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นิพนธ์ต้นฉบับ ORIGINAL ARTICLE

รายงานผู้ป่วยโรคกล้ามเนื้อหัวใจห้องล่างขวาชนิดอะริทโมจีนิก (Arrhythmogenic Right Ventricular Cardiomyopathy; ARVC) ที่มาด้วยภาวะหัวใจเต้นผิดจังหวะ

A Case Report of Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC) Presented With Symptomatic Arrhythmia

ทวิเทพ เลี้ยววัฒนา พ.บ.[1*], สุระ บุญรัตน์ พ.บ.[1], พรวลี ปรปักษ์ขาม พ.บ.[1]

Tawithep Leowattana, M.D.[1*], Sura Boonrat, M.D.[1], Pornwalee Porapakkham, M.D.[1]

[1]กลุ่มงานอายุรศาสตร์หัวใจ สถาบันโรคทรวงอก นนทบุรี

[1] Department of Cardiology, Central Chest Institute of Thailand, Nonthaburi,

*Corresponding author: tomojungbesty@gmail.com

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บทคัดย่อ

ภูมิหลัง: โรคกล้ามเนื้อหัวใจห้องล่างขวาชนิดอะริทโมจีนิก (Arrhythmogenic Right Ventricular Cardiomyopathy; ARVC) เป็นโรคกล้ามเนื้อหัวใจที่ถ่ายทอดทางพันธุกรรม โดยมีผลต่อหัวใจห้องล่างขวาเป็นหลัก ส่งผลให้เกิดการแทนที่ของเนื้อกล้ามเนื้อหัวใจด้วยเนื้อเยื่อพังผืดและไขมัน (fibrofatty replacement) อย่างต่อเนื่อง นำไปสู่การขยายตัวของหัวใจห้องล่างขวาและการทำงานบีบตัวของหัวใจที่ลดลง ผู้ป่วยมักมีอาการทางคลินิกที่หลากหลาย ตั้งแต่ไม่มีอาการไปจนถึงการเสียชีวิตอย่างฉับพลันจากภาวะหัวใจเต้นผิดจังหวะ ดังนั้น การตรวจพบโรคตั้งแต่ระยะเริ่มต้นจึงมีความสำคัญอย่างยิ่ง **รายงานผู้ป่วย:** รายงานผู้ป่วยรายนี้เป็นชายอายุ 65 ปี ซึ่งแม้จะมาด้วยอาการเด่นคือใจสั่นและหัวใจเต้นเร็ว แต่ได้รับการวินิจฉัยว่าเป็น ARVC จากลักษณะทางคลื่นไฟฟ้าหัวใจที่จำเพาะ ได้แก่ ภาวะหัวใจเต้นเร็วจากหัวใจห้องล่างที่มี QRS กว้าง (wide-QRS-complex ventricular tachycardia; VT) การพบ epsilon waves และการกลับของคลื่น T (T-wave inversions) โดยได้รับการยืนยันด้วยการตรวจคลื่นเสียงสะท้อนหัวใจ (echocardiography) และการตรวจภาพหัวใจด้วยคลื่นแม่เหล็กไฟฟ้า (cardiac magnetic resonance imaging; CMR) ซึ่งแสดงให้เห็นการขยายตัวของหัวใจห้องล่างขวา ค่า ejection fraction ที่ลดลง การมี trabeculations ที่เด่นชัด และการสะสมของแกโดลิเนียมล่าช้า (delayed gadolinium enhancement) ผู้ป่วยได้รับการรักษาด้วยยา ได้แก่ ยากลุ่มเบตา-บล็อกเกอร์ (beta-blockers) และอะมิโอดาโรน (amiodarone) และได้รับการฝังเครื่องกระตุกหัวใจด้วยไฟฟ้าอัตโนมัติ (implantable cardioverter-defibrillator; ICD) เพื่อการป้องกันการเสียชีวิตจากภาวะหัวใจเต้นผิดจังหวะเป็นทุติยภูมิ (secondary prevention) บทอภิปรายกล่าวถึงเกณฑ์การวินิจฉัยของ European Task Force ปี ค.ศ. 2023 บทบาทในการก่อโรคของการกลายพันธุ์ของยีนที่เกี่ยวข้องกับเดสโมโซม (desmosomal gene mutations) และแนวทางการรักษาในปัจจุบัน ซึ่งประกอบด้วยการปรับเปลี่ยนพฤติกรรมและวิถีชีวิต การรักษาด้วยยาต้านภาวะหัวใจเต้นผิดจังหวะ (antiarrhythmic therapy) การจี้ด้วยสายสวน (catheter ablation) และการฝังอุปกรณ์ช่วยรักษาจังหวะหัวใจ โดยมีเป้าหมายเพื่อลดความเสี่ยงของการเสียชีวิตจากภาวะหัวใจหยุดเต้นฉับพลันและปรับปรุงผลลัพธ์ทางคลินิก **สรุป:** รายงานผู้ป่วยรายนี้เน้นย้ำถึงความสำคัญของการตรวจพบโรคตั้งแต่ระยะเริ่มต้น การประเมินด้วยการตรวจภาพหัวใจอย่างครอบคลุม และการให้การรักษาอย่างทันท่วงทีในการดูแลผู้ป่วยโรค ARVC

