Cardiac Arrest Associated with Both an Anomalous Left Coronary Artery and KCNE1 Polymorphism

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Summary

A 14-year-old boy collapsed suddenly after a basketball game and was transported to our hospital after recovering from ventricular fibrillation by an automated external defibrillator. He had experienced loss of consciousness twice and has been examined for suspected long-QT syndrome at another hospital. The 12-lead electrocardiogram on admission revealed a prolonged QTc interval of 480 milliseconds. After the patient recovered without any sequelae, computed tomography revealed an anomalous left coronary artery arising from the opposite sinus of Valsalva and coursing between the aorta and the pulmonary artery. Furthermore, genetic testing identified a KCNE1-D85N abnormality. An anomalous coronary artery is one of the major causes of sudden death in young people; therefore, surgical revascularization is recommended for left coronary arteries arising from the contralateral sinus and coursing between the aorta and the pulmonary artery, regardless of myocardial ischemia. Transient myocardial ischemia may have exaggerated the instability from the arrhythmic substrate, even though KCNE1-D85N abnormalities alone are not thought to cause fatal arrhythmias. Besides routine electrocardiography, further examinations, including imaging and genetic testing, can characterize the pathophysiology of fatal cardiac disease.

Key words: Long-QT syndrome, Sudden cardiac death

Here, we present the case of a 14-year-old boy who experienced cardiac arrest associated with both an anomalous left coronary artery (LCA) and polymorphism in the β-subunit of delayed rectifier potassium channel. Coronary artery anomaly (CAA) is one of the major causes of sudden death in young people, so surgical revascularization is recommended for LCAs arising from the contralateral sinus and coursing between the aorta and the pulmonary artery, regardless of myocardial ischemia. Meanwhile, the KCNE1 variants (for LQT5) identified in this patient can modify both KCNQ1- and KCNH2-coded channel currents, but KCNE1-D85N abnormalities alone are not thought to cause fatal arrhythmias.

Case Report

A 14-year-old boy collapsed suddenly after a basketball game and was transported to our hospital after ventricular fibrillation (VF) was successfully treated using an automated external defibrillator (Figure 1A). He had experienced loss of consciousness twice since the age of 12: during jogging two years and after a basketball game one year previously. Moreover, he had often experienced vomiting or palpitations after sports activities. He had no other past histories and no family history of either cardiovascular disease or sudden death. A definitive diagnosis had not been made, although he has been examined for suspected long-QT syndrome (LQTS) at another hospital.

The 12-lead electrocardiogram (ECG) revealed a prolonged QTc interval of 480 milliseconds on admission (Figure 1B). After recovering without any sequelae, he received an implantable cardioverter-defibrillator. Computed tomography (CT) revealed an anomalous LCA arising from the contralateral sinus of Valsalva and coursing between the aorta and the pulmonary artery (Figure 1C). Additionally, coronary angiography demonstrated a pulsating compression of the left main trunk from both arteries (Figure 1D). Treadmill exercise stress testing 13 days after the cardiac arrest showed a marked ST-segment change indicating severe myocardial ischemia (Figure 1E), despite no redistribution with no ST-segment change in exercise scintigraphy with thallium-201 at 35 days. Furthermore, genetic testing identified a KCNE1-D85N abnormality (Figure 2). Finally, the patient underwent unroofing and LCA bypass grafting surgery. After surgery, his vomiting and palpitations after exercise disappeared. He showed no ischemic change on exercise stress test and re-
Figures 1. A: AED monitor showing VF. B: 12-lead ECG showing a prolonged QTc interval (QTc, 480 milliseconds). C: LCA arising from the contralateral sinus and coursing between the aorta and the pulmonary artery. D: Compression of the LMT from both the aorta and the pulmonary arteries between the diastolic and the systolic phase. E: Treadmill exercise test showing ST-segment depression in V4-6, II, III, aVF, and ST elevation in aVR.

Figures 2. Sequence chromatograms of KCNE1-D85N from a case [top: Asp85Asn (GAT>AAT) (red arrow)] and control [bottom: Asp (GAT)].

required no shock therapy from his defibrillator without any medication during the three-year follow-up. Before surgery, the baseline QT interval, QTc interval (corrected QT interval using Bazett’s formula), and QT dispersion (maximum QT interval minus minimum QT interval) were 440, 402, and 65 milliseconds, respectively. The corresponding values at peak exercise were 280, 452, and 60 milliseconds, respectively. After the surgery, the QT interval, QTc interval, and QT dispersion at baseline were 458, 432, and 62 milliseconds, respectively, and those at peak exercise were 272, 460, and 56 milliseconds, respectively.

Discussion

CAA is one of the major causes of sudden death in young people, in addition to hypertrophic cardiomyopathy and myocarditis. Recent advancements in noninvasive imaging facilitated the identification of CAAs before causing serious events. Coronary CT angiogram (CTA) or magnetic resonance angiography provides accurate information regarding the origin, course, destination, and relationship to surrounding structures of CAAs. It is reported that the prevalence of coronary arteries arising from the
opposite sinus on CTA is 1.7%, including 45% of the cases discovered incidentally. Surgical revascularization is recommended for LCAs arising from the contralateral sinus and coursing between the aorta and the pulmonary artery, regardless of myocardial ischemia (Class I recommendation). Meanwhile, LQTS is a disorder that is characterized by repolarization abnormalities in the heart, leading to torsade de pointes, syncope, and sudden death. KCNE1 variants (for LQT5) can modify both KCNQ1- and KCNH2-coded channel currents, but KCNE1-D85N abnormalities alone are not thought to cause fatal arrhythmias. No increase in the heterogeneity of ventricular repolarization evidenced by the QT dispersion exercise testing may support this finding.

This is the first case developing VF due to a CAA accompanied by a polymorphism in the β-subunit of delayed rectifier potassium channel. Similar to this patient, some VF survivors who were diagnosed with LQTS using ECG might develop VF due to another fatal cardiac disease. Conversely, any triggers or additional risk factors for fatal arrhythmia might be latent in patients with CAs who die suddenly, because not all the subjects with CAs die young. In this case, transient myocardial ischemia may have exaggerated the instability from the KCNE1 variant. Furthermore, no myocardial ischemic change was identified at 35 days after the cardiac arrest on exercise stress scintigraphy with thallium-201, despite a marked ST-segment change at 13 days on treadmill exercise stress testing. A temporary negative result of stress examination does not exclude a potentially lethal CAA. Although coronary vasoconstriction has been reported to be associated with coronary anomalies, we did not perform a spasm provocation test.

Both transient myocardial ischemia and arrhythmia substrate might have been associated with VF in the present case. Besides routine ECG, further examinations, including imaging and genetic testing, are needed for VF survivors.

Disclosures

Conflicts of interest: None.

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