Case report

Not letting the left side know what the right is doing!

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Abstract

Arrhythmogenic right ventricular dysplasia (ARVD) encompasses a spectrum of presentations including ventricular tachycardia, sudden cardiac death and heart failure. Complete right ventricular disarticulation was effective in a young athletic male who was refractory to drug therapy and experienced recurrent shock therapies from an implantable cardioverter-defibrillator that were incapacitating. The case highlights the challenging management of ARVD despite over two decades of research and the resurgent interest in ventricular disarticulation.

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1. Introduction

Arrhythmogenic right ventricular dysplasia (ARVD) is a progressive disorder encompassing a clinical and pathological spectrum characterised by fibro-fatty infiltration of the myocardium heart failure and ventricular arrhythmias and was first described over 20 years ago. The disorder affects one in 5000 individuals displaying a familial and male predilection [1]. Although it affects a broad range of the population, it commonly manifests in young athletic adults and can present as sudden cardiac death and recurrent ventricular tachycardia (VT) or fibrillation [2]. Control of symptoms and maintenance of survival still remains a challenge requiring a patient tailored approach. The following case illustrates the difficulties that complicate management.

2. Case presentation

An athletic 46-year-old male was diagnosed with ARVD after presenting with recurrent exercise-induced VT. These episodes were not always associated with haemodynamic instability but in fact the majority were well tolerated. He had normal left ventricular function but the right ventricle (RV) showed progressive dilatation and impairment with sequential trans-thoracic echocardiograms. Stepwise pharmacological therapy (which included beta-blockers, sotalol and amiodarone) failed to control the frequency of VT. He also received an implantable cardioverter-defibrillator (ICD).

The patient pursued an active lifestyle despite medical advice and now presented with recurrent shock therapies during vigorous physical activity. Device analysis revealed appropriate discharges after failed antitachycardia pacing correlating with exercise induced VT. He was not presyncopal prior to the shocks and the repeated device therapies were distressing. Electrophysiological studies elicited multiple morphologies of VT arising from a RV substrate. Nevertheless ablation was attempted on two separate occasions. This did not reduce the frequency of episodes or the burden of successive device therapies. At the patient’s request, despite physician advice, all device tachycardia therapies were eventually turned off. The ICD was programmed to a ‘monitor only’ mode.

The patient remained symptomatic with exercise-induced episodes of slow VT (without syncope). As a consequence of having device therapies turned off, he now carried a high risk of arrhythmia-related sudden cardiac death. Surgical disarticulation of the RV therefore presented an attractive management option. The overwhelming clinical circumstances and potential benefits from the procedure were weighed against the risks. The option was discussed with the patient who accepted the surgical strategy.

The surgical disarticulation was uncomplicated and proved highly successful during subsequent follow-up of...
the patient. It allowed him a symptom-controlled lifestyle without drug therapy and he was able to pursue sporting activities despite episodes of VT documented with endocardial electrograms monitored by the ICD (Fig. 1A). These, however, remained isolated in the RV (Fig. 1B). The ICD was later explanted.

3. Discussion

Control of recurrent VT in the index patient was ineffective with conventional therapy. The decision to opt for a complete surgical RV disarticulation was made after careful deliberation and after following a sequential therapeutic plan.

Surgical disarticulation was described by Guiraudon et al. [3] in 1983 and involves transection of RV myocardium along the free wall extending the natural electrical boundary provided by the fibrous skeleton composed of the pulmonary and tricuspid valve supporting annuli while preserving the RV coronary artery vasculature (Fig. 2). The myocardial involvement is, however, indiscriminate in the disorder, despite identifiable vulnerable segments within the RV; the so-called ‘triangle of dysplasia’ inflow, outflow and apex [3]. These are included within the boundary of incision but atrial and left ventricular and septal involvement in ARVD may compromise the success of the procedure. Pre-surgical imaging studies and documented VT morphologies suggested predominant RV involvement in the index case, hence he was deemed suitable for the procedure. Excellent long-term results following disarticulation have also been documented in well-selected patients [4].

As a consequence of the procedure, the RV free wall is rendered akinetic promoting RV dilatation and contributing to haemodynamic instability [5]. Synchronised pacing has been shown to ameliorate this complication and may also improve heart failure symptoms [6].

Arguably, surgical disarticulation may be the preferred procedure in the late stages of the disease. The severely affected RV wall becomes extremely thin and easily prone to perforation during attempted ablation or defibrillator lead placement [7]. Careful visualised dissection may at this stage be a safer approach.

4. Conclusion

The management of ARVD still remains challenging despite our increased understanding of the disorder. Surgical disarticulation proved effective in the above patient by isolating paroxysms of VT to the RV and should remain a consideration in the management arsenal for this potentially life-threatening and disabling condition.

References