Abstract

Background: Arrhythmogenic right ventricular cardiomyopathy (ARVC) is a hereditary cardiomyopathy that predominantly affects the right ventricle, leading to progressive fibrofatty replacement, dilation, and systolic dysfunction. Patients often present with varied clinical manifestations ranging from asymptomatic to sudden arrhythmic death, making early recognition crucial. **Case Presentation:** Our case illustrates a 65-year-old male who, despite a predominant clinical presentation of palpitations and tachycardia, was diagnosed with ARVC following characteristic electrocardiographic findings (wide-QRS-complex VT, epsilon waves, T-wave inversions) and confirmed by echocardiography and cardiac magnetic resonance imaging (CMR) demonstrating right-ventricular dilation, reduced ejection fraction, prominent trabeculations, and delayed gadolinium enhancement. The patient received pharmacologic therapy with beta-blockers and amiodarone, and underwent implantation of an implantable cardioverter-defibrillator for secondary prevention. The discussion highlights the 2023 European Task Force diagnostic criteria, the pathogenic role of desmosomal gene mutations, and current management strategies—including lifestyle modification, antiarrhythmic therapy, catheter ablation, and device implantation - to reduce sudden cardiac death and improve outcomes. **Conclusions:** This case underscores the importance of early detection, comprehensive imaging, and timely intervention in ARVC management.

Keywords: Arrhythmogenic right ventricular cardiomyopathy (ARVC), Right-ventricular dilation, Epsilon waves, Monomorphic ventricular tachycardia, RV dyskinesia / dyssynchrony, Sudden cardiac death (SCD)

INTRODUCTION

ARVC is an inherited form of cardiomyopathy, predominantly affecting the right ventricle, and mostly seen in young adults. It is characterized histologically by fibrofatty replacement of RV myocardium, resulting in subsequent RV dilation and systolic dysfunction. Due to its wide range of clinical signs and symptoms, varying from being completely asymptomatic to fatal arrhythmia and sudden death, ARVC frequently goes undiagnosed.[1-3] Thus, clinical suspicion is of key importance in diagnosing this condition. We present here a case of ARVC who has presented with VT, and was diagnosed based on ECG and CMR findings.

CASE DESCRIPTION

Our patient is a 65-year-old male with history of hypertension presented to the emergency department with sudden onset palpitations associated with profuse sweating. Further history taking revealed previous episodes of palpitations with constant feeling of exhaustion. He also had a history of nonspecific, non-radiating chest pain, which occurred during exertion and had improvement with rest. Physical examination found no specific findings

except regular tachycardia. Initial clinical findings showed a blood pressure of 103/63 mmHg and a heart rate (HR) of 192 beats per minute. An electrocardiogram (ECG) was subsequently done, revealing a regular wide QRS-complex tachycardia with atrioventricular dissociation, consistent with monomorphic ventricular tachycardia (VT) (Fig. 1). The patient was considered to have stable VT, and was put on maintenance intravenous Amiodarone. Upon arrival at Coronary Care Unit (CCU), he remained tachycardiac and still experienced palpitations and sweating. A light sedative medication was given, by which his heart rate then returned to normal sinus rhythm shortly after. The ECG in sinus rhythm revealed a normal heart axis, an incomplete right bundle block (RBB), and epsilon waves following the QRS in V2-V4, II, III, aVF, as well as negative T waves in the anteroseptoapical leads (Fig. 2).

