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Clinical manifestations of arrhythmogenic right ventricular cardiomyopathy in Korean patients

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Abstract

Background: The clinical manifestations of the Korean patients with arrhythmogenic right ventricular cardiomyopathy (ARVC) are not well known.
Methods: The clinical data of Korean patients who met the Task Force Criteria for ARVC were analyzed.
Results: Thirty-seven patients (41.2±14.8 years old, 19 males) were diagnosed with ARVC. The commonest presenting symptoms were palpitations (30%), syncope/presyncope (30%) and no symptoms (30%). Four patients had a family history of premature sudden death or ARVC. Most patients with no symptoms were evaluated due to ECG abnormalities or asymptomatic ventricular arrhythmias. Ventricular tachycardia, ventricular fibrillation and frequent premature ventricular contractions only were observed in 35%, 5% and 24%, respectively. Wall motion abnormalities of the right and left ventricles were detected in 92% and 41%, respectively. Fatty or fibrofatty infiltration was observed in 26 of the 32 (81%) patients who underwent an endomyocardial biopsy. Two patients had signs of heart failure. Two patients with syncope/presyncope were diagnosed with vasovagal syncope and another was due to side effects from a medication. Most of the patients with ventricular arrhythmias were treated with β-blockers and/or amiodarone. Implantable cardioverter-defibrillators (ICDs) were implanted in 3 patients. During a mean follow-up of 27.4±26.5 months no syncope or sudden death developed except for in one patient with an ICD who suffered from recurrent shocks due to ventricular fibrillation.
Conclusions: ARVC may be an important cause of syncope, ventricular arrhythmias, and ECG and wall motion abnormalities of the ventricles in Koreans. The Korean patients with ARVC exhibited various clinical manifestations.

Keywords: Arrhythmogenic right ventricular cardiomyopathy; Korea

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is a kind of cardiomyopathy characterized by fibro-fatty infiltration of the myocardium, ventricular arrhythmias, sudden death, and heart failure [1,2]. Our previous study also showed that ARVC is an important cause of sudden death among young Koreans [3]. Many previous studies on ARVC exhibited significant differences in the clinical profiles

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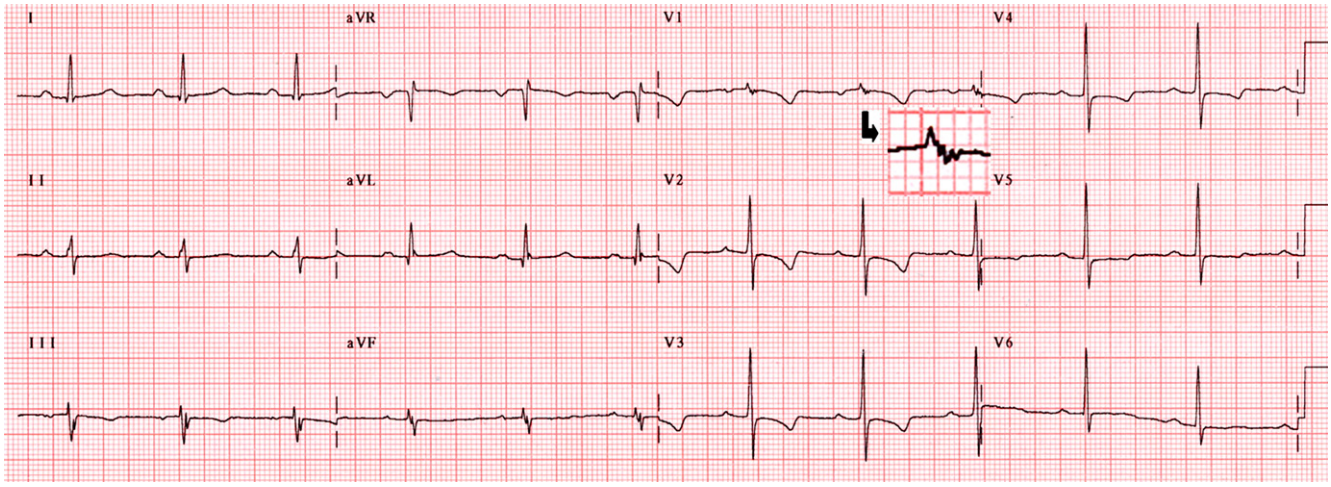

