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Young Male with Recurrent Syncope: A Case Report

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Abstract

Arrythmogenic right ventricular dysplasia/cardiomyopathy (ARVC/D) is an inherited rare cause of cardiomyopathy characterised by fibro-fatty infiltration of right ventricle. ARVC is an under-recognized condition,commonly present in young adults with syncope or sudden cardiac death.Here we present the case of a 20 year old male, presented with history of nausea, chest discomfort and loss of consciousness for 2-3 minutes on exertion. History of similar episodes in the past. ECG revealed T wave inversions in precordial leads with poor R wave progression and ECHO revealed dilated right ventricle (RV) with moderate RV dysfunction. Diagnosis of ARVD was made and treated with antiarrhythmic drugs and AICD (Automated Implantable Cardioverter Defibrillator). It is important to identify the ECG changes of ARVD,there by preventing the potentially lethal consequences.

Keywords: Arrythmogenic Right Ventricular Dysplasia/Cardiomyopathy (ARVD/C); Young Adults; Right Ventricle; T wave Inversions; AICD.

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Introduction

Arrhythmogenic right ventricular dysplasia (ARVD) is a genetic form of cardiomyopathy characterised by fibrofatty infiltration of the right ventricle. It is associated with high incidence of ventricular arrhythmias, including polymorphic non-sustained VT,VF and recurrent sustained monomorphic VT. First manifestation of the disease is unexplained syncope or sudden cardiac death. After HOCM, ARVD is the most common cause for sudden cardiac death in young individuals, especially athletes. It is stated to account for 20% of sudden deaths in all individuals younger than 35 years and 22% of sudden deaths in young athletes.[1] Diagnosis is difficult in most of the cases as clinical presentation may vary. There is no single diagnostic test for ARVD. Diagnosis is based on combination of clinical, electrocardiographic and radiological features (Task Force Criteria).

Case Report

A 20 year old male with no known comorbidities and not on any regular medication presented with history of nausea,chest discomfort followed by loss of consciousness for 2-3 minutes on exertion. History of similar history twice in the past (fivemonths and one year before).No history of tongue bite, tonic-clonic movements, bowel and bladder disturbances, postictal confusion. No history of sudden cardiac death or heart disease in family members.

On Examination: Patient was conscious, oriented with a HR-58bpm, BP-130/80 mm Hg, RR-18cpm. S1,S2 +, no murmurs, NVBS heard in bilateral lung fields, no neurological deficits. ECG (Fig. 1) showed sinus rhythm with a rate 58/min, Twave inversions in V1-V5, poor R wave progression. Echo (Fig. 2) revealed dilated right ventricle, moderate right ventricle dysfunction, normal LV function, RVOT-43 mm Hg, PASP-31 mm Hg. Holter (Fig. 3) showed-ventricular ectopics (bigeminy, couplets, quadrigeminy), Ventricular tachycardia (VT). Diagnosed as ARVC (arrhythmogenic right ventricular cardiomyopathy), treated with sotalol and AICD (Automated Implantable Cardioverter Defibrillator) device placement. After one day of AICD placement, patient had VT, shock delivered by the device, VT still persisting, so started on amiodarone and lignocaine infusion. Patient was monitored for four days and discharged with Tab Sotalol.

