Non-arrhythmic pre-excitation-induced cardiomyopathy

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1. Introduction

Ventricular pre-excitation is a prevalent finding affecting 0.2% of the population [1]. The first report of an accessory pathway connecting the atria to the ventricles was described as a case report by Wilson in 1915 only 13 years after Einthoven recorded the human heart electrical activity with a string galvanometer for the first time [2].

Wolf Parkinson White syndrome (WPW) has been implicated as a cause for syncope, sudden cardiac death, palpitations, and tachycardia-induced cardiomyopathy (TIC). There have been reports of non-arrhythmic dilated cardiomyopathy associated with WPW in the pediatric population [1–3]. We describe a case of accessory pathway-induced dilated cardiomyopathy in an adult with full recovery of left ventricular (LV) function post-ablation.

2. Case

A 44-year-old male was referred to the electrophysiology (EP) service for assessment of WPW. He had a history of syncopal event three years prior, along with infrequent palpitations. He additionally had a history of idiopathic cardiomyopathy, which was incidentally found on an echocardiogram as part of his workup for syncope. He was otherwise asymptomatic, New York Heart Association class I. He was appropriately treated with ACE-inhibitors and beta-blockers. There were no documented arrhythmias. A 48 hour holter monitor showed pre-excited QRS throughout monitored period with no evidence of sustained arrhythmias or any premature ventricular contractions.

His electrocardiogram showed sinus rhythm with evidence of a right free wall accessory pathway (Fig. 1a). An echocardiogram revealed an ejection fraction of 40% calculated using Simpson’s method with inferior and inferolateral wall hypokinesis and left ventricular end diastolic dimension of 5 cm (Fig. 2). Coronary angiography was normal, as well as cardiac biomarkers. He had been on perindopril and carvedilol with no improvement in his EF. Considering his previous syncope and infrequent symptoms of palpitations, he underwent an EP study.

The patient presented to the EP laboratory approximately 9 months after his initial diagnosis of idiopathic cardiomyopathy. Immediate pre-ablation echocardiography confirmed depressed LV function. A right free wall accessory pathway was identified in the EP lab with site of successful ablation at 10-11 o’clock on the tricuspid annulus (Fig. 3). There was no inducible arrhythmia in the lab, antegrade and retrograde effective refractory period of the AP were 400/320 msec and 600/280msec respectively. No complications were noted.

Post-ablation ECG showed resolution of delta waves (Fig. 1b). At one month follow-up, his EF had improved to near normal at 52% measured using Simpson’s method. His LV end diastolic dimension reduced to 4.5 cm and no regional wall motion abnormality could be identified (Fig. 4).

3. Discussion

Ventricular pre-excitation occurs when the ventricular
Fig. 1. a. Sinus rhythm with short PR interval and fully pre-excited QRS with negative delta in V1, positive delta in leads II, aVF, isoelectric delta in lead III suggestive of right anterolateral free wall accessory pathway site of origin. b. Loss of delta wave post ablation with narrow QRS and normal PR interval.
myocardium is partially or completely depolarized through an extranodal atrioventricular connection. The presence of accessory pathway in the WPW syndrome has been associated with an increased risk of sudden cardiac death and arrhythmias [4]. This patient had no significant arrhythmias but had developed cardiomyopathy leading to multiple investigations. He was labeled as idiopathic cardiomyopathy and despite angiotensin converting enzyme inhibitor and beta-blocker therapy his ejection fraction remained depressed until his accessory pathway was ablated.

LV dysfunction in patients with WPW may be secondary to tachyarrhythmias, either symptomatic or asymptomatic, that trigger a TIC. The rate and duration of tachyarrhythmias influence the speed at which TIC develops and LV dysfunction may occur as early as 24 hours after the onset of tachyarrhythmia [5]. Another mechanism maybe similar to that seen with premature ventricular contraction (PVC) induced cardiomyopathy. This entity has been more recognized in recent years and an association between higher PVC burden and cardiomyopathy has been established. Baman et al. in a cohort study found that 24% PVC burden, in otherwise healthy patients, had a sensitivity of 79% and specificity of 78% for diagnosis of PVC cardiomyopathy. Proposed mechanisms for ventricular dysfunction in these cases include ventricular dyssynchrony, increased oxygen consumption, alteration in intracellular calcium and membrane ionic currents, autonomic and hemodynamic impairment [2,5]. Cardiomyopathy was reversible with successful ablation and suppression of PVCs.

With manifest WPW, ventricular activation is dyssynchronous similar to that seen with PVCs. In patients with a manifest accessory

Fig. 2. Pre-ablation ejection fraction measured by Simpsons method and LV dimensions in systole and diastole. A4Cd (apical four chamber in diastole), A4Cs(apical four chamber in systole), A2Cd (apical two chamber in diastole), A2Cs(apical two chamber in systole), PSLd (parasternal long in diastole), PSLs (parasternal long in systole).

Fig. 3. Simultaneous intra-cardiac tracing and fluoroscopic image in left anterior oblique showing ablation catheter at site of successful ablation around the tricuspid annulus at 10-11 o clock (yellow arrow), RV-His catheter positioned with His signal (H) on His mid showing negative HV in first two beats and distal poles at right ventricular apex (dashed arrow), Deflectable decapolar coronary sinus catheter in the coronary sinus (solid black arrow), Surface lead I, II and V2 showing resolution of delta wave and AV signal fusion during ablation (Red arrow). HV 5 msec post ablation.
pathway, ventricular depolarization begins at the site of ventricular insertion of the accessory pathway. While eccentric activation proceeds through the ventricular tissue, activation is also occurring through the atrioventricular node and His Purkinje system. The degree of eccentric activation of the ventricle depends on the relative contribution of ventricular activation via the accessory pathway compared to the AV node. Patients with wider QRS complexes will have a greater degree of eccentric ventricular activation. Our patient’s baseline QRS complex was similar in morphology to a left bundle branch block pattern, with a QRS duration of 186 ms. In our case due to proximity of right sided accessory pathway to the sinus node and AV nodal conduction properties, a higher degree of pre-excitation was present, leading to a wider QRS. It is likely that this degree of pre-excitation led to significant dyssynchronous ventricular activation, similar to that seen in patients with very frequent PVCs.

In the pediatric literature, there is emerging evidence demonstrating an association between the presence of a manifest accessory pathway and dilated cardiomyopathy [1,2,6,7]. The proposed mechanism of dysfunction is that of dyskinesia caused by non-physiologic activation of the ventricular myocardium. Only accessory pathways in the septal, paraseptal and posterior locations have been associated with this phenomenon [1,3,6]. A causal relationship is assumed due to reversal of cardiomyopathy with ablation [1—4,7].

In this case, abnormal ventricular activation secondary to pre-excitation leading to dyssynchrony, contributed to the development of a cardiomyopathy. This process was reversed by ablation of the accessory pathway, which resulted in normalization of LV function and reversal of dilated cardiomyopathy. This case highlights the importance of manifest pre-excitation as an under-recognized cause of idiopathic dilated cardiomyopathy. Although rare, it is an important etiology to consider as it may be reversible with ablation. Larger case series are required to further establish this association.

Disclosures
None.

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