Electro-anatomical mapping in a patient with isolated left ventricular non-compaction and left ventricular tachycardia

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Received 19 February 2009; accepted after revision 26 May 2009

We report the case of a 59-year-old non-Caucasian man with sustained left ventricular (LV) tachycardia and isolated LV non-compaction. An electro-anatomical mapping of the right ventricle and LV with the Carto system was reconstructed. The voltage map excluded the presence of scarred tissue as a possible substrate responsible of the ventricular arrhythmia.

Introduction

Isolated left ventricular non-compaction (IVNC) is a rare congenital cardiomyopathy that has been recently classified as a primary cardiomyopathy of uncertain aetiology. Clinical presentations of IVNC include heart failure (HF), systemic embolism, and malignant ventricular arrhythmias. The mechanisms causing life-threatening arrhythmias are still unknown.

We report the results of an electro-anatomical map in a patient with IVNC and left ventricular tachycardia (LVT) aiming to investigate the presence of scarred tissue or any voltage abnormalities.

Case presentation

A 59-year-old non-Caucasian man was admitted to our Hospital because of sudden onset palpitations associated with intensive weakness and vertigo. The ECG showed a wide QRS tachycardia with left superior axis, right bundle branch block morphology, and the presence of V–A dissociation in several ECG leads (Figure 1) that suggested an LVT. The patient was electrically

Figure 1 Surface ECG showing wide QRS tachycardia with left superior axis deviation, right bundle branch block, and VA dissociation likely arising from infero-septo-apical area of LV.
cardioverted after verapamil infusion failed in terminating the arrhythmia. The patient also referred one previous episode of syncope without prodromes.

Trans-thoracic echocardiography showed a dilated LV with a severe systolic dysfunction (EF < 25%) and the presence of LV apex hypertrabeculation together with a corresponding thinnest area. A cardiac magnetic resonance imaging with gadolinium enhancement confirmed the diagnosis of IVNC as the diastolic ratio of non-compacted/compacted layer was >2 (Figure 2A and B).
Using the Carto system, an electro-anatomical map of the right ventricle (RV) and LV was created. The electrophysiological study showed a normal sinus node and AV node function. Not sustained supraventricular arrhythmias were inducible both at the baseline and during isoproterenol infusion. Programmed ventricular stimulation with up to three extrastimuli following three different pacing trains (drive 600, 500, 430 ms) was performed from RV apex, RV outflow tract, and LV apex without inducing any sustained VT. During isoproterenol infusion, we repeated the same protocols without any VT induction.

The voltage map did not show any abnormalities consistent with scarred tissue (Figure 2C–E). There was no recorded splitting or fractionated electrograms. Even when the mapping catheter was advanced into the apex of LV, a normal signal was recorded (Figure 2F). The pace-mapping from various sites of LV apex did not show any concordance with the clinical tachycardia in the precordial leads. Since we hypothesized an LVT arising from the infero-septo-apical area, we carefully mapped this part of the LV without identifying any distinct ‘Purkinje’ potentials. The attempts of pace-mapping from this site did not result in a close match to the surface ECG of clinical VT. We also paced from RV apex at different cycle lengths in order to unmask an endocardial conduction abnormality on the mapping catheter placed in the LV apex.

Considering the previous episode of syncope and documented VT, the patient was implanted with a dual-chamber implantable cardioverter-defibrillator.

Discussion
Isolated LV non-compaction is a distinct cardiomyopathy1 whose prognosis depends on the progression of HF, the occurrence of thrombo-embolic events, and of life-threatening ventricular arrhythmias.2 The mechanism responsible for malignant ventricular arrhythmias in these patients is still unknown. It has been hypothesized that scarred tissue or the presence of micro-fibrotic areas might be responsible substrates for ventricular arrhythmias.

Studies using positron emission tomography revealed a consistent decrease of coronary flow reserve in non-compacted and compacted layers. Accordingly, an unbalanced coronary micro-circulation distribution between these two areas of the LV has been demonstrated.3

Suspecting a clinical correlation between the LVT and IVNC, we investigated in our patient the presence of a possible substrate causing the arrhythmia. To the best of our knowledge, no previous descriptions of an electro-anatomical map exist in patients with IVNC. We found that the voltage map was completely normal as any voltage points lower than 1.5 mV were recorded. No abnormal electrograms were recorded both in the apex of LV and elsewhere.

In our case, the voltage map does not suggest that VT was due to the presence of scarred tissue or to other predisposing substrates. However, we cannot exclude intramural or epicardial scarred tissue that has been already reported in patients with idiopathic dilated cardiomyopathy and IVNC.4 The poor correlation with the surface ECG of VT when the pace-mapping was performed from LV apex allows to rule out the diseased area as the origin of the arrhythmia. Because of the absence of high-frequency potentials preceding the earliest local ventricular electrogram, a VT originating from the base of posterior papillary muscle (PPM) should be suspected. This idiopathic LVT is due to a focal mechanism. Although the ECG morphology of clinical arrhythmia was suggestive, it is unlikely that the VT arose from the PPM. This clinical syndrome has been mostly described in patients with normal LV function. Furthermore, PPM VTs are exercise-induced and they are easily reproducible with isoproterenol infusion.5

Although VT was not sensitive to verapamil infusion, according to the surface ECG, we cannot exclude an idiopathic LVT (‘Belhassen type’) caused by an inter-fascicular re-entry mechanism.6 These VTs are often associated with structural heart disease. We carefully mapped the infero-septal-apical part of the LV, but we were not able to find any distinct ‘Purkinje’ potentials. Furthermore, during LV mapping, mechanical block due to catheter bumping probably may have prevented VT induction.

The limitation of our case is the coincidental presence of an LVT that is not caused by a predisposing anatomical substrate according to the results of the voltage map. The mechanism of life-threatening arrhythmias in patients with IVNC remains presently unknown and it may involve other mechanisms, such as altered ion channel activity and enhanced intercellular communication. A better mechanism’s understanding of arrhythmias could derive from studies on a larger population of patients with IVNC and inducible VT.

References