Discussion

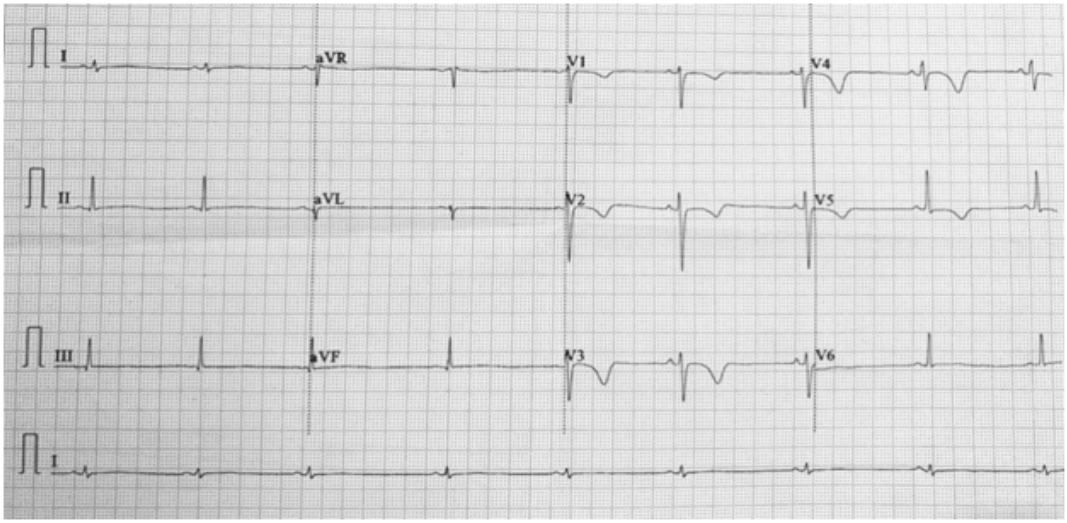


Fig. 1: ECG: T wave inversions V1-V5, poor R wave progression.

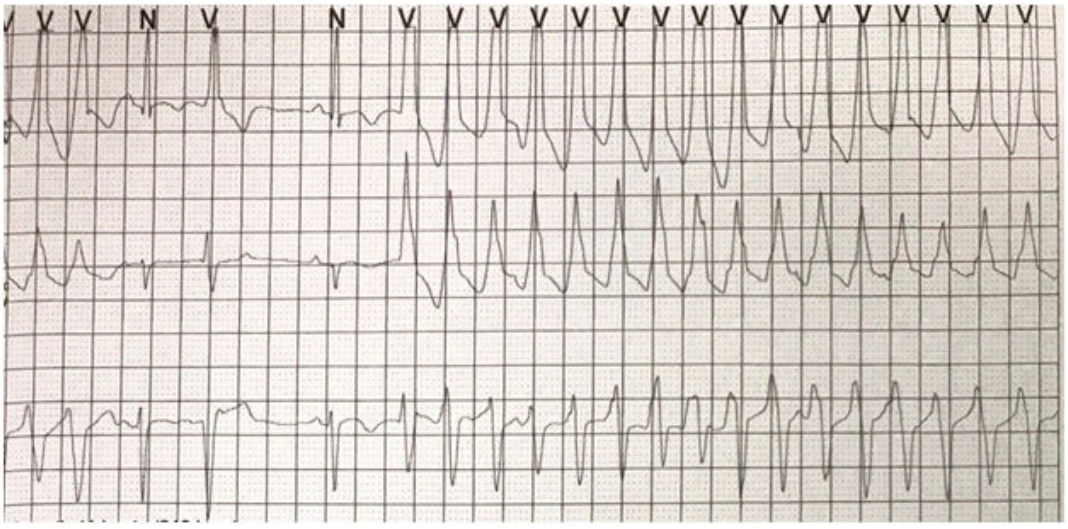
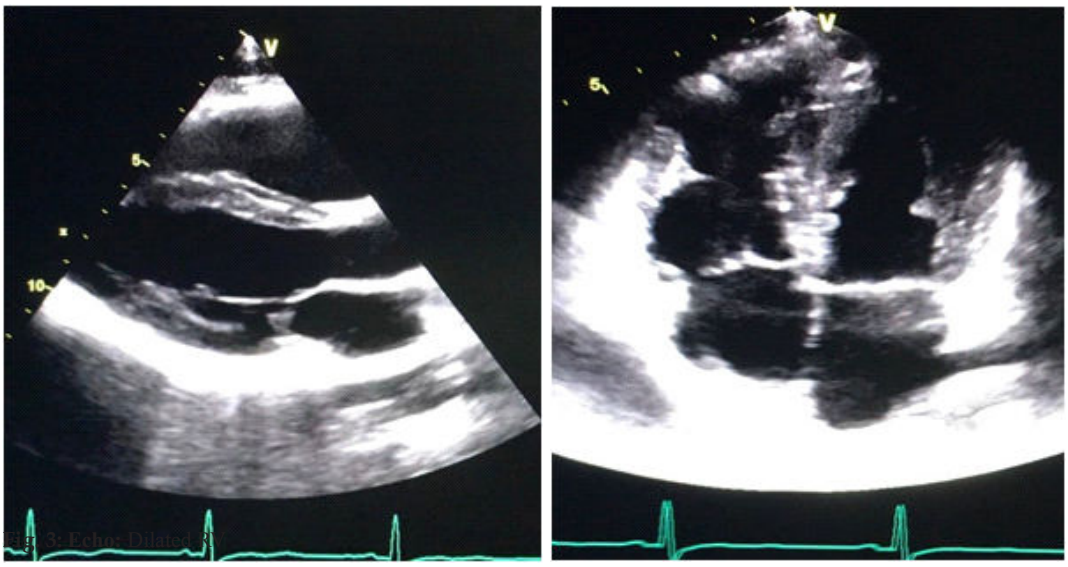


Fig. 2: Holter: Ventricular tachycardia (LBBB morphology).



