Case Report

Electroanatomical Mapping in the Differentiation Between Arrhythmogenic Right Ventricular Cardiomyopathy and Cardiac Sarcoidosis in a Patient With Ventricular Tachycardia Storm

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ABSTRACT

A 36-year-old man with frequent episodes of ventricular tachycardia (VT) was referred to our hospital for ablation due to the suspicion of sarcoidosis and arrhythmogenic right ventricular cardiomyopathy in cardiac magnetic resonance imaging. After implantable cardioverter-defibrillator implantation, the patient presented with VT storm, without any endocardial scar and with an extensive epicardial scar in voltage mapping. Consequently, he underwent epicardial VT ablation. (Iranian Heart Journal 2021; 22(1): 112-116)

KEYWORDS: ARVC, Epicardial ablation, VT storm, ICD, Sarcoidosis

The patient was a 36-year-old man who was referred to us with a diagnosis of right ventricular outflow tract ventricular tachycardia (RVOT–VT) and hemodynamic compromise. The arrhythmia had been terminated with D/C shock. Echocardiography revealed moderate RV dysfunction and mild left ventricular (LV) dysfunction. Accordingly, cardiac magnetic resonance imaging (CMR) was recommended. CMR revealed patchy areas of late gadolinium enhancement (LGE), suggestive of fibrosis in the inflow and outflow tracts of RV and another site of LGE in the mid-posterolateral wall of LV with LV ejection fraction of about 45% and moderate RV dysfunction. The patient was infertile. The first diagnosis in CMR was sarcoidosis, and arrhythmogenic right ventricular cardiomyopathy (ARVC) was considered to be a second diagnosis (Fig. 1 & 2).
An implantable cardioverter-defibrillator (ICD) was implanted for the patient. He refused ablation and was, consequently, referred for a rheumatology consult. Three weeks later, the patient presented with VT storm: 17 episodes of VT, which were subsequently terminated with ICD shock. He was scheduled for ablation and an endocardial biopsy before ablation. Attempts were made to perform the endocardial biopsy under the guidance of 3D voltage mapping. Interestingly, however, no low-voltage area was detected in bipolar voltage mapping, although unipolar mapping revealed a large scar burden. Thus, ARVC with RV epicardial involvement was considered for the patient. An endocardial biopsy was not performed because of the absence of any site of endocardial scarring. Next, for VT ablation, the epicardial approach was adopted. Subxiphoid puncture was done, and 3D electroanatomical mapping of the epicardium revealed multiple sites of patchy scars in RVOT and RV inflow areas. Because of hemodynamic deterioration with VT (Fig. 3 & 4), substrate modifications were carried out for the patient (Fig. 5a & 5b).

Figure 3: VT1, induced by ventricular extra-stimulation, superior axis with negative concordance, and positive I, aVL are illustrated herein.

Figure 4: Figure illustrates wide QRS tachycardia with left bundle branch block morphology, V2 breakthrough, and superior axis, which is compatible with epicardial right ventricular inflow ventricular tachycardia.
Differentiation Between ARVC and Sarcoidosis

Before ablation, the patient underwent coronary angiography to determine the position of the coronary arteries (Fig. 6); the results showed slow flow coronary arteries without stenosis.

All the entrance conducting channels and late potentials in RVOT and RV inflow, as well as all the abnormal epicardial potentials, were ablated (Fig 7 & 8).

**Figure 5a:** Local abnormal ventricular activity is shown in right ventricular epicardium.

**Figure 5b:** Disappearance of late potentials after ablation is illustrated herein.

**Figure 6:** Figure illustrates coronary angiography before epicardial radiofrequency ablation. The epicardial ablation catheter is present.

**Figure 7:** Extensive ablation of right ventricular epicardium by targeting the entrance conducting channels via the local activation time of late potentials is shown herein.
Differentiation B

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Figure 8: Extensive ablation of right ventricular epicardium by targeting the entrance conducting channels via the local activation time of late potentials is illustrated herein.

No arrhythmia was inducible after ablation with 3 ventricular extrasystoles and Isuprel infusion (Fig. 9); hence, ablation was terminated and no arrhythmia was present at 1 month’s and 3 months’ follow-ups later.

Figure 9: No inducible arrhythmia is seen after ablation with ventricular extrasystoles.

DISCUSSION

Scar distribution patterns in CMR may be helpful insofar as they indicate disease-specific patterns of fibrosis in LGE images. Cardiac sarcoidosis is a diagnostic challenge and the workup for it requires biopsy.\cite{1,2,3}

A scar pattern of basal septal RV involvement may be in favor of sarcoidosis, but it does not prove it. In ARVC, the disease process starts in epicardial RV, where fibro-fatty tissue replaces myocardial tissue. In about 50% of patients with ARVC undergoing CMR and voltage mapping, endocardial voltage mapping fails to detect areas of scarring, especially in the inferobasal part of RV.\cite{4,6}

The involvement of LV is present in up to three-quarters of patients with ARVC.\cite{7,9}

An epicardial approach is often necessary to eliminate VT in patients with ARCV. The presence of epicardial scarring can be assessed via unipolar mapping. Low-amplitude electrograms in bipolar mapping are defined in the endocardium by less than 1.5 mv voltage and in the epicardium by less than 1 mv voltage. Endocardial unipolar signals suggesting mid-myocardial or epicardial substrates are defined as less than 5.5 mv in RV and 8.3 mv in LV.\cite{10,11}

In this patient, we performed scar dechanneling with the ablation of the conducting channel entrance sites within the scar, characterized by the earliest late potentials following global ventricular activation. The endpoint of the ablation was the elimination of all the conducting channels into the scar. Following the targeted ablation at the entrance site, the VT was rendered non-inducible via 3 ventricular extra-stimuli with and without Isuprel infusion.

In this patient, ablation was successfully terminated, and intrapericardial triamcinolone was administered before long sheath withdrawn. The procedure was done without any complication.\cite{12,13}

In cases with sarcoidosis, in addition to ablation, immunosuppressive therapy is recommended. Nonetheless, in our patient, electroanatomical voltage mapping showed no endocardial scar, and the extensive epicardial scar pattern was in favor of ARVC.

Electroanatomical mapping could be considered an adjuvant technique for the
Differentiation between cardiac sarcoidosis and ARVC.

CONCLUSIONS

Electroanatomical mapping could be helpful in challenging cases of ARVC and cardiac sarcoidosis.

REFERENCES


