Monomorphic Ventricular Tachycardia, Commonest VT with A Rare Etiology : Case Report

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ABSTRACT

Arrhythmogenic right ventricular dysplasia (ARVD) is an inherited condition characterised by fibrofatty replacement of the right ventricle. clinically by ventricular arrhythmias of right ventricular origin which may lead to sudden death, mostly in young people and athletes.As the presentation is non-specific, ARVD can be a diagnostic challenge leading to delayed treatment. We report two cases along with the review of literature, of a 67-year-old man and a 50-year old man who on presentation had ECG findings of monomorphic VT. Further investigation helped in confirming this rare and potentially fatal cardiac condition.

Introduction:

Originally described as affecting the right ventricle, arrhythmogenic right ventricular dysplasia or cardiomyopathy (ARVD/ARVC), this disorder can affect either or both the ventricles. Patients often present first with ventricular tachycardia. Genetic defects in proteins of desmosomal complex disrupt myocyte functions and adhesion, leading to replacement of myocardium by deposits of fat. Thin ventricular walls may be visualised by echocardiography but are better visualised on MRI. Implantable defibrillators are usually indicated to prevent sudden death.

Case Report

Case 1

A 60-year old male presented with chest discomfort for 4 days, associated with palpitations and profuse sweating. Patient was a known case of type II Diabetes, on treatment for 8 years. Examination revealed extreme tachycardia, while rest of the systemic examination was insignificant. ECG showed ventricular tachycardia ***(Fig.1),*** patient was cardioverted with a DC shock of 120J. Patient reverted to sinus rhythm with ECG showing T wave inversion in leads II, II, aVF and V2, V3, V4 ***(Fig.2).***

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Patient was given Inj. Amiodarone 150 mg stat and started on antiplatelets, ACE-Inhibitor and Beta-Blocker, suspecting an ischemic etiology for the ventricular tachycardia. Patient's lab investigations revealed a deranged LFT and KFT reports, possibly due to hypoxic injury caused by the ventricular tachycardia. Patient subsequently developed bradycardia with occasional VPCs and hence amiodarone and Beta-Blockers were stopped. Trans Thoracic Echocardiogram (TTE) revealed preserved left ventricular (LV) function with no regional wall motion abnormality; but severely impaired and dilated right ventricle (RV) with multiple small outpouchings in right ventricle ***(Fig. 4).*** In view of TTE findings and VT, right heart aetiology, especially ARVD was suspected. Magnified ECG demonstrated characteristic epsilon waves in the anterior leads. Cardiac MRI was supportive of ARVD with thinned out and dilated right ventricle wall and few small outpouchings arising from the right ventricle wall ***(Fig. 5).*** The patient was started on oral phenytoin (Class IB anti-arrhythmic) and subsequently patient's bradycardia resolved. Patient was referred to a higher centre for ICD implantation.

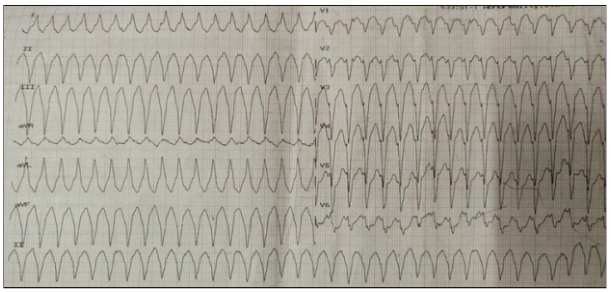

Figure 1 : **ECG of patient on admission showing monomorphic ventricular tachycardia (VT)**

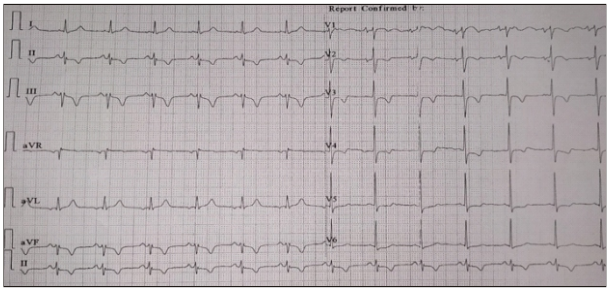

Figure 2 : **ECG after cardioversion showing normal sinus rhythm.**

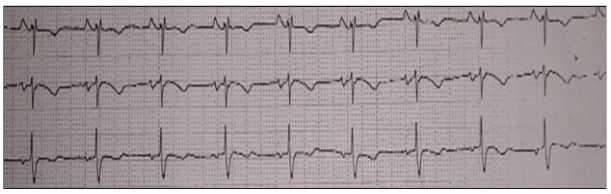

Figure 3 : **ECG with double standardisation showing epsilon waves in V1, Prolonged S wave duration, T wave inversion in V1-V3. (Characteristic of ARVD)**

