International Journal of Health Sciences and Research
www.ijhsr.org ISSN: 2249-9571

Case Report

Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia (ARVC/D): A Rare Case Report

Dr. Kushal Kalvit[1], Dr. Rajesh Debbarma[2]

[1]Department of General Medicine, Agartala Government Medical College, Agartala, Tripura India.
[2]Associate Professor, Department of General Medicine, Agartala Government Medical College, Agartala, Tripura, India.

Corresponding Author: Dr. Kushal Kalvit

ABSTRACT

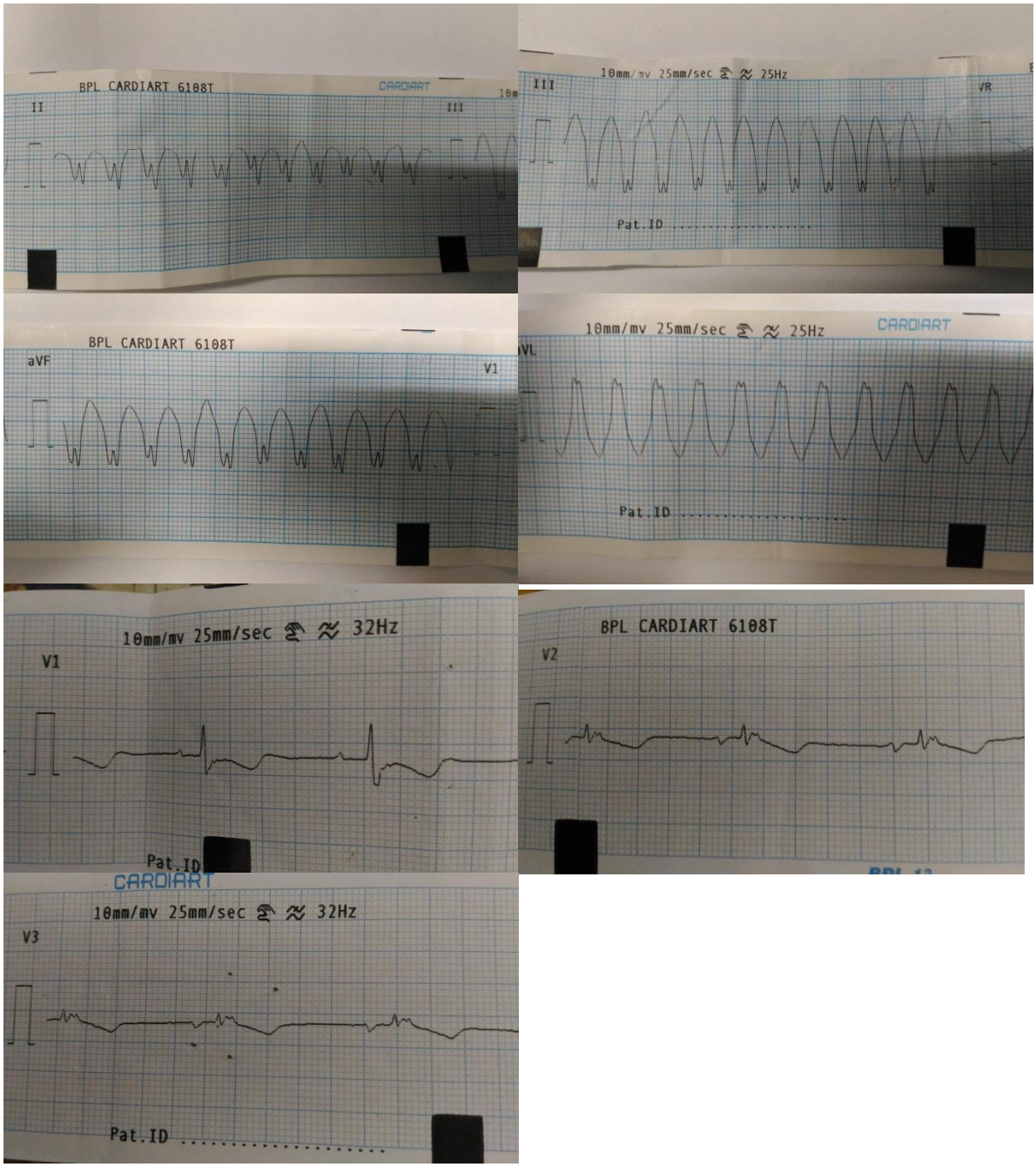
Arrhythmogenic right ventricular cardiomyopathy/dysplasia (ARVC/D) is a genetic form of cardiomyopathy primarily affecting the right ventricle. The natural history is highly variable with a wide spectrum of clinical presentations. Presentation may vary from asymptomatic individuals to overt ventricular arrhythmias. Individuals are at an increased risk of sudden cardiac death (SCD) especially on exertion. We report a case of ARVC/D in a 25 year old healthy male with a strong family history of sudden unexplained death.

