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Case Report

Arrhythmogenic right ventricular cardiomyopathy or dysplasia: a case report

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ABSTRACT

Arrhythmogenic cardiomyopathy (ACM) is a progressive genetic disease of the myocardium characterized by loss of myocardial cells and replacement by fibrofatty tissue in the right and/or left ventricle (RV/LV), clinically manifested by syncope, palpitations, heart failure, or sudden cardiac death (SCD). We presented a case where the patient suffered from palpitations for the previous three to four years, which resolved spontaneously each time and lately presented with persistent palpitations and chest discomfort. ECG showed a widening of the QRS complexes during sinus rhythm and premature complexes (PVCs) of various patterns. After initial evaluation cardiac MRI was done which revealed typical features of arrhythmogenic right ventricular dysplasia/ cardiomyopathy. The patient was continuing amiodarone 100 mg daily and doing well.

Keywords: Arrhythmogenic, Right ventricle, Cardiomyopathy, Dysplasia

INTRODUCTION

Arrhythmogenic cardiomyopathy (ACM) is a progressive genetic disease of the myocardium characterized by loss of myocardial cells and replacement by fibrofatty tissue in the right and/or left ventricle (RV/LV), clinically manifested by syncope, palpitations, heart failure, or SCD.[1] Originally, described as arrhythmogenic right ventricular dysplasia/cardiomyopathy (ARVD/C) due to involvement of the right ventricle. A case series of twenty-four patients were reported in 1982 by Mercus et al.[2] At that time it was thought that myocardium had not developed in the right ventricle and coined the term dysplasia. But with further study and follow-up of this group of new patients, it was revealed that there is a progressive loss of right ventricular myocardium and replacement by fibrofatty tissues, and then the term cardiomyopathy was introduced. Still, the term arrhythmogenic right ventricular dysplasia/cardiomyopathy (ARVD/C) is used. Later, increasing recognition of left ventricular (LV) involvement has recently led to the adoption of the term ACM. There is an estimated prevalence of ACM from 1:2000 to 1:5000 and is an important cause of SCD among young individuals, including athletes.[3] The disease is more malignant in men than in women either due to the influence of sex hormones on phenotypic expression or differences in the intensity of physical exercise in women and men. Approximately 50% of affected patients have a positive family history, but both incomplete penetrance and limited phenotypic expression are common. The mode of inheritance follows a Mendelian autosomal dominant pattern, where disease-causing mutations occur in more than 13 genes that have been identified.[4]

CASE REPORT

A 60-year-old man presented in the emergency department at midnight with a complaint of palpitation and chest

discomfort for one day. Examination revealed an irregular pulse with frequently dropped beats. ECG monitor showed frequent ventricular premature complexes (PVC) in the pattern of couplets, bigeminy, and trigeminy along with isolated PVCs; (Figure 1) and the patient was immediately shifted to the ICU for lifesaving management. Treatment started with intravenous amiodarone and morphine; the arrhythmia then gradually abated. The next morning, he was feeling well and underwent a coronary angiogram which was uneventful with normal coronary arteries. He was discharged in the evening with bisoprolol 5 mg daily. Two days later, he again developed palpitations and was readmitted to the hospital. ECG monitor showed frequent PVCs than the treatment was revised, and bisoprolol was replaced with amiodarone 100 mg daily to continue.