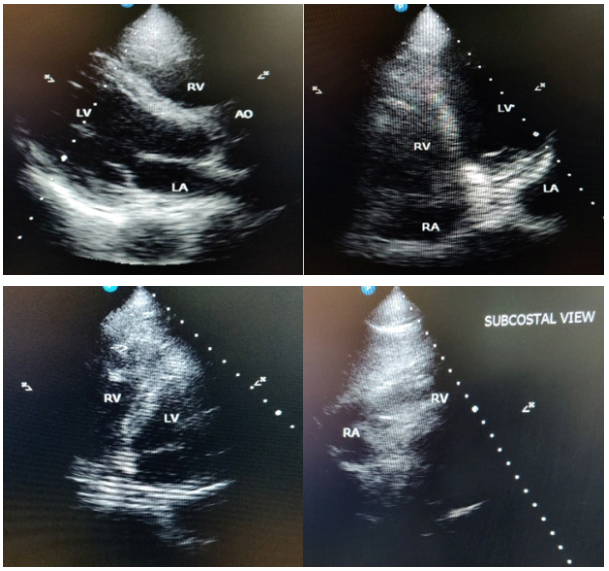

Figure 4 : **Echocardiogram images showing dilated RV with outpouching in RV (arrow)**

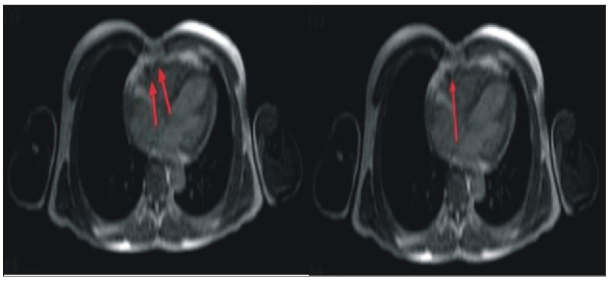

Figure 5 : **Cardiac MRI of patient showing a dilated and thinned out RV with few small outpouchings (arrows)**

Case 2 -

A 50-year old male presented to the emergency department with complaint of palpitations for a duration of 7 hours. There was no history of chest pain, breathlessness or syncope. Patient's past medical history was insignificant. Patient has extreme tachycardia, apart from which rest of his systemic examination was normal and so were his lab reports. Patient's ECG showed ventricular tachycardia ***(Fig. 6).*** Patient was sedated with IV Diazepam and was cardioverted with a DC shock of 50 J, after which the patient's rhythm reverted to normal sinus rhythm ***(Fig. 7).*** Patient was given IV Amiodarone 150 mg stat. An hour later, patient again developed palpitations, with cardiac monitor showing ventricular tachycardia. This time patient was given a DC shock of 200 J and patient reverted to normal sinus rhythm. Patient was the started on continuous IV infusion of Amiodarone for 24 hours and oral Amiodarone thereafter. Transthoracic Echocardiogram (TTE) revealed a dilated right ventricle (RV) with prominent trabeculae and outpouching in RV free wall ***(Fig. 8),*** while the left ventricle (LV) was normal, with an ejection fraction (LVEF) of 60% and no RWMA. With the TTE findings and ECG findings pointing towards a diagnosis of ARVD, cardiac MRI of the patient was planned. Cardiac MRI was suggestive of a dilated right ventricle (RV) with global RV hypokinesia, RV wall appeared to be thinned out with irregular appearance ***(Fig. 9),*** consistent with the diagnosis of

arrhythmogenic right ventricular cardiomyopathy / dysplasia (ARVC/ARVD). Coronary angiography was done and revealed no abnormality. Patient consented for ICD insertion and was referred to higher centre for the same.