Keywords: ARVC/D, Sudden cardiac death (SCD), ICD.

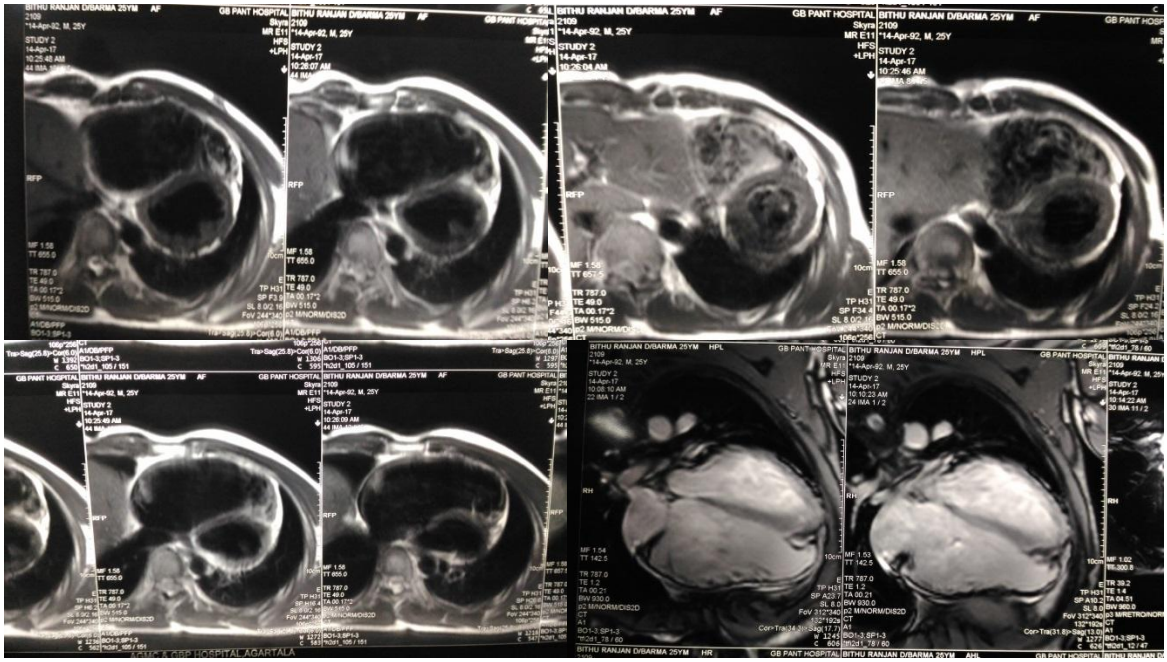
CASE REPORT

A 25 year old male presented with palpitations and shortness of breath while playing football. The symptoms did not subside even after one hour of rest. He had never experienced similar episodes in the past. There was no history of syncope, fever, cough or chest pain. He had no history of diabetes mellitus, hypertension, cardiac disease or any other chronic illness. He had a family history of sudden unexplained death of his father and elder brother few years back. He denied abuse of alcohol or any recreational drugs. On examination, his GCS was 15/15, temperature was 98.7^0F, pulse 165/minute regular rhythm, BP 80/50mmHg. Examination of cardiovascular system revealed a diffuse apical impulse in the left 6^{th} intercostal space anterior axillary line. There was no presence of parasternal heave, thrill or any murmur. Examination of respiratory system, abdomen and CNS was unremarkable. ECG was done which revealed sustained monomorphic ventricular tachycardia (VT) with LBBB morphology with superior axis. Immediate electrical cardioversion was done and patient was stabilised. A repeat ECG after cardioversion showed sinus rhythm, right axis deviation and epsilon waves with inverted T waves in right precordial leads (RVI-RV4) [Figures 1-7]. Routine blood investigations showed Haemoglobin=12.2 grams/dl, TLC=5800/mm^3, platelet count= 2.6 lakh/mm^3, Sodium = 142mmol/L, Potassium = 4.2mmol/L, Blood urea nitrogen = 17 mg/dl, Creatinine = 1.15 mg/dl, Bilirubin = 0.9 mg/dl and Random blood glucose = 110mg/dl. Chest roentgenogram was done which showed cardiomegaly. 2D Echocardiography showed a dilated right

