

Arrhythmogenic Right Ventricular Dysplasia Presenting as Regression of Left Ventricular Dysfunction: A Case Report

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Abstract

Arrhythmogenic right ventricular dysplasia is considered to be a slowly progressive disease in which left ventricular dysfunction and congestive heart failure usually appear at the end stage. The initial clinical presentation of this 56-year-old Japanese woman was left-sided heart failure, and the diagnosis was dilated cardiomyopathy, but her left ventricular size and ejection fraction regressed during 10 years of treatment, whereas her right ventricular parameters showed no change.

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Key Words

Cardiomyopathies, other(arrhythmogenic right ventricular dysplasia) Heart failure
Quality improvement(regression)

INTRODUCTION

Congestive heart failure(predominantly right-sided)and sustained ventricular tachycardia are common clinical manifestations associated with arrhythmogenic right ventricular dysplasia (ARVD)¹⁾. Left-sided heart failure could also occur in the end-stage of the disease. We report a case of ARVD in a Japanese woman who was first treated under a diagnosis of congestive heart failure with dilated cardiomyopathy.

CASE REPORT

A 56-year-old Japanese woman who had been treated for hypertension was admitted to a hospital

due to cerebral infarction in August 1989. Coronary angiography and right and left ventriculography were performed. The diagnosis was dilated cardiomyopathy. She was referred to our hospital complaining of exertional dyspnea and systemic edema in May 1990. There was no increase in serum level of creatinine kinase. Electrocardiography showed normal sinus rhythm with low voltage in all leads and Q waves in leads I and aF . Epsilon waves could be seen in leads V_1 and V_2 as at the previous admission(**Fig. 1 - left**). Left and right ventriculography revealed marked dilation of both ventricles with depressed ventricular ejection fraction(**Fig. 2** : Right ventricular volume was calculated by the methods based on Simpson's rule²⁾). There were

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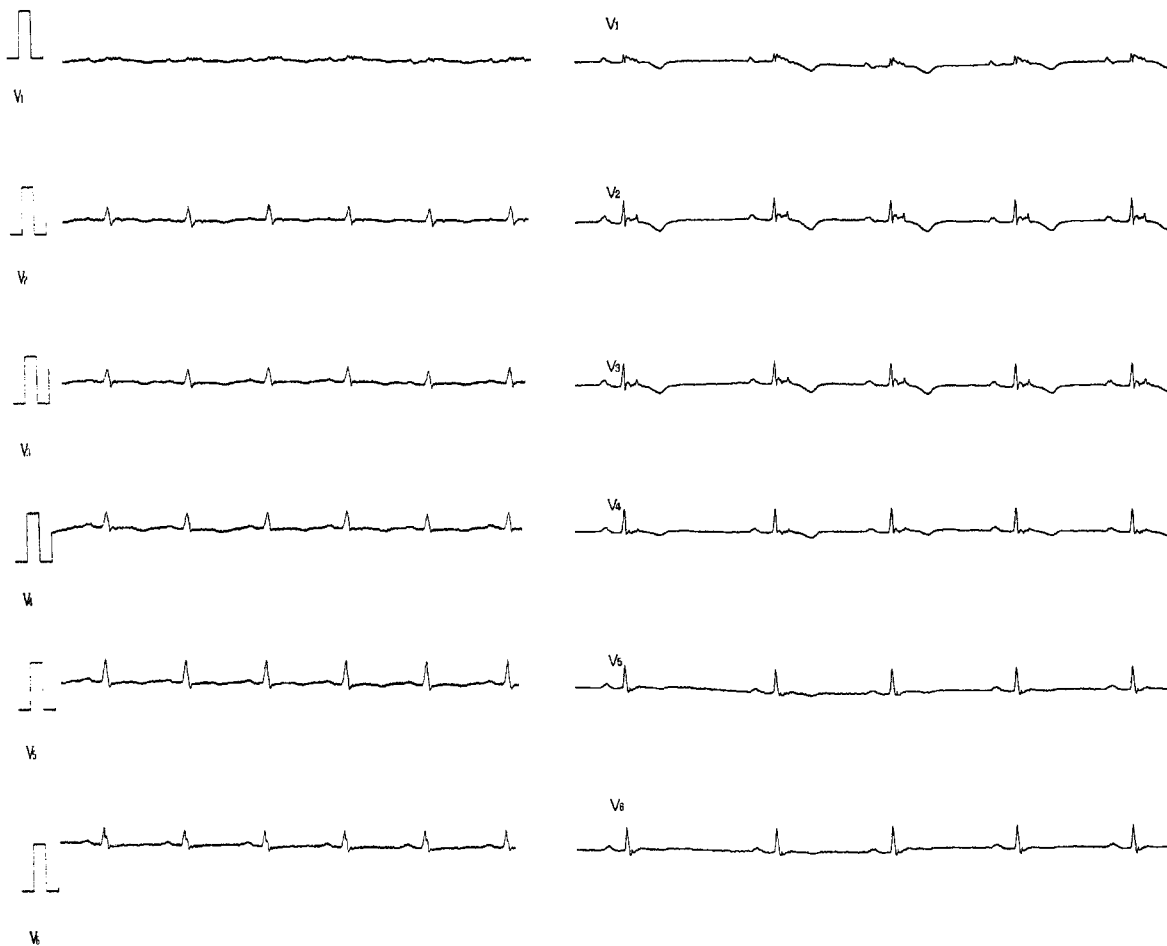