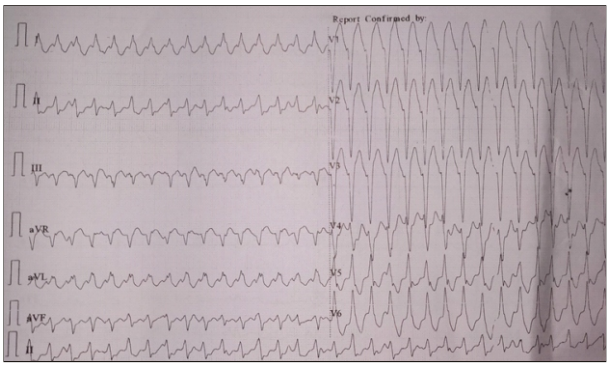

Figure 6 : **ECG of patient on admission showing Monomorphic ventricular tachycardia (VT)**

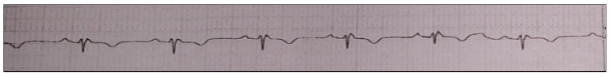

Figure 7 : **ECG of patient post DC shock reverted to normal sinus rhythm with characteristic epsilon wave.**

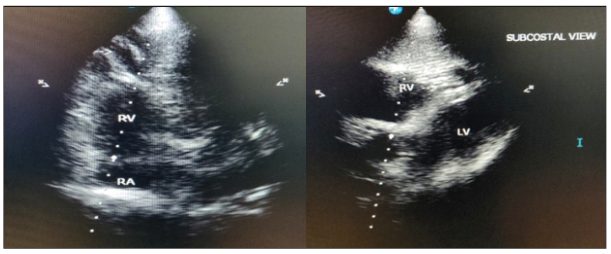

Figure 8 : **2d Echo imaging of the patient showed dilated right ventricle (RV) with prominent trabeculae and outpouching in RV free wall (arrow).**

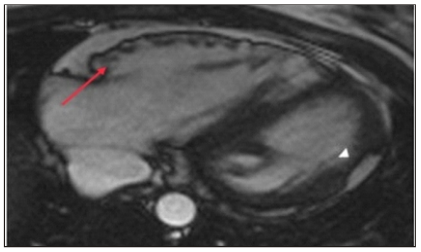

Figure 9 : **Cardiac MRI of the patient showed a dilated right ventricle with irregular and thinned out margin suggestive of ARVD (arrow)**

Discussion :

Ventricular Tachycardia (VT) is a broad complex tachycardia originating in the ventricles. There are several different varieties of VT the most common being Monomorphic VT. Ventricular tachycardia may impair cardiac output with consequent hypotension, collapse, and acute cardiac failure. This is due to extreme heart rates and lack of coordinated atrial contraction. The presence of pre-existing poor ventricular function is strongly associated with cardiovascular compromise.

Decreased cardiac output may result in decreased myocardial perfusion with degeneration to VF. Prompt recognition and initiation of treatment (e.g. electrical cardioversion) is required in all cases of VT.

Morphologically Ventricular tachycardia can be classified into the following types

- Monomorphic
- Polymorphic VT
- Torsades De Pointes
- RVOT
- Fascicular Tachycardia
- Bidirectional VT
- Ventricular Flutter
- Ventricular Fibrillation

Monomorphic VT can be identified by

- Regular rhythm.
- Originates from a single focus within the ventricles.
- Produces uniform QRS complexes within each lead each QRS is identical (except for fusion / capture beats).

Among the various causes of monomorphic VT the important ones are myocardial infarction, dilated cardiomyopathy, hypertrophic cardiomyopathy and arrhythmogenic right ventricular dysplasia (ARVD).

Arrhythmogenic right ventricular dysplasia (ARVD) is an inherited condition characterised by fibrofatty replacement of the right ventricle. It causes sudden death in 30% of young adults and in

5% of those less than 65 years of age. True incidence of this disease is not known but the prevalence is approximately 0.020.1% in the general population.