ventricle with regional dyskinesia, hyper-reflective moderator band along with fractional area change of right ventricle (FAC) 17% and parasternal long axis right ventricular outflow tract (PLAX RVOT) 38mm (end diastole). Cardiac MRI was also done and showed marked dilation and thinning of right ventricle, RV dyskinesia and no impairment of left ventricle [Figures 8-11]. Based on the above findings, a diagnosis of arrhythmogenic right ventricular cardiomyopathy/dysplasia was made according to the Revised Task Force Criteria 2010. The patient was prescribed beta blockers and was advised to undergo ICD (implantablecardioverter-defibrillator) implantation for the prevention of sudden cardiac arrest. He was also advised to avoid participation in competitive sports.



Figures 1-8: ECG showing Monomorphic sustained VT with superior axis. Right precordial leads (RV1-RV3) show T wave inversion and presence of epsilon waves.



Figures 8-11. Cardiac MRI shows marked thinning and dilation of right ventricle with normal left ventricle.

DISCUSSION

The natural history of ARVC/D is highly variable. [1] ARVC/D accounts for 11%-22% of cases of SCD in the young athlete patient population. [2,3] In approximately 30-50% of cases it is transmitted with an autosomal dominant pattern of inheritance, with incomplete penetrance and variable expression. Seven genes have been identified that are associated with ARVC/D: plakoglobin, desmoplakin, plakophilin-2, desmoglein-2, desmocollin-2, transforming growth factor beta-3 and TMEM43. [4] It is characterized pathologically by fibrofatty replacement of the right ventricular myocardium. ARVC/D is diagnosed according to the 2010 Task Force Criteria which involves electrocardiography, echocardiography, cardiac MRI, family history, myocardial biopsy, right ventricular angiography.[4]Sotalol and amiodarone have been proposed as effective treatment of sustained VT or ventricular fibrillation (VF) as adjunctive therapy to implantable cardioverter-defibrillator (ICD) or in patients with ARVC/D that are not candidates for ICD implantation. [5] Patients with episodes of sustained VT or VF, unexplained syncope, non-sustained VT on noninvasive monitoring, familial history of sudden death, extensive disease including those with LV involvement and good functional status are potential candidates for ICD implantation even in the absence of ventricular arrhythmias. [6]

REFERENCES

1. Jorge Romero, Eliany Mejia-Lopez, Carlos Manrique, Richard Lucariello. Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC/D): A Systematic Literature Review. Clin Med Insights Cardiol. 2013; 7: 97-114.
2. Dalal D, Nasir K, Bomma C, et al. Arrhythmogenic right ventricular dysplasia: a United States experience. Circulation. 2005; 112(25):3823-32.
3. Corrado D, Basso C, Schiavon M, Thiene G. Screening for hypertrophic cardiomyopathy in young athletes.N Engl J Med. 1998 Aug 6;339(6):364-9.
4. Marcus FI, McKenna WJ, Sherrill D, et al. Diagnosis of arrhythmogenic right ventricular cardiomyopathy/dysplasia: proposed modification of the Task Force Criteria. Circulation. 2010; 121(13): 1533-41.
5. Zipes DP, Camm AJ, Borggrefe M, et al. ACC/AHA/ESC 2006 guidelines for management of patients with ventricular arrhythmias and the prevention of

sudden cardiac death. J Am Coll Cardiol. 2006;48(5):e247-346.
6. Corrado D, Calkins H, Link MS, et al. Prophylactic implantable defibrillator in patients with arrhythmogenic right ventricular cardiomyopthy/dysplasia and no prior ventricular fibrillation or sustained ventricular tachycardia. Circulation. 2010;122(12):1144-52.

How to cite this article: Kalvit K, Debbarma R. Arrhythmogenic right ventricular cardiomyopathy/dysplasia (ARVC/D): a rare case report. Int J Health Sci Res. 2019; 9(9):248-251.

******