A following echocardiography showed an enlarged right ventricle (RV) with poor systolic function (RV FAC 12.9%), with mildly dilated tricuspid annulus and mild tricuspid regurgitation. The left ventricle (LV) was non-dilated, with good systolic function (LVEF at 68%) (Fig. 3). Due to high suspicion of ARVC, the patient underwent a subsequent magnetic resonance imaging (MRI) scan, which revealed a markedly dilated RV with impaired systolic function (RVEF 25%), being in concordance with echocardiography findings. There were also prominent, thickened trabeculae within the RV and abnormal RV wall motion abnormalities, including regional dyssynchronous contraction (Accordion sign) in the sub-tricuspid region, dyskinesia and paradoxical movement most notable at the anterior free wall and the right ventricular apex. Right ventricular microaneurysms were also noted. Diffuse delayed gadolinium enhancement (DGE) was seen involving the RV free wall, RV inflow and outflow, and RV apex. DGE was also visible in RA wall. Left ventricle appeared normal from the study (Fig.4). .Based on the clinical presentation and work—up, a definite diagnosis of ARVC was made. The patient was treated with β-blockers and underwent automated implantable cardioverter defibrillator (AICD) implantation as secondary prevention.

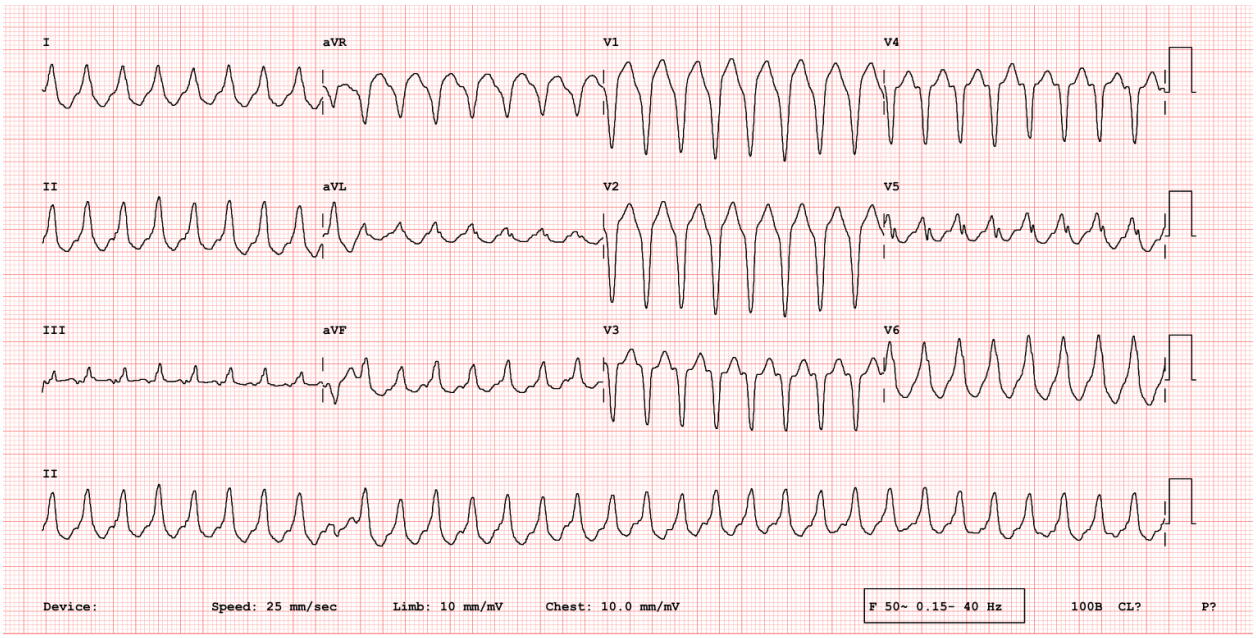


Figure 1. 12-lead EKG showing ventricular tachycardia with left bundle branch block pattern and positive QRS in inferior leads.

