

Asymptomatic Presentation in Arrhythmogenic Right Ventricular Cardiomyopathy: a Case Report.

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Abstract

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is an inherited cardiac condition leading to one of the common cause for sudden cardiac death in young individuals. It is characterized by fibrofatty replacement of the cardiac myocytes predominantly the right ventricle. As the presentation can be varied and very nonspecific, it often leads to delay in diagnosis and specific treatment. Here, we present a 30 year male who was asymptomatic and detected to have electrocardiograph (ECG) and echocardiographic abnormalities suggestive of ARVC. Further, a cardiac imaging with cardiac MRI confirmed the diagnosis of ARVC highlighting the importance of multimodality imaging.

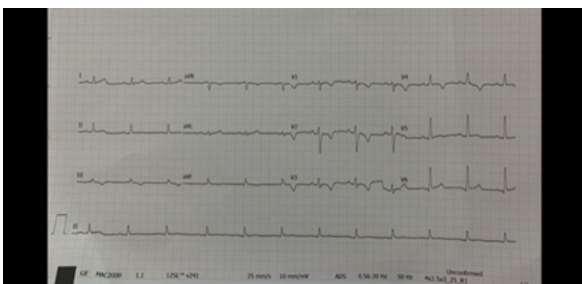
Keywords: Arrhythmogenic Right ventricular Cardiomyopathy; Asymptomatic Presentation.

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Case

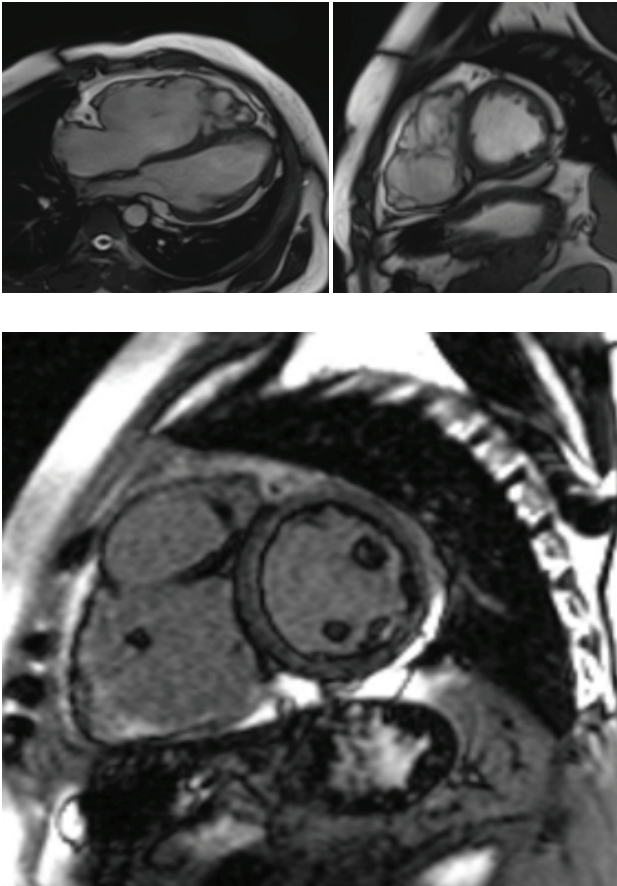
A 30 year old male teacher by occupation presented for a routine health examination at our institute. He had no symptoms of breathlessness, chest pain, dizziness or palpitation. His clinical examination was unremarkable with normal vital parameters. There was no significant history of cardiac disease or any sudden cardiac death in the family. He had been active and no any comorbidities till date. His routine electrocardiograph (ECG) showed repolarization abnormalities in form of T wave inversion in leads V1-V4 and lead III (major). (Image 1)

Image 1: ECG showing T wave inversion in V2-V4.