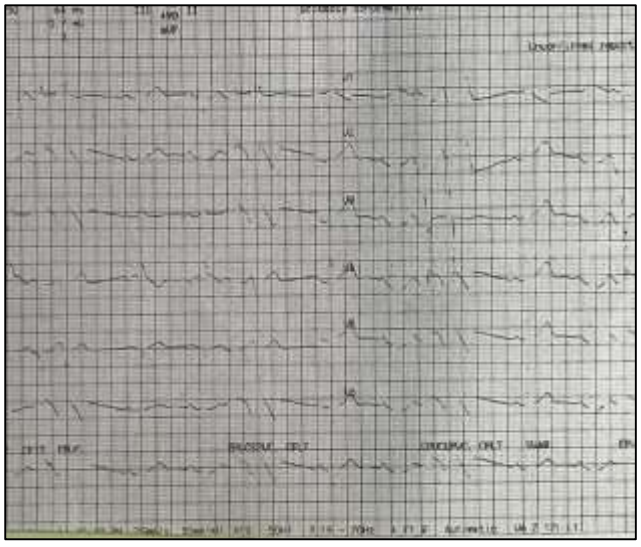

Figure 1: ECG shows PVCs in the pattern of bigemini, trigemini, and isolated form. QRS complexes in sinus rhythms are widened. (Initial ECG while admitted to the hospital).

PVCs disappeared gradually and he was discharged from the hospital the next day. During the hospital stay routine investigations were unremarkable. But ECG showed a widening of the QRS complexes during sinus rhythm and PVCs of various patterns (Figure 3).

The cardiac silhouette was normal in the chest X-ray, and the echocardiography study was normal. No other systemic abnormalities were detected. He was a normotensive, nondiabetic, and nonsmoker. After a thorough evaluation (both physical and investigational) the cause of the arrhythmia remained undetected. The patient was continuing amiodarone 100 mg daily and doing well. He admitted that he had episodes of palpitations that occurred intermittently for the previous three to four years, which resolved spontaneously each time. He noticed that palpitations occurred especially after taking meals. Besides several episodes of palpitations, he had also suffered from prostatic symptoms for several years and then he underwent a TURP operation for benign prostatomegaly one year before this presentation. He has oral lichen planus for which he uses topical steroids intermittently. He had also gastro-oesophageal reflux disease and was taking PPI intermittently. Later, he went abroad for evaluation of the arrhythmia though he was doing well with amiodarone.

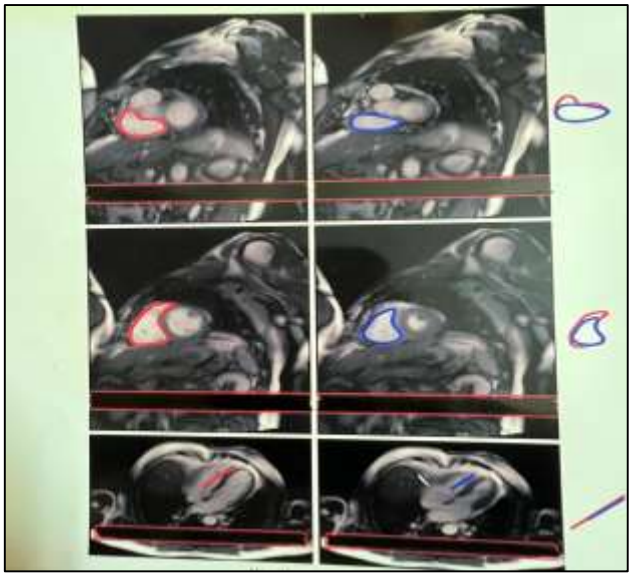

Figure 2: Cardiac MRI shows mild dilatation of the right atrium and the right ventricle, thinning of the free wall of the RV, and patchy areas of fibrofatty infiltration in the RVOT.

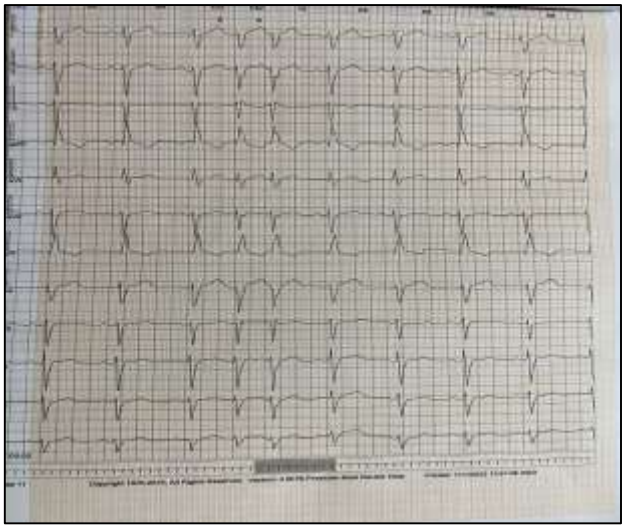

Figure 3: Recent ECG showing couplet of PVC with prolonged TAD.

