INTRODUCTION

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is a clinical entity characterized by ventricular arrhythmias and a specific right ventricular pathology (1). On the other hand isolated left ventricular non-compaction is a sporadic or familial cardiomyopathy characterized by prominent trabeculae and deep intertrabecular recesses in left ventricle (2). Although both of these cardiomyopathies are rare disorders, coexistence of these cardiomyopathies at one patient is extremely rare, a review of literature revealed no reported case of ARDC co-existing with isolated left ventricular non-compaction. We present a rare case of ARVC coexisting with isolated left ventricular non-compaction who had incessant ventricular tachycardia (VT) originating from right ventricular outflow tract (RVOT) that has been ablated by electro anatomic mapping successfully.

Keywords: Arrhythmogenic right ventricular cardiomyopathy (ARVC), right ventricular outflow tract (RVOT), ventricular tachycardia

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CASE REPORT

A male patient 36 years of age presented for frequent palpitations and lightheadedness but he did not have any genuine syncope. In his resting ECG there was incomplete right bundle branch block and negative shallow T waves at all precordial leads (figure 1). Because some of his relatives had the diagnosis of ARVC, a cardiovascular MRI test ordered which revealed a non-compacted myocardial layer at apical anterior and apical lateral segments with an epicardial to endocardial myocardial ratio greater than two, additionally interatrial septal lipomatous hypertrophy was detected (Figure 2, 3).

Fig 1: Baseline ECG of the patient
Fig 2: MR image of the patient that shows noncompaction of the left ventricle, the ratio of non-compacted to compacted myocardium is greater than 2.3. Long axis view

Fig 3: Short axis view showing non-compacted to compacted ratio is greater than 2
But right ventricular volumes and wall motion was normal. To investigate concealed brugada an ajmalin test was ordered. At 15th second of ajmalin test the patient had a VT episode with a left bundle and inferior axis ECG morphology. But no ST elevation happened at right precordial leads during fifteen minutes of the test. Thereafter at intensive care unit he continued to have bursts of sustained VT episodes of the same morphology which finally became incessant VT resistant to multiple antiarrhythmic agents and synchronized DC. Then he was transferred to electrophysiology room for catheter ablation to eliminate this electrical storm. By careful moving of ablation catheter in left and right ventricle; RVOT area just below pulmonary valve found to be the earliest activated site during ventricle tachycardia. This site was suspected to be the origin of this incessant VT and at this site multiple radiofrequency energies were given. But during ablation procedure we observed two different left bundle VT morphologies which suggested the possibility of ARVC. At that time the tachycardia was successful ablated and the tachycardia were terminated (Fig 4, 5).

Fig 4: incessant ventricular tachycardia of RV origin
The patient sent back to intensive care unit. But a few hours later the same VT reappeared and a coronary angiogram performed to rule out an ischemic etiology but revealed normal coronaries. Next morning an ablation of VT reattempted but this time using a nonfluoroscopic catheter-based electro anatomic mapping system. After a long session of ablation finally arrhythmia was terminated completely. This patient met one major and three minor criteria of current Task Force criteria of ARDC (T wave inversion in V1-6 derivations as major criteria, VT of RV outflow configuration and, more than 500 ventricular extra systoles per 24 hours by holter and ARVC confirmed by current task criteria in a second-degree relative. As minor criteria)

**DISCUSSION**

ARVC is an under-recognized but an important cause of sudden cardiac death accounting approximately 11 percent of cases overall. Many patients however remain clinically silent and
asymptomatic for decades. 30 percent of cases are familial. Patients usually present with arrhythmias of right ventricular origin and/or sudden death between ages of 10 and 50 years with a mean age of diagnosis approximately 30 years. The most common ventricular arrhythmia is sustained or nonsustained monomorphic VT that originates in the RV and therefore has a left bundle branch block pattern. VT may originate from any part of right ventricle but if it originates RV outflow tract it may be difficult to distinguish ARVC from idiopathic RV outflow tract tachycardia, But different left bundle VT morphologies suggests the possibility of ARVC(3-4). It is important to distinguish RVOT tachycardia from VT due to ARVC. A potential source of confusion is that RVOT tachycardia, like ARVC, may be associated with right ventricular outflow dilatation. On the other hand RVOT tachycardia arise typically from a very narrow area just inferior to the valve in the anterior aspect of the RVOT and occurs exclusively in young to middle aged patients without structural heart disease. Distinguishing an idiopathic VT syndrome from other monomorphic VT syndromes is important given the far better prognosis, greater array of antiarrhythmic drug options, and amenability to cure with ablation (5). But VT's of ARVC and of isolated left ventricular non-compaction are candidates for ICD implantation. Non-compaction cardiomyopathy is a recently recognized disorder, based on an arrest in endomyocardial morphogenesis. The disease is characterized by heart failure, systemic emboli and ventricular arrhythmias (6). But in the case we presented here the coexisting left ventricular non-compaction is a coincidental finding which has no active role in this patient’s arrhythmias. All arrhythmias were originated from right ventricle. Since in early stages of ARVC, imaging findings might be negative. On the other hand non-compaction demonstrated by MRI might be silent.
CONCLUSION
This case demonstrates the importance of diagnostic evaluation by various invasive and noninvasive techniques and detailed analysis of clinical history and presentation to establish the presence and type of heart disease and the origin and pathology causing lethal arrhythmias. Reliance to only imaging findings may result in misdiagnosis of the origin of arrhythmias.
REFERENCES


