CASE REPORT
VENTRICULAR TACHYCARDIA AS A FIRST PRESENTATION OF ARRHYTHMOMGENIC RIGHT VENTRICULAR CARDIOMYOPATHY (ARVC) IN A PREVIOUSLY ASYMPTOMATIC SEXAGENARIAN

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Abstract: Arrhythmomgenic right ventricular dysplasia/ cardiomyopathy (ARVD/C) is characterised by morphological and histological changes in the ventricles. Progressive myocyte loss and fibrofatty tissue replacement, producing islands of scar, can lead to reentrant ventricular tachycardia and sudden cardiac death. It usually presents as chest pain, palpitations, heart failure, or syncope. The majority of cases are seen before the age of 40. Ventricular arrhythmia as a first presentation in the elderly is seen infrequently. We present a case of a previously asymptomatic 62-year-old gentleman who had an episode of ventricular tachycardia as the first manifestation of ARVC without having any positive family history for this disease. He was managed with amiodarone and was later planned for an implantable cardiac defibrillator (ICD).

Keywords: Arrhythmomgenic right ventricular dysplasia, ventricular tachycardia, elderly

INTRODUCTION

Arrhythmogenic right ventricular dysplasia/ cardiomyopathy (ARVD) is a rare disease involving the right ventricle and very rarely the left ventricle. Its prevalence has been estimated to be around 1 in 5000 in the western population.1 Histologically, the cardiac tissue is replaced with fibro-fatty tissue. The disease can manifest in a variety of ways, from being asymptomatic to causing symptoms including chest pain, palpitations, syncope, heart failure, tachyarrhythmia (most frequently ventricular tachycardia), and even sudden cardiac death (SCD). Left ventricular (LV) involvement can occur in a very small number of patients, and it is linked with the severity of the disease.2 The majority of cases are present before the age of 40, while late manifestation includes heart failure, which typically appears later in life.3 Ventricular tachycardia as the first presentation in the older age group is an infrequent occurrence. Due to the complexity associated with this disease, Task Force Criteria have been laid out for ARVD diagnosis, initially in 1994 and then revised in 2010. The cardiovascular magnetic resonance (CMR) modality is an essential part of the diagnosis, including both major and minor criteria, that helps in assessing the anatomy as well as the function of the right ventricle, including the presence of fibrofatty infiltration.4

To date, there is only limited data available in the form of a few case reports in the Asian population.5 We present a case of a previously asymptomatic 62-year-old gentleman who had a VT episode as the first manifestation of ARVC without having any positive family history for this disease.

CASE REPORT

A 62-year-old medical administrator of a local hospital, having no premorbid, presented initially to a secondary health care hospital with a history of chest pain and palpitations for a few hours followed by a sudden collapse. On presentation, he had a BP of 80/50 mmHg, a heart rate of 170 bpm, and a respiratory rate of 29 breaths per minute. Physical examination showed clear lungs and no murmurs on auscultation. The ECG revealed a wide complex tachycardia with a ventricular rate of 180 beats per minute, atypical left bundle branch morphology with no RS complex in any of the chest leads, VA dissociation, and a left inferior axis. He was immediately cardioverted successfully with a 150 J biphasic shock. In sinus rhythm, the ECG revealed T wave inversions in all chest leads v1-v6 (Figure 1). The biochemical profile revealed a complete blood picture; renal function tests and liver function tests were all in the normal range. Troponin I was markedly elevated at 471 ng/L (normal range 1–29 ng/L). Based on chest pain and venicular tachycardia, it was decided to take the patient to an emergent cardiac catheterization, which revealed mild ectasia in the proximal and mild narrowing in the distal left anterior descending artery.

He was loaded with intravenous amiodarone. His history was negative for sudden cardiac death in the...
family. An echocardiogram showed good left ventricular function with an ejection fraction of 55%. The right ventricle appeared dilated and poorly contracting. The moderator band was thickened and hypertrophied. Signal average ECG (SAECG) was positive for late potentials (QRS duration=160 msec, RMS last 40 msec=1.705 microvolts, duration under 40 microvolts=90 msec). The 24-hour tape did not reveal any further arrhythmias.

**DISCUSSION**

Arrhythmogenic right ventricular dysplasia/cardiomyopathy (ARVD/C) is a hereditary cardiomyopathy represented by fibrofatty replacement of the myocardial tissue of the right ventricle and, in rare cases, the left ventricle, providing a substrate for reentrant dysrhythmias. The infiltration of the right ventricle's myocardium by fibroadipose tissue differs significantly from age-related fatty replacement of the myocardium.\(^6\)

There is a genetic predisposition, with 50% of the cases being autosomal dominant. However, a recessive mode of inheritance has also been reported. Commonly, ARVD/C presents as ventricular arrhythmia between the second and fifth decades of life. Other presentations can be chest pain, fatigue, syncope, palpitations, heart failure, and sudden cardiac death. With the aid of genetic testing, the disease is now more frequently recognised among the elderly population with the aid of genetic testing.\(^7\) There are a few reported cases in the literature who presented with ventricular tachycardia in their late sixties.\(^8\) It has been suggested that late manifestation in some patients is related to some unidentified genetic mutations, and these subgroups usually do not harbor a pathogenic mutation.\(^9\) Individuals who present late also have fewer ECG features, including precordial T-wave repolarization, and also represent less ventricular ectopy, which interestingly do not correlate with the risk of arrhythmic events compared to younger cohorts. Some of the features that predict arrhythmic outcome in later stages of life include male gender, family history negative for ARVC, gross involvement of RV, and pathogenic mutation.\(^10\) Our patient also had the majority of these features.