Patients with ARVC may have a wide range of presenting symptoms, varying from fatigue, atypical chest pain, syncope or acute coronary syndrome. Main presenting features include :

1. ***Arrhythmias :*** Generally, with left bundle branch block morphology, as they are originating from the RV.
2. ***SCD :*** ARVC is responsible for SCD for approximately 5% of patients under the age of 65 years and for 34% deaths in young athletes. The annual mortality rate varies from 3% in the untreated to 1% in those with pharmacological medical treatment (not including ICD treatment). The cause of SCD is commonly VT degenerating into ventricular fibrillation (VF). The fibrofatty deposits form the substrate required to trigger these events induced by adrenergic stimulation.
3. ***Heart Failure :*** Patients may develop isolated RV failure or left-sided failure. This is generally seen in the fourth or the fifth decadesand is due to dilatation and thinning of the RV.
4. In addition, patients may present with frequent ectopics.

Diagnosis :

Approximately 90% of patients present with ECG abnormalities. The important ones include T wave inversions in leads V1-V3, which are seen in about 54% of the patients (in the absence of RBBB). It is important to remember that this can be a normal variant in children and in young, trained athletes and is therefore useful only in patients over the age of 12 years. In addition, complete RBBB is seen in 15% and incomplete in 18% of patients with ARVD and with the presence of QRS complexes of >50 ms in leads V1-V3 as compared with V6 forms a major criterion. Epsilon waves, which are postexcitation electrical potentials that occur at the end of QRS complex are seen in 30% of all cases of ARVD. They are very specific of ARVD and are due to delayed RV activation. In addition, presence of prolonged QRS duration of >110 ms in precordial leads and presence of RBBB with QRS precordial dispersion of >50 ms is a predictor of arrhythmic events. Late action potentials on signal averaged ECG, especially on the left precordial leads are a minor criterion for diagnosis of ARVD. Several studies have also shown a relation between ARVC and Brugada syndrome.

The major echocardiographic findings include RV dilation, localised aneurysms, enlarged left atria (LA), dilated RV outflow tract, increased reflectivity of moderator band, prominent trabeculations of RV apex and inferobasal dyskinesis. Other important parameters include RV end-systolic and end-diastolic diameters and ratio of RV to LV end-diastolic diameters. A ratio of >0.5 has a sensitivity of 86%, specificity of 93% and a positive predictive value of 86% in confirming the diagnosis of ARVC.

MRI is considered to be the one of the important and accurate non-invasive tests available to diagnose ARVC. It helps to analyse RV anatomically, morphologically and functionally. It is useful in demonstrating intramyocardial fat deposits, wall thinning, hypertrophy, trabeculations, right ventricular outflow tract (RVOT) enlargement, presence of RV aneurysms, RV dilation, failure of systolic thickening and impaired global RV function. In addition, MRI has been used for diagnosing ARVC using anatomical, morphological and functional criteria including : presence of high intensity areas indicating fat deposits, RVOT ectasia, dyskinesia, RV dilatation and right atrial (RA) enlargement. However, this technique may be limited because of thin RV free wall resulting in difficulty in assessing RV thickness and in identifying intramyocardial fat in the presence of epicardial and pericardial fat deposits.

Management :

Despite the importance of antiarrhythmic drugs in ARVC there are no clinical trials focusing on their efficacy in these patients. -blockers and amiodarone are considered useful for patients with asymptomatic ARVC to suppress adrenergically stimulated arrhythmias. Studies that have been carried out on sotalol prove it to be more effective than other -blockers and amiodarone for inducible

and non-inducible VTduring programmed stimulation in 68% but its efficacy in preventing sudden death is unknown. In some cases, verapamil can also be considered. In a recent study of patients with ICDs, amiodarone proved to be more effective than â-blockers and sotalol.

The recommendations for ICD implantation for the prevention of SCD in patients with ARVD/ARVC include documented sustained VT or VF for those who are receiving optimal medical therapy and have a reasonable expectation of survival. For primary prevention, ICD implantation can be considered in patients with extensive disease, including those with LV involvement, with one affected family member with SCD, or when VT or VF has not been excluded as the cause of syncope, and who are receiving optimal medical therapy and have a reasonable expectation of survival.

As ARVC has been recognised an important cause of death in young athletes, mostly secondary to malignant ventricular arrhythmias caused by adrenergic stimulation, young patients with this condition are prohibited from vigorous athletic sports. This restriction applies even after ICD implantation, as most devices are programmed to deliver a maximum of six shocks only, which may not be enough to terminate malignant ventricular fibrillation.

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