Case Report

ARRHYTHMOGENIC RIGHT VENTRICULAR DYSPLASIA WITH BIVENTRICULAR DYSFUNCTION CONFIRMED BY GATED MULTI DETECTOR COMPUTERIZED TOMOGRAPHY SCAN

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ABSTRACT

Arrhythmogenic right ventricular dysplasia (ARVD) is a familial cardiomyopathy characterized by fibro fatty replacement of right ventricle. Frank left ventricular dysfunction is a rare occurrence in ARVD. Diagnosis is confirmed by magnetic resonance imaging (CMR) and /or biopsy but both are limited by logistic and technical factors. Multi detector computerized tomography (MDCT) scan is a useful and readily available non-invasive diagnostic alternative of CMR. We report a case of ARVD presenting with ventricular tachycardia (VT) and severe biventricular dysfunction confirmed by MDCT.

KEYWORDS: Arrhythmogenic right ventricular dysplasia, Biventricular dysfunction, Multi detector computerized tomography (MDCT) scan.

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INTRODUCTION

Arrhythmogenic right ventricular dysplasia (ARVD) is a poorly understood and underdiagnosed form of familial cardiomyopathy which is characterized by fibro-fatty replacement of right ventricular myocardium leading to a wide spectrum of anatomical, morphological, functional and electrical derangements. These can manifest in young males as minor symptoms of palpitations to severe life threatening arrhythmias, congestive cardiac failure and death.

Latent subclinical left ventricular involvement has been reported frequently (50-67%) in ARVD but frank biventricular dysfunction is quite rare and occurs very late in the course of disease in patients who are prevented from sudden death by implantation of internal cardioverter defibrillator (ICD)[1].

Cardiac magnetic resonance imaging (CMR) is historically considered to be a non-invasive tool of choice for confirming the diagnosis of ARVD but lack of its wide availability and technical factors make it less viable in many institutes. MDCT is a useful and readily available non-invasive diagnostic alternative to MRI.[2] We report a case of ARVD presenting with VT and severe biventricular dysfunction confirmed by gated MDCT scan.

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CASE REPORT

A twenty years old male was admitted to hospital with the complaint of palpitations and dizziness for 15 days. He reported similar episode about five years back and was treated with Aspirin, Diltiazem and a â-blocker but then he lost follow up. He also had history of shortness of breath on mild to moderate exertion and pedal edema but denied any history of orthopnea or paroxysmal nocturnal dyspnea. There was no family history of heart disease or sudden death. He had no history of smoking, illicit drug use or alcohol consumption.

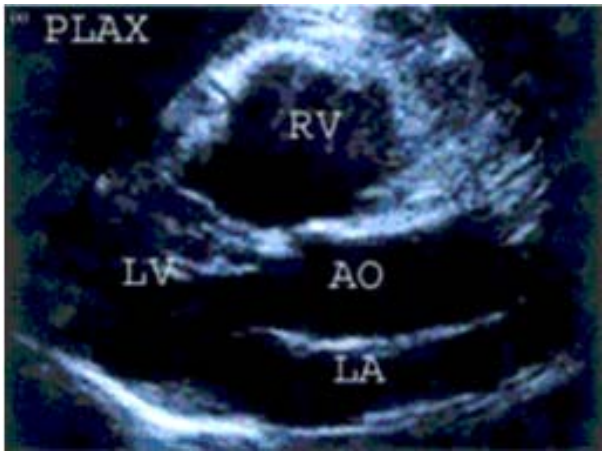
On admission he was haemodynamically stable and was not in overt heart failure. Blood pressure was 110/70 mmHg and heart rate was around 180 b/min. Jugular vein was distended. The precordial examination revealed gallop rhythm with no appreciable murmur. Chest examination revealed no abnormal finding. ECG showed sustained ventricular tachycardia of LBBB pattern (1a).

Ventricular tachycardia failed to respond to intravenous Amiodarone and had to be cardioverted with synchronized DC shock. His post conversion 12-lead ECG showed sinus rhythm and inverted T-waves in leads V1–V3 with rippling of ST segment in V1 and a 'P' pulmonale. Frontal plane

axis was normal. His cardiac biomarkers were normal as were other routine laboratory tests. Chest X-ray showed globally enlarged cardiac silhouette with no significant lung parenchymal changes. Echocardiography showed markedly dilated right ventricle with global hypokinesia. The right ventricular wall was abnormally echogenic and thin suggesting myocardial replacement by fibro fatty tissue (Figure 1). Surprizingly, the left ventricular systolic function was also severely reduced with normal dimensions. Cardiac valves were normal as were estimated pulmonary artery pressure and pulmonary capillary wedge pressure. As CMR was not available at our hospital, MDCT scan was done which showed markedly dilated RV with apical aneurysm and markedly impaired biventricular function in cine imaging. Attenuation pattern, though not conclusive, but was quite suggestive of fatty replacement of right ventricular myocardium (Figure 2).

