ANDERSEN-TAWIL SYNDROME: A NOVEL APPROACH FOR THE MANAGEMENT OF VENTRICULAR ARRHYTHMIAS

Poster Contributions
Poster Hall B1
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Background: Andersen-Tawil syndrome (ATS) is a rare autosomal dominant or sporadic disorder defined by a triad of periodic muscle paralysis, ventricular arrhythmias (long QT-7) and dysmorphic features. The underlying mutation is in the KCNJ2 gene encoding the inward rectifier potassium channels (Kir2.1) present in both skeletal and cardiac muscles. Ventricular arrhythmias are precipitated by adrenergic stimuli like exercise or intense emotion, which accounts for the use of beta-blockers as standard therapy.

Case: A 58-year-old female with a lifelong history of hyperkalemic periodic muscle paralysis and bi-directional ventricular tachycardia. No known history of coronary artery disease. She had tried multiple antiarrhythmic agents without significant suppression of premature ventricular contractions (PVCs) and was currently on propranolol. An implantable cardiac device was inserted for sudden cardiac death (SCD) prevention. She was on acetazolamide for control of periodic paralysis. There was no family history of SCD. Physical examination was significant for regularly irregular cardiac rhythm and the presence of craniofacial dysmorphic features. EKG showed frequent PVCs in a trigeminy pattern and a corrected QT interval of 508 msec. A PVC burden of 35,000 per day was noted on holter monitoring.

Decision Making: Due to the presence of the typical triad, there was a suspicion for ATS. Genetic testing confirmed she was heterozygote for the KCNJ2 gene mutation. Ventricular arrhythmias had not been responsive to multiple antiarrhythmic agents or PVC ablation. There was a recent case report from Japan suggesting atrial pacing was successful in reducing PVC burden after findings of increased PVC frequency after meals or during sleep. An atrial lead was implanted to provide continuous overdrive pacing at 80 beats per minute. There was a reduction in the PVC burden in the post-operative period. One year later, she continues to have a low PVC burden (24 per day) with improvement in quality of life.

Conclusion: This is the second case report to our knowledge that shows atrial overdrive pacing may control ventricular arrhythmias in ATS, particularly when PVCs are noted in low adrenergic states.