After initial evaluation cardiac MRI was done which revealed typical features of arrhythmogenic right ventricular dysplasia/ cardiomyopathy. The right atrium and the right ventricle are mildly dilated. The anterior wall of the right ventricle showed patchy areas of thinning with small aneurysmal pockets in the apical area and patchy areas of fibrofatty scars were found in the right ventricular

outflow tract (RVOT) (Figure 2). The right ventricular ejection fraction was 51%, while the LV ejection fraction was 64.1%; the concluding remark was mild ARVD with right ventricular dysfunction.

DISCUSSION

ACM is a progressive genetic disease of the myocardium characterized by loss of myocardial cells and replacement by fibrofatty tissue in the RV and/or LV (RV/LV) clinically manifested by syncope, palpitations, heart failure, or SCD.[1] Originally, described as arrhythmogenic right ventricular dysplasia/cardiomyopathy (ARVD/C) due to involvement of the right ventricle. A case series of twenty-four patients were reported in 1982 by Mercus et al.[2] At that time it was thought that myocardium had not developed in the right ventricle and coined the term dysplasia. But with further study and follow-up of this group of new patients, it was revealed that there is a progressive loss of right ventricular myocardium and replacement by fibrofatty tissues, and then term cardiomyopathy was introduced. Still, term arrhythmogenic right ventricular dysplasia/ cardiomyopathy (ARVD/C) is used. Later, increasing recognition of LV involvement has recently led to the adoption of the term ACM. There is an estimated prevalence of ACM from 1:2000 to 1:5000 and is an important cause of SCD among young individuals, including athletes.[3] The disease is more malignant in men than in women either due to the influence of sex hormones on phenotypic expression or differences in the intensity of physical exercise in women and men. Approximately 50% of affected patients have a positive family history, but both incomplete penetrance and limited phenotypic expression are common. The mode of inheritance follows a Mendelian autosomal dominant pattern, where disease-causing mutations occur in more than 13 genes that have been identified.[4] Desmosome, a cell-to-cell adhesion structure composed of multiple different protein molecules exerts tissue strength in the myocardium. In ACM, mutations in genes encoding desmosomal proteins: PKP2 (plakophilin-2), DSP (desmoplakin), DSC2 (desmocollin-2), JUP (junction plakoglobin), and DSG2 (desmoglein-2) occur with variable expressions and penetrance.[5] The mutant protein weakens the cell-to-cell adhesion in the myocardium resulting in progressive loss of myocardial cells, especially after stressful activities which are replaced by fibrofatty scar tissues. The fibrofatty scars progress from the epicardium towards the endocardium and predominantly involve the right ventricular free wall resulting in wall thinning and aneurysmal dilatation (Figure 8).[6] The pathological changes are typically localized in the inflow tract (sub tricuspid region), outflow tract (infundibular region), and the apex of the right ventricle commonly described as a "triangle of dysplasia." Initially, only the right ventricular involvement was identified, but the advent of modern diagnostic methods like ECG, MRI, endomyocardial biopsy, and molecular genetic testing led better understanding of the disease, and the pathological changes are being increasingly identified in other areas of the heart especially the left ventricle (LV). In recent reports, biventricular, predominant LV, or even only the LV involvement were described.[7] Histopathological study revealed the right ventricular myocardium is replaced by fibrofatty tissue in variable extent depending upon the severity of the disease when biopsied (Figure 4).

