CASE REPORT

A Rare Case of Arrhythmogenic Right Ventricular Cardiomyopathy Co-existing with Isolated Left Ventricular Non-compaction

NS Yelgeç, AT Alper, AI Tekkeşin, C Türkkan

ABSTRACT

Arrhythmogenic right ventricular cardiomyopathy is a rare heart-muscle disorder characterized by progressive replacement of right ventricular myocardium by fibrofatty tissue. Noncompaction of the ventricular myocardium is also rare congenital cardiomyopathy, characterized by an arrest in intrauterine endomyocardial morphogenesis. We present an extremely rare patient who presented with incessant ventricular tachycardia and who had both of these two cardiomyopathies at the same time.

Keywords: Arrhythmogenic right ventricular cardiomyopathy, electroanatomic mapping, right ventricular outflow tract.

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 the left ventricle (2). Although both of these cardiomyopathies are rare disorders, their co-existence in one patient is extremely rare. A review of the literature revealed no reported case of ARVC co-existing with isolated left ventricular non-compaction. We present a rare case of ARVC co-existing with isolated left ventricular non-compaction in a patient who had incessant ventricular tachycardia (VT) originating from the right ventricular outflow tract (RVOT), which was successfully ablated by electroanatomic mapping.

CASE REPORT

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

The RV volumes and wall motion were normal. To investigate the concealed Brugada, an ajmaline test was done. At the 15th second of the ajmaline test, the patient had a VT episode with a left bundle and an inferior axis ECG morphology. However, no sinus tachycardia elevation appeared at the right precordial leads during the 15 minutes of the test. Thereafter, at the 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 direct current. Then, he was transferred to the electrophysiology room for catheter ablation to eliminate this electrical storm. By the careful movement of the ablation catheter in the left and right ventricles, the RVOT area just below the pulmonary valve was found to be the earliest activated site during the VT. This site was suspected to be the origin of this incessant VT, and at this site, multiple radiofrequency energies were given.

From: Department of Cardiology, Research and Training Hospital, İstanbul, Turkey.

Correspondence: Dr NS Yelgeç, Department of Cardiology, Research and Training Hospital, Tıbbiye Cad. No: 13 Kadiköy/Istanbul 34668, Turkey. Email: yelgec@gmail.com

 DOI: 10.7727/wimj.2015.281

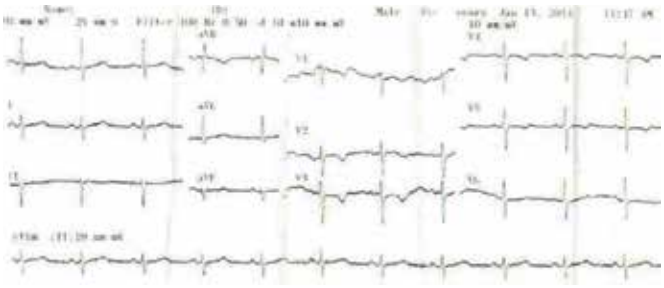

Fig. 1: Baseline ECG of the patient.

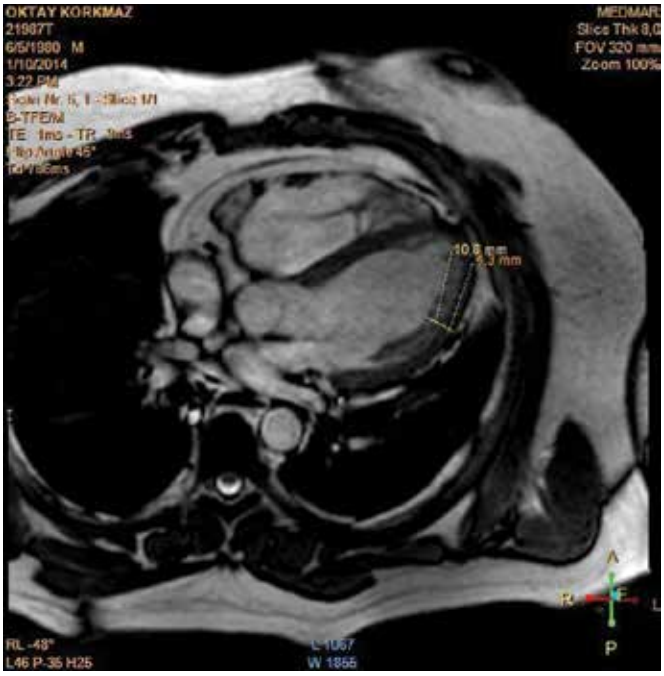

Fig. 2: Long axis view MR image of the patient shows non-compaction of the left ventricle, and the ratio of non-compacted to compacted myocardium is greater than 2.3.

During the ablation procedure, we observed two different left bundle VT morphologies which suggested the possibility of ARVC. At that time, the tachycardia was successfully ablated and the tachycardia was terminated (Figs. 4 and 5).

