Catecholaminergic Polymorphic Ventricular Tachycardia: A Rare Cause of Cardiac Arrest Following Blunt Chest Trauma

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Catecholaminergic polymorphic ventricular tachycardia (CPVT) is an electrophysiological disorder of a physically normal heart that occurs in children when the body is subjected to intense emotional or physical stress that causes adrenergic discharge. This troubling disease can be sporadic (spontaneous) or familial (genetic/inherited). Unfortunately, its associated ventricular tachycardia may cause sudden death, so early diagnosis of CPVT is very important. Treatment modalities include medical treatment, implantation of a cardioverter defibrillator, or surgical sympathectomy; but the implantable cardioverter defibrillator (ICD) should be the first choice in patients with a history of cardiac arrest. We herein present the case of a patient diagnosed with CPVT after a successful cardiopulmonary resuscitation triggered by blunt chest trauma. We implanted an implantable cardioverter defibrillator and started oral B-blocker treatment. During the course of follow-up, flecainide was added to his treatment depending on the patient’s status regarding recurrent ICD shock. The patient has now continued follow-up without recurrent ICD shock since flecainide treatment was initiated. In conclusion, in patients with syncope and sudden cardiac arrest secondary to physical stress or blunt chest trauma, CPVT should be considered and an implantable cardioverter defibrillator must be implanted. Additionally, flecainide therapy should be considered to decrease recurrent ICD shock.

Key Words: Blunt chest trauma • Cardiac arrest • CPVT

INTRODUCTION

Catecholaminergic polymorphic ventricular tachycardia (CPVT) is a form of tachycardia that can be triggered by adrenergic stimuli, such as emotional and/or bodily stress, in children with a structurally normal heart. The condition may be isolated or familial; its diagnosis carries considerable importance because it can cause sudden death. Medical treatment, and surgical sympathectomy are among the treatment options, although patients who have experienced cardiac arrest should have an ICD implanted. We present here the case of a patient who experienced cardiac arrest following blunt chest trauma, who was diagnosed with CPVT after cardiopulmonary resuscitation, and in whom an ICD was implanted.

CASE REPORT

Previously, a nine-year-old male patient who had no history of prior cardiac disease experienced sudden syncope after being hit in the chest by a ball while playing in the schoolyard. He was brought to the nearest hospital, which was five minutes away. Cardiac arrest was diagnosed at the private hospital, and cardiopulmonary resuscitation...
resuscitation (25 minutes) and 50 J with defibrillation were applied to the patient due to the observed ventricular fibrillation. He was monitored in the hospital’s intensive care unit during which time an amiodarone infusion was administered. The patient thereafter received mechanical ventilation and medical treatment and his condition improved. Subsequent to extubation, he was discharged from the hospital four days after admission. However, the patient experienced effort-related syncope twice during further follow-up, and was referred to our facility. The patient’s physical examination and electrocardiography (ECG) were normal (heart rate: 77 beats per minute, PR: 140, QRS: 80, QT: 390, and QTc: 441 ms) (Figure 1A), as was his echocardiography. Bidirectional polymorphic ventricular tachycardia was found upon effort ECG (Figure 1B). After considering the patient’s history of cardiac arrest and defibrillation, a single-chamber transvenous ICD was implanted. During follow-up, flecainide was added to his treatment regimen depending upon whether recurrent ICD shock occurred (Figure 2). Currently, the patient is being followed-up, and he is under treatment with a beta-blocker (propranolol, 3 mg/kg/day) and flecainide (80 mg/m²) without recurrent ICD shock to date.

Figure 1. (A) The patient’s first electrocardiography. (B) Catecholaminergic polymorphic ventricular tachycardia seen during the stress test.
DISCUSSION

Catecholaminergic polymorphic ventricular tachycardia was first described in 1975 by Reid et al., and is known as a cause of ventricular tachycardia, syncope, and sudden cardiac death. It is rarely seen in children without cardiac disease and with normal QT values. Approximately 30% of cases have a familial history of sudden death before age 40. Hereditary transmission is either autosomal dominant or recessive, and generally with high penetrance. Genes leading to CPVT have been identified on chromosome 1. Mutations have been identified on the calcium release channel cardiac ryanodine receptor (RyR2) and calsequestrin 2 (CASQ2) genes. Following a mutation of these two genes, calcium — liberated from the sarcoplasmic reticulum through the unstable RyR2 Ca++ channel — increases in the cell, causing a delay in the post-depolarization triggering activity and consequent bidirectional polymorphic ventricular tachycardia.

Diagnosis is difficult in the absence of symptoms, and with normal ECG and echocardiographic findings. In the case of suspected disease, a Holter ECG recording or — when considering a possible triggering from exercise — an effort test should always be performed. Provocative (epinephrine test) testing should be considered in cases that cannot be diagnosed with an exercise test. A loop recorder can be used when a stress and provocative test concludes without results. The typical constellation of three criteria for diagnosing CPVT consists of: a) an exercise-triggered or emotionally-triggered severe ventricular arrhythmia, b) a normal resting ECG together
with polymorphic ventricular tachycardia (VT) on stress ECG, and c) a structurally normal heart. Arrhythmia-producing conditions (for example, right ventricular dysplasia, prolonged QT, Brugada syndrome, pre-excitation, and Andersen-Tawil syndrome), which are part of the differential diagnosis of syncope and sudden death in children, must also be convincingly excluded.4

Even though beta-blockers used in treatment do reduce arrhythmia frequency and mortality, they cannot entirely eliminate the risk. Left sympathetic cardiac denervation combined with ICD implantation may be an effective treatment, especially in patients not controlled by medical treatment for arrhythmia. Of late, flecainide, which had been discovered to directly depress the RyR2 channels, has become an additional treatment possibility.7

Even though the mechanism by which VT develops following blunt trauma to the chest is not entirely clear, it has been speculated that a blunt trauma during the 30-millisecond period of ventricular repolarization may cause polymorphic ventricular tachycardia.4,5 Blunt trauma may increase the possibility of and lead to VT after longer ventricular repolarization by a delay in the post-depolarization in CPVT. The case presented represents a rare occurrence of cardiac arrest following blunt chest trauma, which has not yet been reported in the publications.

Although a rare event, CPVT should be considered in cases of syncope and cardiac arrest triggered by exercise or blunt chest trauma, and an ICD should be implanted to avoid early sudden death. Flecainide therapy should be considered to decrease recurrent ICD shock.

REFERENCES