Case Report

A Case of Arrhythmogenic Right Ventricular Cardiomyopathy with Left Ventricular Involvement Presenting as Recurrent Ventricular Tachycardia

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Abstract

Background : Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC) is condition of genetic inheritance. Pathologically characterized by fibrofatty replacement of cardiac muscle mainly in right ventricular myocardium. Ventricular arrhythmias of right ventricular origin may lead to sudden cardiac death in young adults and athletes. Usually, male patients with age less than 40 years present with recurrent palpitation later develop progressive right heart or biventricular failure. Here we report a case of Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC) with left ventricular involvement with recurrent ventricular tachycardia.

Case Presentation : A 39-year-old male patient with no comorbidities presented with palpitations and giddiness. He was hemodynamically unstable and ECG showed ventricular tachycardia with Left Bundle Branch Black (LBBB) morphology. Patient was managed with cardioversion and anti arrhythmic drugs amiodarone and betablockers. His 2D echo and cardiac MRI done and diagnosis of arrhythmogenic right ventricular cardiomyopathy with left ventricular involvement was made based on 2020 international criteria. He was adviced Implantable Cardioverter-Defibrillator (ICD) implantation. As patient refused ICD implantation he was continued on amiodarone and beta blockers.

Conclusions : ARVC is a rare disorder, and proper diagnosis is important to avoid unnecessary delay in managing the patients. 2020 International criteria helps to diagnose ARVC. Although ICD implantation is the treatment of choice for these patients, medical management with beta-blockers and amiodarone are helpful to prevent recurrent episodes of ventricular tachycardia in these patients.

Key words : Arrhythmogenic Right Ventricular Cardiomyopathy, Recurrent Ventricular Tachycardia, LV Involvement.

Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC) is a rare inherited disorder with autosomal dominant inheritance. Pathologically characterized by fibrofatty replacement of cardiac muscle mainly in right ventricular myocardium[1]. As per literature review, prevalence of ARVC is 2 to 5 persons per 10,000 persons and most of them were asymptomatic and diagnosed before the age of 40 years. Severe manifestation of ARVC leads to sudden death especially in young Athletes[2].

Clinically ARVC present with recurrent ventricular tachycardia or supraventricular arrhythmias with morphological manifestation in Left Bundle Branch Block (LBBB), with typical findings of right pericardial ECG changes[2,3]. However, because of nonspecific nature and broad spectrum of variation of disease, diagnosis of ARVC is often difficult. Hence several investigations like 2D echo, Cardiovascular MRI and 2020 International guidelines may help to correctly diagnose this disorder[4].

Herewith we report a case of ARVC with left ventricular involvement presenting as recurrent ventricular tachycardia.

Case Report

A 39-year-old male patient with no comorbidities was admitted in CCU with palpitations and giddiness. His ECG showed ventricular tachycardia with LBBB morphology (Fig 2). His BP was low, and rhythm got converted to sinus rhythm after synchronized cardioversion with 200 J. He was given bolus amiodarone injection. He had similar complaints in the past for which he was admitted in another hospital and received cardioversion and started on amiodarone. His amiodarone was continued.

On physical examination no significant positive findings. On cardiovascular examination his heart sounds were normal and no murmurs and additional sounds were detected. After cardioversion his resting ECG showed T inversion in V1 to V4 (Fig 1). On further work up his blood investigations were normal. 2D echocardiography showed dialted RA, Right Ventricle (RV). RV was dilated with RV apex aneurysm and hypokinesia of RV free wall with RV dysfunction TAPSE 11 mm. There was low pressure moderate TR. Left Ventricle (LV) appeared normal on 2D echocardiography.

His MRI was done which showed dilated RV with thinning of walls and fibrofatty infiltration seen more severe near

Editor's Comment :

- ARVC is a rare cardiac life threatening disorder presents with ventricular tachycardia.
- 2020 International criteria helps to diagnose ARVC.
- Cardiac MRI is helpful in diagnosing involvement of left ventricle.
- ICD implantation is the treatment choice for these patients.
- Medical management with beta-blockers and amiodarone are helpful to prevent recurrent episodes of ventricular tachycardia.

