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Case Report

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Successful ECPR in a teen with brugada syndrome presenting as unexplained cardiac arrest

Abstract

This case report presents the successful management of a 16-year-old male patient who initially presented with unexplained cardiac arrest (UCA) and was subsequently diagnosed with Brugada syndrome. The patient, who was on propranolol and propafenone for supraventricular tachycardia, developed in-hospital hypotension, bradycardia, and cardiac arrest. Following 110 minutes of prolonged cardiopulmonary resuscitation (CPR), extracorporeal cardiopulmonary resuscitation (ECPR) was implemented, and the patient recovered without neurological sequelae. Initially suspected to be drug intoxication, genetic testing revealed Brugada syndrome as the underlying etiology. This case highlights the importance of timely ECPR application in unexplained cardiac arrest. This report underscores the significance of proper patient selection, timing, and logistical coordination for ECPR, supporting the potential of ECPR in Brugada syndrome.

Keywords: Extracorporeal membrane oxygenation, cardiopulmonary resuscitation, brugada syndrome, cardiac arrest, sudden cardiac arrest

INTRODUCTION

Unexplained cardiac arrest (UCA) is a medical emergency characterized by sudden cardiac arrest, either in-hospital or out-of-hospital, where the underlying cause is not immediately apparent. Extracorporeal membrane oxygenation (ECMO) is a therapeutic modality used to provide mechanical support in severe respiratory and/or cardiac failure, thereby gaining time for further intervention. When ECMO circulation is applied in conjunction with cardiopulmonary resuscitation (CPR), it is termed extracorporeal cardiopulmonary resuscitation (ECPR). ECPR supports vital functions by providing hemodynamic and respiratory mechanical support in cases of cardiac arrest refractory to conventional CPR, allowing time for the identification and treatment of the underlying etiology. However, the effective management of prolonged ECPR, particularly in UCA, remains a subject of debate. This report discusses a case initially presenting as UCA, subsequently diagnosed with Brugada syndrome, and successfully treated with ECPR.

CASE REPORT

A 16-year-old male patient, followed up with a diagnosis of supraventricular tachycardia and using oral Propranolol 20 mg BID and Propafenone 150 mg BID for the last 2 weeks, presented to the emergency department with complaints of nausea, pallor, weakness, and chest pain. An intravenous line was established, and blood tests were ordered to evaluate for possible infection, liver and kidney dysfunction, and electrolyte imbalance. The electrocardiography (ECG) showed a nodal rhythm with a wide

QRS (240 milliseconds) at 66 beats/minute (Figure 1). The patient was monitored, and hydration was provided with 500 cc of intravenous saline. Biochemical tests revealed normal renal and liver functions and no electrolyte disturbances. Transthoracic echocardiography showed no cardiac pathology other than patent foramen ovale. Blood gas analysis showed normal oxygenation and no acidosis. The patient, who was hypotensive (80/60 mm Hg), underwent a computed thomography (CT) scan for possible brain pathology. While being examined in the radiology clinic, the patient developed convulsions followed by cardiac arrest. A code blue was activated, and the patient was immediately intubated, and CPR was initiated. The patient did not have a defibrillable heart rhythm. Based on the known history of antiarrhythmic drug use, drug intoxication was suspected. The patient, who could not achieve a heart rhythm despite 40 minutes of CPR, was transferred to the intensive care unit, and a decision was made to apply ECMO circulation. Since there was no ready ECMO set in the hospital at that moment, a heart-lung pump was placed via right femoral artery and vein cannulation under CPR. When the extracorporeal pump circulation reached full flow, which was 4 lt/min according to his weight, mechanical cardiac massage was stopped. A total of 110 minutes of CPR was applied to the patient until pump circulation was established. During CPR, an intravenous pacemaker device was inserted, but despite pacemaker stimuli being observed on the monitor, there was no cardiac or hemodynamic response to these impulses. Continuous and effective CPR was maintained until the initiation of extracorporeal circulation cannulation. CPR was performed with uninterrupted chest compressions and mechanical ventilator support. During the cannulation procedure, chest compressions were briefly interrupted (3-5 seconds) due to surgical manipulations. Adrenaline 1 mg was administered intravenously every 3 minutes. Chest compressions were administered at a rate of 100-110 per minute, with a depth sufficient to maintain a systolic arterial pressure above 80 mmHg, as monitored arterially. Complete chest wall recoil was allowed after each compression, and the compression and relaxation durations were approximately equal. At the end of CPR, the patient's body temperature was measured to have dropped spontaneously to 35 degrees Celsius. No additional hypothermia was applied. Two hours after the start of pump circulation, an ECMO set was obtained, and the pump device was replaced with veno-arterial ECMO circulation. For the first 2 hours after CPR, the patient had a heart rhythm of 20-25 beats/minute with a prolonged QT interval, despite high-dose inotropic support. Four hours after the start of ECMO circulation, a wide QRS rhythm of 40 beats/minute continued. In the following hours, the QRS interval returned to the normal range, and the heart rate reached normal speed. The patient, who woke up without neurological sequelae 1 day after the procedure, was extubated. Inotropic drug doses were gradually reduced, and oral food was started. On the 2nd day, the ECMO circulation flow was reduced with hemodynamic control. On the 3rd day, ECMO circulation was stopped, and cannulas were removed. The patient was mobilized on the 4th day and then transferred to the ward. Control echocardiography showed normal ventricular functions, and the QRS interval on the ECG was 90 ms and returned to the normal range. There were complaints of pain in the left pectoral region due to CPR and hemoptysis findings lasting 2 days due to possible intraalveolar hemorrhage. Recovery was achieved with symptomatic treatment. At the end of the 1st week, the patient was discharged home in a condition where he could mobilize on his own without neurological sequelae. In the follow-up examinations, neuropathic pain and hyperesthesia symptoms developed in the right lower extremity peroneal region where extracorporeal circulation cannulas were inserted. Neurological examination revealed entrapment neuropathy findings in the right peroneal nerve and 4/5 strength loss. The patient was admitted to an outpatient physical therapy and rehabilitation program. Genetic testing revealed mutations consistent with Brugada syndrome, establishing the etiology of the cardiac arrest.

