Ablation of Ventricular Tachycardia Originating from the Right Ventricle Associated with Scleroderma Cardiomyopathy

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A 49-year-old male was referred to the hospital because of syncope caused by ventricular tachycardia (VT) with a pulse of 240 bpm and QRS morphology with a left bundle branch block (LBBB) configuration and superior axis. The patient had been on a long-term regimen of steroids to treat his scleroderma. Satisfying 2 major criteria (QRS widening and an epsilon wave in the right chest leads) and 3 minor criteria (a slight enlargement and akinesis of the right ventricle, positive late potential in the signal averaged electrocardiogram and left bundle branch block-type VT) he was diagnosed with arrhythmogenic right ventricular cardiomyopathy. A voltage map of his right ventricle (RV) during sinus rhythm was obtained using an electroanatomical mapping system (CARTO, Biosense-Webster, Diamond, CA, USA). Two islet-like low voltage areas were found and linear double potentials were recognized between areas in the lateral wall of the right ventricle (RV) very close to the tricuspid annulus. The earliest activation of the double potential line during VT was 70 msec prior to the QRS onset. We applied radiofrequency energy at that point during the VT and it successfully slowed and terminated the VT. Thereafter the VT could not be induced by any stimulation from multiple RV sites.

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Case Report

The patient was a 49-year-old male. He had been taking oral steroids since 1999 to treat scleroderma (esophagitis and pulmonary fibrosis). In August of 2004 he lost consciousness and was taken to the emergency department of another hospital. The surface electrocardiogram revealed a ventricular tachycardia (VT) with a left bundle branch block (LBBB) and superior axis at a rate of 240 bpm.
A venous injection of 100 mg of lidocaine was ineffective, however, after a venous injection of 2 mg of propranolol the patient’s tachycardia terminated. A twelve-lead electrocardiogram recorded on admission revealed a left axis shift with an incomplete right bundle branch block and epsilon wave immediately following the QRS complex in lead, V2 (Figure 1). A venous injection of 100 mg of lidocaine was ineffective, however, after a venous injection of 2 mg of propranolol the patient’s tachycardia terminated. A twelve-lead electrocardiogram recorded on admission revealed a left axis shift with an incomplete right bundle branch block and epsilon wave immediately following the QRS complex in lead, V2 (Figure 1). The echocardiography revealed moderate enlargement of both ventricles (LVDd = 59 mm), and hypokinesia of the entire left ventricle (EF = 30%, FS = 14%), especially the posterior wall. The signal averaged electrocardiogram exhibited positive late potentials. From these findings arrhythmicogenic right ventricular cardiomyopathy was suspected. Angiography did not show any significant stenoses in the left or right coronaries. Right ventriculography showed a slight enlargement of the right ventricle and slight hypokinesis (RVEDV = 253 ml, RVEF = 25%). Left ventriculography revealed hypokinesis of the posterior wall and a slight hypokinesis in the apical area (LVEDV = 266 ml, LVEF = 51%). A myocardial biopsy of the tissue taken from the septal wall of the right ventricle was normal. Multi-detector computed tomography (MDCT) revealed an enlargement of the right ventricle but no other abnormalities, such as fatty degeneration.

Electrophysiological Study

From the morphology of the 12 lead electrocardiogram during the VT, we suspected that the VT originated from the right ventricle. An electroanatomical mapping system (CARTO, Biosense-Webster, Diamond Bar, CA, USA) was used to conduct voltage mapping of the RV during sinus rhythm since the patient’s vital signs were unstable and we were unable to construct an activation map during VT. Islets of low voltage were noted at the opposite wall just under the tricuspid valve, and a linear configuration of double potential sites was found between the islets (Figure 3). No VT was induced by programmed stimulation from the right ventricular apex during the baseline or after administering isoproterenol. However, VT was induced with burst stimulation conducted from the most electrically fragmented part of the right ventricular wall. At this site a satisfactory pace map was generated. On the surface electrocardiogram taken during the VT, the V wave preceded the QRS complex by 70 msec, so an attempt was made to ablate that site with a 4 mm tip Navistar™ (Biosense-Webster, Diamond Bar, CA, USA). The current was delivered but the temperature did not rise, so another attempt was made with an 8 mm tip, resulting in successfully slowing the pulse and reversing the VT (Figure 3). Thereafter, the VT could not be induced even under an isoprotelenol infusion and thus, the treatment was terminated.

Discussion

Scleroderma is a connective tissue disease in which lesions and fibrosis in the microvasculature cause damage to the skin, esophagus and respiratory system. Reports indicate that the heart is affected in about 25% of such patients. Clinical manifestations include conduction defects, pericarditis, angina pectoris and acute myocardial infarction, which are sometimes confirmed by post-sudden-death autopsy. In the present report we described a patient undergoing treatment for scleroderma exhibiting changes consistent with arrhythmicogenic right ventricular myocardosis. The patient satisfied 2 of the major criteria (QRS widening and an epsilon wave in
the right chest leads) and 3 of the minor criteria (slight enlargement and akinesis of the right ventricle, a positive late potential in the signal averaged electrocardiogram and a left bundle branch block-type VT) of the criteria of scleroderma reported by Mckenna WJ et al.3) There are some reports in the literature of myocarditis-like changes with scleroderma as in the present case.4) The cardiac damage

Figure 2 The 12-lead electrocardiogram at absolute rest.
A 12-lead surface electrocardiogram at admission showed a left axis shift with an incomplete right bundle branch block and an epsilon wave immediately following the QRS complex in lead V₂.

Figure 3
An electroanatomical mapping system was used to conduct voltage mapping of RV during sinus rhythm. Islets of low voltage were noted at the opposite wall just under the tricuspid valve, and a linear configuration of double potential sites were found between the islets (Right: RAO view, Left: Right lateral view).
that occurs in scleroderma is considered to be a result of reduced coronary perfusion, damage to small vessels and fibrosis.\textsuperscript{4,5} The involvement of the right ventricle in scleroderma has been reported to be the result of the RV dilatation following pulmonary hypertension or a myocardial infarction due to coronary vasospasms.\textsuperscript{4,5}

In the present case areas with islets of low-voltage were measured in the voltage map from the right ventricle taken from the lateral region just below the tricuspid valve, and radiofrequency ablation was successful at the fractionated potential sites between the islets. Since the patient’s vital signs were unstable, we were unable to create an activation map during the VT, and were unable to verify the VT circuit. There have been two cases reported in the literature describing patients undergoing treatment for scleroderma that developed VT that was verified to have originated from the right ventricle by electoraanatomical mapping as in the present case.\textsuperscript{5}

In that report, RV septal scarring was associated with a reentrant substrate in the first case, and in the second case, the LV and RV free walls close to the tricuspid valve were associated with two clinical VTs.

We experienced a case with arrhythmogenic right ventricular myocarditis-like changes and VT undergoing treatment for scleroderma. We were successful with radiofrequency ablation applications aimed at fractionated potential sites just below the tricuspid valve. Although we were able to successfully ablate the origin of the VT, we decided it was better to implant an ICD after the ablation since scleroderma was a progressive disease. However, we were unable to get the patient’s consent.

References