

Discussion

To our knowledge, this is the first report on spontaneous ST-segment elevation preceding an episode of VF in LQTS. Generally, cardiac events in LQTS occur either during exercise and emotional stress (LQT1 and LQT2 types) or at rest (LQT3), whereas ST-segment elevation may precede episodes of VF in patients with vasospastic angina or stress-induced cardiomyopathy (so-called Takotsubo syndrome [TTS]). Some clinical features of our patient may resemble TTS. First, the syncopal event was triggered by emotional stress. Second, ECG at admission showed ST elevation and negative T waves with QT prolongation, and coronary angiography showed no organic stenosis. Finally, wall motion abnormalities detected in echocardiography resolved within 2 weeks. On the contrary, apart from the fact that TTS usually affects older women in their 6th decade of life, the echocardiogram performed in our patient after the syncopal episode presenting with ST elevation was normal, and wall motion abnormalities appeared only after coronary spasm induced at coronaryography. Moreover, the pattern of echocardiographic abnormalities observed in our patient was not typical for TTS, which presents usually with hypokinesis of apex and hyperkinesis of basal LV segments. Recently, Sasaki et al described a case of a young woman with LQTS in whom TTS developed after a syncopal attack caused by torsade de pointes. It is possible that, in LQTS patients, a release of catecholamines in response to emotional stress may serve not only as a trigger for ventricular tachyarrhythmias, but by activation of myocardial β-adrenoreceptors, it may lead to catecholamine-mediated myocardial stunning. Catecholamines may also produce coronary vasoconstriction in susceptible patients; however, the clear relation between vaso-
spastic angina and LQTS has not been proven. Interestingly, Aizawa et al demonstrated that intracoronary acetylcholine infusion induces QT prolongation in LQTS patients, suggesting a theoretical possibility of provoking torsade de pointes by coronary vasospasm.

The rationale for treatment of the patient presenting with LQTS and coronary vasospasm remains uncertain, because β-blockers, which are the mainstay of LQTS therapy (particularly in LQT1 type), by leaving the vasoconstricting effects of α-receptors unopposed, may produce or aggravate vasospasm in susceptible patients. It seems probable that in some LQTS patients an emotional stress may induce both vasospasm and arrhythmias; thus, the administration of β-blockers might be beneficial, despite any possible adverse effect on coronary arteries.

Figure 1. The ECG before ventricular fibrillation, showing ST-segment elevation in leads I, II, III, aVF, and V3–V6, with ST-segment depression in leads aVR, aVL, and V1.
Figure 2. The ECG after 10-month follow-up, showing negative T wave in the III and aVF leads and pathological Q wave in the aVL lead. QT, 440 ms; QTc, 550 ms.
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Disclosures
None.

References


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