Fig. 1 Electrocardiograms recorded in 1990 and 1999
Left: In 1990, epsilon waves could be seen in leads V_1 and V_2 .
Right: In 1999, epsilon waves became apparent in leads V_1 to V_4 .

no coronary lesions or congenital defects. Therefore, the diagnosis of dilated cardiomyopathy was confirmed. Treatment with drugs such as digoxin, diuretics, and angiotensin converting enzyme inhibitor were continued. She was readmitted to our hospital in July 1999 with sustained ventricular tachycardia and left bundle branch block configuration. After ventricular tachycardia was converted to sinus rhythm, an epsilon wave was apparently detected in leads V_1 to V_4 (**Fig. 1 - right**). Right ventriculography indicated marked dilation of the right ventricle and depressed right ventricular ejection fraction that was almost the same as that in 1990. Biopsy findings of the right ventricular septum showed fibrofatty replacement and degenerative changes or moderate fibrosis of myocytes (**Fig. 3**). The diagnosis of ARVD was based on the diagnostic criteria suggested by

McKenna *et al.*³⁾ During the 10 years since her original treatment, the dilated left ventricle had shrunk, and both cardiac index and left ventricular ejection fraction had improved. However, the right ventricular size and ejection fraction were unchanged (**Table 1**).

DISCUSSION

Congestive heart failure (predominantly right-sided) and sustained ventricular tachycardia are common clinical conditions in patients with ARVD⁴⁾. About 18% of reported ARVD cases include left ventricular involvement⁴⁻¹³⁾, but patients presenting with left-sided heart failure are rare. ARVD is considered a slowly progressive disease^{6,14)}, with a natural course that can be schematically distinguished into four phases¹⁵⁾: Concealed phase, overt electrical disorder, right ventricular

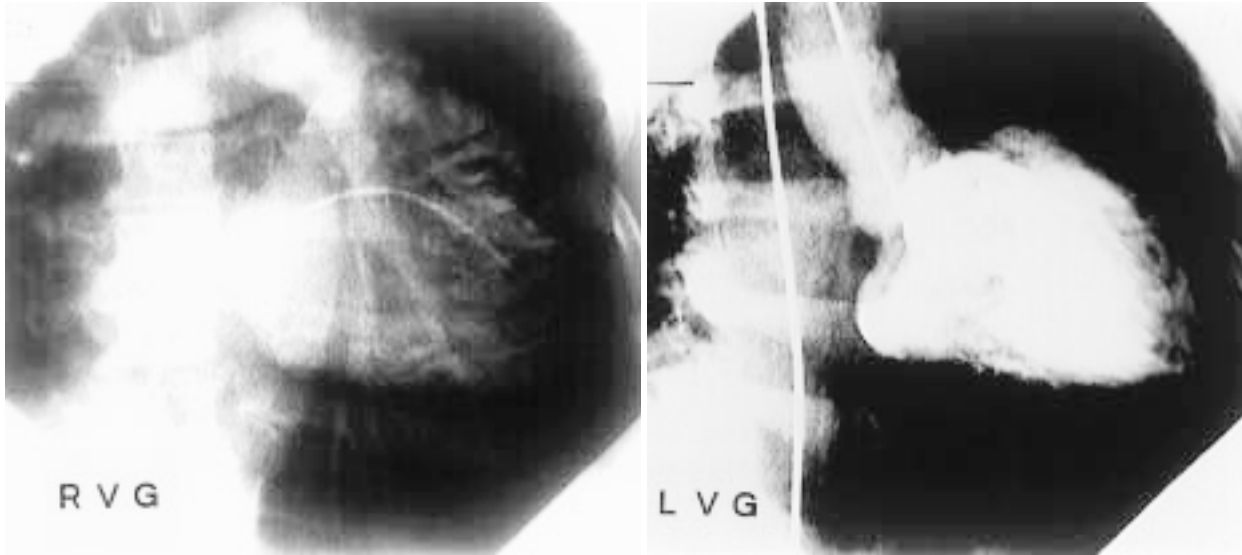


Fig. 2 Right and left ventriculograms in 1990

Left: Right ventriculogram (RVG) showing marked right ventricular dilation with decreased ejection fraction (right ventricular end-diastolic volume index 145 ml/m^2 , right ventricular end-systolic volume index 127 ml/m^2 , right ventricular ejection fraction 21%; 30° right anterior oblique view, end-diastolic phase).

Right: Left ventriculogram (LVG) demonstrating marked left ventricular dilation with decreased ejection fraction and cardiac index (left ventricular end-diastolic volume index 161 ml/m^2 , left ventricular end-systolic volume index 111 ml/m^2 , cardiac index 1.54 l/min/m^2 , left ventricular ejection fraction 31%; 30° right anterior oblique view, end-diastolic phase).

