A fatal combination in a young lady: Long QT syndrome and coronary artery anomaly

Emir Karacaglar,[1] Alp Aydinalp,[1] Mehmet Coskun,[2] Haldun Muderrisoglu[1]

[1]Cardiology Department; [2]Radiodiagnostic, Baskent University School of Medicine, Bahcelievler/Ankara, Turkey

Abstract

Anomalous origin of coronary arteries is also a relatively rare congenital malformation and has been reported as the cause of angina pectoris and arrhythmia. Long QT syndrome (LQTS) is a rare inherited arrythmogenic disease characterized by susceptibility to life-threatening arrhytmias andsudden cardiac death. We present a 36-year-old patient in whom two rare anomalies coexist and treated succesfully with β-blocker therapy.

Introduction

Long QT syndrome (LQTS) is a rare inherited arrhythmogenic disease characterized by susceptibility to life-threatening arrhythmias and sudden cardiac death. Anomalous origin of coronary arteries is also a relatively rare congenital malformation and has been reported as the cause of angina pectoris, arrhythmia, syncope and fatal myocardial infarction.[1,2] We present a patient in whom two rare anomalies coexist.

Case Report

A 36-year-old female was admitted with palpitation and hypertension. She has normal functional capacity. Her blood pressure was 145/79 mmHg on admission. She had no medication. The only relevant feature in her history was palpitation episodes especially during swimming for a few years and she was unaware of her grade 1 hypertension. There was no sudden death or cardiac arrest or syncope both in her and her family's medical history. On her electrocardiogram (ECG) we determined long QT pattern with QTc prolongation of 482 msn (Figure 1A). In addition we detected no QT interval shortening during the treadmill exercise test. She was given 50 mg metoprolol daily and underwent 24 h Holter ECG monitorization which revealed no arrhythmia record. No structural abnormalities were found on her transthoracic echocardiography. According to her medical history and QT prolongation pattern we classified this case as Romano-Ward Long QT Syndrome type 1.

We planned a six month follow up visit but 2 months after the first visit she was admitted to our emergency room with palpitation and chest pain during sleep.

On her admission ECG there was no ischemic changes and no arrhythmia but we hospitalized the patient because of typical angina. Because of dynamic T wave changes and typical angina we performed a coronary computed tomography angiography (CTA) (Figure 1B) to rule out coronary artery disease. We did not prefer diagnostic coronary angiography because there were no risk factors and troponin levels were negative during follow up. Coronary CTA showed an anomalous origin of right coronary artery with mechanical compression of the anomalous right coronary artery between the aorta and pulmonary root, which causes a moderate stenosis. Because of this moderate stenosis and angina we performed a myocardial perfusion scintigraphy and there were no ischemic changes at maximum heart rate. We continued β-blocker therapy for the patient and did not plan ICD because there was no previous cardiac arrest, syncope and / or documented ventricular tachycardia, also her QTc was under 500 ms. We offered family screening with further genetic analyses but the family refused genetic analyses. ECG recording from the mother (67-year-old) and the son (5-year-old) of the patient also revealed LQTS (Figures 1C and 1D). But these relatives had no history of arrhythmic episodes.

Discussion

LQTS is an inherited arrhythmogenic disease characterized by susceptibility to life-threatening arrhythmias, sudden cardiac death. Two major forms of LQTS have been identified, one transmitted as an autosomal dominant trait (Romano Ward Syndrome), the second is an autosomal recessive disease (Jervell and Lange-Nielsen Syndrome).[3] The incidence ranges from 0.01-0.02%.[4]

Therapy for LQTS is directed toward reduction of the incidence of syncope and sudden death. The data to guide management comes from large registries and referral centers with a bias toward patients with severe disease. Current therapeutic options involve the use of β-blockers and ICDs. The pacemaker function is also used in those with pause-dependent or bradycardia-induced ventricular tachycardia. An important clinical dilemma has been deciding with certainty who should or should not have a defibrillator. The American College of Cardiology/American Heart Association/European Society of Cardiology have issued guidelines, which include the use of ICDs, for the management of LQTS.[5] Prophylactic b-blockade has a Class I indication for all individuals with abnormal prolongation of the QT interval, regardless of symptoms. ICDs have a Class I indication in secondary prevention for those surviving cardiac arrest, Class IIa for those with symptoms or syncope while taking b-blockers, and Class IIb for primary prevention in those with possible high-risk characteristics. If the patient has syncope despite full dose β-blockade ICD implantation should be considered with the final decision being based on the individual patient characteristics (age, sex, previous history, genetic subgroup including sometimes mutation-specific features, presence of ECG signs – including 24 h Holter recordings – indicating high electrical instability). For this case we did not plan ICD because there was no previous cardiac arrest, syncope and/or documented ventricular tachycardia in her medical history, also measured QTc was under 500 ms.