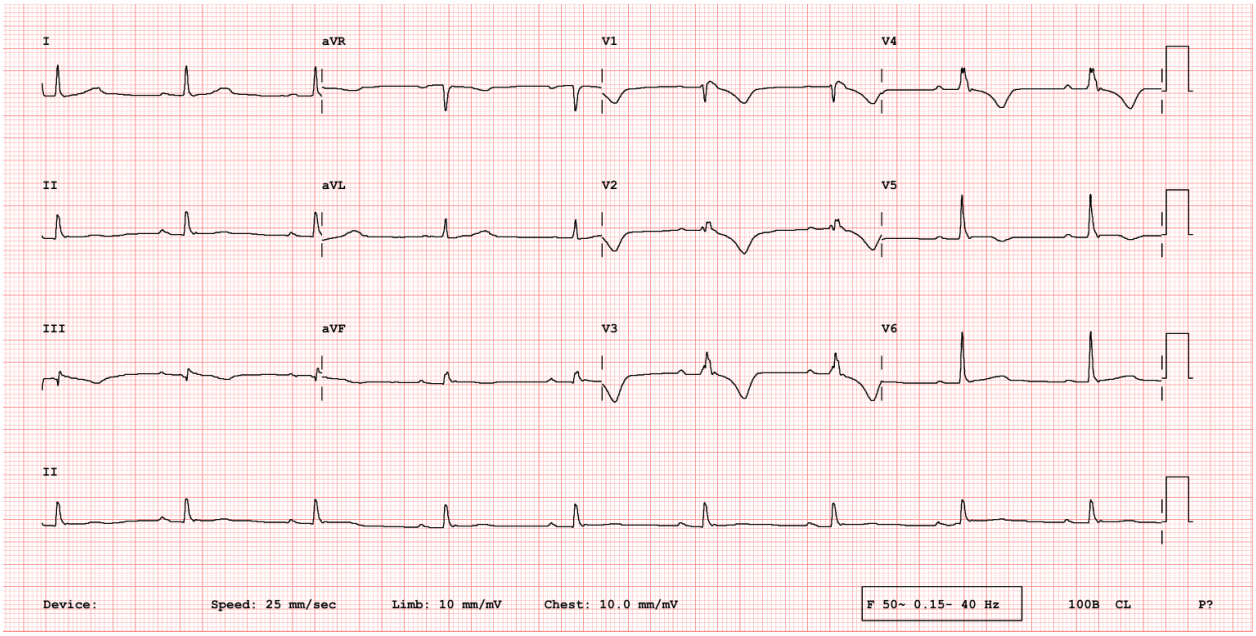


Figure 2. Resting 12-lead EKG showing symmetric T-wave inversion in right precordial leads (V1, V2 and V3) (black arrows). Epsilon waves can be seen in inferior and right precordial leads (red arrows).