The patient was sent back to the intensive care unit. A few hours later, the same VT reappeared and a coronary angiogram was performed to rule out an ischaemic aetiology which revealed normal coronaries. The following morning, an ablation of VT was reattempted, but, this time, using a non-fluoroscopic catheter-based electroanatomic mapping system. After a long session of ablation, finally the arrhythmia was terminated completely. This patient met one major and three minor criteria of the current Task Force criteria of ARVC (T-wave inversion in V1-6 derivations as major criteria, VT of RV outflow configuration, and more than 500 ventricular extra

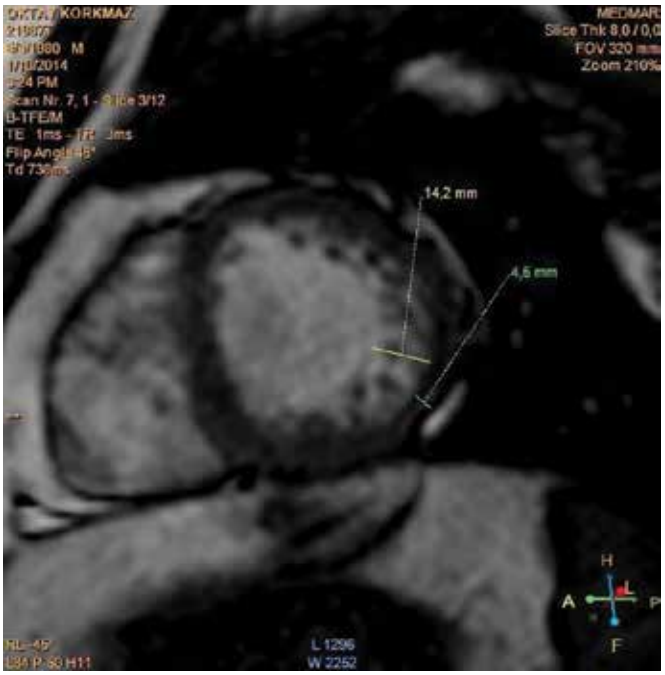

Fig. 3: Short axis view shows non-compacted to compacted ratio is greater than 2.

systoles per 24 hours by Holter and ARVC confirmed by current task criteria in a second-degree relative, as minor criteria).

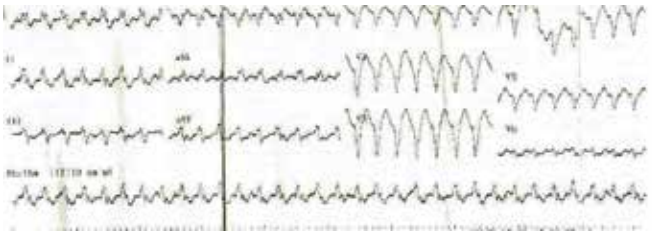

Fig. 4: Incessant ventricular tachycardia of RV origin.

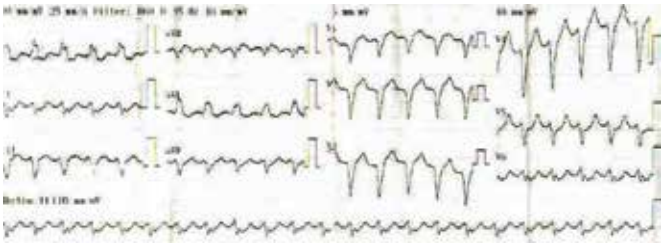

Fig. 5: Different VT morphologies of RV origin of the same patient.

DISCUSSION

Arrhythmogenic right ventricular cardiomyopathy is an under-recognized but an important cause of sudden cardiac death accounting approximately 11% of the cases overall. Many patients, however, remain clinically silent and asymptomatic for decades. A total of 30% of

the cases are familial. The patients usually present with arrhythmias of RV origin and/or sudden death between the ages of 10 and 50 years with a mean age of diagnosis of approximately 30 years. The most common ventricular arrhythmia is sustained or non-sustained monomorphic VT that originates in the RV, and therefore has a left bundle branch block pattern. Ventricle tachycardia may originate from any part of the RV. However, if it originates from the RVOT, it may be difficult to distinguish ARVC from the idiopathic RVOT tachycardia. Different left bundle VT morphologies suggest 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 RV outflow dilatation. On the other hand, the RVOT tachycardia arises typically from a very narrow area just inferior to the valve in the anterior aspect of the RVOT and occurs exclusively in the young to the 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). Ventricular tachycardias of ARVC and isolated left ventricular non-compaction are the candidates for implantable cardioverter defibrillator implantation. The 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). However, in the case we presented here, the co-existing left ventricular non-compaction is a coincidental finding which has no active role in this patient's arrhythmias. All arrhythmias originated from the RV, since in the early stages of ARVC, imaging findings might be negative. On the other hand, the non-compaction demonstrated by the MRI might be silent.

CONCLUSION

This case demonstrates the importance of diagnostic evaluation by various invasive and non-invasive techniques and detailed analyses of clinical history and presentation to establish the presence and type of heart disease and the origin and pathology causing lethal arrhythmias. The reliance on only imaging findings may result in the misdiagnosis of the origin of arrhythmias.

REFERENCES

1. Gemayel C, Pelliccia A, Thompson PD. Arrhythmogenic right ventricular cardiomyopathy. J Am Coll Cardiol 2001; **38:** 1773.
2. Ritter M, Oechslin E, Sütsch G, Attenhofer C, Schneider J, Jenni R. Isolated noncompaction of the myocardium in adults. Mayo Clin Proc 1997; **72:** 26.
3. Sen-Chowdhry S, Lowe MD, Sporton SC, McKenna WJ. Arrhythmogenic right ventricular cardiomyopathy: clinical presentation, diagnosis, and management. Am J Med 2004; **117:** 685.
4. Nava A, Bauce B, Basso C, Muriago M, Rampazzo A, Villanova C et al. Clinical profile and long-term follow-up of 37 families with arrhythmogenic right ventricular cardiomyopathy. J Am Coll Cardiol 2000; **36:** 2226.
5. Brooks R, Burgess JH. Idiopathic ventricular tachycardia: a review. Medicine (Baltimore) 1988; **67:** 271.
6. Aragona P, Badano LP, Pacileo G, Pino GP, Sinagra G, Zachara E. Isolated left ventricular non-compaction. Ital Heart J Suppl 2005; **6:** 649–59.