COMPLEX CASE STUDY

Ventricular Tachycardia Substrate Voltage Map and Radiofrequency Ablation in a Patient with Fabry’s Disease Cardiomyopathy

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ABSTRACT. Ventricular tachycardia (VT) substrate voltage mapping and radiofrequency ablation in non-ischemic cardiomyopathy patients with Fabry’s disease is not well described. We present a 49-year-old male patient with multiple implantable cardioverter-defibrillator (ICD) shocks due to sustained VT (cycle length 461 ms, right bundle branch block (RBBB) morphology, superior axis), refractory to antiarrhythmic drug therapy with amiodarone. Electroanatomic three-dimensional (3D) voltage mapping and ablation of the VT site of origin is presented to characterize the endocardial involvement of this glycosphingolipid deposition disorder. Comparison is made with known pathologic findings in autopsy series, showing a consistent posterolateral endocardial involvement near the basal mitral valve annulus.

KEYWORDS. Fabry’s disease, non-ischemic cardiomyopathy, ventricular tachycardia, radiofrequency ablation, voltage substrate mapping.

Introduction

Fabry’s disease is a disorder of glycosphingolipid metabolism caused by X-linked deficiency in the lysosomal enzyme alpha-galactosidase A. Cardiac involvement is common, occurring in up to 60% of patients, often resulting in atrial fibrillation, heart block, and ventricular tachycardia (VT) associated with sudden cardiac death.1 The pattern of cardiac infiltration and subsequent scar formation with creation of potential VT circuits has been characterized in autopsy studies. Concentric left ventricular hypertrophy with severe left ventricular systolic and diastolic dysfunction, as well as thinning of the left ventricular posterior wall, are observed with glycosphingolipid accumulation within myocytes.2 Although VT is frequently observed in patients with Fabry’s cardiomyopathy, endocardial voltage substrate mapping is poorly characterized. We present a case of sustained monomorphic VT with voltage mapping and successful radiofrequency ablation showing excellent correlation to known histologic cardiac deposition patterns in this rare disease.

Case

A 49-year-old man with Fabry’s disease for 18 years treated with fabryzyme, complicated by cardiac involvement, presented with sustained VT and multiple implantable cardioverter-defibrillator (ICD) shocks despite amiodarone and carvedilol therapy. His cardiomyopathy is characterized by atrial fibrillation initially refractory to cardioversion, complete heart block, and left ventricular systolic dysfunction with New York Heart Association (NYHA) Class III congestive heart failure (left ventricular ejection fraction by two-dimensional (2D) echocardiography 25–30% with moderate concentric hypertrophy, stage II diastolic dysfunction, and posterolateral akinesis). A Medtronic (Minneapolis, MN) biventricular ICD model 7304 was implanted in 2008. Device interrogation revealed multiple treated sustained VT episodes over 2 months at 460-ms cycle length with acceleration by antitachycardia pacing therapy, followed by ICD shock rescue at 25 joules. The patient presented in sustained VT with right bundle branch block (RBBB) pattern QRS, superior axis, and R-wave transition at V4, with fusion complexes and atrioventricular dissociation (Figure 1).
The patient was taken to the electrophysiology laboratory in sinus rhythm with complete atrioventricular block and biventricular paced rhythm. A His bundle and right ventricular apical quadripolar catheter were placed and used to exclude bundle branch re-entry VT (BBR-VT). Arterial access was obtained. A Thermocool (Biosense Webster, Diamond Bar, CA) F/J 3.5 mm saline irrigated map and ablation catheter was inserted into the left ventricle through a 45-cm Arrow stainless steel sheath (Teleflex Medical, Limerick, PA) via retrograde aortic approach. Using CARTO-III (Biosense Webster), a three-dimensional electroanatomic voltage map was created of the left ventricular endocardium. Voltage <0.5 mV was labeled as low voltage scar, and >1.5 mV was classified as normal. Gray point tags indicate sites of catheter contact, but local electrogram (EGM) <0.25 mV, with no pacing capture, consistent with dense scar (Figure 2). T with cycle length 460 ms was elicited by programmed stimulation. Pace mapping confirmed a posterolateral site of origin near the mitral annulus with a 12/12 pace map by surface electrocardiogram (ECG). Radiofrequency ablation at 40–45 W for 90 s with
30 ml/min saline flow was performed at the VT site of origin and at regions of low voltage isthmus channels felt to represent potential VT circuits. Ablation lesions are noted in red. Pacing sites with 12/12 surface ECG pace map match are marked with a yellow tag. The white point tag indicates a protected isthmus site with termination of VT during radiofrequency delivery (Figure 3). Post ablation, there was no inducible VT with triple extra-stimulus pacing at 350 ms/400 ms and 600 ms drive trains. At the site of successful VT termination, isolated mid-diastolic potentials were noted with local EGM to QRS onset of 56 ms (Figure 4). Three months later, the patient remains clinically stable without VT recurrence.

**Discussion**

We report a case of recurrent monomorphic VT in a patient with Fabry’s disease and cardiac involvement manifest by atrial fibrillation, heart block, and congestive heart failure with concentric left ventricular hypertrophy. Initial descriptions of cardiac infiltration from glycosphingolipid...
accumulation in myocytes demonstrate concentric hypertrophy, and restrictive physiology mimicking hypertrophic cardiomyopathy.\textsuperscript{3,5} Recurrent monomorphic VT and sudden cardiac arrest from ventricular fibrillation have been reported and suggest that significant arrhythmic death risk is present when cardiac deposition occurs.\textsuperscript{6} Autopsy studies show a consistent pattern of posterolateral left ventricular thinning and scar formation towards the base of the left ventricle, often adjacent to the basal mitral valve region (Figure 5).\textsuperscript{2} In our case, three-dimensional (3D) electroanatomic voltage mapping identified a characteristic posterobasal site scarring in the left ventricular endocardium, with several low voltage isthmus channels and fractionated late systolic potentials during sinus rhythm. Pace mapping and successful radiofrequency ablation of the VT circuit occurred at a location with isolated mid-diastolic potentials during VT near the mitral valve annulus, consistent with a protected critical isthmus for VT propagation. This pattern of involvement may account for slow monomorphic VT circuits arising along channels parallel to the mitral annulus, as is frequently seen at electrophysiology study or with cardiac magnetic resonance imaging in other forms of non-ischemic cardiomyopathy, such as cardiac sarcoidosis\textsuperscript{7,8}.

References