A Case of Arrhythmogenic Right Ventricular Dysplasia with Atrial Flutter

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SUMMARY

A 57-year-old male who had arrhythmogenic right ventricular dysplasia (ARVD) with recurrent atrial flutter (AF) is reported. The patient had more frequent episodes of AF than of ventricular arrhythmias. Magnetic resonance imaging, echocardiography and right ventriculography revealed dilatation of the right ventricle and endomyocardial biopsy specimens from the right ventricle showed findings which were compatible with ARVD. The left ventricular specimen, however, also revealed a loss of myocytes and interstitial fibroelastic changes. The present case demonstrates an overlap of post-inflammatory or primary endomyocardial fibroelastic changes with ARVD. (*Jpn Heart J* 35: 689-694, 1994)

Key words: Right ventricular dysplasia Fatty infiltration Endocardial fibroelastosis

ARRHYTHMOGENIC right ventricular dysplasia (ARVD) is a disorder characterized by fatty and fibrotic replacement of the right ventricular myocardium in association with ventricular tachycardia (VT).¹⁾ In a few patients with ARVD, pathologic lesions may involve the left ventricle or the atrium.²⁻⁴⁾ In addition, other arrhythmias, including sinus node dysfunction and atrioventricular block, have been reported.^{5,6)} Supraventricular arrhythmias often occur in ARVD despite antiarrhythmic therapy.⁷⁾ We report a case of ARVD with slow atrial flutter and infrequent episodes of ventricular arrhythmias.

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CASE REPORT

A 57-year-old male was admitted to our hospital with a chief complaint of recurrent palpitations. The patient had suffered from this symptom for 10 years despite treatment with several antiarrhythmic drugs. The patient's brother had died suddenly during athletic exertion at 26 years of age. At the time of admission, the patient's blood pressure was 144/88 mmHg, and the heart rate was 80 beats per minute (bpm). No abnormal physical findings were noted. Laboratory examination was unremarkable. Chest roentgenogram showed a cardiothoracic ratio of 49%.

The electrocardiogram (ECG) showed regular sinus rhythm with a rate of 72 bpm. Tall R waves in lead V₁, inversion of T waves in leads II, III, aVf, and V₁-V₆, intraventricular conduction disturbance, and prominent post excitation (Epsilon) waves were noted (Figure 1A). During a palpitation attack, slow atrial flutter with various degrees of ventricular conduction was observed (Figure 1B). Holter ECG revealed sporadic ventricular extrasystoles, suggestive of a right ventricular origin, but no sustained VT was documented on repeated recordings. A signal-averaged ECG showed a markedly prolonged, filtered QRS duration (197 msec) and decreased root mean square voltage (11.2 μ V).

Echocardiography demonstrated marked dilatation of the right ventricle

Figure 1. A: Prominent post-excitation waves are seen during the asymptomatic ECG. B: Atrial flutter was observed on the ECG during a palpitation attack.

Figure 4. A: Endocardial biopsy from the RV revealed moderate fibroelastic thickening of the endocardium and severe fibrosis of the subendocardium with partial fatty infiltration (Hematoxylin-Eosin stain, $\times 150$). B: This biopsy specimen from the left ventricle showed prominent interstitial fibrosis (Hematoxylin-Eosin stain, $\times 150$).

Delayed potentials in the apex and outflow tract of the right ventricle were noted (Figure 3). Sinus node function and atrioventricular conduction time were within normal limits. Double ventricular premature stimuli from the right ventricular apex and outflow tract failed to induce any repetitive response. Atrial premature stimulation was also unable to induce any clinical tachycardia at the time of study.

Endocardial biopsies were performed on both the right and left ventricles. Specimens from the right ventricle showed moderate to severe fibroelastic thickening of the endocardium and severe fibrosis of the subendocardium with partial fatty infiltration (Figure 4A). In the posterior wall of the left ventricle, a loss of myocytes and prominent interstitial fibrosis were observed (Figure 4B).

After administration of pilsicainide (150 mg/day), the incidence of atrial

flutter was markedly decreased.

DISCUSSION

In most cases of ARVD, VT is commonly observed,¹⁾ and other clinical arrhythmias are also seen.²⁻⁴⁾ However, few descriptions of supraventricular arrhythmias in ARVD exist.⁷⁾ Tonet et al⁷⁾ have reported the incidence of supraventricular tachycardia in ARVD seen in 17 of 72 patients (24%) in spite of antiarrhythmic therapy. In addition, right atrial enlargement was found in 5 of 12 patients (42%) examined by echocardiogram. They have suggested that enlargement of the right atrium or a right atrial abnormality, or both, might be the mechanism of supraventricular arrhythmias in ARVD.

In our case, atrial flutter presented as the main symptomatic arrhythmia. Although a biopsy of the atrium was not obtained, atrial involvement with the lesions noted above could be the genesis of atrial flutter in this patient. Secondary changes brought on by enlargement of the right atrium could also play a role in the development of atrial arrhythmias as suggested by Tonet et al.⁷⁾ Although the sporadic occurrence of extrasystoles originating from the right ventricle was observed in this case, sustained VT was not documented by repeated Holter recordings. It is not surprising that several patients with ARVD have had only ventricular premature beats.⁸⁾ Sustained VT may eventually develop with progression of the pathologic lesions.

Mild fatty infiltration with endomyocardial fibroelastosis compatible with ARVD was seen in this case. The endomyocardial fibroelastosis with hypertrophy was also noted in the left ventricle. Fibroelastic lesions of both ventricles could occur as the result of diffuse inflammation or as a primary endomyocardial fibrosis. Lobo et al⁹⁾ have described the pathologic features of 13 cases of ARVD, including 10 necropsy hearts, 1 explant heart, and 2 partial right ventricular resections. They found three patterns of histologic change: (1) right ventricle markedly thinned, epicardium and endocardium contiguous with virtually no intervening tissue; (2) normal wall thickness or slightly thinned, myocardium almost totally replaced by fat; and (3) normal or thin wall, myocardium largely replaced by fat with scattered residual myocardial cells and fibrous tissue. Although endocardial fibroelastosis was present in only eight cases; it was found in each of these three patterns and occasionally in the left ventricle. In this case, the presence of endocardial fibroelastosis in the LV could explain the extensive degree of ARVD; however, it remains unclear whether the endocardial fibroelastosis means the coexistence of ARVD and post-inflammatory changes. It is difficult to make a definite diagnosis at present only by the limited biopsy materials. Further experience, including examination of autopsy materials, is nec-

essay for clarifying the pathogenesis of ARVD.

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