Genetic analysis is also usefull for life style recommendations. If genetic analysis is not available it is best to avoid very strenuous exercise and keep stress at bay. Breathing exercises, meditation, and yoga are all ways to help manage stress. Eating foods high in potassium, such as bananas, or take potassium supplements may also be suggested. All patients must be informed about QT prolonging drugs. These patients must stay away from stressful jobs. Symptomatic patients must also stay away from jobs that might threaten the lives of other people (pilot, bus driver etc.)

Anomalous origin of coronary arteries has been reported as the cause of angina pectoris,

Correspondence: Emir Karacaglar, Fevzi Cakmak Street. No. 45 Baskent University School of Medicine, Cardiology Department, Postal Code: 06490. Bahcelievler/Ankara, Turkey.
Tel: +90.312.223.20.85 - Fax: +90.312 223 73 33.
E-mail: dremirkaracaglar@hotmail.com

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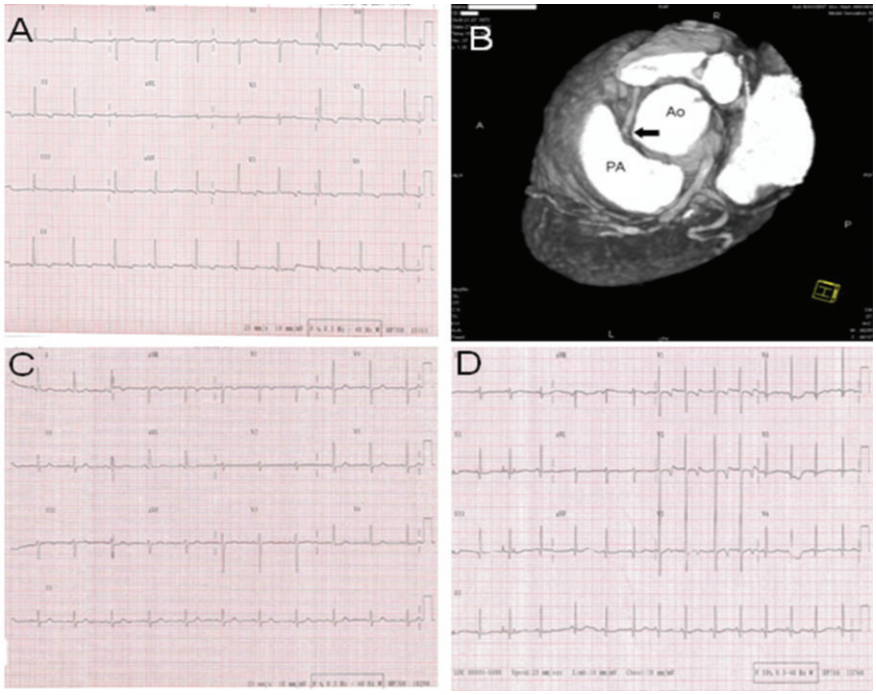

Figure 1. A) Electrocardiogram (ECG) recording of the patient; B) Right coronary artery with anomalous origin from the aorta partially running between the aorta and pulmonary artery where moderate stenosis is seen (arrow). Ao, aorta; PA, pulmonary artery; C) ECG recording from the mother of the patient; D) ECG recording from the son of the patient.

arrhythmia, syncope and fatal myocardial infarction.[1,2] Its incidence ranges from 0.61% to 1.3 %.[6] The incidence of anomalous origin of the right coronary artery (RCA) out of the right sinus of Valsalva ranges from under 0.01-0.09%.[7] The usually described variance is RCA arising from the wrong sinus of Valsalva. The exact mechanism, associated to cardiac events, is unclear in cases of coronary artery with anomalous origin and no obvious obstructive lesion. It might be related to mechanical compression of the anomalous coronary artery between the aorta and pulmonary root or great vessels, especially during exercise.[8,9]

Surgical repair is recommended, especially with anomalous origin of the left coronary artery (LCA). However, there is controversy concerning the treatment of anomalous right coronary artery (RCA) with interarterial course due to its relatively high incidence and the fact that it leads to few, if any, clinical problems.

We describe for the first time a patient with coronary anomaly and LQTS. All two entities are associated with an increased risk for developing malignant tachyarrhythmia and sudden cardiac death. Generally T wave pattern changes are expected in LQTS patients during follow up. This may be conflicting enough to overlook coronary artery disease or LQTS itself mimicking coronary artery disease. Further-more this case emphasizes that these two conditions may coexist in the same patient. If angina or ECG changes occur, one of coronary imaging techniques must be chosen to investigate coronary artery disease according to the patient's risk factors and coronary CT angiography seems like the best option in low risk group. Surgical repair or ICD implantation were not necessary for our patient but detection of coronary artery disease or coronary artery anomalies causing ischemia may change medical treatment as life saving.

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