SHORT SERIES

Electroanatomic mapping and ablation of ventricular tachycardia associated with systemic sclerosis

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Abstract Two cases of systemic sclerosis with sustained ventricular tachycardia (VT) are presented. The first patient received hydroxychloroquine for skeletal muscle disease coexisting with cardiac involvement. In both cases, 3D-electroanatomic mapping showed low-voltage areas in the right ventricle. In the first patient the tachycardia was mapped and a protected isthmus suggesting reentry was delineated and ablated. Other substrate locations were indirectly identified by pacemapping on the right and left ventricular endocardium in the second patient. VT did not reoccur during follow-up. Radiofrequency catheter ablation is safe and effective and electroanatomic mapping may be helpful in patients with systemic sclerosis. © 2004 The European Society of Cardiology. Published by Elsevier Ltd. All rights reserved.

KEYWORDS electroanatomic mapping; hydroxychloroquine; scleroderma overlap syndrome; systemic sclerosis; ventricular tachycardia

Introduction

Systemic sclerosis (SS) is a connective tissue disease of autoimmune nature that affects the skin, the gastrointestinal, respiratory and cardiovascular systems. Skin and organ changes in SS are the results of widespread damage to small blood vessels and fibrosis. Cardiac involvement has been shown to be frequent at post-mortem examination [1] and conduction disturbances as well as supraventricular tachycardias or ventricular extrasystoles commonly occur during Holter monitoring [2]. Nonetheless, reports of sustained ventricular tachycardia (VT) remain scarce [3–7]. Pharmacological therapy by beta blockers, sotalol or amiodarone is usually limited by severe Raynaud’s phenomenon and pulmonary interstitial fibrosis. There is a single report about the use of an implantable defibrillator (ICD) in the available literature [8]. Against its use are the risk of difficult wound healing and the care to preserve venous accessibility in case of future haemodialysis. Therefore,
direct ablative techniques may represent an alternative option for these patients [6,7]. We report two observations of VT ablation with the use of a 3D-electroanatomic mapping system in SS.

Case report 1

A 35-year-old man presented with a 5-year history of SS. The manifestations of the disease included sclerodactyly, Raynaud’s phenomenon and pulmonary interstitial fibrosis. At the age of 34, the patient complained of palpitations related to ventricular ectopy, the initial echocardiogram revealed mild enlargement of the right ventricle (RV) with a 30-mm end-diastolic diameter, and left ventricular (LV) function was normal. Holter monitoring showed >10,000 ventricular premature beats per day with some episodes of non-sustained VT from 3 to 5 consecutive beats. At this time, elevated plasma level of creatine kinase was found up to 2800 IU/L with electromyogram results consistent with myositis. This skeletal myopathy led to the diagnosis of overlap syndrome [9]. The preceding drug therapy which consisted of nifedipine and penicillamine was replaced by losartan and aspirin. Hydroxychloroquine was commenced at a dose of 400 mg daily. Three months later, the patient was admitted for VT which was well tolerated despite a rate of 205 bpm. Electrolytes and troponin levels were normal, sinus rhythm with a normal QT interval (QTc: 451 ms) was restored by an external DC shock under general anaesthesia (Fig. 1A). Short runs of non-sustained VT identical to the sustained pattern persisted after cardioversion. Echocardiography showed a deterioration of RV function with a 43 mm end-diastolic diameter, the interventricular septum was hypokinetic and the LV ejection fraction was reduced at 45%. Radioisotope RV ejection fraction was 25%. Due to strong limitations in the possible use of antiarrhythmic drugs, ablation was considered. At baseline electrophysiological testing, the clinical VT with normal axis and left bundle QRS configuration with cycle lengths between 290 and 310 ms was reproducibly induced by two extrastimuli. Following a previously described methodology [10], endocardial 3D-mapping was performed with a 7F catheter tipped by a 4-mm electrode (Navistar™, Biosense Webster, Figure 1

Figure 1  A 12-lead electrocardiogram before ablation in the two cases reported. (A) First patient. (B) Second patient.
Johnson & Johnson). RV mapping was performed during both sinus rhythm and VT (Fig. 2). The reconstruction involved the septal aspect of the RV, covering the endocardium from the tricuspid annulus toward the apex and the pulmonary annulus. Low-voltage electrograms were found in the RV outflow tract extending caudally to the level of the His bundle. An electroanatomic activation map during VT defined a small isthmus with the exit site in the immediate vicinity of delayed potentials, this area was 20 mm ahead of the His bundle and had no anatomical boundaries. The earliest activation was 30 ms prior to QRS onset and the total activation time was 170 ms (i.e. 57% of the 300 ms cycle length). Entrainment manoeuvres in the isthmus or outside were not attempted. The activation pattern was consistent with a small reentrant circuit. Radiofrequency energy (RF) was applied for 60 s in a temperature-controlled mode at 60 °C with a power limited to 50 W. Four RF pulses were delivered, VT stopped (Fig. 3) and remained non-inducible from multiple RV sites. The patient was discharged without antiarrhythmic drugs, hydroxychloroquine was withdrawn. Follow-up Holter monitoring showed persisting ventricular ectopy but disappearance of repetitive premature beats. Later he was treated by glucocorticoid and immunoglobulins, he has remained free of VT recurrence for the subsequent 25 months.

