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Case Report

Efficacy of bilateral thoracoscopic sympathectomy in a patient with catecholaminergic polymorphic ventricular tachycardia

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ABSTRACT

A 27-year-old woman with frequent implantable cardioverter defibrillator (ICD) shocks related to catecholaminergic polymorphic ventricular tachycardia (VT) experienced aborted sudden death due to incessant polymorphic VT despite the administration of beta-blockers, verapamil, and flecainide. Catheter ablation failed to suppress the polymorphic VT. Based on the temporary efficacy of the local anesthetic administered at the left and right cervical sympathetic nerves to suppress VT under an isoproterenol infusion, stepwise, bilateral thoracoscopic sympathectomy was performed. Postoperatively, no further VT or syncopal episodes were documented under ICD telemetry. Bilateral thoracoscopic sympathectomy may be an alternative for patients with drug-refractory catecholaminergic polymorphic VT.

1. Introduction

Catecholaminergic polymorphic ventricular tachycardia (CPVT) was first reported in 1975 and is characterized by exercise- or emotional stress-induced polymorphic ventricular tachyarrhythmias, syncope, or sudden cardiac death [1,2]. Although β-blockers, verapamil, and Na-channel blockers have been used to suppress VT in these patients, their symptoms persist and prognosis remains poor [3]. We describe a case of aborted sudden death due to catecholaminergic polymorphic VT in a patient who had received treatment with antiarrhythmic agents and an implantable cardioverter defibrillator (ICD) in whom VT could be successfully suppressed after bilateral thoracoscopic sympathectomy.

2. Case report

A 27-year-old woman diagnosed with CPVT, who did not have a family history of sudden cardiac death, and was implanted with a dual chamber ICD at another institution 11 years previously for recurrent syncopal episodes during exercise and emotional stress was followed up by our hospital. She had experienced frequent (nearly 5 times per year) ICD shocks (Secura™, Medtronic, Minneapolis, USA) related to ventricular fibrillation or atrial tachycardia (AT) with a rapid ventricular response under the oral administration of antiarrhythmic agents, including bisoprolol 5 mg/day, verapamil 240 mg/day, and flecainide 150 mg/day (3 mg/kg), which were her maximally tolerable doses because of general fatigue due to hypotension. During the follow-up, she was resuscitated from aborted sudden death due to incessant polymorphic VT documented on electrocardiography. Deep sedation with intratracheal intubation was effective in suppressing VT, and fortunately, no brain damage occurred after hypothermia therapy. The initial premature ventricular contractions (PVCs), which triggered the polymorphic VT, were confirmed to have mainly two morphologies: left bundle branch block with a superior axis (PVC1) and right bundle branch block with an inferior axis (PVC2) under an isoproterenol infusion (ISP) at 1.5 μg/min. We confirmed that the maximum beat runs of VT could be suppressed to only couplets of PVCs by a left stellate ganglion blockade using lidocaine as the local anesthesic, under the same dose of the ISP

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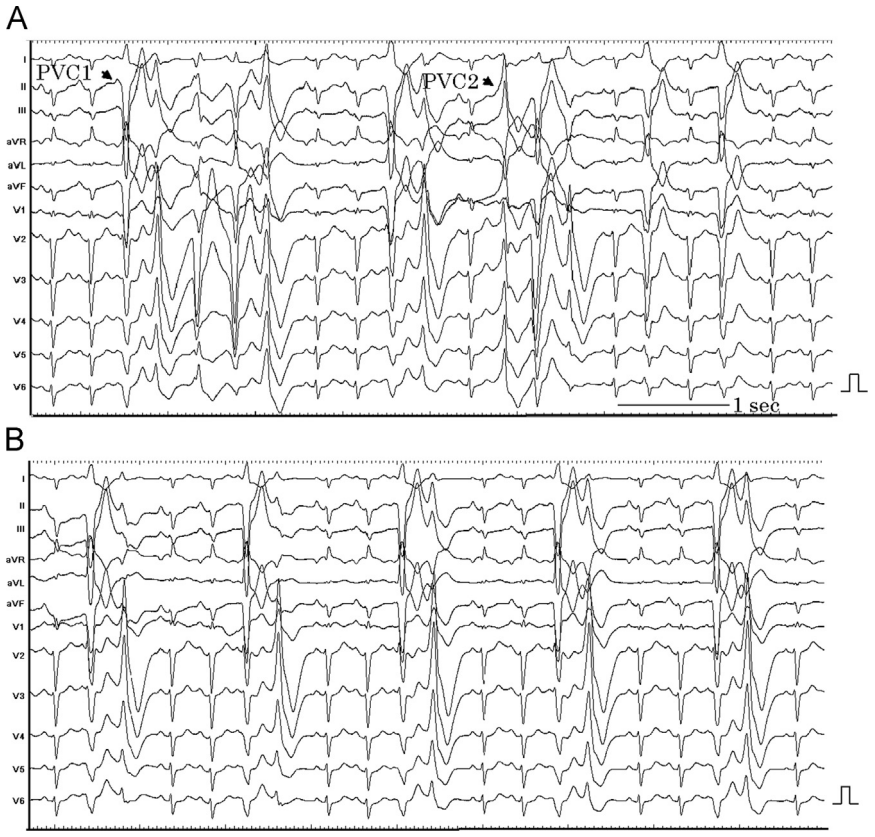

