Sudden cardiac arrest in a young professional athlete

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Cardiac arrest is an acute medical emergency characterized by the absence of cardiac activity, circulatory failure, abnormal respiration, and unresponsiveness. Historically, the term sudden cardiac death (SCD) has been used to describe all cardiac arrests. Current guidelines suggest that the label SCD should only be applied when attempts at resuscitation have failed and death occurs. The term sudden cardiac arrest (SCA) is now used to describe those individuals whose cardiac activity has returned either spontaneously or following interventions such as cardio-pulmonary resuscitation (CPR) and/or defibrillation.

Cardiac arrest is a common occurrence throughout the world. In Canada, a cardiac arrest occurs every 12 minutes with a yearly incidence of approximately 45,000. Cardiac arrest can be thought of as an end event of any of the following four conditions: ventricular tachycardia, pulseless electrical activity, ventricular fibrillation, and asystole. The causes by which these conditions/arrhythmias are generated can be broadly classified into those associated with overt structural heart disease (diagnosed on coronary angiography, echocardiography or resting electrocardiogram (ECG)) and those without (Table 1).

In general, ischemic heart disease is the most common cause of cardiac arrest, although less common causes are found more frequently in younger patients. This case will highlight a rare cause of SCA in a young professional athlete.

CASE

Mr. C is a previously healthy 25-year old professional hockey player who presented to hospital following SCA while swimming with friends. At the time of his arrest, he received CPR and was defibrillated three times by an automated external defibrillator for presumed ventricular fibrillation or ventricular tachycardia. Upon admission, Mr. C was intubated and transferred to the intensive care unit (ICU) where he underwent therapeutic hypothermia. Following several days in the ICU, Mr. C was transferred to the coronary care unit (CCU) for further investigations and management.

Once in the CCU, Mr. C remained stable and the efforts of the medical team turned towards investigating the etiology of Mr. C’s cardiac arrest. An echocardiogram revealed a normal left ventricular ejection fraction and mildly decreased right ventricular function with moderate dilatation of the right ventricle. Cardiac magnetic resonance imaging found no evidence of myocardial infarction, fibrosis, scarring, or findings suggestive of arrhythmogenic right ventricular cardiomyopathy. Coronary angiography showed normal anatomy with no obstructive coronary disease. Interestingly, while resting ECGs appeared normal, subsequent stress ECG testing revealed an abnormal QT interval. Genetic testing confirmed the diagnosis of congenital long QT syndrome (LQTS) type 1. Prior to discharge, Mr. C received an implantable cardioverter-defibrillator device (ICD). The remainder of his convalescence was unremarkable and he will be followed by the electrophysiology service.

DISCUSSION

First described in the 1957, our understanding of LQTS has grown enormously with the advancement of molecular biology and genetics. LQTS is a rare disorder characterized by delayed myocardial repolarization that predisposes these individuals to a form of polymorphic ventricular tachycardia known as torsades de pointes. This dangerous arrhythmia can quickly develop into ventricular fibrillation and subsequent cardiac arrest. The pathophysiology of LQTS relates to mutations in the genes responsible for producing the cardiac ion-channel proteins that regulate the flux of sodium and potassium ions during myocardial contraction. To date, more than 150 different mutations in 7 genes have been implicated in LQTS. The location and type of mutation, as well
CASE REPORT

Table 1: Major causes of cardiac arrest

| Cardiac arrest with overt structural heart disease | Coronary Disease | Ischemic heart disease |
| Cardiomyopathies                                |                 | Anomalous coronary circulation |
| Other                                           |                 | Ischemic cardiomyopathy |
| Cardiac arrest without overt structural heart disease | Primary electric | Long-QT syndromes |
| Metabolic imbalance                             |                 | Short-QT syndromes |
| Noncardiac                                      |                 | Brugada syndromes |
|                                                |                 | Hyperkalemia/hypokalemia |
|                                                |                 | Hypocalcemia |
|                                                |                 | Acidosis |
|                                                |                 | Pulmonary embolus |
|                                                |                 | Intracranial hemorrhage |
|                                                |                 | Drug induced |

as variable genetic penetrance, help to explain the broad range of clinical presentations that are observed in this syndrome.\(^9\)

The diagnosis of LQTS is based on history, 12-lead ECG, exercise and sympathomimetic drug provocation, and genetic testing. On history, patients may report palpitations, syncope, seizures, or no symptoms at all. It is important to inquire about a family history of cardiac arrest as well as several gene-specific symptom triggers including exercise, emotional states, loud noise, and sleep related events.\(^10\)

The 12-lead ECG is an important tool in the diagnosis of LQTS (Figure 1). When interpreting the QT interval, it must be corrected for heart rate. This is achieved through the use of Bazett’s formula (QTc=QT/√R-R [seconds]) which is most accurate for heart rates between 60-100bpm.\(^11\) Corrected QT intervals >440ms in males and >460ms in females are considered abnormally long.\(^4\) Particular attention should also be given to the presence of characteristic T-wave morphology which has shown to be associated with certain types of LQTS.\(^12\) The presence of a normal resting ECG does not exclude the diagnosis of LQTS. As exemplified in this case report, exercise or drug provocation may be required to bring out repolarization abnormalities in individuals who exhibit normal QT intervals at rest.\(^13\)

The management of LQTS is complex and multi-faceted. Patient education, genetic counseling, and family screening are important to consider when caring for these patients. With regards to medical therapy, beta-blockers are first-line medications as they have been shown to reduce cardiac events or ICD implantation should be strongly considered in high risk individuals.\(^15\)

In summary, establishing a definitive diagnosis following SCA in the absence of overt structural heart disease remains challenging. The recommended approach is to obtain a comprehensive clinical history and employ sequential non-invasive and invasive investigations including provocative testing and advanced imaging techniques. Unfortunately, despite these efforts, the underlying cause in nearly half of these patients will remain unknown and a diagnosis of idiopathic ventricular fibrillation will be given.\(^17\)

REFERENCES


