Successful Surgical Treatment of Heart Failure and Ventricular Tachycardia in a Patient With Arrhythmogenic Right Ventricular Dysplasia With Cardiomyopathy

Hidenori Sako, MD; Tetsuo Hadama, MD; Shinji Miyamoto, MD; Hirofumi Anai, MD; Tomoyuki Wada, MD; Naohiko Takahashi, MD*; Hironobu Yoshimatsu, MD*

A 70-year-old male patient with arrhythmogenic right ventricular dysplasia/cardiomyopathy demonstrating frequent attacks of ventricular tachycardia (VT) as well as heart failure underwent surgical treatment. Although the patient had severe regurgitation at the mitral and tricuspid valves, the contractility of the right and left ventricles was almost maintained. Annuloplasty of both valves abolished the regurgitation and very effectively controlled heart failure. Surgical cryoablation was performed on the lesion showing the earliest potential before the ORS complex during VT and the arrhythmia was terminated. However, a cardioverter defibrillator was implanted to prevent new VT caused by disease progression. (Circ J 2005; 69: 996–999)

Key Words: Arrhythmia surgery; Heart failure; Right ventricle; Tricuspid valve

Case Report

A 70-year-old, 52-kg, 167-cm man was admitted as an emergency because of palpitations and syncope. Chest X-ray on admission showed severe dilation of the right atrium (RA), as well as an increased cardiothoracic ratio (77%) (Fig 1B), and sustained VT on the electrocardiogram (ECG) (Fig 2A). Although the sustained VT was terminated by intravenous administration of lidocaine hydrochloride (100 mg), nonsustained episodes was observed several times after admission.

There was a family history of arrhythmia; his mother had died suddenly in her 40s, and both his elder brother and the brother’s daughter have received pacemakers. When the patient was 62 years old, he was diagnosed as having sinus arrest, at which time echocardiography demonstrated dilation of the RA and RV, as well as 2+ tricuspid regurgitation. An endomyocardial biopsy demonstrated fatty replacement of the cardiac muscle (Fig 2B). When he was 66 years old, he underwent implantation of a VVI pacemaker (Fig 1A), and at 69 years old, he underwent an electrophysiological study and sustained VT was induced by burst pacing. Based on these findings, he was diagnosed as having ARVD. Although an ICD was recommended at that time, he refused.

On the present admission, echocardiography demonstrated aggravation of the disease: a severely dilated RA and RV, and 3+ mitral and 3+ tricuspid regurgitation (Fig 3A,B). Contractility of the left ventricle (LV) was moderately decreased and the calculated LV ejection fraction was 33%. Laboratory data and abdominal echography showed mild congestive hepatic dysfunction. Because the disease had advanced despite medical therapy, and the RV and LV contractility had been maintained, surgical therapy was selected to treat the valvular regurgitation.

Surgical Procedure

Both the RA and RV were remarkably dilated (Fig 1C) and although there was neither contraction nor fibrillation of the RA, the contractility of the RV was almost maintained. Before starting cardiopulmonary bypass (CPB), VT was induced by programmed pacing and epicardial mapping confirmed that the earliest potential before the ORS complex during VT was located on the posterior wall of RV. After CPB was initiated, the RA was opened and endocardial mapping was performed, with the same result as the epicardial mapping. Cryoablation was performed transmurally from the endocardial side on the posterior wall of the RV, including the region of the earliest potential (−60°C, 3 min). After cross-clamping the ascending aorta, the left atrium was opened and a mitral annuloplasty was
performed with a 27 mm Duran ring because the valve had not prolapsed. The tricuspid regurgitation was controlled by annuloplasty with a 29 mm Duran ring. Because the intravenous VVI pacing lead may have worsened the tricuspid regurgitation it was removed. Transesophageal echocardiography after weaning from CPB showed no regurgitation of the mitral and tricuspid valves (Fig 3C,D). The ICD patch electrode (Medtronic 6721-L) was placed on the inferior wall and fixed to the diaphragm, and the bipolar VVI epicardial lead (Medronic 4968) was sutured to the anterior wall of RV. Both leads were connected to the generator (Medtronic GEM II VR 7229Cx) in the existing subcutaneous pocket in the left pectoral region through the left third intercostal space. The median sternotomy was closed.

**Postoperative Course**

During hospitalization, VT occurred once and was termi-
nated by the ICD. The cycle length and the QRS duration of this episode differed from the preoperative VT. Bepridil hydrochloride (150 mg/day) controlled the VT and the patient was discharged. One year after surgery, during a bout of severe acute enteritis, sustained VT occurred and was immediately terminated by the ICD. The antiarrhythmic agent was changed to sotalol hydrochloride (80 mg/day) and VT was again well controlled and no further VT episodes occurred during the follow-up period. The patient was alive and well at the latest follow-up, 2 years after surgery.

**Discussion**

ARVD is a cardiac muscle disease of unknown cause that predominantly affects the RV. The diagnosis is based on the presence of certain major and minor diagnostic criteria stipulated by the international task force. The management strategy consists of achieving clinical diagnosis using the genetic, ECG, arrhythmic, morphofunctional, and histopathologic findings (Table 1). The diagnosis is established by the presence of 2 major criteria or 1 major criterion plus 2 minor criteria or 4 minor criteria. The present case fulfilled 2 major criteria (structural abnormalities in the RV, abnormal myocardial tissue in the RV with fatty infiltration confirmed by endomyocardial biopsy) and 2 minor criteria (sustained VT, mild segmental dilatation of the RV), and so was classified as definitive ARVD.

The development of heart failure in ARVD is rare, but often leads to death. The mechanism of the RV dysfunction is dilation, wall thinning and loss of contractile force because of progressive atrophy of the myocardium. Therefore, many cases of end-stage heart failure caused by ARVD mimic the dilated cardiomyopathy of other causes leading
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to congestive heart failure. In the present case, there was severe dilation of RA, but the contractile ability of RV and LV was almost maintained, so the main cause of the heart failure was regurgitation of the mitral and tricuspid valves, which was treated successfully by annuloplasty and the heart failure was well controlled postoperatively.

Ablation techniques have been used for the treatment of VT in cases of ARVD that are resistant to drug therapy. However, the acute success rate of catheter ablation of the VT reentry circuit varies widely (40–90%) and VT frequently recurs (<60% of cases). Relapse is closely related to the development of new arrhythmogenic zones because of the progressive nature of the underlying disease. Although a surgical approach to treating the VT in ARVD patients is uncommon, several methods were reported before the advent of catheter ablation. Surgical cryoablation was performed in some cases, and the success rate of cryoablation applied from the endocardial side was reported to be better than that from the epicardial side. In the present case, cryoablation terminated the induced VT during surgery and VT was well controlled postoperatively. However, a defibrillator was implanted to prevent recurrence or new VT. In this case, there was a postoperative relapse and antiarrhythmic agents were needed. An ICD provides life-saving protection in ARVD patients by effectively terminating life-threatening ventricular arrhythmias and improves the long-term prognosis.

References