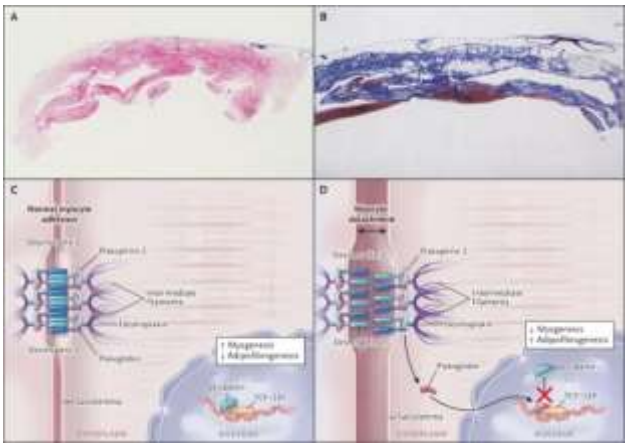

Figure 4: Histopathological features and myocyte detachment in arrhythmogenic right ventricular cardiomyopathy/dysplasia.[26]

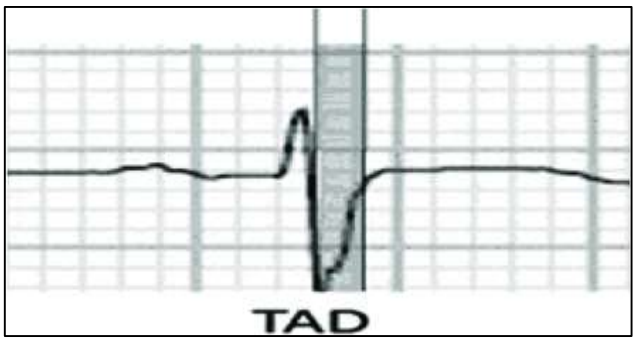

Figure 5: ECG showing prolonged terminal activation duration, a sign of depolarisation abnormality. (TAD is measured from the nadir of the S wave to the end of all depolarization deflections and is prolonged if _>55 ms in any of the V1-V3 leads in the absence of complete right bundle branch block.)

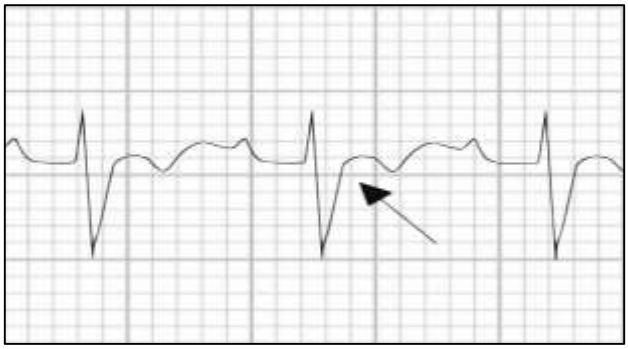

Figure 6: ECG showing prolonged terminal activation duration.

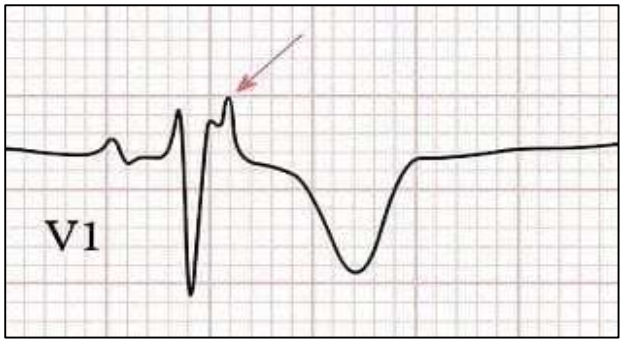

Figure 7: ECG showing epsilon wave.