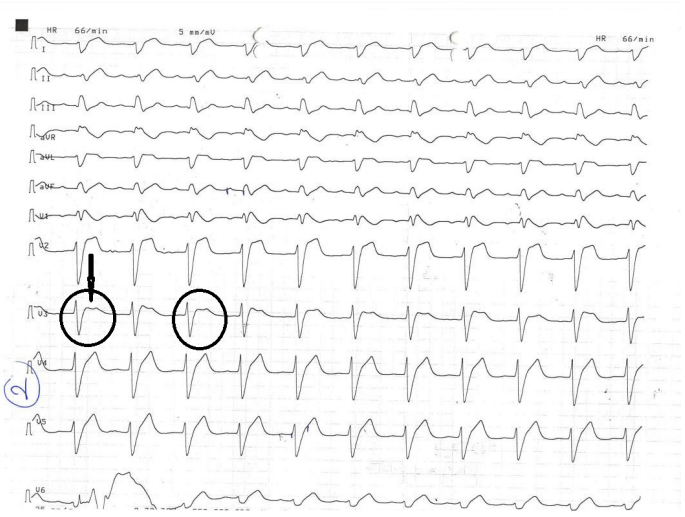

Figure 1. ECG on arrival to the emergency unit. A saddleback-shaped ST elevation was observed in lead V3

DISCUSSION

In this article, we present the case of a 16-year-old patient who, while using therapeutic doses of propranolol and propafenone for supraventricular tachycardia, developed unexplained hypotension, bradycardia, and in-hospital cardiac arrest, and subsequently recovered without central neurological sequelae following 110 minutes of prolonged CPR and ECPR.

UCA is the inability to determine the cause of cardiac arrest at the time of the event. This condition, which can occur in young, relatively healthy individuals, can be associated with rhythm disturbances such as ventricular fibrillation (VF), cardiac asystole, or pulseless ventricular tachycardia (VT). According to the American Heart Association (AHA), the underlying cause cannot be determined in 25% of out-of-hospital cardiac arrest cases (1).

Initially, drug intoxication was suspected due to the patient's medication history, prolonged QRS interval on the initial

ECG, and lack of cardiac response to pacemaker impulses during CPR. However, detailed history revealed adherence to prescribed medication doses and ruled out suicidal intent through psychological evaluation. Blood propafenone levels, obtained during CPR, were within the therapeutic range (1520 µg/L). During the intensive care unit course, brain CT imaging and subsequent biochemical and systemic evaluations were conducted. These investigations yielded no pathological evidence to explain the etiology of the cardiac arrest. Thus, the initial cause of cardiac arrest remained undetermined, and the case was considered UCA. Subsequent investigations, including genetic testing, identified Brugada syndrome as the underlying etiology.

Brugada syndrome is an autosomal dominant genetic disorder caused by mutations in sodium channel genes. Defects in these channels lead to reduced cellular sodium influx, prolonged action potential duration, and delayed contractions. It can precipitate ventricular tachyarrhythmias, ventricular fibrillation, and sudden cardiac death, particularly in young males (2).Genetic testing has identified two gene mutation in SCN gene in Brugada Syndrome; SCN5A and SCN10A.

ECG changes in Brugada syndrome are primarily classified into three types: Type 1: Characterized by a "coved-type" ST-segment elevation of ≥2 mm in at least two of leads V1, V2, or V3, followed by a negative T-wave. This morphology is considered diagnostic for Brugada syndrome. Type 2: Demonstrated by a "saddleback-shaped" ST-segment elevation of ≥2 mm in lead V1 and/or V2. And Type 3: May exhibit either Type 1 or Type 2 morphology, but with an ST-segment elevation of <2 mm (3).

