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Shon Thomas
Corewell Health South, Saint Joseph, MI, shonmthomas@gmail.com

Jaclynn Jackubowski
Corewell Health South, Saint Joseph, MI

Kevin Ismair
Corewell Health South, Saint Joseph, MI

Jeremiah Chang
Corewell Health South, Saint Joseph, MI

Jerome Kuhnlein
Corewell Health South, Saint Joseph, MI

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CASE REPORT

Arrhythmogenic Right Ventricular Cardiomyopathy Presenting With VT Electrical Storm

Shon Thomas*, Jaclynn Jackubowski, Kevin Ismair, Jeremiah Chang, Jerome Kuhnlein

Corewell Health South, Saint Joseph, MI, USA

Abstract

Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC) is a right ventricular disease caused by desmosomal gene mutations leading to fibro-fatty replacement of the myocardium causing ventricular arrhythmias such as ventricular tachycardia (VT). A 59-year-old female presented with new onset VT manifesting as shortness of breath and chest discomfort. Diagnostic workup revealed right ventricular dilation/dysfunction on echocardiogram, VT with left bundle branch block (LBBB) and diffuse T wave inversions (TWIs) on EKG. She was discharged with a diagnosis of ARVC. Later, she developed VT storm, treated with epi-endocardial ablation for complete resolution of recurrent VT. Our case demonstrates the rare presentation of ARVC as ventricular storm and the various management strategies such as anti-arrhythmics, automated implantable cardioverter-defibrillator (AICD) and ablation procedures.

Keywords: Arrhythmogenic right ventricular dysplasia, Arrhythmogenic right ventricular cardiomyopathy, Ventricular tachycardia, Electrical storm

1. Introduction

ARVC is an inherited arrhythmogenic condition, caused by progressive replacement of fibrofatty deposits within the myocardium.[1] Prevalence of ARVC varies from 1 in 2500 in some European countries like Italy and Germany to 1 in 5000 in the general population, accounting for 5–10% of sudden cardiac deaths (SCD) in age below 65.[3,11] Distinct pathological features of ARVC are deposition of fibrous and fatty tissue along the free wall of the right ventricle (RV), leading to aneurysmal dilation among inflow and outflow tract and apex.[4] Measurement of RV strain on echocardiogram has been identified as a useful marker in determining patients at higher risk for progression of ARVC.[1] ARVC is a leading cause of SCD in young athletes.[4] The enlargement of the RV predisposes patients for lethal ventricular arrhythmias such as VT. This case highlights the uniqueness of ARVC management, including pharmacological and advanced procedural interventions (i.e. AICD, endo-epicardial ablation) to control fatal ventricular arrhythmias. Since ARVC patients are 23 times more likely to develop SCD, they need to be risk stratified for SCD and evaluated for AICD implantation.[14]

2. Case presentation

A 59-year-old female without significant past medical history presented to the emergency room (ER) with sudden onset shortness of breath and chest discomfort. She used an albuterol inhaler (from a previous bronchitis) the previous night and woke up with deep severe chest heaviness lasting 2 h. She endorsed palpitations when turning sideways. Additionally, she reported chills, rigor, abdominal discomfort, and severe malaise/weakness.

She reported symptoms started 9 days ago, starting with nausea, vomiting, and mid abdominal pain/cramp which worsened overnight with new chest heaviness. Symptoms resolved after waking up the next day. However, symptoms progressively worsened daily. She denied chest pain, paroxysmal nocturnal dyspnea, leg swelling, dizziness, or lightheadedness. Initial vitals were Temperature 98.2 deg F, Blood Pressure 89/62 mmHg, Heart Rate 200 bpm, Respiratory Rate 24 breaths per minute, SpO2

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* Corresponding author at: 3425 Lakeshore Drive, Unit 138, Saint Joseph, MI 49085, USA.
E-mail address: shonmthomas@gmail.com (S. Thomas).