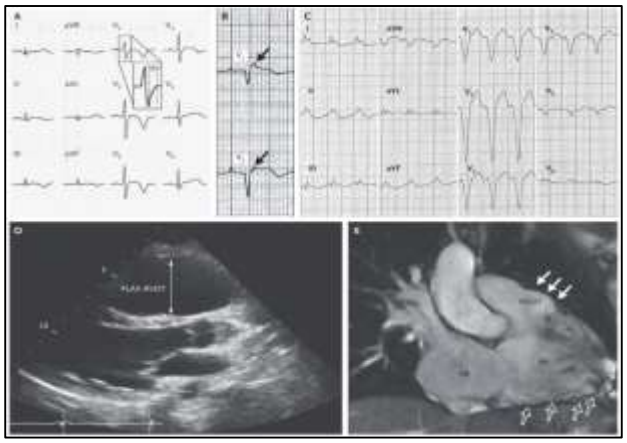

Figure 8 (A-E): ECG showing TAD, ECG showing epsilon wave, ECG showing frequent PVC, echocardiogram and MRI shows mild dilatation of the right atrium and the right ventricle thinning of the free wall of the RV with aneurysm and patchy areas of fibrofatty infiltration in the RVOT.[26]

ARVC typically becomes clinically apparent between the second and fourth decades of life. The case presented here is a late presentation; around 60 years of age, i.e., a late phenotypic expression. SCD may be the first clinical manifestation of the disease. The most common clinical presentation is palpitations or effort-induced syncope in an adolescent or young adult, with T-wave inversion in the right precordial leads (V1 through V4) on the electrocardiogram, ventricular arrhythmias with a left bundle-branch block pattern, and right ventricular abnormalities on imaging tests. Electrocardiographic depolarization abnormalities, which reflect defective conduction through the diseased right ventricular myocardium, may also be present. The clinical features of ARVC are related to the RV electrical instability due to the presence of fibrofatty scars which are associated with lethal ventricular arrhythmias including ventricular tachycardia and fibrillation. The electrocardiogram (ECG) has a central role in diagnosis since it comprises two major and two minor criteria in the diagnostic criteria published by task force (Table 1). Lipomatous or lipofibromatous composition of right ventricle (RV) tissue is responsible for the delay in depolarization and the development of re-entrant ventricular arrhythmias and SCD. ECG expression of this depolarization delay was first described in 1978, when Fontaine named these tiny signals found during endocardial mapping (and also in the surface ECG) located right after the QRS complexes in leads V1-V3, as 'epsilon waves' (Figure 7). The ECG patterns that may be found are the following: Normal electrocardiogram pattern, Delay of terminal activation time, (Figure 6) Right bundle branch block pattern (20-40% cases), presence of epsilon wave, and repolarisation abnormalities. Repolarization abnormalities are inverted T waves ST segment alteration. Arrhythmias: ventricular tachyarrhythmias.

Table 1: Revised task force criteria (2010) for the diagnosis of ARVD/C.[26]

Category	Major criteria	Minor criteria
Global or regional dysfunction and structural alteration		
On 2-dimensional echo-cardiography	Regional RV akinesia, dyskinesia, or aneurysm and one of the following (end diastole): PLAX ROVT ≥32 mm (≥19mm/ m^2 when corrected for body-surface area), PSAX RVOT ≥36 mm (≥21 mm/ m^2 when corrected for body-surface area), or fractional area change of ≤33%	Regional RV akinesia/dyskinesia and one of the following (end diastole): PLAX RVOT 29 <32 mm (16-19 mm/ m^2 when corrected for body-surface area), PSAX RVOT 32-<36 mm (18-<21 mm/m^2 meter when corrected for body-surface area)/fractional area change of 34-40%
On MRI	Regional RV akinesia or dyskinesia or dyssynchronous RV contraction and one of the following ratios of RV end-diastolic volume to body-surface area ≥ 110 ml/m^2 (male patients) or ≥ 100 ml per square meter (female patients), or RV ejection fraction ≤0%	Regional RV akinesia or dyskinesia/ dyssynchronous RV contraction and one of following: ratio of RV end-diastolic volume to body-surface area 100-110 ml/m^2 (male patients)/90-<100 ml per square meter (female patients)/RV ejection fraction 41-45%
RV angiography	Regional RV akinesia, dyskinesia/aneurysm	
Tissue characterization	<60 % residual myocytes on morphometric analysis (or <50%, if estimated) and fibrous replacement of the RV free-wall myocardium, with or without fatty replacement of tissue, in at least one endo- myocardial-biopsy sample	60 to 75% residual myocytes, on morphometric analysis (or 50 to 65%, if estimated) and fibrous replacement of the RV free-wall myocardium, with or without fatty replacement of tissue, in at least one endomyocardial-biopsy sample

Continued.