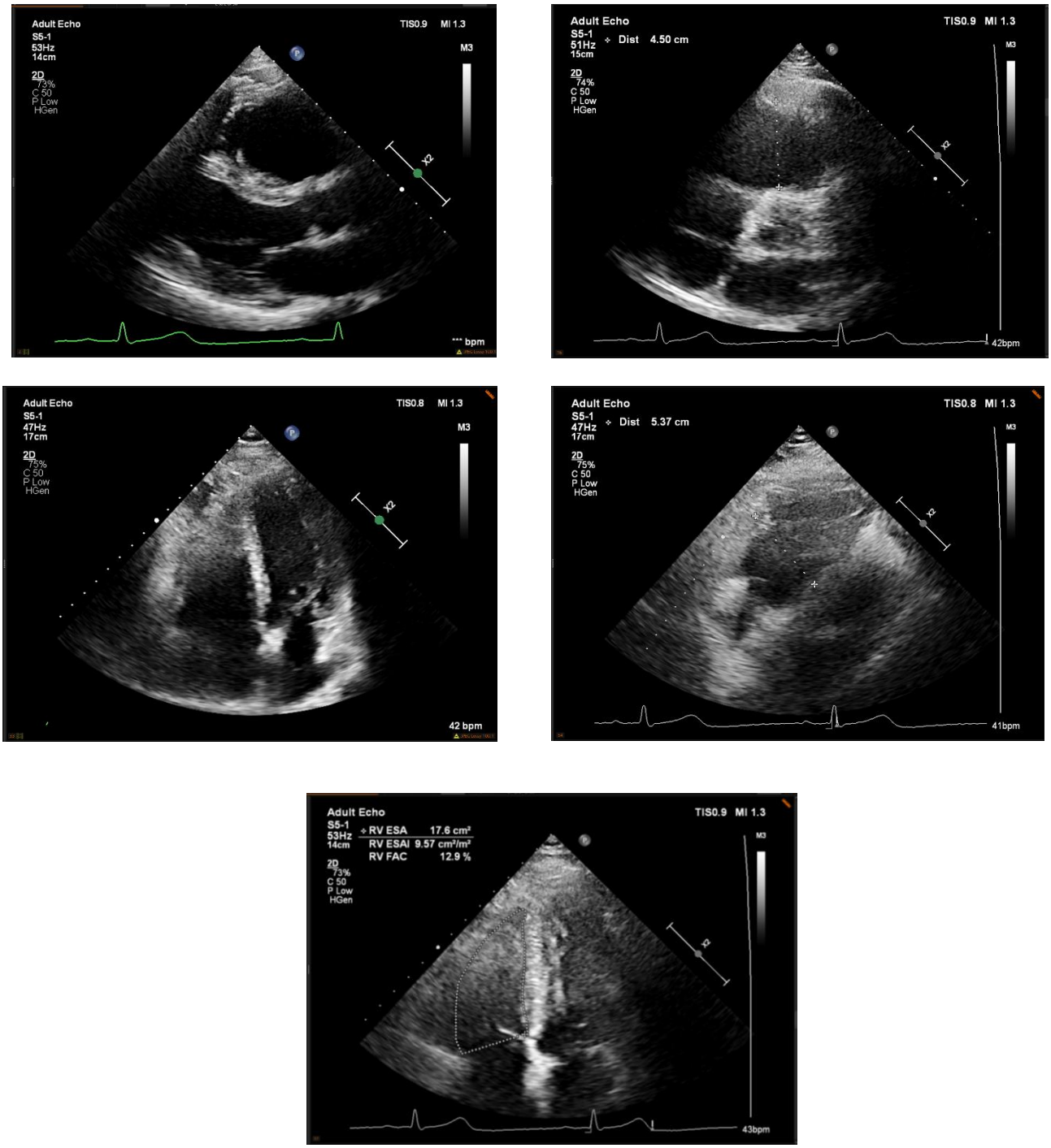


Figure 3. Echocardiography showing right ventricular dilatation and impaired right ventricular systolic function.

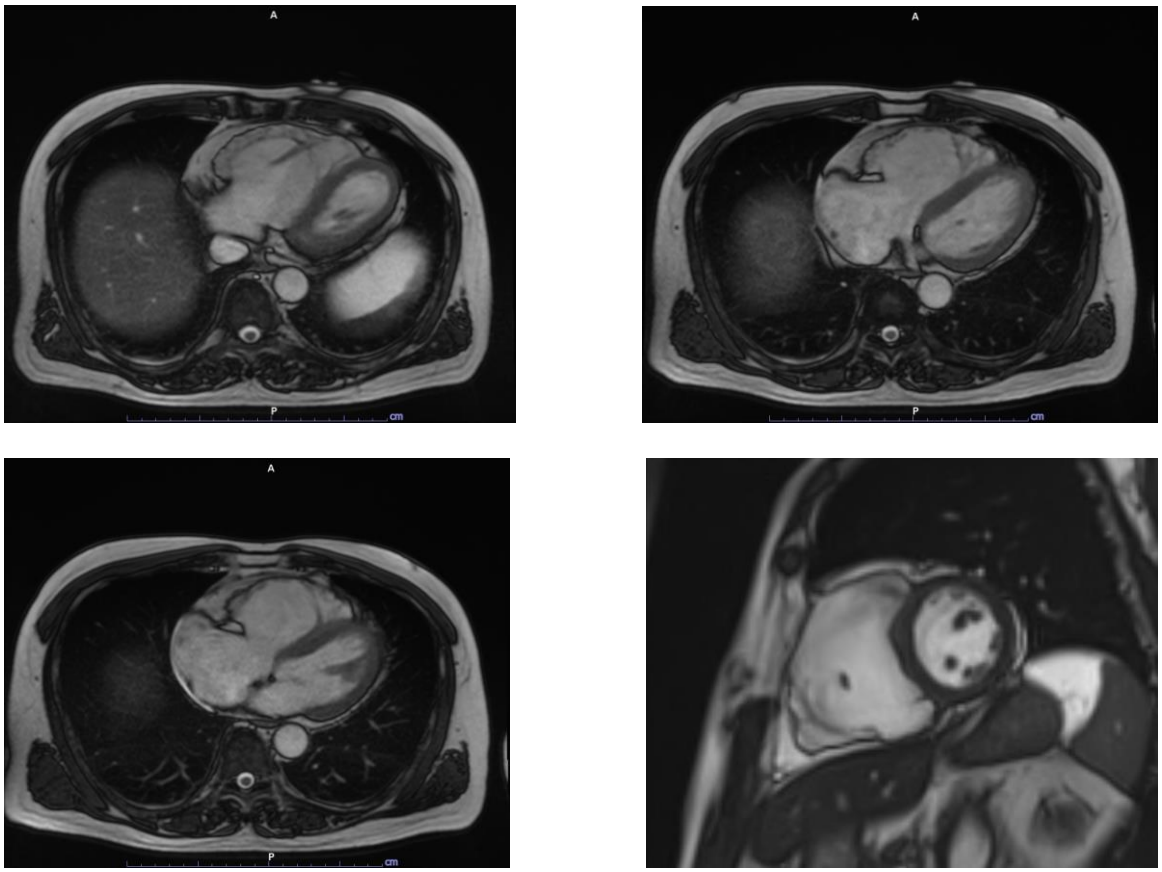


Figure 4. Cardiac MRI sequence Cine-MRI showing marked RV dilatation. There are prominent, thickened trabeculae within the RV (white arrow) and abnormal right ventricle wall motion abnormalities including regional dyssynchronous contraction (Accordion sign) in the subtricuspid region, dyskinesia and paradoxical movement most notable at the anterior free wall and the right ventricle apex (red arrow).

