CASE REPORT

Arrhythmogenic Right Ventricular Dysplasia: Clinical Study

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Arrhythmogenic right ventricular dysplasia (ARVD) is a complex arrhythmogenic cardiomyopathy, characterized by a partial or total replacement of the right ventricular myocytes by fatty and fibrous tissue. In this study, we present a case of ARVD in 17 year old girl, who was admitted to the hospital after syncope with ventricular arrhythmia. The echocardiography did not demonstrate structural cardiac abnormalities but the magnetic resonance recently showed thinning of the right ventricular wall. The girl was treated with the lidocaine, amiodarone and next, after radiofrequency catheter ablation she was receiving metoprolol. The girl has remained asymptomatic for four years of follow-up.

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CASE STUDY

In this study we present the case of ARVD in 17 year old girl, who was very active in sports (swimming, skiing). The girl had experienced two episodes of syncope, and she was admitted to the hospital after the last syncope. The medical examination included history collection, physical examination, electrocardiogram (ECG), Holter monitoring, 2D and Doppler echocardiography and magnetic resonance (MR). The girl was diagnosed based on the criteria proposed by McKenna et al.¹ She has been experiencing arrhythmia-related symptoms such as palpitations since age 14. On Holter ECG, single premature ventricular complexes with left bundle branch block (LBBB) morphology were recorded. At the time of admission to the hospital the girl was asymptomatic, but she had abnormal ECG findings. She had longer QRS duration in lead V₁ (120 ms) than V₆ (QRS = 80 ms) with the T wave inversion in leads V₁. Numerous premature ventricular extrasystoles (with LBBB pat-

tern) with right axis deviation were recorded on the resting electrocardiogram (ECG) and on Holter (Fig. 1). The echocardiography did not identify structural cardiac abnormalities of right ventricle but MR showed thinning of the right ventricular free wall indicating focal dysplasia (Fig. 3). After three days of admission, the girl reported of feeling very weak and then, she became syncope. She had four repeated syncopal attacks, associated with ventricular fibrillation, which was recorded on Holter monitoring. The onset of ventricular fibrillation appeared to be preceded by premature ventricular beats (Fig. 2). She was treated with lidocaine, amiodarone, intravenous infusion of magnesium and potassium and after that, the arrhythmia was converted to the sinus rhythm. Radiofrequency catheter ablation (RFCA) combined with electroanatomical mapping system (CARTO) was attempted in the girl. Endocardial mapping during ventricular tachycardia demonstrated a site of earliest ventricular activation in right ventricular outflow tract (anteriolateral wall). After

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Figure 1. On the resting electrocardiogram (ECG) irregular rhythm and premature ventricular extrasystoles with left bundle branch block (LBBB) morphology with right axis deviation were recorded.

Figure 2. ECG recorded on a Holter monitor, which reveals ventricular fibrillation from the episode during repeated syncopal attacks (associated with ventricular fibrillation). The onset of ventricular fibrillation appears to be preceded by premature ventricular beats.

Figure 3. Magnetic resonance (MR) shows thinning of the right ventricular free wall.

successful ablation, the girl was receiving metoprolol, and magnesium and potassium orally. She has remained asymptomatic for four years of follow-up, but has been recommended to stop intense physical exercise and competitive sport. All family members underwent history collection, physical examination, electrocardiogram and 2D and Doppler echocardiography. No sudden deaths, undiagnosed syncope and/or features of ARVD have occurred in this family.

DISCUSSION

The term of arrhythmogenic right ventricular dysplasia was defined in 1994,¹ then in 1996, the WHO and the International Society and Federation of Cardiology introduced the name ARVD since the disease, affected mainly the right ventricle and led to arrhythmia.² Clinical symptoms including chest pain, palpitation, syncope and sudden death, are related to ventricular arrhythmia.^{4–6} Young adults are more frequently affected and symptoms include syncope or sudden death.^{3,6,7} The girl showed clinical features typical of ARVD, according to the diagnostic criteria of McKenna et al.¹ She was symptomatic (palpitation accompanied by syncopal episodes) when she was 14 year old. During the 24-hour ambulatory ECG monitoring, single ventricular complexes with left bundle branch block (LBBB) morphology were recorded. The site of origin was probably the right ventricular outflow tract. She had not earlier been treated by any antiarrhythmic drugs. Three years later she had syncopal attacks, associated with ventricular fibrillation, but

she was successfully resuscitated. Corrado et al. have shown that eight patients with ARVD have suddenly died while taking antiarrhythmic drugs to treat ventricular arrhythmias.⁶ The main characteristics of ARVD are the reverted T waves in the right precordial leads (V_1 – V_3) and prolonged QRS duration (QRS >110 ms).¹ The girl presented in this study had longer QRS duration in lead V_1 (120 ms) than in V_6 (QRS = 80 ms) with T wave inversion in leads V_1 . Corrado et al. have reported similar ECG abnormalities in 36 young victims with ARVD, and they showed that ST segment elevation in V_1 – V_3 commonly found in ARVD.⁸ The sudden death was frequently related with the exercises. Furlanello et al. have showed that ARVD was manifested in 6%–27% of competitive athletes with arrhythmias,⁹ and Heidbuchel et al., at 59% of young cyclists.¹⁰ They have speculated that endurance sport by itself may also lead to right ventricle structural damage, that might not develop without any activity.¹⁰ The girl has also been very active in sports (swimming, skiing), but at present she is discouraged from participating in competitive sports. Radiofrequency catheter ablation (RFCA) combined with electroanatomical mapping system (CARTO) is an attractive alternative to pharmacological treatment of ARVD arrhythmias and the prevention of sudden cardiac death. Ablation can be useful as adjunctive therapy in management of patients with ARVD with recurrent ventricular arrhythmias despite optimal antiarrhythmic drug therapy.¹¹ Radiofrequency catheter ablation in ARVD can achieve a good short-term success rate. Recurrences become increasingly common during long-term follow-up, possibly as a result of the progressive nature of ARVD.^{4,12} RFCA may be indicated for some patients either as a primary therapy or as an adjunct to ICD implantation. The use of implantable cardioverter/defibrillator (ICD) is the treatment of choice for many ARVD patients with ventricular arrhythmias and left ventricle involvement and one or more affected family member with sudden cardiac death or undiagnosed syncope.¹¹ The diagnosis in children is very difficult. Cardiac MR has allowed the identification of focal structural changes, which could not be recognized with echocardiography.⁵ In the girl of our interest, the echocardiography has not demonstrated structural cardiac abnormalities of the right ventricle. In pediatric patients this examination does not provide structural cardiac abnormalities, but syncope with idiopathic right ventricular ectopy could be an early

manifestation of arrhythmogenic right ventricular cardiomyopathy. The cardiac magnetic resonance and genetic studies (in future) have the potential to play a central role in the diagnosis of ARVD.

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