His echocardiogram revealed dilated right atrium (RA) and right ventricle (RV) with dyskinetic RV apex along with dilated right ventricle outflow tract (RVOT):(parasternal long axis (PLAX) dimension-51mm, parasternal short axis dimension (PSAX)-53mm, moderate tricuspid regurgitation with mild pulmonary hypertension Tricuspid annular plane systolic excursion (TAPSE)-19mm, RV systolic tissue doppler velocity (RV s TDI) 9.9mm and normal Left Ventricle (LV) dimension and systolic function (major). He further underwent a cardiac MRI (plain and gadolinium enhanced) which showed a markedly dilated right ventricle and markedly hypokinetic free walls of the right ventricle with associated thinning of the RV wall and patchy fatty areas noted in the right ventricular wall. Late gadolinium enhancement scan showed patchy enhancement of the right ventricle free wall. Right ventricle apex and mid free wall showed hypertrabeculated appearance. (Major) These findings with compared to the Task Force criteria (2010) for Arrhythmogenic right ventricular cardiomyopathy (ARVC) fulfilled for the definite diagnosis of ARVC with 2 major (both Echo and cardiac MRI/ECG). The index patient was totally asymptomatic and hence was offered no active treatment but with close follow up with monitoring of any new symptoms. However, all first degree relatives of the index case have been advised for a screening echocardiographic examination.

Image 2: Cardiac MRI showing dilated right ventricle with thinned right ventricular wall with features of fatty infiltration and late gadolinium enhancement .



Discussion

In 1978 Frank and Fontain first described ARVD (arrhythmogenic right ventricular dysplasia) in six patients presenting with ventricular tachycardia (VT).¹ It was defined as “total or partial replacement of right ventricular muscle with fibro-fatty tissues associated with left bundle branch (LBBB) morphology arrhythmias”. True incidence of this disease is not known but the prevalence is approximately 0.02–0.1% in the general population.¹

The diagnosis of ARVC is made in 80% of cases in patients below the age of 40 years. Men are more commonly affected. Hence, it should be suspected in young patients presenting with unprecedented history of syncope, cardiac arrest or arrhythmias. The natural history of ARVC depends on the rate of progression of RV dysfunction and the clinical presentation. The mortality rate has been reported to be in the range of 4–20%. It seems to be equal in men and women, though peaking in the fourth decade. It is stated to account for 5% of deaths in young adults and 25% deaths in athletes.²

Patients with ARVC may have a wide range of presenting symptoms, varying from fatigue, atypical chest pain, syncope or acute coronary syndrome. The main presenting features include arrhythmias, sudden cardiac death or even heart failure. 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).³ Epsilon waves, which

are post excitation electrical potentials that occur at the end of QRS complex are seen in 30% of all cases of ARVC. They are very specific of ARVC and are due to delayed RV activation.⁴

Major echocardiographic findings include RV dilation, localized aneurysms, enlarged left atria (LA), dilated RV out flow tract, increased reflectivity of moderator band, prominent trabeculations of RV apex and inferobasal dyskinesia. MRI is considered to be one of the important and accurate non-invasive tests available to diagnose ARVC. It helps to analyse RV anatomically and functionally. It also identifies intramyocardial fat deposits, wall thinning, hypertrophy, RVOT enlargement and RV dilatation and aneurysm .

β-blockers and amiodarone are considered useful for patients with asymptomatic ARVC to suppress adrenergically stimulated arrhythmias. Intracardiac defibrillators (ICD) should be implanted in for the prevention of SCD in patients with ARVD/ARVC include documented sustained ventricular tachycardia or fibrillation for those who are receiving optimal medical therapy and have a reasonable expectation of survival (Class I recommendation). For primary prevention of ICD implantation can be considered (Class II a recommendation) in patients with extensive disease, including those with LV involvement, with one affected family member with SCD, or undiagnosed syncope when Ventricular Tachycardia (VT) or Ventricular Fibrillation (VF) has not been excluded as the cause of syncope, and who are receiving optimal medical therapy and have a reasonable expectation of survival.⁵

Conclusion

ARVC is an important cause of sudden cardiac death in young individuals. It is an inherited condition characterized by fibrofatty replacement of the right ventricle. Diagnosis should always be suspected in young patients who have history of resuscitated cardiac arrest or aborted sudden cardiac death, syncope and ventricular arrhythmias in the past. The clinical presentation can be varied from asymptomatic to life threatening arrhythmias. This case highlights the role of basic 2D echocardiography to identify suspected ARVC case along with multimodality imaging with cardiac MRI to make a definite diagnosis of ARVC.

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