Category	Major criteria	Minor criteria
Repolarization abnormalities	Inverted T waves in right precordial leads (V1, V2, and V3) or beyond in patients older than 14 years of age (in the absence of complete right bundle -branch block, QRS ≥ 120 msec)	Inverted T waves in leads V1 and V2 in patients older than 14 years of age (in absence of complete right bundle-branch block)/in V4, V5, or V6 inverted T waves in leads V1, V2, V3, and V4 in patients older than 14 years. of age (in presence of complete right bundle-branch block)
Depolarization and conduction abnormalities	Epsilon wave (reproducible low-amplitude signals from end of QRS complex to onset of the T wave) in the right precordial leads (V1, V2 and V3)	Late potentials on signal-averaged ECG in at least one of three parameters in the absence of a QRS complex duration of ≥ 110 msec on the standard ECG; filtered QRS complex duration,≥ 114 msec; duration of terminal QRS complex < 40 µV (low-amplitude signal duration), ≥38 msec, root-mean-square voltage of terminal 40 msec, ≤20 µV; TAD of QRS complex,≥ 55 msec, measured from the nadir of the S wave to the end of the QRS complex, including R, in V1,V2,or V3 in absence of complete right bundle-branch block.
Arrhythmias	Non-sustained or sustained ventricular tachycardia with l left bundle -branch block and superior axis pattern (negative or indeterminate QRS complex in leads II, III and aVF and positive QRS complex in lead aVL	Non-sustained or sustained ventricular tachycardia of RV outflow configuration with a left bundle-branch block and inferior axis pattern (positive QRS complex in leads II, III, and aVF and negative QRS complex in lead aVL) or unknown axis, or > 500 ventricular extrasystoles per 24 hr (on Holter monitoring)
Family history	ARVC confirmed in a 1st-degree relative who meets current task force criteria, ARVC confirmed pathologically at autopsy or surgery in a 1st-degree relative, or identification of a pathogenic mutation categorized as associated or probably associated with ARVC in the patient under evaluating	History of ARVC in a 1st degree relative in whom it is not possible or practical to determine whether current task-force criteria are met, premature sudden death (at <35 year of age) due to suspected ARVC in a 1st degree relative, or ARVC confirmed pathologically/by current task-force criteria in a second-degree relative.

There is no single diagnostic test for ARVC/D. The diagnosis is made based on major and minor clinical, electrical, and imaging criteria that have been devised by expert consensus of the task force criteria (TFC) originally proposed in 1994 and further revised in 2010.

Echocardiography is the first-line investigation and may demonstrate a dilated, hypokinetic right ventricle with prominent apical trabeculae and dilatation of the RV outflow tract (Figure 8). cardiac MRI, which can accurately demonstrate structural and functional features of ARVD such as fibrofatty infiltration and thinning of the RV myocardium, RV aneurysms, RV dilatation, regional wall motion abnormalities, and global systolic dysfunction. Histological diagnosis, either via endomyocardial biopsy or at autopsy, provides a definitive diagnosis.[8]

Genotyping is mainly used to identify disease-causing mutations among family members. Patients with multiple desmosomal gene mutations are likely to have a more severe phenotype and may have an increased lifetime risk of malignant arrhythmia and SCD.[9] During the clinical course of ARVC SCD may occur due to ventricular arrhythmia, and biventricular systolic dysfunction can lead to death from heart failure. In ARVC, mortality rates vary in studies and range from 0.08 to 3.6% per year. The prognosis of ARVC depends largely on the severity of arrhythmias and ventricular dysfunction. The most important predictors of life-threatening arrhythmic events include previous history of cardiac arrest due to ventricular fibrillation and sustained ventricular tachycardia.[3] And other major risk factors include unexplained syncope, nonsustained ventricular tachycardia on ambulatory monitoring or exercise testing, and severe systolic dysfunction of the right ventricle, left ventricle, or both. The goals of ARVC management are to reduce the risk of SCD and improve the quality of life. Appropriate treatment of arrhythmia and heart failure will alleviate symptoms like palpitations, syncope, and dyspnoea. Avoidance of strenuous exercise is important to prevent malignant arrhythmia. Beta-blockers are recommended for all clinically diagnosed patients to prevent arrhythmia and reduce right ventricular wall stress. Ventricular arrhythmias are treated with antiarrhythmic drugs like amiodarone with or without beta-blockers and sotalol. Catheter-based radiofrequency ablation can be used in selected patients who have episodes of sustained

