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Case Report

Arrhythmogenic Right Ventricular Dysplasia/Cardiomyopathy Involving Isolated Ventricular Septum: Case Report

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Abstract: Arrhythmogenic right ventricular dysplasia (ARVD) is a disorder in which normal myocardium is replaced by fibrofatty tissue. This disorder usually involves the right ventricle, but the left ventricle and septum also may be affected. Patients with ARVD are usually men younger than 35 years who complain of chest pain or rapid heart rate. In some cases, sudden cardiac death is the first presentation. Further confirmation of the diagnosis includes noninvasive studies, such as echocardiography and magnetic resonance imaging of the heart, and invasive studies such as ventricular angiography and endomyocardial biopsy. Patients with ARVD are treated initially with antiarrhythmic agents with serious consideration for automatic implantable cardioverter-defibrillator placement. We are reporting a case of arrhythmogenic Right Ventricular Dysplasia/ Cardiomyopathy involving isolated ventricular septum.
Keywords: Arrhythmogenic right ventricular dysplasia/Cardiomyopathy (ARVD/C), Cardioverter-defibrillator placement, RBBB pattern

INTRODUCTION

Arrhythmogenic right ventricular dysplasia (ARVD), also called arrhythmogenic right ventricular cardiomyopathy (ARVC) or arrhythmogenic right ventricular dysplasia/cardiomyopathy (ARVD/C), is an inherited heart disease. Arrhythmogenic right ventricular dysplasia, which was first described in 1977, is a poorly understood yet potentially lethal cause of cardiac disease [1, 2]. Once thought to be rare, ARVD has been shown to have an incidence of six per 10,000 persons in certain populations [3]. ARVD accounts for 3 to 4 percent of deaths in sports and 5 percent of sudden cardiac deaths in persons younger than 35 years [4]. Given the relative frequency of this disorder and its potential for disastrous outcomes, it is important for primary care physicians to be familiar with its presentation, diagnosis, and management.

CASE REPORT

48yrs male patient was presented with giddiness, multiple episodes lasting for about 1 min. Denies history of palpitations, chest pain, loss of consciousness; with similar history 3 months ago. His medical history included type 2 diabetes mellitus since 1 yr on oral hypoglycemic agents and Dyslipidemia since 1 yr on statin therapy. His blood investigations were within normal limits.

ECG showed sinus rhythm with RBBB pattern and illsustained VT of LBBB pattern with inferior axis (Fig. 1, 2). ECHO showed mild dilatation of RV.TMT test showed Ventricular ectopics (Quadrigeminy) during test and (Bigeminy) during recovery.24 hrs holter was connected which showed Ventricular Bigeminy, Trigeminy and Illsustained Ventricular Tachycardia. MRI showed diffuse thinning and irregularity in the interventricular septum at the basal and mid level suggestive of scar tissue (infract) (Fig. 4, 5).

CAG done showed normal epicardial coronaries.MRI report of scar tissue secondary to ischemia was ruled out. EP Study using 3D CARTOS Mapping system showed two different morphologies of VT noted. Successful RF Ablation of the VT focus along with linear ablation of low voltage area in proximal RVOT and Posteroseptal aspect in the Peritricuspid area.

In view of ARVD and recurrent fast NSVT and Sustained VT and for long term treatment, Patient underwent AICD Implantation.

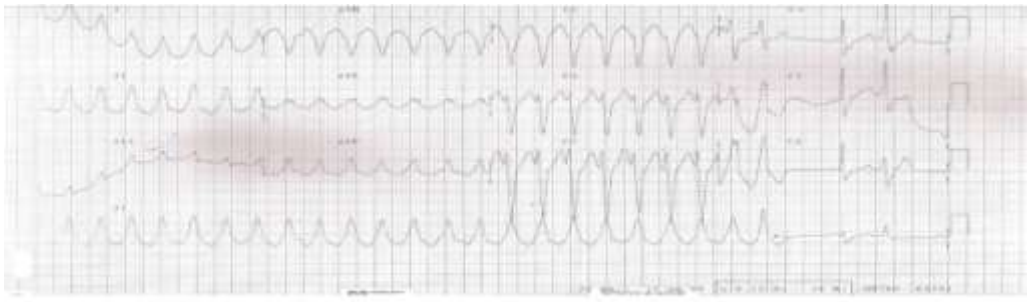

Fig. 1: ECG showing RBBB pattern and illsustained VT of LBBB pattern with inferior axis.