The ECG of 90% of ARVD patients with symptoms is abnormal, with frequent nonspecific changes such as T wave inversions in the right chest leads with or without right bundle branch block. In 30% of patients, chest leads show epsilon waves. These are essentially low-amplitude potentials detected at the terminal end of the QRS complex, indicating delayed activation of RV myocytes.\(^11\) The patient in our case showed T inversions in chest leads with no evidence of RBBB and classical Epsilon waves were also not observed.

Patients can have arrhythmias as well, and they can present either as asymptomatic premature ventricular beats or as ventricular tachycardia (VT), which can sometimes lead to sudden cardiac death. The tachycardias usually arise from the right ventricle and, as a result, are of left bundle branch (LBBB) morphology. However, left ventricle involvement can produce right bundle branch morphology. These arrhythmias are easily provoked by catecholamines and therefore are easily inducible with isoprenaline. The left variant usually involves the posterolateral wall with midmyocardial or subepicardial infiltration, and LV involvement sometimes precedes RV dysfunction. There is minor LV damage in the initial phase and deformation imaging may play a better role compared to CMR in such patients. Due to variability in phenotype expression, LV involvement can either occur in isolation or in combination with the right ventricle with predominant infiltration.\(^12,13\)

Echocardiography can show features pathognomonic of ARVD. These include RV dilatation; segmental RV dilatation with or without dyskinetic segments
(aneurysms or bulging); RV bulging in diastole; inferobasal free wall; and apical dyskinesia during systole; exaggerated trabecular pattern in the RV; hypertrophy; and increased reflectivity of the moderator band. Our patient did show some of these echocardiographic changes, including dilation of the RV and inferobasal dyskinesia.\textsuperscript{14}

Cardiac MRI is considered an important imaging tool for making a diagnosis. It detects fatty infiltration in the right ventricle, dilatation, and aneurysmal transformation along with regional wall motion abnormalities. It is also helpful in finding myocardial fibrosis and scarring.\textsuperscript{15} On MRI, features suggestive of this disease were found in this patient that included diffuse thinning of the RV free wall, subtricuspid dilatation with dyskinesia, and a right ventricular ejection fraction of 40\% (Figure 2). ARVD diagnosis is made on the criteria laid out by the Taskforce as shown in table 1.\textsuperscript{16} Our patient had 2 major and 3 minor criteria, which made him a definite case of ARVC.

![Figure 2: MRI showing diffuse thinning of RV wall (blue arrowhead) and aneurysmal dilatation of subtricuspid RV free wall (red arrow) suggestive of ARVC](image)

The management outlines medical, radiofrequency ablation, and ICD. Since ventricular arrhythmias are responsive to amiodarone, it was already started before he reported it to us.\textsuperscript{17} ICD was offered since he was considered a high-risk patient for SCD, but the patient was not willing to undergo the procedure on the same admission.

Table 1: Diagnostic criteria (Major and Minor) based on 2010 modified task force criteria for diagnosing ARVC adopted from Marcus et al.\textsuperscript{16}

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<tr>
<th>Major Criteria</th>
<th>Depolarization Abnormalities (ECG)</th>
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<tr>
<td>Repolarization Abnormalities (ECG)</td>
<td>Inverted T waves in leads V1, V2, and V3 or beyond in individuals &gt;14 years of age without having RBBB</td>
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<th>Depolarization Abnormalities (ECG)</th>
<th>Presence of Epsilon wave</th>
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<tr>
<td>Arhythmias</td>
<td>Nonsustained or sustained VT of LBBB with superior axis</td>
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| Tissue characterization of wall               | Endomyocardial biopsy showing fibrous replacement of the RV free wall myocardium in ≥1 sample, with or without fay replacement and with residual myocytes ≤60% |

| Global or regional dysfunction / Structural changes | Regional RV akinesia, dyskinesia, or aneurysm and 1 of the following (end diastole), PLAX RVOT ≥32 mm, PSAX RVOT ≥36 mm, Fractional area change ≤33\%, Ratio RVEDV/BSA ≥110 mL/m\(^2\) (male), ≥100 mL/m\(^2\) (female), RVEF ≤40\% |

| Family History                                | Family member 1st degree with confirmed ARVC, or confirmed at autopsy or pathogenic mutation related to ARVC identified |

| Family History                                | Premature sudden death (<35 years of age) due to suspected ARVC in a first-degree relative, ARVC confirmed pathologically in 2nd degree relative |

| Minor criteria                                | Inverted T waves in leads V1 and V2 in individuals >14 years of age without RBBB or Inverted T waves in leads V1, V2, V3, and V4 in individuals >14 years of age with complete RBBB |

In conclusion, ARVD/C is a genetic disease with a wide range of manifestations. It primarily affects young people, with the vast majority of those affected under the age of 40. It is rarely found among the elderly. Any individual, regardless of age, who has LBBB morphology ventricular arrhythmia and ECG and symptoms of right heart disease, or even heart failure, should be investigated for ARVC/D.
AUTHORS’ CONTRIBUTION
MA and QHK: Concept and design, data acquisition, interpretation, drafting, final approval, and agree to be accountable for all aspects of the work. MA, TBN, and QHK: Data acquisition, interpretation, drafting, final approval and agree to be accountable for all aspects of the work.

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REFERENCES

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