monomorphic ventricular tachycardia. Subsequent recurrences of ventricular tachycardia are common because of the progressive nature of the disease which leads to the development of multiple arrhythmogenic foci over time.[10] Defibrillators can prevent SCD and are used in patients with a previous history of sustained ventricular tachycardia and fibrillation. A defibrillator is effective in preventing SCD and is safe. Defibrillator therapy is indicated in patients who had an episode of ventricular fibrillation or sustained ventricular tachycardia. It is not clear whether defibrillator therapy is helpful for the primary prevention of SCD in patients with one or more risk factors and no prior major arrhythmic events. Prophylactic defibrillator therapy is not indicated in asymptomatic patients with no risk factors and in healthy gene carriers.[11] There is also a risk of significant device or electrode-related complications during long-term follow-up. Heart failure should be treated with standard pharmacological agents like angiotensin-converting enzyme inhibitors, angiotensin receptor blockers, beta-blockers, and diuretics. Oral anticoagulant is indicated in patients with atrial fibrillation and thromboembolic complications.[12] Cardiac transplantation is reserved for people with untreatable arrhythmias and refractory heart failure.

Literature review

Arrhythmogenic right ventricular cardiomyopathy is a rare inherited heart-muscle disease that is a cause of sudden death in young people and athletes. Causative mutations in genes encoding desmosomal proteins have been identified and the disease is nowadays regarded as a genetically determined myocardial dystrophy. The left ventricle is so frequently involved as to support the adoption of the broad term ACM.[13] The wide clinical spectrum of arrhythmogenic right ventricular cardiomyopathies/ dysplasia appears to be the result of one or possibly two factors: 1. replacement of most of the right ventricular myocardium by fat and 2. genetic susceptibility to environmental agents (myocarditis).[14] The term "dysplasia" was used to define an entity that was thought to be the result of a developmental defect of the right ventricular myocardium. A better understanding of clinical manifestations as well as morphologic findings do not support the theory of a congenital absence of the myocardium, but is in keeping with non-ischaemic, ongoing atrophy of the right ventricular myocardium, most likely genetically determined, which becomes symptomatic in adolescents and young adults.[15] It is well known that a large majority of patients with ARVD have histological evidence suggestive of inflammation. It may be that if myocarditis is superimposed on the background of myocardium interspersed by fat, strands of cardiomyocytes involved in myocarditis will produce fibrosis and will transform the purely fatty to the fibrofatty form. Activation of neutrophils induced by myocarditis will enhance the risk of arrhythmogenesis. Main differentials could be, idiopathic dilated cardiomyopathy, isolated myocarditis, or adipomatosis cordis.[16] Alike the present study, depolarization abnormalities in right ventricular dysplasia are thought to reflect slowed propagation in the right ventricular free wall, as opposed to altered conduction in the bundle branch. Symptomatic patients without clear evidence of disease expression may be offered an implantable loop recorder. This option is of particular value when palpitation, dizzy spells, or syncopal episodes are sporadic and unlikely to be captured by standard ambulatory electrocardiographic monitoring. Ascertaining whether such symptoms have an arrhythmic etiology enables appropriate management of patients with underlying cardiac disease and reassurance of others.[17] According to a study, the most common electrocardiographic abnormalities were localized right QRS prolongation, poor r-wave progression in the right precordial leads, incomplete right branch bundle block, prolonged S-wave upstroke in V_1 to V_3, parietal block, ST-segment elevation in V_1 to V_3, inversion of T waves beyond V_2, and epsilon wave. Low QRS voltages in the precordial leads were frequently present in all patients with ARVC compared with a group of 120 healthy subjects.