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CASE REPORT

ADVANCED

CLINICAL CASE

Sudden Cardiac Arrest in the Postpartum Period Due to Long QT Syndrome and Dilated Cardiomyopathy

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ABSTRACT

We describe the case of a previously healthy patient presenting with sudden cardiac arrest in the postpartum period as a result of concomitant congenital type 1 long QT syndrome and *BAG3* dilated cardiomyopathy. This case highlights the increased rate of cardiac events for patients with long QT syndrome in the postpartum period. (**Level of Difficulty: Advanced.**)

HISTORY OF PRESENTATION

A 19-year-old woman presented with sudden cardiac arrest (SCA) 5 weeks after premature delivery. She collapsed in her kitchen and had bystander cardiopulmonary resuscitation for 20 minutes before the arrival of emergency medical services (EMS). She was in ventricular fibrillation and received 2 defibrillations by EMS before the return of spontaneous circulation. On hospital admission, the initial physical examination showed an intubated and sedated patient with a benign cardiopulmonary examination.

LEARNING OBJECTIVES

- To identify the increased rate of cardiac events in the postpartum period in patients with LQTS.
- To recognize the importance of CMR in the work-up of SCA.
- To describe the appropriate treatment of patients with LQTS.

PAST MEDICAL HISTORY

The patient was 5 weeks post partum from delivery at 24 weeks of gestation resulting from placental abruption. This was her second gestation, with a previous uncomplicated term pregnancy. A few weeks after her delivery she had a motor vehicle accident while driving in which she suddenly lost consciousness. She did not seek evaluation by medical personnel after the accident. Pertinent family history included the sudden cardiac death of her mother in her late 20s. The cause of her mother's death was reportedly an unnamed "cardiomyopathy," and no autopsy information was available. Otherwise, the patient was healthy and took no medications.

DIFFERENTIAL DIAGNOSIS

The initial differential diagnosis for her SCA included hereditary cardiomyopathy, type 2 long QT syndrome (LQTS), peripartum cardiomyopathy, stress cardiomyopathy, SCA, spontaneous coronary artery dissection, and anomalous coronary artery, with

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ABBREVIATIONS AND ACRONYMS

CMR = cardiac magnetic resonance

DCM = dilated cardiomyopathy

EMS = emergency medical services

LGE = late gadolinium enhancement

LQTS = long QT syndrome

LVEF = left ventricular ejection fraction

SCA = sudden cardiac arrest

highest concern for hereditary cardiomyopathy given her family history of sudden cardiac death.

INVESTIGATIONS

Initial laboratory investigation revealed an elevated troponin I level, which peaked at 3.07 ng/mL (<0.02 ng/mL) and signs of end-organ dysfunction with elevated creatinine at 1.1 mg/dL (0.6-1.1 mg/dL) from baseline 0.5 mg/dL and lactic acid of 3.3 mmol/L (0.5-2.2 mmol/L). Magnetic resonance imaging of her brain revealed diffuse hypoxic ischemic insults throughout the parieto-occipital cortex and basal ganglia. An initial electrocardiogram before therapeutic cooling revealed a prolonged corrected QT interval at 534 milliseconds by using the Bazett formula (**Figure 1**). The transthoracic echocardiogram showed severely reduced systolic function, with a left ventricular ejection fraction (LVEF) of <15% with otherwise normal wall thickness and valve structure. A cardiac magnetic resonance (CMR) scan obtained 1 week after admission showed an improvement in LVEF to 47% with mildly dilated left ventricular cavity size and a basal to midwall septal stripe late gadolinium enhancement (LGE) consistent with nonischemic dilated cardiomyopathy (DCM) (**Figures 2A and 2B**). Genetic testing revealed a pathogenic heterozygous sequence variant in both *KCNQ1*, indicating congenital type 1 LQTS, and *BAG3*, which is associated with DCM.