DISCUSSION

ARVC is a cardiac muscle disease which is characterized by prominent, severe ventricular arrhythmias, especially right ventricle. The updated 2023 European Task Force Criteria was used for diagnosis (Table 1). Similar to previous criteria[4,15] (e.g. Revised Task Force criteria in 2010 – Table 2), the new European TF diagnostic criteria distinguish three consequent steps in the diagnostic process including (1) ventricular involvement; (2) phenotypic definition; (3) etiology and classification, as well as maintain the same "six-category structure" as the previous ones and focus on the specific involvement of right, left or both ventricles using a multiple step diagnosis process. At least 2 major criteria, or 1 major and 2 minor criteria, or 4 minor criteria, are required for definite diagnosis.

Our patient's clinical findings were consistent with definite diagnosis of ARVC according to 2023 European Task Force Criteria, which includes the following:

(1) **Morphological criteria**
- Major criterion: Regional RV dyskinesia or dyssynchronous
  - RV EDV/BSA 110 ml/m2 (male)

- RV ejection fraction 25%

(2) Repolarization abnormalities criteria

- Major criterion: Inversion of T-waves in right precordial leads (V1-V3) in the absence of complete RBBB and not preceded by J-point/ST-segment elevation

(3) Depolarization abnormalities criteria

- Minor criterion: Epsilon waves in right precordial leads (V1-V3)

(4) Arrhythmias criteria

- Minor criterion: sustained ventricular tachycardia of LBBB morphology with inferior axis

Definite diagnosis was made according to 2 major criteria being met.

Category	Criteria for RV involvement	Criteria for LV involvement
1. Morpho-functional ventricular abnormalities	*Major* • Regional RV akinesia, dyskinesia, or aneurysm plus one of the following: - global RV dilatation (increase of RV EDV according to the imaging test specific nomograms for age, sex and BSA) *or* - global RV systolic dysfunction (reduction of RV EF according to the imaging test specific nomograms for age and sex) *Minor* • Regional RV akinesia, dyskinesia or aneurysm of RV free wall	*Minor* • Global LV systolic dysfunction, with or without LV dilatation (increase of LV EDV according to the imaging test specific nomograms for age, sex, and BSA)
2. Structural myocardial abnormalities	*Major* • Fibrous replacement of the myocardium in ≥1 sample, with or without fatty tissue, at histology *Minor* • Unequivocal RV LGE (confirmed in 2 orthogonal views) in ≥1 RV region(s) (excluding tricuspid valve)	*Major* • "Ring-like" LV LGE (subepicardial or midmyocardial stria pattern) of ≥3 segments (confirmed in 2 orthogonal views) *Minor* • LV LGE (subepicardial or midmyocardial stria pattern) of 1 or 2 Bull's Eye segment(s) (in 2 orthogonal views) of the free wall, septum, or both (excluding patchy, focal or septal junctional LGE)
3. ECG repolarization abnormalities	*Major* • Negative T waves in right precordial leads (V1, V2, and V3) or beyond in individuals ≥14-year-old (in the absence of complete RBBB and not preceded by J-point/ST-segment elevation) *Minor* • Negative T waves in leads V1 and V2 in males ≥14-year-old (in the absence of RBBB and not preceded by J-point/ST-segment elevation) • Negative T waves beyond V3 in the presence of complete RBBB • Negative T waves beyond V3 in individuals <14-year-old	*Minor* • Negative T waves in left precordial leads (V4–V6) (in the absence of complete LBBB)
4. ECG depolarization and conduction abnormalities	*Minor* • Epsilon wave (reproducible low-amplitude signals between end of QRS complex to onset of the T wave) in the right precordial leads (V1 to V3) • Terminal activation duration of QRS ≥55 ms measured from the nadir of the S wave to the end of the QRS, including R', in V1, V2, or V3 (in the absence of complete RBBB)	*Major* • Low QRS voltages (<0.5 mV peak to peak) in all limbs leads in the absence of other causes (e.g., cardiac amyloidosis, obesity, emphysema, or pericardial effusion)
	ular tachycardia of LBBB morphology with non-inferior axis *Minor* • Frequent ventricular extrasystoles (>500 per 24 h), non-sustained or sustained ventricular tachycardia of LBBB morphology with inferior axis ("RVOT pattern") • History of cardiac arrest due to ventricular fibrillation or sustained ventricular tachycardia of unknown morphology	a RBBB morphology or multiple RBBB morphologies (excluding the "fascicular pattern") • Non-sustained or sustained ventricular tachycardia with a RBBB morphology (excluding the "fascicular pattern") • History of cardiac arrest due to ventricular fibrillation or sustained ventricular tachycardia of unknown morphology
6. Family history/genetics	*Major* • Identification of a pathogenic ACM-gene variant in the patient under evaluation • ACM confirmed in a first-degree relative who meets diagnostic criteria • ACM confirmed pathologically at autopsy or surgery in a first-degree relative *Minor* • Identification of a likely-pathogenic ACM-gene variant in the patient under evaluation • History of ACM in a first-degree relative in whom it is not possible or practical to determine whether the family member meets diagnostic criteria • Premature sudden death (<35 years of age) due to suspected ACM in a first-degree relative • ACM confirmed pathologically or by diagnostic criteria in second-degree relative	

