Electrical Storm Late after Surgery for a Double-Chambered Right Ventricle, Aortic Regurgitation and a Ventricular Septal Defect: A Case of Successful Catheter Ablation

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ABSTRACT
An electrical storm is defined as multiple occurrences of ventricular tachycardia/fibrillation (VT/VF) within a single day; this is a medical emergency and a poor prognostic marker in patients with an implantable cardioverter-defibrillator (ICD). We report here on the occurrence of electrical storms in a 35-year-old man with a repaired DCRV and ICD. He had recurrent VT and electrical storms that were refractory to amiodarone and β-blocker. A cardiac electrophysiologic study was performed 11 months after the ICD was implanted and two forms of VT were induced. After the catheter ablation of the VTs, the monomorphic VT became non-inducible. The frequency of the VT decreased from 35 per month before the catheter ablation to 1.1 times per month after the procedure. Catheter ablation of VT could be an effective treatment for patients with electrical storms refractory to antiarrhythmic drugs.

KEY WORDS: Ventricular tachycardia; Catheter ablation.

Introduction
An electrical storm (ES) is defined as the occurrence of more than 2 episodes of sustained ventricular tachycardia/fibrillation (VT/VF) within 24 hours, and this can occur in patients with an implantable cardioverter-defibrillator (ICD). It portends a poor prognosis in these patients. This malady can be managed by antiarrhythmic drugs, transvenous pacing or catheter ablation depending on the nature of the ventricular tachycardia/fibrillation and the type of underlying heart disease. We report here on a successful catheter ablation to treat ES that was refractory to amiodarone and β-blocker in an ICD recipient who underwent surgery for a double-chambered right ventricle and the associated severe aortic regurgitation and ventricular septal defect.

Case
A 36-year-old man with an ICD presented with multiple ICD shocks over 2 days. He was diagnosed as having a double-chambered right ventricle (DCRV), severe aortic regurgitation and ventricular septal defect (VSD) at the age of 14. He underwent open heart surgery at the age of 17, and this involved a resection of the muscular bundles in the right ventricle, aortic valve replacement and closure of a VSD. He presented at the age of 35 with sustained monomorphic VT with hypotension, which was restored to sinus rhythm by direct current cardioversion (Fig. 1). His physical examination was unremarkable except for a laterally displaced apical impulse and a mechanical valve click sound. The blood tests, including the serum electrolytes and cardiac enzymes, were within normal limits. Two dimensional and Doppler echocardiography revealed a dilated and globally hypokinetic left ventricle (end-systole dimension: 7.8 cm, end-diastole dimension: 9.0 cm and the left ventricular ejection fraction: 35%). The bi-leaflet tilting disc was well-functioning and there was no evidence of any obstruction of the right ventricular outflow tract or an intracardiac shunt. His coronary angiogram was normal. After written informed consent was obtained from the patient, he underwent implantation of a single chamber ICD (Atlas™ VR V-199, St. Jude Medical, St. Paul, MN, USA) with a screw-in defibrillator lead (RIATA™ 1588, St. Jude Medical, St. Paul, MN, USA) with acceptable...
parameters related to the sensing, pacing and defibrillation. He had an ES one day after this procedure. Twelve episodes of VT with a cycle length (CL) of 455 to 465 ms were successfully terminated by antitachycardia pacing (ATP). He began taking 800 mg of amiodarone and a β-blocker thereafter in addition to an angiotensin-converting enzyme (ACE) inhibitor, diuretics and warfarin. He continued to have recurrent episodes of VT after the ICD implantation. The frequency of the VT episodes decreased after a loading dose of amiodarone was given (Fig. 2). A decrease in the amiodarone dose from 300 to 200 mg per day due to the elevation of his liver transaminase level was followed by an increase in the frequency of the VT episodes. Most of the episodes of slow VTs (CL: 375-500 ms) were terminated by ATP (Fig. 3), and those of fast VTs (CL: 355-375 ms)

Fig. 1. Standard 12-lead electrocardigrams at a paper speed of 25 mm/s. Ventricular tachycardia (A). Sinus rhythm (B).