Fig. 1. (A) Polymorphic VT (maximally five beat runs) was frequently induced by an ISP infusion (1.5 μg/min). The heart rate (P–P interval during sinus rhythm) was 134 bpm. We immediately discontinued the ISP infusion because we were concerned about the occurrence of life-threatening sustained polymorphic VT. (B) VT could be suppressed up to only couplets of PVCs after the local left stellate ganglion blockade under the same dose of ISP infusion. The heart rate was similar to that mentioned above.

infusion (Fig. 1). First, we attempted catheter ablation targeting those initial PVCs. The right ventricular inferior wall under the tricuspid valve (PVC1: the local bipolar ventricular potential preceded the onset of the PVC by 20 ms without any discrete pre-potentials, and a good pacemap of the QRS configuration was obtained) and left ventricular outflow tract immediately under the aortic valve of the left coronary cusp (PVC2: a good pacemap of the QRS configuration was obtained) were considered as the origins of the PVCs, and ablation at those sites with 25–30 W suppressed the PVCs under the same ISP infusion dose of 1.5 μg/min. However, ablation at the above sites was ineffective under an infusion of ISP of 3.0 μg/min because a maximum of 8 beat runs of a non-sustained polymorphic VT (NSPVT) with an average cycle length of 237 ms was induced. In addition, treadmill exercise testing (10.2 Mets) after the ablation revealed the occurrence of bidirectional polymorphic VPCs. Those PVCs were mainly of two types, such as a left bundle branch block with a superior axis and an inferior axis type, and the morphologies were different from those before the ablation. Although VT did not recur clinically, the catheter ablation was considered insufficient to prevent any future VT recurrence. Therefore, after written informed consent was obtained, the patient underwent left thoracoscopic sympathectomy from the lower one-third of the left stellate ganglion to the Th4 level under general anesthesia (Fig. 2) based on the effectiveness of the left stellate ganglion local blockade and according to a previous report [4]. After that operation, VT was no longer inducible until 10.2 Mets of exercise via treadmill exercise testing, but was easily induced over 11.2 Mets of exercise. We next evaluated the efficacy of the local right stellate ganglion blockade, and found that this dramatically suppressed the polymorphic VT under an ISP 3.0 μg/min infusion. Based on this result, she underwent an additional right thoracoscopic sympathectomy from the lower one-third of the right stellate ganglion to the Th5 level. After the operation, VT could no longer be induced for up to 11.2 Mets of exercise (Table 1: each number after NSPVT indicates maximum beat runs). The heart rate was lower (93 bpm vs. 103 bpm) during the same 10.2 Mets of exercise, and AT inducibility was suppressed to a greater extent during the exercise (11.2 Mets vs. 12.9 Mets) after right thoracoscopic sympathectomy than after left thoracoscopic sympathectomy alone. Complications such as left ptosis and decreased sweating of the left hand (Horner's syndrome) persisted during the follow-up after left thoracoscopic sympathectomy, but the symptoms could be well tolerated without any complaints of severity. The patient was discharged from our hospital under the oral administration of bisoprolol 5 mg/day and verapamil 240 mg/day. Nine months after bilateral sympathectomy, she felt an ICD shock immediately after bathing but did not experience any syncope. ICD telemetry revealed an inappropriate shock for an AT with a rapid ventricular response and some PVCs without any polymorphic VT (Fig. 3B), but no ICD shocks have been recorded after that last event up to the 12 months of follow-up.

3. Discussion

CPVT is an inherited arrhythmia syndrome, characterized by polymorphic ventricular tachycardia induced by adrenergic stress and caused by mutations of the cardiac ryanodine receptors or calsequestrin gene without any structural heart disease. Beta blockers are currently a class I indication for the treatment of clinically diagnosed patients [5]. Watanabe et al. discovered that flecainide prevents arrhythmias in a mouse model of CPVT by inhibiting the cardiac ryanodine receptor-mediated Ca^{2+} release and directly targets the underlying molecular defect. In addition,