ACM, arrhythmogenic cardiomyopathy; BSA, body surface area; ECG, electrocardiogram; EDV, end diastolic volume; EF, ejection fraction; LBBB, left bundle branch block; LGE, late gadolinium enhancement; LV, left ventricle; RBBB, right bundle branch block; RV, right ventricle; RVOT, right ventricular outflow tract. Adapted from [13].

Table 1: European Task Force criteria for diagnosis of arrhythmogenic cardiomyopathy.[4]

Major Criteria	Minor Criteria
I. Imaging	
By 2D echo: Regional RV akinesia, dyskinesia, or aneurysm And 1 of the following (end diastole): PLAX RVOT ≥32 mm (corrected for body size [PLAX/BSA] ≥19 mm/m^2) PSAX RVOT ≥36 mm (corrected for body size [PSAX/BSA] ≥21 mm/m^2) Fractional area change ≤33% By CMR: Regional RV akinesia or dyskinesia or dyssynchronous RV contraction And 1 of the following: Ratio of RV end-diastolic volume to BSA ≥110 ml/m^2 (male) or ≥100 ml/m^2 (female) RV ejection fraction ≤40% By RV angiography: Regional RV akinesia, dyskinesia, or aneurysm	By 2D echo: Regional RV akinesia or dyskinesia And 1 of the following (end diastole): PLAX RVOT ≥29 to <32 mm (corrected for body size [PLAX/BSA] ≥16 to <19 mm/m^2) PSAX RVOT ≥32 to <36 mm (corrected for body size [PSAX/BSA] ≥18 to <21 mm/m^2) Fractional area change >33% to ≤40% By CMR: Regional RV akinesia or dyskinesia or dyssynchronous RV contraction And 1 of the following: Ratio of RV end-diastolic volume to BSA ≥100 to <110 ml/m^2 (male) or ≥90 to <100 ml/m^2 (female) RV ejection fraction >40% to ≤45%
II. Endomyocardial Biopsy	
Residual myocytes (60% by morphometric analysis or 50% if estimated), with fibrous replacement of the RV free wall myocardium in ≥1 sample, with or without fatty replacement of tissue on endomyocardial biopsy	Residual myocytes (60% to 75% by morphometric analysis or 50% to 65% if estimated), with fibrous replacement of the RV free wall myocardium in ≥1 sample, with or without fatty replacement of tissue on endomyocardial biopsy
III. Repolarization Abnormalities	
Inverted T waves in right precordial leads (V_1, V_2, and V_3) or beyond in individuals >14 years of age (in the absence of complete right bundle-branch block QRS ≥120 ms)	Inverted T waves in leads V_1 and V_2 in individuals >14 years of age (in the absence of complete right bundle-branch block) or in V_4, V_5, or V_6 Inverted T waves in leads V_1, V_2, V_3, and V_4 in individuals >14 years of age in the presence of complete right bundle-branch block
IV. Depolarization/Conduction Abnormalities	
Epsilon wave (reproducible low-amplitude signals between end of QRS complex to onset of the T-wave) in the right precordial leads (V_1 to V_3)	Late potentials by SAECG in ≥1 of 3 parameters in the absence of a QRS duration of ≥110 ms on the standard ECG Filtered QRS duration (fQRS) ≥114 ms Duration of terminal QRS <40 μV (low-amplitude signal duration) ≥38 ms Root mean square voltage of terminal 40 ms ≤20 μV Terminal activation duration of QRS ≥55 ms measured from the nadir of the S-wave to the end of the QRS, including R′, in V_1, V_2, or V_3, in the absence of complete right bundle-branch block
V. Arrhythmias	
Nonsustained or sustained ventricular tachycardia of left bundle-branch morphology with superior axis (negative or indeterminate QRS in leads II, III, and aVF and positive in lead aVL)	Nonsustained or sustained ventricular tachycardia of RV outflow configuration, left bundle-branch block morphology with inferior axis (positive QRS in leads II, III, and aVF and negative in lead aVL), or of unknown axis >500 ventricular extrasystoles per 24 h (Holter)
VI. Family History	
ARVC confirmed in a first-degree relative who meets current TFC ARVC confirmed pathologically at autopsy or surgery in a first-degree relative Identification of a pathogenic mutation categorized as associated or probably associated with ARVC in the patient under evaluation	History of ARVC in a first-degree relative in whom it is not possible or practical to determine whether the family member meets current TFC Premature sudden death (<35 years of age) due to suspected ARVC in a first-degree relative ARVC confirmed pathologically or by current TFC in a second-degree relative