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100% on room air. On exam, she appeared fatigued, tender to palpation of periumbilical region, tachycardic, lungs were clear to auscultation and peripheral pulses were normal. She endorsed drinking 1–2 glasses of wine weekly and denies smoking. She had no significant family history of cardiac diseases. Labs remarkable for glucose 197 mg/dl, BUN 50 mg/dl, creatinine 1.4 mg/dl, EGFR 43 ml/min, sodium 131 mmol/L, potassium 3.9 mmol/L, magnesium 2.4 mg/dl, CO2 17 mmol/L, anion gap 16 mmol/L, AST 517 U/L, ALT 443 U/L, ALK 64 U/L, bilirubin 0.4 mg/dl, lipase 11 U/L and mag 2.6 mg/dl. High-sensitivity troponin was elevated to 148 ng/L and down trended to 133 ng/L on 4 h repeat. Initial EKG (Fig. 1) showed ventricular tachycardia with HR 209 with LBBB.

3. Investigations

CT angiogram done in the ER revealed small pericardial effusions, bilateral pleural effusions and no pulmonary embolisms.

Cardiology was consulted and she underwent left heart catheterization (LHC) which showed normal coronaries. LHC also confirmed mild-to-moderate left ventricular dysfunction (EF 40–45%) with moderate anterolateral and apical hypokinesis. Takotsubo cardiomyopathy was a possibility due to the apical hypokinesis and regional wall motion abnormality expanding beyond a single epicardial vascular territory in the absence of coronary artery disease, but difficult to determine initially as it's a diagnosis of exclusion.[15] Echocardiogram showed mildly decreased LVEF 45–50%, mild global hypokinesis, moderately dilated RV, and reduced RV systolic function. Of note, patient's prior exercise stress echocardiography done 2 years ago for an episode of chest pain associated with an upper respiratory illness was unremarkable with EF 55–60%.

Patient met 3 major criteria for ARVC (TWIs in V2–V6, RV wall motion abnormality and dilation/dysfunction, and VT with LBBB pattern). Of note, endomyocardial biopsy is a rarely used diagnostic criteria due to sampling error; it was not done in this patient.[9]

4. Management

After the initial EKG, she was given 150 mg amiodarone IV, 100 mg lidocaine IV, 20 mEq potassium PO and 2 g magnesium IV without any effect. Her blood pressure subsequently decreased from 113/82 to 90/74. Given her unstable vitals, she was cardioverted with 150 J electrically once to normal sinus rhythm (NSR). Post cardioversion EKG (Fig. 2) demonstrated new anterolateral and inferolateral TWIs.

Electrophysiology (EP) was consulted and planned to insert a dual chamber AICD. Guideline directed medical therapy (GDMT) was initiated with lisinopril and carvedilol for new onset non-ischemic systolic heart failure with mid-range EF (HFmrEF). Of note, the role of SGLT2-I was not realized then. She was discharged after AICD placed and device check completed, to be followed up as outpatient with a cardiac MRI and genetic testing.

Cardiac MRI (Fig. 3) done 2 months after discharge noted severe dilated RV, moderately reduced RV systolic function and hypokinesis of basal to mid RV segments. The RV is severely dilated by four chamber RV measurements (RV basal diameter 51 mm and RV mid diameter 42 mm). RV systolic function is moderately reduced by visual estimation. There is hypokinesis of the basal to mid segments of the RV with preserved apical contractility. This met