[18] Most of the ECG findings were relatable to the current study in the diagnosis of the disease. Another study stated that QRS duration in $(V1 + V2 + V3)/(V4 + V5 + V6) \geq 1.2$-called localized right precordial QRS prolongation-was present in 261/265 patients (98%) and represents the essential finding. Right precordial epsilon potentials were found in 23% of standard and in 75% in highly amplified and modified recording techniques. Right precordial T wave inversions were present in 143 cases (54%) and ST-segment elevation of different types in 66 patients (25%). Localized prolongation of inferior QRS complexes could be found in 58 cases (22%), complete right bundle branch block with T inversions beyond V2 in most cases in 17 patients (6%), incomplete right bundle branch block in 38 cases (14%), pseudo-incomplete right bundle branch block in 8 patients (3%), and right precordial R wave reduction in 14 cases (5%).[19] Epsilon potentials in right precordial leads are reliable diagnostic electrocardiographic (ECG) criteria of arrhythmogenic right ventricular dysplasia-cardiomyopathy (ARVD/C), which was seen in the present study.[20] In another study, T-wave inversions in V_1 through V_3 were observed in 85% of ARVD/C patients in the absence of RBBB compared with none in RVOT and normal controls, respectively ($P<0.0001$); epsilon waves were seen in 33%, and a QRS duration ≥110 ms in V_1 through V_3 was present in 64% of patients. Among those without RBBB, their newly proposed criterion of "prolonged S-wave upstroke in V_1 through V_3" ≥55 ms was the most prevalent ECG feature (95%) and correlated with disease severity and induction of VT on the electrophysiological study. This feature also best distinguished ARVD/C (diffuse and localized) from RVOT.[21] There are five therapeutic options in patients with ARVD/C: (1) antiarrhythmic agents, (2) radiofrequency ablation, (3) implantable cardioverter-defibrillator therapy, (4) heart failure treatment, and (5) surgical treatment.[22] The most important objectives of clinical management of ARVC/D patients include (i) reduction of mortality, either by arrhythmic SCD or death

from heart failure; (ii) prevention of disease progression leading to RV, LV, or biventricular dysfunction and heart failure; (iii) improvement of symptoms and quality of life by reducing/abolishing palpitations, VT recurrences, or ICD discharges (either appropriate or inappropriate); and (iv) limiting heart failure symptoms and increasing functional capacity. Patients with ARVC/D should undergo lifelong clinical follow-up to periodically evaluate new onset or worsening of symptoms, progression of morphological and/or functional ventricular abnormalities, and ventricular arrhythmias to reassess the risk of SCD and optimize the treatment. Cardiac evaluation of affected patients including resting 12-lead ECG, echocardiography, 24-h Holter monitoring, and exercise testing (for detection of effort-induced ventricular arrhythmias) should be performed regularly (every 1-2 years) depending on the age, symptoms, and disease severity.[12,23,24] Moreover, in a recent study, inducible ventricular tachycardia by programmed ventricular stimulation (PVS) predicted clinical sustained ventricular arrhythmia (VA) during the 5-year follow-up. This will help to take appropriate measures to prevent malignant arrhythmias and SCD.[25]

CONCLUSION

The case presented above is a late presentation around sixty years of age and diagnosed with cardiac MRI findings. Recurrent episodes of palpitations were the only presentation, but the last episode was persistent and forced him to ICU admission. Intravenous amiodarone abated the attack. Thereafter he has been on oral amiodarone 100 mg daily. He is now on follow-up for the last five years and remains symptom-free. ARVC is a progressive disease leading to malignant arrhythmias and SCD and/or refractory heart failure also causes cardiac death. Early detection and appropriate treatment may cause symptomatic improvement and prevent SCD. Genetic testing may be helpful to identify and follow up on the healthy carriers.

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