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CASE REPORT

CLINICAL CASE

Ventricular Tachycardia Due to Arrhythmogenic Right Ventricular Cardiomyopathy in the Setting of Acute Coronary Syndrome

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ABSTRACT

A 55-year-old man presented with sustained monomorphic ventricular tachycardia with a left bundle QRS complex pattern in the setting of non–ST-segment elevation myocardial infarction. The electrocardiographic morphology of the ventricular tachycardia and echocardiographic findings of significant right ventricular dysfunction led to the diagnosis of arrhythmogenic right ventricular cardiomyopathy. (J Am Coll Cardiol Case Rep 2023;28:102092) © 2023 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 55-year-old man presented with palpitations, dizziness, midsternal chest discomfort, and extreme tachycardia while exercising. Vital signs revealed a heart rate of 237 beats/min and blood pressure of 83/62 mm Hg. Electrocardiogram (ECG) showed a regular wide complex tachycardia with a left bundle (LB) left superior axis QRS complex morphology consistent with sustained monomorphic ventricular tachycardia (SMVT) (**Figure 1**). He was initially treated with intravenous (IV) adenosine 6 and 12 mg and IV diltiazem 20 mg with no effect. A 150-mg IV bolus of amiodarone converted him to sinus rhythm. ECG postconversion showed sinus rhythm with right bundle branch block and diffuse ST-segment depression with no ST-segment elevation or ventricular preexcitation. Physical examination findings were unremarkable. Amiodarone infusion at 1 mg/min was initiated, and he was transferred to the coronary care unit.

LEARNING OBJECTIVES

- To understand 12-lead ECG localization of VT exit sites.
- To understand the differences of VT in acute coronary syndrome and ARVC.
- To understand the evaluation and treatment of VT and ARVC.

PAST MEDICAL HISTORY

The patient had no past medical history. His family history was significant for a paternal grandmother who died of sudden death at age 55 years.

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ABBREVIATIONS AND ACRONYMS

ARVC = arrhythmogenic right ventricular cardiomyopathy

ECG = electrocardiogram

ICD = implantable cardioverter-defibrillator

IV = intravenous

LAD = left anterior descending artery

LB = left bundle

LV = left ventricle

RV = right ventricle

SMVT = sustained monomorphic ventricular tachycardia

TTE = transthoracic echocardiography

VT = ventricular tachycardia

INVESTIGATIONS

Laboratory test results revealed normal electrolytes and a high-sensitivity troponin I level that peaked at 4,813 ng/mL. Transthoracic echocardiography (TTE) revealed an ejection fraction of 40% to 45% with anterior apical wall motion abnormality and a severely dilated right ventricle (RV) with moderate dysfunction. Given concerning clinical findings for acute coronary syndrome, left heart catheterization was performed, which revealed 90% stenosis of the proximal to mid left anterior descending artery (LAD) (**Figure 2**). He underwent percutaneous coronary intervention with placement of a drug-eluting stent. Following successful coronary revascularization, questions regarding the etiology of his SMVT remained. Further analysis of the VT on the 12-lead ECG demonstrated an LB pattern in V_1 with a superiorly directed axis and near-complete negative precordial concordance of the QRS complexes, which was consistent with a VT origin from the RV apex. Myocardial ischemia caused by high-grade stenosis in his LAD could not explain the VT or the RV abnormalities seen on TTE and raised suspicion for an alternative etiology. Further investigation with cardiac magnetic resonance demonstrated severely dilated RV, end-diastolic volume index of 148 mL/m^2, and RV ejection fraction of 34% with basal to apical anterior dyskinesia and late gadolinium enhancement of the affected segments. The left ventricle (LV) size was normal, and the end-diastolic volume index was 86 mL/m^2 with an ejection fraction of 50%. (Video 1). The patient was diagnosed with definite arrhythmogenic RV cardiomyopathy (ARVC) after fulfilling 2 major criteria of the 2010 Revised Task Force Criteria because of the abnormal cardiac magnetic resonance findings and LB SMVT. Electrophysiology study was performed before implantable cardioverter-defibrillator (ICD) insertion to assess the inducibility of nonclinical VTs and effectiveness of antitachycardia pacing therapy. Programmed ventricular stimulation was positive for a single induced VT with the same QRS complex morphology and rate as the clinical VT (**Figure 3**). Voltage mapping or ablation was not performed. The paced QRS complex morphology from the catheter placed into the RV apex showed similar QRS complex morphology to the induced VT, which was successfully terminated with overdrive RV pacing.

MANAGEMENT

A dual-chamber ICD was inserted for secondary prevention. The lead was implanted into the midseptal segment of his RV with adequate sensing and pacing. Metoprolol succinate 25 mg daily was initiated.