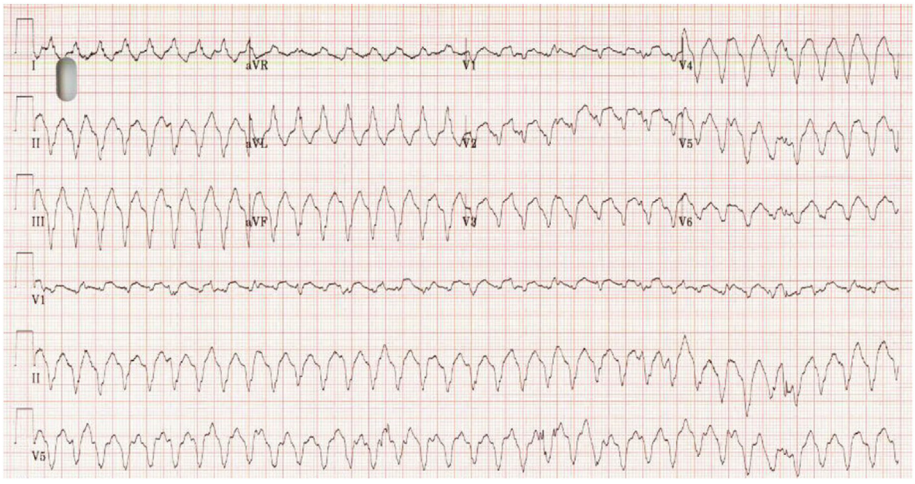

Fig. 1. Initial EKG 10/23/22. Monomorphic Ventricular Tachycardia with HR 209. QRS 126 with left bundle branch morphology.

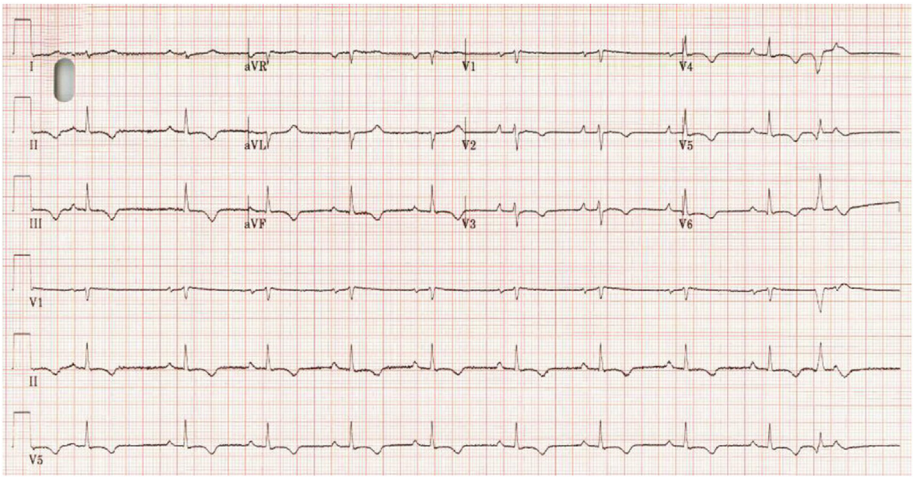

Fig. 2. Post cardioversion EKG 10/23/22 with sinus bradycardia HR 55 and new T wave inversions in leads II, III, aVF, V2–V6. No acute ST changes.

one major imaging criteria for ARVC per the 2020 International Task Force Consensus (ITFC) for ARVC (i.e. RV dilation and systolic dysfunction).[9,10] Furthermore, outpatient genetic testing reported 3 genes of undetermined significance including DSG2, KCNH2 and TPM1.

Patient later presented to the ER, 2.5 months after discharge, with multiple monomorphic VT (i.e. VT storm) with HR 190s and eight AICD shocks; she eventually cardioverted in the ER. She endorsed prodromal symptoms such as lightheadedness and presyncope without syncope. EP was consulted and she was started on amiodarone and lidocaine continuous IV infusion, later transitioned to oral amiodarone and mexiletine. Labs showed normal troponin levels. She remained in NSR without further VT episodes; she was then discharged with oral amiodarone 200 mg BID, mexiletine 150 mg TID, and increased carvedilol 6.25 mg BID. Patient followed up with EP outpatient and completed VT ablation with endo-epicardial approach one month after the second admission. Spironolactone 12.5 mg daily and empagliflozin 10 mg daily were also started at this time for HFmrEF. She was discharged and, after 1 year, she had recurrent VT requiring cardioversion and repeat endo-epicardial ablation. Patient has not had any VT recurrence per device checks since ablation to date.