ARVD is a hereditary progressive cardiomyopathy-first suggested by Fontaine and co-workers in 1977. The best clinical description of the disease originates from Marcus group.[2] Prevalence ranges between1:2000-1:5000. ARVD has a strong family predilection; it is commonly inherited as autosomal dominant trait with various degrees of penetrance. Familial ARVD accounts for 30%–50% of all cases,[3] although penetrance in some families is estimated to be <30%. History of palpitations or syncope at young age or family history of sudden cardiac death at young age should raise suspicion for ARVD.

Six genes have been implicated in the pathogenesis of the disease, four encode major desmosomal proteins and two nondesmosomal genes have been associated with specific types of ARVD.[4] It is characterized by progressive replacement of the right ventricular myocardium by fibrofatty tissue.[5] Left ventricle and septum may be involved in more extensive cases. The most common location for this tissue transformation is between the anterior infundibulum, right ventricular apex, and inferior or diaphragmatic aspect of the right ventricle, the so-called "triangle of dysplasia"[2]

Clinical presentation of the disease is highly variable, patients may present with palpitations, syncope, fatigue, dizziness, ventricular arrhythmias, ventricular ectopics, heart failure or cardiac arrest. Non-specific complaints include abdomen pain or mental confusion. Symptoms usually occur during exertion and mortality is 19%.[6] Pregnant women may present with palpitations and shortness of breath during second trimester.[7] Physical examination may be normal in most of the patients. Widely split S2,S3/S4 heart sound, murmur or asymmetry of chest wall may be seen in few patients. ECG usually shows sinus rhythm, epsilon wave(terminal deflection within or at the end of qrs complex) in V1-V3 or T wave inversions in right precordial leads. ARVD is characterised by LBBB pattern ventricular arrhythmia, as they are originating from RV. Fibrofatty islands generate macro-reentries, thus forming the arrhythmogenic substrate. Echo reveals dilatation and reduction of right ventricle(RV) function , right ventricle aneurysms. Right ventricular aneurysms, regional thinning and dilation of RV, failure of systolic thickening, impaired RV function is visualised on cardiac MRI.

Diagnosis is challenging and critically important. Marcus et. al.[8] have proposed the 2010 revised Task Force Criteria. Task Force criteria includes six categories:

1) Global and/ or regional dysfunction and structural alterations by MRI or ECHO or aniography.
2) Tissue characterization of wall (fibrous replacement and percentage of residual myocytes in right ventricle).
3) Repolarization abnormalities on ECG (T wave inversion in V1, V2 and V3)
4) Depolarization/conduction abnormalities on ECG (epsilon wave in V1, V2 and V3)
5) Arrhythmias (VT with LBBB morphology and superior axis)
6) Family history (AVRC in a first degree relative confirmed with Task Force criteria or at autopsy).

Management of arrhythmias and prevention of sudden cardiac death are the major treatment goals. Therapeutic options include life style modification, Antiarrhythmic drugs, implantable cardiac

defibrillator (ICD) insertion, Radiofrequency ablation, surgery. Lifestyle modification is recommended for all patients because of association with exercise and arrhythmias.[9] Antiarrhythmic medications such as sotalol, amiodarone or beta blockers are used to abolish the arrhythmias. ICD treats arrhythmias by delivering shocks and prevents sudden cardiac death. Indications for ICD placement are symptomatic VT, cardiac arrest due to VT/VF, drug refractory VT,LV involvement,younger age of onset.[10] Indications for radiofrequency ablation are drug refractory VT, tachycardia after ICD placement.[11] It eliminates the conducting pathways causing arrhythmias. Recurrence occurs due to disease progression.In patients with end stage heart failure and refractory ventricular arrhythmias heart transplantation should be considered.

First degree relatives should be screened with ECG,ECHO or cardiac MRI if needed.

Conclusion

ARVD is a cause of sudden cardiac death due to ventricular arrhythmias in young adults. Clinical course is nonspecific, so high index of suspicion is required for diagnosis in any adult presenting with dizziness, syncope or palpitations. It is critically important to identify the ECG changes of ARVD and to evaluate further with ECHO/Cardiac MRI and to initiate treatment. This case was presented to create awareness among physicians about the lethal condition ARVD.

Conflicts of interest: No conflict of interest

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