Fig. 1. Routine ECG in a patient with arrhythmogenic right ventricular cardiomyopathy exhibiting T-wave inversion in the right precordial leads and an ϵ-wave (arrow) in lead V_1.

[1,2,4–11]. The clinical manifestations of the Korean patients with ARVC are not well known. This prospective study was designed to describe the clinical manifestations of the Korean patients with ARVC.

1. Methods

From 1998 to 2006, 175 patients with syncope/presyncope, ECG abnormalities, nonischemic wall motion abnormalities, and/or left bundle branch block pattern ventricular arrhythmias were prospectively evaluated. The study protocol was approved by the institutional ethics committees and written informed consent was obtained from each patient. The clinical data of the patients who met the Task Force Criteria for ARVC [12] were analyzed. The evaluation included a detailed history taking and physical examination. Noninvasive tests, including a 12-lead routine ECG, 2-dimensional (D) echocardiography, signal-averaged ECG (SAECG), Holter monitoring and exercise-ECG were performed in all patients. A cardiac MRI was not routinely performed as our experience with using an MRI for the diagnosis of ARVC was limited. A T-wave inversion beyond V_1, QRS duration >110 ms in the right precordial leads, and an ϵ-wave were searched from the routine ECGs (Fig. 1). Two-D echocardiography with a 2.5 MHz transducer (GE, Norway) was performed with a specific emphasis on the right ventricle in order to determine the presence of findings such as global or regional right ventricular dilation and/or dysfunction. The SAECGs were obtained using a MAC 15 system (Marquette, USA) with a high-gain amplification and bidirectional Butterworth filters (40–250 Hz). Late potentials were considered present if ≥2 of the following criteria were met: (1) a filtered QRS duration >114 ms, (2) a low-amplitude signal duration >38 ms, and (3) a root mean square voltage <20 μV [13].

Invasive studies, including programmed ventricular stimulation (n=33), coronary angiography (n=33), selective biplane right and left ventriculography (n=33) and endomyocardial biopsy (n=32), were performed in most patients. The ventricular stimulation was performed with basic cycle lengths of 500 and 400 ms with up to two extrastimuli from the right ventricular apex and outflow tract. Isoproterenol was used when the arrhythmia was non-inducible in the baseline state. Right and left ventriculography was performed by the usual methods. Endomyocardial biopsies were performed from the septal–apical region of the right ventricle using 5.5 Fr forceps (Cordis, USA) until approximately five to seven specimens of an adequate size were obtained.

2. Results

Thirty-seven native Korean patients were diagnosed with ARVC (Table 1). Their mean age at the clinical presentation was 41.2±14.8 (17–72) years old and 19 were males. Four (11%) patients had a family history of ARVC or premature (age<40 years) sudden death including one autopsy-proven case of ARVC. Asymptomatic younger brothers of three patients died suddenly while watching television at the age of 39, during sleep at the age of 32, and during dinner at the age

Table 1
Demographic data and presenting symptoms in the study group

Age (years)	41.2±14.8
Sex (M/F)	19/18
Presenting symptoms	
Palpitations	11 (30%)
Syncope/presyncope	11 (30%)
Atypical chest pain	3 (8%)
Cardiac arrest	1 (3%)
Sudden death in a family member	1 (3%)
None	10 (27%)
Family history	4
Diagnosed ARVC	2*
Premature sudden death	2

ARVC: arrhythmogenic right ventricular cardiomyopathy, *: including 1 case of autopsy-proven sudden death.