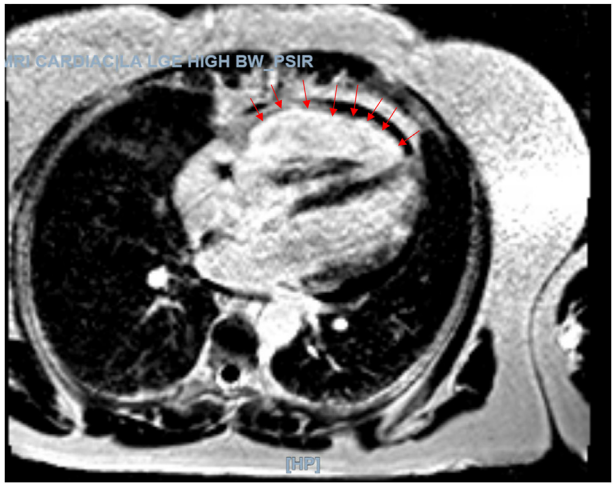

Fig. 3. Cardiac MRI 01/12/23 with severely dilated right ventricle (red arrows) with moderately reduced systolic function by visual estimation. In conjunction with hypokinesis of the basal to mid right ventricular segments, findings fulfill one major imaging criteria for the diagnosis of arrhythmogenic right ventricular cardiomyopathy.

5. Discussion

ARVC, formerly known as Arrhythmogenic Right Ventricular Dysplasia, is a right ventricular disease which has fibro-fatty replacement of the myocardium causing ventricular arrhythmia. Ventricular dilation can occur at apex, outflow tract or inflow tract, commonly sparing the septum. The disease initially starts with regional wall motion abnormalities and progresses to global abnormalities, causing RV dilation. Genetic causes of ARVC are multifactorial but have been associated with desmosomal gene mutations.[2]

The ITFC developed an algorithm to stratify the risk of SCD in patients with ARVC.[5] The degree of SCD risk (Class I (high), II a and b (intermediate), III (low)) would be determined based on genetic mutation (proband > carrier), sex (males > female), EKG (sustained VT > non-sustained VT, >/= 3 precordial TWI), echocardiogram (severe > moderate LV and/or RV dysfunction) and symptoms (aborted cardiac arrest > syncope).

Management strategies for ARVC include GDMT, antiarrhythmics, AICD and, in refractory cases of ARVC associated VT, radiofrequency catheter ablation (RFCA).[6] AICD is indicated for Class I and possibly Class IIa/b; however, AICD isn't indicated for Class III. An AICD is recommended for primary prevention in high-risk ARVC patients with family history of SCD, extensive disease, or syncope, and secondary prevention.[7] Early implantation of AICD has mortality benefits.[13] Medical management with antiarrhythmics can be trialed prior to AICD, but it does not reduce the risk of SCD.[12]

Antiarrhythmics such as amiodarone and sotalol may reduce VT or ventricular fibrillation (VF) and are used in patients unable to tolerate an AICD placement. RFCA can be used as an adjunctive therapy in ARVC since long term efficacy of RFCA in ARVC is limited. Specifically, combined endo-epicardial ablation has a lower risk of VT recurrence and subsequent mortality than endo only VT ablation in patients with scar-related VT.[8]

Our case highlights an older female patient with ARVC, as opposed to the typical younger athletic male, presenting with VT storm. She presented with RV dysplasia on echocardiogram, monomorphic VT with LBBB pattern on Fig. 1 and NSR diffuse TWIs on Fig. 2. The typical epsilon wave wasn't present in her case; however, she met other diagnostic major criteria, including echocardiogram, cardiac MRI and EKG findings. Because ARVC associated VT increases risk for SCD in our patient (Class I), antiarrhythmic therapy, AICD and adjunctive endo-epicardial ablation was used to treat her VT storm. Recognizing the various diagnostic criteria for ARVC is important in managing these high-risk structural heart diseases successfully.

6. Conclusion

ARVC remains a rare but well recognized cause of right sided heart failure and appropriate evaluation and management with AICD and/or ablation (specifically endo-epicardial ablation) may reduce mortality and morbidity associated with ventricular arrhythmias secondary to ARVC. Our case study highlights the appropriate evaluation and diagnostic criteria as highlighted by the 2020 ITFC for ARVC, utilizing EKG, echocardiogram, cardiac MRI, family history, and genetic testing.

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Ethics information

None.

Conflict of interest

There are no conflict of interest.

Disclaimer

None.

Disclosures

None.

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