Patient was treated with intra venous Amiodarone and oral â blockers and registered for insertion of an ICD as early as possible.

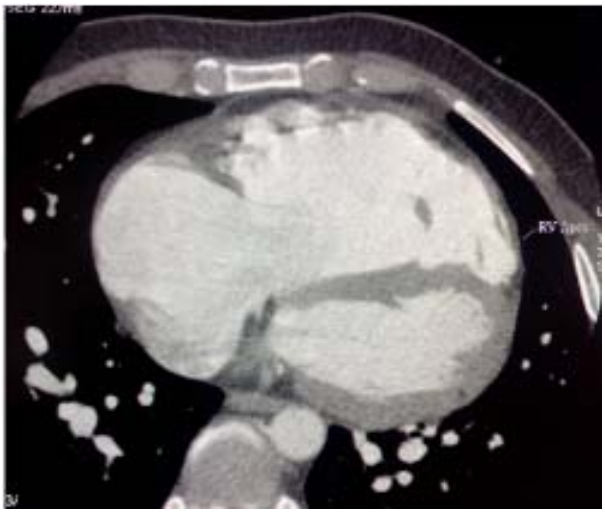
Figure 1: Echocardiography (parasternal long axis view), showing huge right ventricular out flow tract (measuring 56mm) along with abnormal echogenicity



DISCUSSION

Arrhythmogenic right ventricular dysplasia (ARVD) is a common cause of sudden death in young adults.[3] Typical patient is a male in the third decade of life.[3] ARVD is primarily inherited in autosomal dominant fashion but autosomal recessive variants also occur. Culprit genes are those encoding for desmoplakin and plakoglobin, the proteins for cell to cell adhesion in myocardium and ryanodine receptor, RYR2 involved in ion channels.[4]

Figure 2: Contrast enhanced MDCT image showing dilated RV with thinning of RV free wall. Note apical aneurism (arrow) and hypo attenuation suggestive of fatty tissue. Cine CT showed severe biventricular dysfunction



Diagnosis of ARVD is made on echocardiography and usually confirmed by CMR or myocardial biopsy both revealing fatty or fibrofatty replacement of right ventricular myocardium. Major treatment goal is to prevent sudden death with implantation of an ICD.

In our patient, marked right ventricular dilatation and dysfunction along with echogenic thin wall were diagnosed by echocardiography and confirmed by gated MDCT scan which additionally showed an RV apical aneurysm and abnormal attenuation suggestive of fatty tissue. Cardiac MRI and biopsy would have been confirmatory.

Another surprising finding in our patient was significant left ventricular systolic dysfunction which is indeed a rare phenomenon in these patients and confers a poor prognosis. Jain et al[5] demonstrated reduced peak systolic regional circumferential strain, calculated by tagged MRI, both in definite and probable ARVD. Moreover, recent studies on genetic aspects of ARVD point to the possibility of left ventricular involvement in its early stages. Because most important genes responsible are the ones encoding different component proteins of the cardiac desmosome, and the fact that both human ventricles are similar with respect to desmosomal structure and gene expression, the phenotypic expression of those genes should in-

volve both ventricles concomitantly though differentially.[6] Our finding of LV dysfunction can be explained on the basis of supposed common genetic substrate between the two ventricles and it should prompt for the revision of left ventricular description in the contemporary Task Force criteria which state that significant LV dysfunction argues against the diagnosis of ARVD.

Magnetic resonance imaging is uniquely suited to evaluate ARVD because of its excellent three dimensional depiction of RV structure, morphology and function and ability to demonstrate intra myocardial fat which is the pathological hallmark of ARVD.[2] However it is not widely available and is limited by technical factors like motion artifacts and is more operator dependent. Many patients with ARVD already have an ICD implanted before a firm diagnosis is made; it also precludes the use of this exceptionally specific diagnostic modality.

Wide spread availability of multi detector CT scanners and less dependence on technical factors have made CT imaging popular compared to MRI in cardiovascular examinations. CT imaging has the capability of providing tissue characterization of myocardium to some extent. The ability to depict fatty tissue along with cardiac morphology makes CT imaging an option for evaluation of ARVD.[2]Dery et al[7], were the first to demonstrate dilated hypokinetic right ventricle using CT imaging in a patient with ARVD. However the ability of conventional CT to detect intra myocardial fat was first reported by Villa et al.[8] in a series of seven patients of ARVD, and subsequently Sotozono et al.[9] provided biopsy confirmation of CT findings. Hamada et al[10] evaluated four patients with ARVD where volume-mode CT clearly depicted an enlarged right ventricle with a scalloped surface of the free wall, conspicuous trabeculations with low attenuation, and abundant epicardial adipose tissue, which are characteristics of ARVD.

CONCLUSION

Arrhythmogenic right ventricular dysplasia is a potentially lethal condition. Left ventricle may also be involved in patients with ARVD and should be diagnosed early to prevent biventricular failure. Gated MDCT scan is a useful alternative to CMR for evaluation and diagnosis of these patients.

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