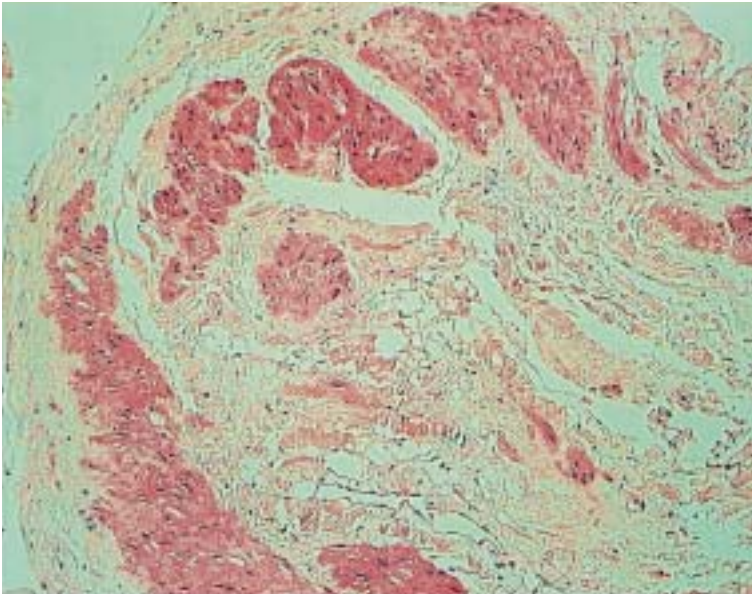


Fig. 3 Photomicrograph of the endomyocardial biopsy specimen from the right ventricular septum

Fibrofatty replacement and degeneration of myocytes are present (hematoxylin-eosin stain, original magnification $\times 100$).

failure, and congestive heart failure. Congestive heart failure usually appears during the end stage of the disease. Many studies have focused on the role of ventricular arrhythmias, so there is little data about the incidence and prognostic significance of heart failure in ARVD. Five of six patients with ini-

tial congestive heart failure subsequently died⁴. The cumulative survival rate of ARVD patients with congestive heart failure was significantly lower than that in patients with arrhythmias or no symptoms⁴. Therefore, the association of congestive heart failure with ARVD would be an adverse prog-

Table 1 Serial changes of cardiac parameters detected by cardiac pool scintigraphy, echocardiography and cardiac catheterization

	1990	1996	1998	1999
Cardiac pool scintigraphy				
RVEF(%)	11	(-)	13	13
LVEF(%)	12	(-)	27	41
Echocardiography				
RVDd(mm)	50	48	46	51
LVDd(mm)	51	44	38	40
LVDs(mm)	46	38	45	30
LVEF(%)	27	38	48	51
Cardiac catheterization				
RVEDVI (ml/m ²)	145	(-)	(-)	155
RVESVI (ml/m ²)	127	(-)	(-)	121
LVEDVI (ml/m ²)	161	(-)	(-)	(-)
LVESVI (ml/m ²)	111	(-)	(-)	(-)
CI (l/min/m ²)	1.54	(-)	(-)	1.84

RV = right ventricular; LV = left ventricular; EF = ejection fraction; Dd = diastolic dimension; Ds = systolic dimension; EDVI = end-diastolic volume index; ESVI = end-systolic volume index; CI = cardiac index.

nostic sign.

Our patient had a unique clinical picture. The initial clinical presentation was left-sided heart failure and she was treated for dilated cardiomyopathy. During the 10-year treatment period, the left ventricular size reduced, and both cardiac index and left ventricular ejection fraction had improved without concomitant right ventricular change (**Table 1**). Hypertensive heart disease or myocarditis should also be considered as the cause of left ventricular dysfunction and heart failure in this case. However, we have no evidence of apparent left ventricular hypertrophy or elevation of cardiac enzyme levels. In general, drugs such as diuretics

or angiotensin converting enzyme inhibitor are useful for promoting preload and/or afterload reduction as part of the treatment for congestive heart failure. In our patient, the left ventricular size could be reduced with such treatment. However, patients with ARVD who present with left-sided heart failure involving dilation of both ventricles and depressed cardiac function do not usually benefit from drug therapy.

A few cases have mimicked dilated cardiomyopathy like our case^{16,17}. All patients underwent heart transplantation or died. Our patient is unique in demonstrating regression of the dilated and depressed left ventricle.

要 約

左心機能が改善した不整脈源性右室異形成症の1例

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症例は56歳，女性．不整脈源性右室異形成症は緩徐進行性の心筋疾患で，右室の拡大と右室心筋の脂肪変性，心室頻拍を特徴とするが，終末像として左心室機能低下とうっ血性心不全をきたすこともある．本症例は左心不全で発症し，心機能や形態から拡張型心筋症と診断されて経過観察されていた．しかし，その後の10年の治療経過で，左心室のサイズは縮小し左室駆出率は改善したが，右心系には変化がなかった．

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