Fig. 2. Clinical course of the patient. The upper panel summarizes the daily dose (mg) of amiodarone and lower graph summarizes the monthly episodes of ventricular tachycardia (VT) after the implantation of the ICD. After the catheter ablation of the VT, the frequency of the VT episodes markedly decreased from 35 to 1.1 per month. ICD: implantable cardioverter-defibrillator.
were terminated by ATP or 5 J electrical shocks.

He presented with 7 electrical shocks at 11 months after the ICD implant. Interrogation of the ICD revealed 110 episodes of VTs over one month (60 episodes of VTs with a CL of 415 to 460 ms in two days), and most of these were terminated with ATP. There was neither any fast VT nor ventricular fibrillation. However, there were 7 episodes of 30 or 36 J electrical shocks after the maximal time to fibrillation (MTF) timer had expired. There was no evidence of any electrolyte abnormalities or myocardial ischemia. Amiodarone was infused intravenously with a daily dose of 1200 mg without any further recurrence of the VT.

An electrophysiological study was performed 7 days after the electrical storm with the patient in the fasting state after written, informed consent was obtained from him. Two multipolar electrode catheters were introduced from the femoral vein and they were positioned in the His-recording area and right ventricular apex or outflow tract. The bipolar intracardiac electrograms were filtered between 30-500 Hz. All the data was digitally recorded using a CardioLab™ System (GE, Prucka, Houston, TX.

Fig. 3. Stored electrograms from episodes of ventricular tachycardia (VT). Electrogram from true ventricular bipole, marker channel, and electrogram from ICD can to ring were arranged from top to bottom. There was sudden rate acceleration and configuration change of ventricular electrograms, which were consistent with VT. Upon detection of VT, antitachycardia pacing (ATP) began. However, VT did not terminated by the first ATP (A). The second ATP terminated VT (B). ICD: implantable cardioverter-defibrillator.
USA). The basic interval measurements were the following: sinus cycle length: 1320 ms, QRS width: 100 ms, AH interval: 130 ms and HV interval: 80 ms. Ventricular tachycardias with two morphologies were reproducibly induced by programmed electrical stimulation, including burst pacing and ventricular extrastimulation. The first VT (type I VT) had the same morphological configuration as the clinical VT, with a CL of 360 to 385 ms and a right superior axis (Fig. 4A). The second VT (type II VT) had a CL of 520 ms and right axis deviation (Fig. 4B). No His deflection or right bundle potential was apparent during the VTs despite a stable position of the His recording catheter. However, the VTs were not sustained even after an infusion of isoproterenol at 2 μg/min due to the effects of the previous infusion of amiodarone. We decided to perform an ablation of the VTs under the guidance of pace-mapping. During the ablation procedure, the ICD was programmed to ‘therapy off’. Access to the left ventricle was achieved antegradely across the mitral valve after transseptal catheterization. Systemic anticoagulation was achieved with heparin, and sedation was done with midazolam and fentanyl. The left ventricular mapping and ablation were performed with a 7F quadrupolar, steerable catheter with a 4-mm distal tip electrode. Radiofrequency (RF) current (500 kHz) was applied under the temperature control method at 65°C and a power of 70 W for 60 seconds at the sites where the QRS morphologies of the 11 or 12 leads during pacing were identical to those of the targeted VT (Fig. 4C, D). No fascicular or diastolic potentials were recorded from the distal electrode of the mapping catheter at each of the best pace-match sites during sinus rhythm. The sites with the best pace-map of each VT were localized to the apicolateral and mid-lateral walls of the left ventricle (Fig. 4E, F). Booster RF energy applications were delivered circumferentially around the sites of the best pace-map. The total number of RF deliveries was 31. After the applications of the RF energy, the monomorphic VT was no longer inducible by burst pacing down to 2 to 1 ventricular capture or by ventricular triple extrastimuli with/without an infusion of isoproterenol at 2 μg/min. The polymorphic VT induced by the triple ventricular extrastimuli was defibrillated. The total procedural and fluoroscopic durations were 240 and 39 minutes, respectively. The procedure was finished without any complications.