DISCUSSION

VT caused by myocardial ischemia is often polymorphic and associated with an increased risk of ventricular fibrillation. The underlying mechanism of

FIGURE 1 Initial Electrocardiogram Demonstrating Monomorphic Ventricular Tachycardia With Left Bundle Superior-Axis QRS Complex

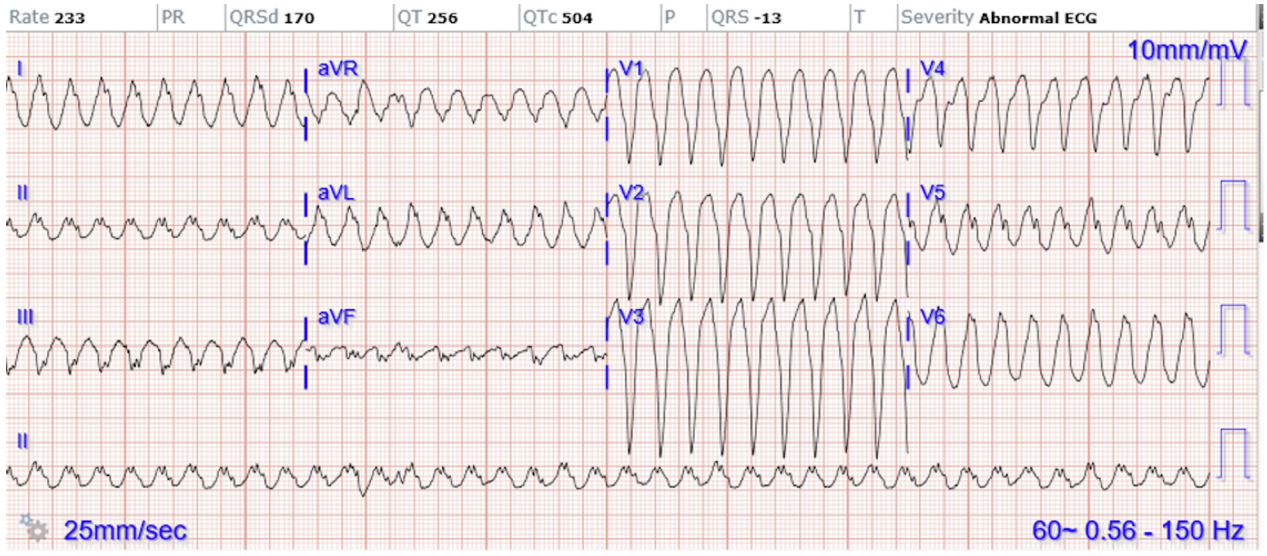
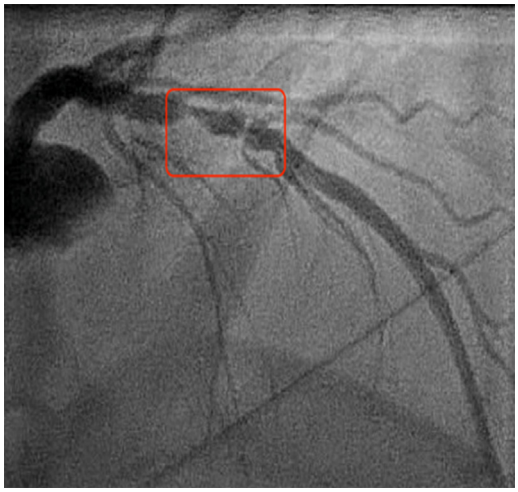


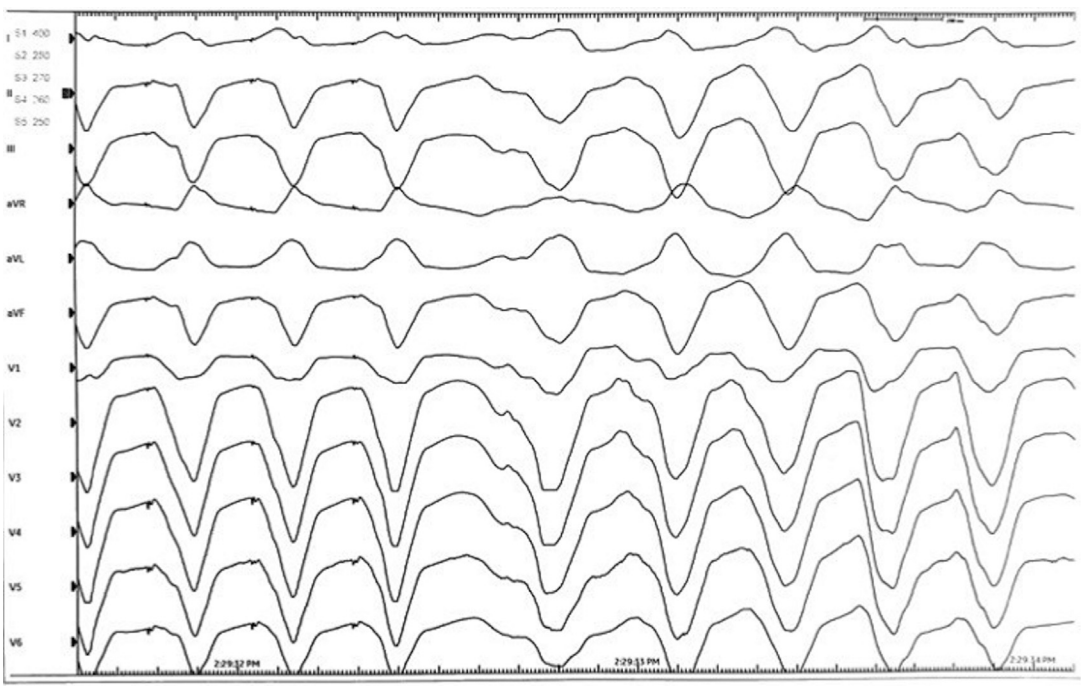
FIGURE 2 **Left Heart Catheterization Demonstrating Proximal to Mid Left Anterior Descending Artery Lesion With ~90% Stenosis**