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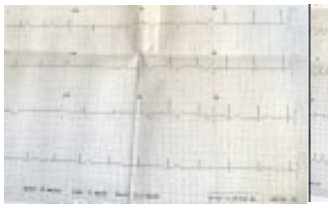

Fig 1 — Resting ECG showing T inversion in V1, V2, V3.

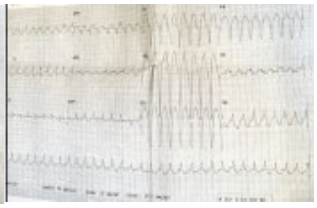

Fig 2 — ECG during Ventricular tachycardia showing LBBB morphology.

apical region. Right ventricular ejection was 38%. MRI also revealed patchy involvement of left ventricle with mildly reduced ejection fraction to 40%.

The definitive diagnosis of ARVC was made as per 2020 International criteria. He was advised ICD implantation as he meets the criteria of class 1 indication for ICD implanatation. He was not willing for ICD implanation. During hospital stay he had 2 recurrent episodes of venricular tachycardia which required DC cardioversion.

As he was not willing for ICD impalntation we continued his amiodarone and started betablockers with metoprolol and ramipril in view of left ventricle involvement. His genetic testing was adviced, but he was not willing for the testing and could not be done.

There were no further recurrences of tachycardia and he was discharged with amiodarone and betablockers and ramipril.

Discussion

Arrhythmogenic Right Ventricular Cardiomypathy (ARVC) also called arrhythmogenic right ventricular dysplasia is an inherited heart disease with autosomal dominant inheritance with variable expression. ARVC was first described in 1977, is a poorly understood, yet lethal cardiac disease.

ARVC is characterised by fibrofatty replacement of the right ventricular myocardium. It is more commonly seen in males. It can be isolated or familial with autosomal dominant pattern of inheritance. Genetic variations have been found in desmosomes responsible for cell to cell binding[5]. The clinical onset is delayed to adolescence or early adulthood. Clinical manifestations vary with age and stage of disease[6]. In early stage of the disease changes are subtle or absent and confined to localised region of the right ventricle typically of the inflow tract, outflow tract and apex of the RV, the triangle of dyplasia[1,4]. LV involvement in ARVC has been described with a prevalence of 16% to 76% with increase in use of cadiac MRI. It can affect inter ventricular septum but more often involves LV free wall with a predilection for posteriolateral area[4].

The diagnosis was based on 2020 International criteria[7], our patient met two major and one minor criteria hence definitive diagnosis of ARVC done.

Our patient had T wave inversion in V1-V4 on baseline ECG and during ventricular tachycardia LBBB morphology suggesting origin of VT from right ventricular out flow tract. His 2D echocardiography revealed dilated right ventricle with hypokinesia of right free wall and apex. His MRI showed involvement of right ventricle along with left ventricle. Based on MRI findings we categorized patient into fourth stage of disease.

Specific management options like life style changes, pharmacological treatment, catheter ablation, ICD placement or heart transplantation are recommended for the patients with ARVC.

In this patient ICD placement was a class I recommendation, but could not be done as patient was not ready for the procedure. ICD placement is recommended in ARVC patients who are at high risk of sudden death, experienced one or more episodes of hemodynamically unstable sustained VT or VF (Class I) or severe systolic dysfunction of RV,LV or both irrespective of arrhythmias (Class I).

We treated our patient with beta-blockers and amiodarone and angiotensin converting enzyme inhibitors in view of left ventricle dysfunction on cardiac MRI. Patient was advised to avoid strenuous activities that can precipitate VT or VF.

Conclusions

ARVC is a rare disorder, and correctly diagnosing is important to avoid unnecessary delay in managing the patients. 2020 International criteria helps to diagnose ARVC. Cardiac MRI is helpful in diagnosing involvement of left ventricle. Although ICD implantation is the treatment choice for these patients, medical management with beta-blockers and amiodarone are helpful to prevent recurrent episodes of ventricular tachycardia.

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