He was continued on medical treatment that included amiodarone (100 mg qd), a β-blocker, an ACE inhibitor, diuretics and warfarin. He had only 3 episodes of VTs with a CL of 465 to 490 ms, and all 3 VT episodes were successfully terminated by ATP over a period of 10 months. The patient discontinued his amiodarone
by himself and an ES (17 VTs with a CL of 455-490 ms, all of which were terminated by ATP) occurred 11 months later. He remained free of any VT recurrence under the same drugs, including amiodarone, during the next 6 months follow-up period. The frequency of the VT episodes decreased from 35 to 1.1 episodes per month after the catheter ablation of the VTs.

Discussion

We report here on a case of successful ablation of ventricular tachycardias that presented with an ES in a patient with a repaired DCRV, a mechanical aortic valve and an ICD.

An ES is not uncommon and this is a poor prognostic marker in patients with an ICD. Catheter ablation of ventricular tachycardia can be used as a primary therapy or adjunctive therapy in addition to an ICD depending on the presence of structural heart disease. Catheter ablation is one of the treatment options in selected patients with an ES.

An ES may occur in patients with repaired congenital heart disease and an ICD. A DCRV is a form of a subvalvular right ventricular outflow tract obstruction by anomalous muscular bundles that divide the right ventricle (RV) into a high-pressure proximal chamber and a low-pressure distal chamber. Left bundle branch block morphology VT rarely occurs in patients with unrepaired DCRV and the origin of the VT was localized to the RV. The VT did not recur after corrective surgery in those cases. On the contrary, this present case presented with VT 18 years after the corrective surgery for the DCRV, and the VTs in this case had a morphology with an atypical right bundle branch block (RBBB), suggesting the VT origin was from the left ventricle (LV). The hemodynamic overload in the LV that was due to the severe aortic regurgitation, along with ventricular septal defect before the corrective surgery, might have contributed to the development of the VT from the LV.

Bundle branch reentrant ventricular tachycardia (VT) is a type of VT that can develop early in patients after an aortic valve replacement, as well as in patients with dilated cardiomyopathy. The prolonged HV interval in association with a dilated left ventricle should raise the possibility of bundle branch reentry as the mechanism of the VT. However, we excluded bundle branch reentrant VT as a mechanism of the VTs in this patient on the grounds 1) that the atypical RBBB patterns of the induced VTs were different from the typical left bundle or right branch block pattern of bundle branch reentrant VT 2) in Fig. 3A, when the first ATP was ineffective in terminating the VT, the postspacing interval from the defibrillator lead positioned in the right ventricular apex was measured at 620 ms, whereas the mean tachycardia cycle length was 445 ms and the resulting difference was 175 ms, which excludes bundle branch reentry and 3) good efficacy was noted for the endocardial ablation at the apicortal and mid lateral walls of the left ventricle where the bundle branch potential was not recorded. Fascicular VT after aortic valve replacement has also been reported. However, a different configuration of the VT from that of the above mentioned case and the absence of fascicular potentials at the target site during sinus rhythm excludes fascicular VT as the mechanism of our patient’s VT. The reproducible induction of VT and the proven efficacy of overdrive pacing in terminating the VT favor reentry or triggered activity as the mechanism of the VT in our case. A recent study suggested reentry as the main mechanism of VT in patients with prior valve replacements. However, the exact mechanism and circuit of the VTs could not be fully evaluated due to the non-sustaining nature of the VTs in our case.

Pace-mapping has been used for the mapping of VTs in patients with and without structural heart disease. Even though pace-mapping has its own limitations, it might be a last resort for guiding the ablation when the VT is noninducible, or activation and entrainment mapping cannot be performed due to non-sustained VT.

There are several limitations in our study. First, the mechanism of the VT could not be fully elucidated due to the nonsustained nature of the VT. If we had delayed the ablation several days or weeks with the patient off the amiodarone, then the VT might have been sustained. However, that might have caused unnecessary shock deliveries for the VT in this patient during the waiting period. Second, electroanatomic mapping would have facilitated defining the mechanism and reentrant circuits of the patient’s VTs.

In conclusion, we report here on successful ablation of VT in a patient with a repaired DCRV and ICD. Catheter ablation of ventricular tachycardia that’s guided by pace-mapping could be a valuable adjunct to antiarrhythmic drugs in patients with an ICD and who are suffering with an electrical storm.

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