2D = 2-dimensional; ARVC = arrhythmogenic right ventricular cardiomyopathy; aVF = augmented voltage unipolar left foot lead; aVL = augmented voltage unipolar left arm lead; BSA = body surface area; CMR = cardiac magnetic resonance; ECG = electrocardiogram; echo = echocardiography; PLAX = parasternal long-axis view; PSAX = parasternal short-axis view; RV = right ventricle/ventricular; RVOT = right ventricular outflow tract; SAECG = signal averaged electrocardiogram; TFC = Task Force Criteria.

Table 2: 2010 Revised Task Force Criteria for Diagnosis of ARVC[14]

ARVC is a familial disease but genetic penetrance can be low and hence disease expression is highly variable. The underlying pathogenesis is presumed to be the mutation of genes encoding desmosomes, which are intercellular adhesion complexes in cardiac myocytes. Malfunctioning of these desmosomes result in myocyte detachment and cellular death. This leads to inflammation of affected myocyte, subsequently resulting in apoptosis and fibro-fatty replacement of the myocardium.[5-6]

ARVC management aims to slow the progression of the disease, minimize the risk of sudden cardiac death, and enhance the quality of life. The management of ARVC involves a range of approaches, such as clinical management, pharmacological treatment, catheter ablation, implantation of a cardiac defibrillator. Clinical management primarily involves restricting physical activity, because exercise can trigger arrhythmias and accelerate disease progression, which may

lead to sudden cardiac death.[7] Pharmacological treatment for ARVC aims to suppress arrhythmia and prevent heart failure. Cardio-selective beta-blockers such as metoprolol are recommended for all clinically affected ARVC patients to prevent arrhythmias and reduce the stress on the RV wall. Sotalol (a non-selective beta-blocker) may be used to treat arrhythmia. Amiodarone (class III antiarrhythmic) or adjunctive therapy with flecainide (class Ic antiarrhythmic) may be considered for patients with persistent ventricular arrhythmias despite the use of sotalol. However, flecainide should be avoided when the left ventricular ejection fraction is reduced (<40%).[8-10] Other medications, including diuretics, ACEi/ARB, and MRA, can be used as the standard therapy for heart failure.[11] Radiofrequency catheter ablation can be employed in patients who suffer from refractory or incessant ventricular tachycardia. An ICD can be implanted to prevent sudden cardiac death.[12-13]

Our patient was first given IV Amiodarone to control his stable VT, by which a beta-blocker was subsequently added. An Electrophysician (EP) specialist was consulted, and the patient proceeded for an ICD implantation as secondary prevention for future ventricular arrhythmia. Guideline-directed medical therapy (GDMT) was prescribed for benefit of poor RV function.

Conclusion

ARVC is a hereditary cardiomyopathy and poses a high risk of ventricular arrhythmias and SCD. Our patient's clinical presentation of recurrent palpitations raised suspicion of cardiac arrhythmia, whereby the following EKG and echo findings led to the discovery of a potentially life-threatening disease and subsequent ICD implantation. Therefore, it is very crucial to recognize the possible EKG findings of ARVC so to be able to pursue further investigations and implement proper therapies. Timely management of ventricular arrhythmias help reduce mortality and improve patient prognosis. The clinical courses of ARVC vary greatly among patients, and sustained VT/VF can occur at any time during the disease course, hence long term follow up is crucial. Prognosis tends to be favorable for high-risk patients who receive ICDs for prevention of SCD.

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