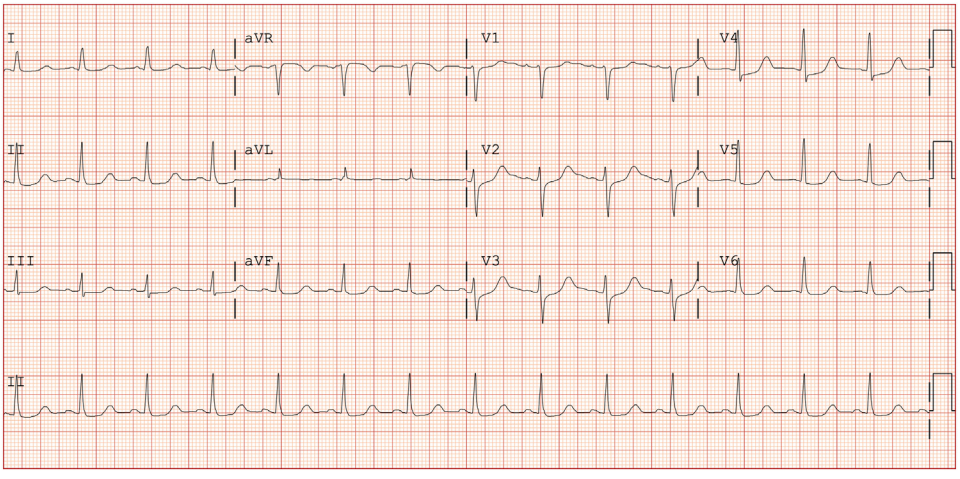
MANAGEMENT

Therapeutic hypothermia was initiated following cardiac arrest according to institutional protocol. She was initially given an indefinite prognosis of neurologic recovery by neurology. She had a tracheostomy and percutaneous endoscopic gastrostomy tube placement while awaiting neurologic recovery. Given the diagnosis of congenital type 1 LQTS, she was started on prophylactic nadolol. Her initial low LVEF was presumed to be the result of her postshock state given the rapid improvement in LVEF. She had an implantable cardioverter-defibrillator placed for secondary prevention.

DISCUSSION

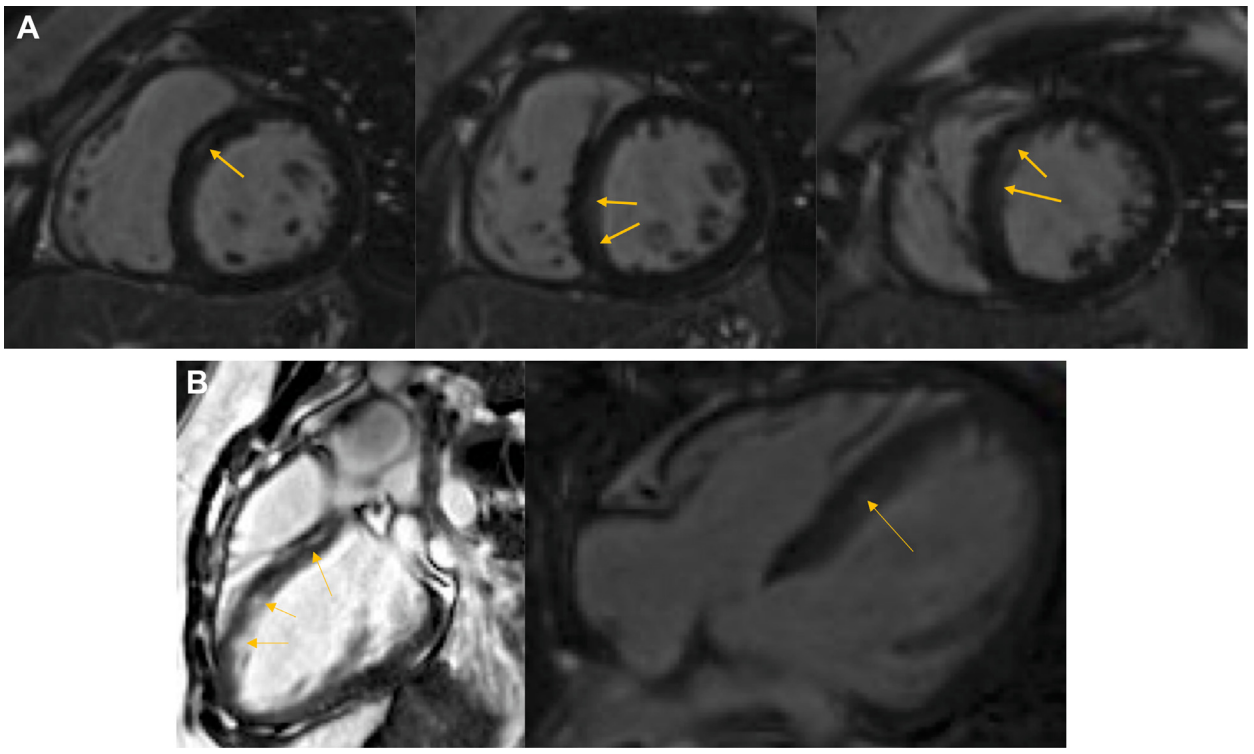
LQTS can be either congenital or acquired.[1] Among congenital LQTS cases, 13 genes have been implicated, with the *KCNQ1* gene, indicative of type 1 LQTS, the most common variant.[1] Patients with type 1 LQTS have been shown to have a higher risk of cardiac events with sympathetic activation, especially exercise and stress.[2] There has been particular attention paid to the effects of pregnancy on the risk of arrhythmias. Initial studies showed an increased risk of cardiac events for patients with LQTS.[3] Later investigations revealed that the preponderance of cardiac events occurring post partum was in patients with type 2 LQTS rather than type 1 LQTS (16% vs <1%, respectively).[4,5] A follow-up study specifically examining the risk of cardiac events in patients