ischemic-driven ventricular arrhythmia is triggered activity related to delayed after-depolarizations from intracellular calcium overload. SMVT is an uncommon presentation of acute myocardial ischemia and, if present, should lead to further investigation into an underlying cardiomyopathy with abnormal ventricular substrate.[1]

Ischemic cardiomyopathy is the most common cause of SMVT caused by myocardial re-entry related to scar from prior myocardial infarction.[2] VTs with an LB pattern on ECG originate from the RV or interventricular septum, and VTs with a right bundle pattern originate from the LV. The precordial transition is seen later if the origin is closer to the ventricular apex.[3] In this case, the LB pattern in V_1 with the superiorly directed axis with late precordial transition indicated an RV apical exit site, which would not have been expected from acute LAD ischemia. These discordant results led to further investigation with cardiac magnetic resonance and

FIGURE 3 **Electrophysiology Study Demonstrating Inducible Sustained Monomorphic Ventricular Tachycardia With a Left Bundle Superior-Axis QRS Complex**



electrophysiology study, which confirmed the diagnosis of ARVC.

ARVC is an inherited disease characterized by fibrofatty infiltration and replacement of the RV myocardium that presents clinically by causing ventricular arrhythmias of RV origin.[4] It is a progressive disease that leads to RV failure and can lead to LV failure in later stages of the disease.[5] Diagnosis of ARVC is made by clinical findings that fulfill either the major or minor criteria of the revised Task Force Criteria of 2010.[6] The most common ECG abnormality is T-wave inversion in precordial leads V_1 to V_3 not in the setting of a right bundle branch block pattern.[5] TTE often reveals a dilated RV with dysfunction and dyskinetic segments. Cardiac magnetic resonance provides excellent visualization of the RV for diagnosing wall motion abnormalities and the presence of scar or fat.[7] Electrophysiology study is typically performed to confirm the diagnosis of VT and to further evaluate causes of syncope.

The main treatment goals are to prevent sudden cardiac death and slow the progression of heart failure. Risk stratification of patients with ARVC who are at increased risk of ventricular arrhythmias and sudden cardiac death continues to remain a challenge. An ARVC risk calculator has been developed as a tool to aid in assessing arrhythmic risk. Electrophysiology study has recently been shown to predict the development of ventricular arrhythmia over a 5-year period and shows promise as a risk stratification tool.[8]

ICD insertion in patients with ARVC can be challenging; understanding areas to avoid based on cardiac magnetic resonance and local signal mapping are important factors to consider during implant.

FOLLOW-UP

The patient presented to our device clinic 3 months after discharge. ICD interrogation showed a 4-hour episode of SMVT with a heart rate of 164 beats/min, which was slower than his clinical VT. The ICD was reprogrammed to detect and treat the slower clinical VT, and sotalol was initiated. Genetic testing detected 2 variants of unknown significance in the *ALPK3* gene heterozygous for p.M765R and the *DSP* gene heterozygous for p.R1184W. Genetic counseling was provided to the patient, and family screening was performed.

CONCLUSIONS

Understanding the mechanisms and 12-lead ECG interpretation of SMVT are crucial parts of the clinical evaluation of VT. In this case, the etiology of the VT may have been mistakenly attributed to acute myocardial ischemia, and coronary revascularization alone would not have been adequate treatment. Further investigation to explain the discordant findings of LB VT morphology with RV anomalies ultimately led to the correct diagnosis and management of ARVC.

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KEY WORDS arrhythmogenic, ARVC, cardiomyopathy, palpitations, right ventricle, tachycardia

APPENDIX For a supplemental video, please see the online version of this paper.