RED GUMMY BEAR OR RED HERRING?
WHEN CARDIAC ARREST ETIOLOGY ISN’T WHAT IT SEEMS

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INTRODUCTION

• We present a case of a 4-year-old male with history of one prior cardiac arrest attributed to choking on a foreign body, later determined to have Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT) after a second cardiac arrest prompted a genetic workup.

CASE DESCRIPTION

• This is a single patient case study of a 4-year-old male with two cardiac arrest events. Initial cardiac arrest was attributed to respiratory arrest due to story of possible choking on a red gummy bear.

• His rhythm at the scene showed coarse polymorphic ventricular tachycardia which reverted to normal sinus rhythm after multiple defibrillations.

• Approximately 3 months after the initial cardiac arrest, he was found unresponsive and pulseless. CPR was initiated and the patient was transported to our Emergency Department.

• The patient’s cardiac rhythm showed polymorphic ventricular tachycardia, requiring multiple shocks and magnesium sulfate boluses for return of spontaneous circulation.

• Initial electrocardiogram demonstrated prolonged QTc that resolved the following day. Echocardiogram showed normal anatomy and function.

CASE DESCRIPTION (CONT.)

• Telemetry showed intermittent ventricular ectopy, trigeminy, bigeminy, and couplets.

• A genetic workup revealed heterozygosity for a variant of the CASQ2 gene, c.538A>C (p.Lys180Gln), a variant of uncertain significance that is associated with CPVT.

• To prevent future cardiac arrests, a joint decision was made with the family for a sympathetic ganglion denervation.

Pathology

Mutations in the RyR2 and CaSQ2 proteins lead to Ca leak from the sarcoplasmic reticulum in diastole, causing delayed afterdepolarizations.

CONCLUSION

• Standardized cardiac arrest workup including genetic testing in young children with initially unexplained cardiac arrest may play a valuable role in diagnosis and management.

• Even in seemingly straightforward cases, it is essential to avoid premature closure and to follow a standardized investigational approach including genetic testing.

REFERENCES