Case report 2

A 60-year-old woman with SS was admitted for VT. The disease was diagnosed at the age of 45, with terminal renal failure treated by haemodialysis since the age of 56. She suffered from pulmonary interstitial fibrosis, severe Raynaud’s phenomenon, and hypertension. Haemodialysis was performed through the right subclavian vein via a permanent catheter. Current therapy comprised aspirin and glucocorticoids. A poorly tolerated VT at 200 bpm with hypotension occurred during haemodialysis with right bundle branch block pattern and extreme right QRS axis deviation. Sinus rhythm was restored by intravenous lignocaine. Body surface ECG showed a normal QRS complex with first degree AV block and a normal QT interval (Fig. 1B). Echocardiography showed concentric LV hypertrophy with preserved ejection fraction, the RV
function seemed to be normal. Since venous access was extremely limited in this patient precluding an ICD implantation, RF ablation was considered. Only short runs (<5 beats) of the clinical VT were induced by a combination of two extrastimuli and isoprenaline (Fig. 4A). Mapping was therefore conducted in sinus rhythm in the LV with the use of a 25-electrode mapping catheter able to record near-field electrograms up to 8 mm from the endocardial surface (Qwikstar™, Biosense Webster, Johnson & Johnson). An anatomical reconstruction was built using points acquired during stable and moving catheter locations. Voltage and activation in sinus rhythm were normal in the LV free wall (Fig. 5), pacemapping identified an area with a satisfactory 12-lead QRS match with the VT configuration (Fig. 4B). Our interpretation was in favour of a focal non-macro-reentrant mechanism and RF energy was applied at this site in a temperature-controlled mode at 60 °C with power limited to 50 W. After delivery of 6 RF pulses nonsustained VT was no longer observed with extrastimulation plus isoprenaline but a different VT with a left bundle branch block pattern and left superior axis QRS deviation was induced. It was sustained, poorly tolerated with a cycle length of 255 ms but terminated spontaneously (Fig. 4C). Its inducibility was not reproducible. RV was mapped in sinus

Figure 3  (A) A 12-lead surface ECG of the index ventricular tachycardia mapped in Fig. 2. (B) Endocardial bipolar (ABL 1-2) and unipolar (ABL WCT-1) recordings at the exit site preceding the QRS onset by 30 ms with a “QS” pattern in the unipolar mode, tachycardia cycle length is 300 ms. (C) Tachycardia termination within 10 beats after onset of radiofrequency energy application (+).
rhythm (Fig. 5), a low-voltage area was found in the outflow tract. Another area with normal voltage was demonstrated in the RV free wall very close to the tricuspid annulus where pacemapping was satisfactory in mimicking the second VT configuration (Fig. 4D). Again, a non-macro-reentrant mechanism was hypothesized and 7 RF pulses were delivered at this site in a temperature-controlled mode at 60°C with power limited to 50 W. Non-inducibility was confirmed but was not informative because induction at baseline was not reproducible. The patient was discharged without antiarrhythmic drugs, glucocorticoids were maintained, and Holter monitoring was normal. She has remained free of VT recurrence for the following 14 months.

Discussion

Both observations underline that right-sided cardiac involvement may be the site for possible VT substrate location in patients with SS [1,6]. SS may lead to VT by numerous processes for example RV dilation following pulmonary hypertension or myocardial infarction due to coronary vasospasm. In the first patient, RV septal scarring was associated with a probable reentry substrate.
Electroanatomic mapping permitted the identification of low-voltage areas in sinus rhythm and direct identification of a protected isthmus in VT. Entrainment with concealed fusion was not attempted in order to prove the reentrant mechanism: this is a limitation of this observation. The case also illustrates the previously reported link between skeletal and cardiac muscle involvement in SS and the possible risk of proarrhythmia with hydroxychloroquine. Indeed, sustained arrhythmias are thought to be more frequent when the two types of muscle damage coexist [11]. When myositis is confirmed hydroxychloroquine is sometimes proposed as an adjunctive therapy even though this drug has the proarrhythmic potential of Class 1 agents [12].

In the second observation, VT occurred during haemodialysis but was unmappable due to the induction of only short non-sustained episodes despite the use of isoprenaline. Additional arrhythmogenic factors only present during haemodialysis like ionic potassium or calcium changes may have accounted for this discrepancy. Hypertension with LV hypertrophy in the setting of terminal renal failure is a possible cause for this left-sided VT rather than myocardial fibrosis directly related to SS [13]. The lack of reproducible VT initiation has already been reported, and non-reentrant mechanisms have also been hypothesized [4,6]. In support of the non-reentrant hypothesis is the absence of low-voltage or fragmented electrograms identified in sinus rhythm at the LV endocardium in this patient.

These two cases illustrate the interest of VT mapping and ablation in patients with SS. Electroanatomical 3D-mapping was contributory in identifying a critical protected isthmus in the first patient. However, it was also of some help to delineate low-voltage areas and to guide pacemapping in the second case in which the clinical VT was not inducible.
References