FIGURE 1 Electrocardiogram on Presentation



Corrected QT interval prolonged at 534 milliseconds.

FIGURE 2 **Cardiac Magnetic Resonance**



(A) Short-axis views demonstrating late gadolinium enhancement **(arrows)** in the basal to midseptum. **(B)** The 3- **(left)** and 4-chamber views **(right)** demonstrating basal to midseptal late gadolinium enhancement **(arrows).**

with type 1 LQTS in the peripartum period showed a 1% rate of cardiac events during pregnancy and 2% rate of cardiac events post partum.[6]

In addition to type 1 LQTS, our patient also had a mutation in *BAG3*, which is a known cause of DCM.[7] *BAG3* is present in up to 6.7% of patients with DCM.[8] Among subtypes of LQTS, type 3 LQTS has shown the closest association with DCM.[9,10] In patients with mutations in *KCNQ1*, there have been case reports of concomitant hypertrophic cardiomyopathy and noncompaction cardiomyopathy.[11] Our patient is the only known case of concomitant *KCNQ1* and *BAG3* mutations. In a recent study, 15.6% of Kazakh patients with DCM who presented with ventricular tachycardia had a mutation of *KCNQ1*.[12] This finding suggests an association between channelopathies, such as type 1 LQTS, and the presence of ventricular tachycardia in patients with DCM.

The diagnosis of DCM in our patient was made because of the *BAG3* mutation coupled with CMR findings of a dilated left ventricle and nonischemic LGE. CMR is recommended in the work-up of SCA and has important diagnostic and prognostic implications.[13] In patients with SCA, CMR findings such as decreased LVEF and extent of LGE have been associated with an increased frequency of major adverse cardiac events.[14]

The mainstay of therapy in LQTS is β-blockers, which decrease mortality from 21% in 1 year to 1% in 15 years.[1,6] Preferred agents are propranolol and nadolol over other β-blockers.[1] This therapy is effective in both pregnancy and the postpartum period.[3-6] There is debate about whether implantable cardioverter-defibrillator placement is appropriate for primary prevention in patients with LQTS; however, the consensus is that device placement is indicated for secondary prevention, as performed in our patient.[1]

FOLLOW-UP

She was weaned from sedation and experienced progressive neurologic recovery. Her LVEF normalized to 65% to 70% on repeat transthoracic

echocardiogram. Her QTc interval remained prolonged at >500 milliseconds on repeat electrocardiograms throughout hospitalization. She was counseled on the risk of cardiac events in future pregnancies and was started on contraception. Genetic counseling was scheduled for her children and siblings. She was discharged to an acute rehabilitation facility 36 days after the initial event.

CONCLUSIONS

This case is a rare example of a previously asymptomatic patient with SCA in the postpartum period resulting from concomitant congenital type 1 LQTS and *BAG3* DCM. It highlights the increased risk of peripartum events in patients with LQTS. Additionally, it emphasizes the role of CMR in the work-up of SCA.

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The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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