In our patient's ECG, a saddleback-shaped ST elevation was observed in lead V3 (Figure 1). Based solely on these ECG findings, a definitive diagnosis of Brugada syndrome cannot be established, but it can be suspected. This particular ECG pattern was only be considered a supportive finding, particularly in conjunction with genetic testing results.

The patient’s genetic testing confirmed the presence of a specific SCN5A mutation, consistent with Brugada syndrome. Because the diagnosis of Brugada syndrome was confirmed by genetic testing, pharmacological testing (such as Ajmaline) was not required.

CPR, the initial treatment for cardiac arrest, provides oxygenated blood to vital organs. Cardiac output during CPR is approximately 15-25% of normal (4). Prolonged CPR refers to resuscitation efforts exceeding 20 minutes. Effective chest compressions and ventilation are essential during this time, but the efficacy of prolonged CPR diminishes over time, necessitating additional interventions like ECMO circulation. Studies demonstrate that ECMO-supported prolonged CPR significantly improves survival rates in in-hospital cardiac arrest. ECPR has been shown to increase survival by 20-45% in these cases (5,6). According to the 2022 Extracorporeal Life Support Organization (ELSO) Registry Report, survival rates for ECPR were reported as 41% in adults and 58% in pediatric patients. Concurrently, the number of centers utilizing ECMO has significantly increased, rising from 150 in 2008 to 500 in 2021, a surge partly attributed to the recent pandemic (7).

Cohort studies indicate that extracorporeal circulation initiation within 20-29 minutes of CPR is associated with excellent survival and minimal neurological damage. Ischemic organ damage prior to ECMO circulation initiation significantly impacts prognosis, with survival decreasing by 25% every 10 minutes after 30 minutes (8). However, it is not yet known how long resuscitation efforts should be continued in arrests of unknown cause and when ECMO should be considered in such patients. There is no absolute time limit, and successful ECPR outcomes have been reported after 60 minutes. Early ECMO circulation initiation is critical to maximizing survival chances.

Our patient, who experienced in-hospital cardiac arrest, received rapid interventions, including intubation and CPR. After 40 minutes of unsuccessful CPR, cardiovascular surgery was consulted for extracorporeal circulation support. Since there was no ECMO device available, a decision was made to provide circulatory support with a heart-lung pump as a bridge treatment until the device was provided. Thus, the total CPR time reached 110 minutes until femoral cannulation and extracorporeal circulation were provided. Throughout these 110 minutes and until full flow circulation was achieved with the heart-lung machine, uninterrupted CPR was maintained. The success in this patient underscores the importance of effective and continuous CPR.

Beyond the time factor, patient selection and systemic organ condition are critical for ECPR effectiveness. Pre-extracorporeal circulation evaluation of renal, hepatic, and neurological functions is essential. ECPR efficacy diminishes in patients with multiple organ failure or poor life expectancy. ECPR is most effective when the underlying pathology is treatable, and the event is a "matter of time." A meta-analysis by Maier et al. reported a 50.9% survival rate with ECPR in pharmacological and non-pharmacological poisoning-induced cardiac arrest (9). Wanscher et al. documented a 100% survival rate in seven victims of a Danish Fjord accident with an average body temperature of 18.48°C, where the average time to ECMO circulation was 226 minutes.ECMO circulation is now considered the gold standard for hypothermic cardiac arrest (10). The successful outcome in our patient can be attributed to several factors: in-hospital cardiac arrest with immediate CPR initiation, the initial presentation as UCA, the patient's young age, and the absence of other organic diseases. The persistent refractory asystole during CPR raised suspicion of drug intoxication or suicide attempt, necessitating time to allow for substance clearance. ECMO circulation support was thus initiated to bridge this critical period.

Extracorporeal circulation is a valuable tool for gaining time in cardiac arrest due to transient or treatable pathologies. In retrospect, earlier ECMO circulation initiation might have been beneficial. As clinical experience grows, the decision to

implement ECMO circulation in appropriate patients can be expedited. The timely transfer to an ECMO-capable center is also crucial. Even when an ECMO device is not immediately available, the heart-lung pump as a bridge to definitive ECMO circulation should be considered.

Case reports of ECPR use in Brugada syndrome-induced cardiac arrest are limited (11,12). This case demonstrates that Brugada syndrome can result in sudden cardiac arrest and that ECMO circulation can be successfully employed in these patients.

CONCLUSION

Initially, drug intoxication was suspected as the cause of cardiac arrest in this case; however, Brugada syndrome was subsequently diagnosed. ECPR is a vital intervention that enhances survival rates in cardiac arrest requiring time for definitive treatment. Optimal patient selection, timing, and logistical coordination are essential for maximizing ECPR effectiveness. In cardiac arrest with reversible causes, rapid transport to an ECMO-capable center with ongoing high-quality CPR and readily available ECMO circulation systems are paramount. The potential of ECPR as a life-saving option in Brugada syndrome should be recognized. Future randomized controlled trials are needed to refine treatment protocols and address knowledge gaps in this area.

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