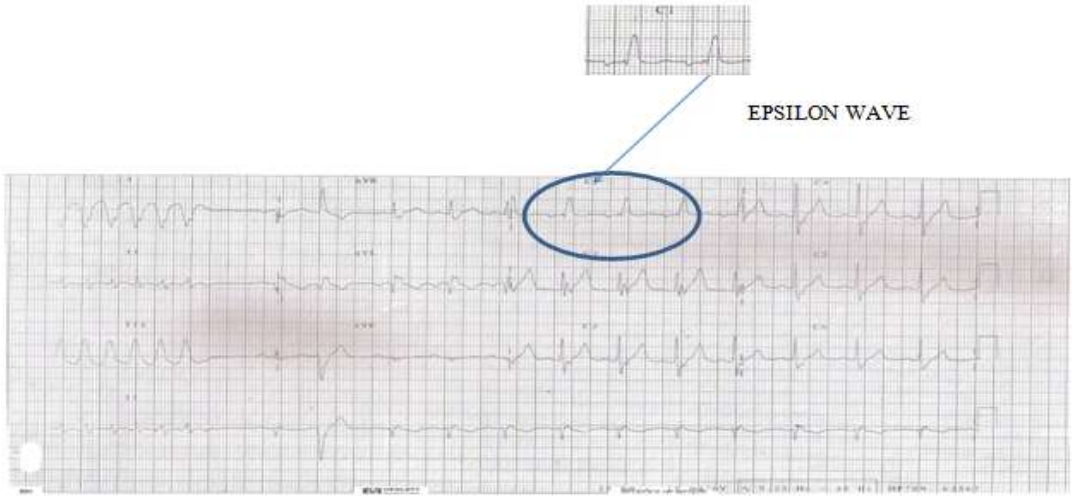

Fig. 2: ECG showing sinus rhythm with RBBB pattern and EPSILON WAVE

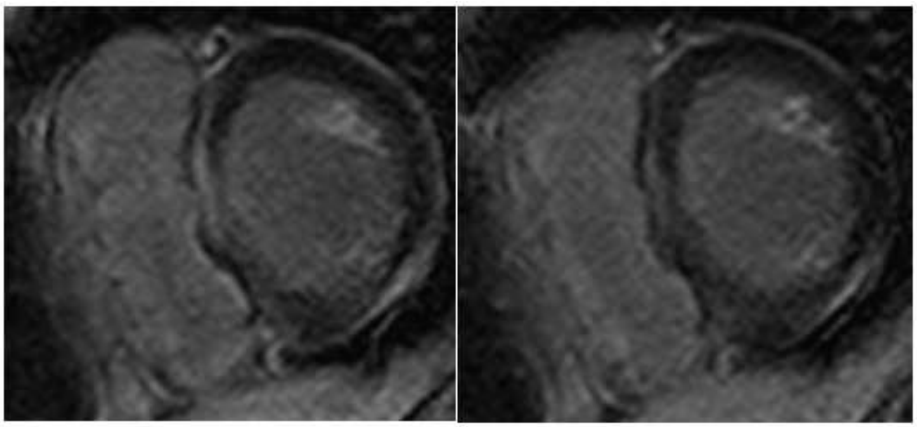

Fig. 4 &5: MRI showed diffuse thinning and irregularity in the interventricular septum at the basal and mid level suggestive of scar tissue (infract)

DISCUSSION

Arrhythmogenic right ventricular dysplasia/cardiomyopathy is genetically determined, progressive cardiomyopathy characterized histopathologically by fibrofatty replacement of the right ventricular myocardium [5]. An estimated prevalence in united states is 1:5000. It is more commonly seen males.It can be isolated or familial with autosomal dominant pattern. Genetic variations have been found in desmosomes that are responsible for cell to cell binding [6]. The clinical onset is delayed to adolescence or early adulthood. Clinical manifestations vary with age and stage of the disease [7]. In early stage of the disease, changes are subtle or absent and confined to a localized region of the right ventricle, typically the inflow tract, outflow tract or apex of the RV, the triangle of dysplasia [8]. Progression leads to more diffuse RV involvementand also LV. Predominant LV disease is also recognized [9]. LV involvement in ARVD/C has been described with a prevalence of 16% to 76% [10]. It can affect septum but more often involves the LV free wall with a predilection for

posterolateral area [11]. The diagnosis is made based on the criteria established by the task force for ARVD/C, limitation is that it does not include LV involvement. These patients are at increased risk for arrhythmias and SCD in early stages and biventricular failure in late stages.

CONCLUSION

ARVD/C presents a diagnostic challenge in patients with predominantly LV or septal involvement due to lack of diagnostic criteria. CMR plays an important role in the diagnosis of ARVD/C. SCD can be prevented with help of an AICD.

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