Journal of Cardiology Cases 10 (2014) 216–220

Contents lists available at ScienceDirect

Journal of Cardiology Cases

journal homepage: www.elsevier.com/locate/jccase

Case Report

A case of arrhythmogenic right ventricular cardiomyopathy presenting with progressive right ventricular failure and recurrent multifocal monomorphic ventricular tachycardia during 15 years of follow-up

Kensuke Kuwabara (MD)*, Kazuhito Hirata (MD), Minoru Wake (MD), Takanori Takahashi (MD), Jun Nakazato (MD), Nobuhito Yagi (MD), Asako Fukuyama (MD)

The Division of Cardiology, Okinawa Chubu Hospital, Okinawa, Japan

ARTICLE INFO

Article history:
Received 6 March 2014
Received in revised form 29 July 2014
Accepted 31 July 2014

Keywords:
Arrhythmogenic right ventricular cardiomyopathy
Right ventricular failure
Ventricular tachycardia

ABSTRACT

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is a multigenic form of cardiomyopathy characterized by myocardial loss and fibrofatty replacement mainly in the right ventricle. Progressive right ventricular dysfunction, ventricular arrhythmias, and sudden cardiac death are the clinical picture of this disease. Despite its clinical importance as a cause of sudden death, ARVC is likely to be under-recognized. In case reports about ARVC, disease characteristics such as arrhythmias, images, and genes are described in fragments. Little is reported about the long-term course of ARVC in the same patient. In this report, we present a case of a 68-year-old male who was diagnosed with ARVC after his first episode of ventricular tachycardia. Both mechanical and electrical progression were seen during the 15 years of follow-up, requiring the modification of disease management. This report could help improve the understanding of this rare disease, and the way of its management.

<**Learning objective:** Arrhythmogenic right ventricular cardiomyopathy (ARVC) is a multigenic form of cardiomyopathy characterized by fibrofatty degeneration mostly in right ventricular myocardium. Despite its importance as a cause of sudden death, ARVC is likely to be under-recognized. This paper describes the progressive course of ARVC confirmed both clinically and pathologically during 15 years of follow-up, which could help improve understanding of this disease.>

Introduction

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is characterized by fibrofatty degeneration mainly in the right ventricular myocardium [1]. The prevalence of ARVC is reported to be 0.02–0.1% [2,3]. Progressive right ventricular dysfunction, ventricular arrhythmias, and sudden cardiac death are the clinical features. Little has been reported on the long-term course of this rare disease. This paper describes the progressive course of ARVC confirmed both clinically and pathologically during 15 years of follow-up.

Case report

A 68-year-old male was admitted to our hospital (Fig. 1) because of monomorphic ventricular tachycardia (VT).

His initial presentation dates back 15 years, when he first developed sustained monomorphic VT requiring cardioversion (Figs. 1 and 2A). At that time he was hospitalized and evaluated (Fig. 1). There was no family history of specific cardiovascular disease. On physical examination, jugular venous pressure was not elevated, and there was no leg edema. No cardiac murmur was heard on auscultation. Chest radiograph showed normal cardiothoracic ratio (CTR) of 49% and no signs of lung congestion. Electrocardiogram (ECG) obtained during normal sinus rhythm showed ε-waves on leads V1 and V2 (Fig. 3A). Echocardiography revealed dilated right ventricle (RV), while left ventricular (LV) size and systolic function were maintained (Fig. 3C). Cardiac catheterization showed normal pressure in each cardiac chamber (Fig. 4A), and coronary angiography was normal. Endomyocardial biopsy showed mild fibrosis and disarray of myocardial fibers (Fig. 4B).

* Corresponding author at: The Division of Cardiology, Sendai Kousei Hospital, 4-15 Hirosemachi, Aoba Ward, Sendai, Miyagi 980-0873, Japan. Tel.: +81 22 222 6181; fax: +81 22 223 8442.
E-mail address: kuwabarakenten@gmail.com (K. Kuwabara).

http://dx.doi.org/10.1016/j.jccase.2014.07.014
1878-5409/

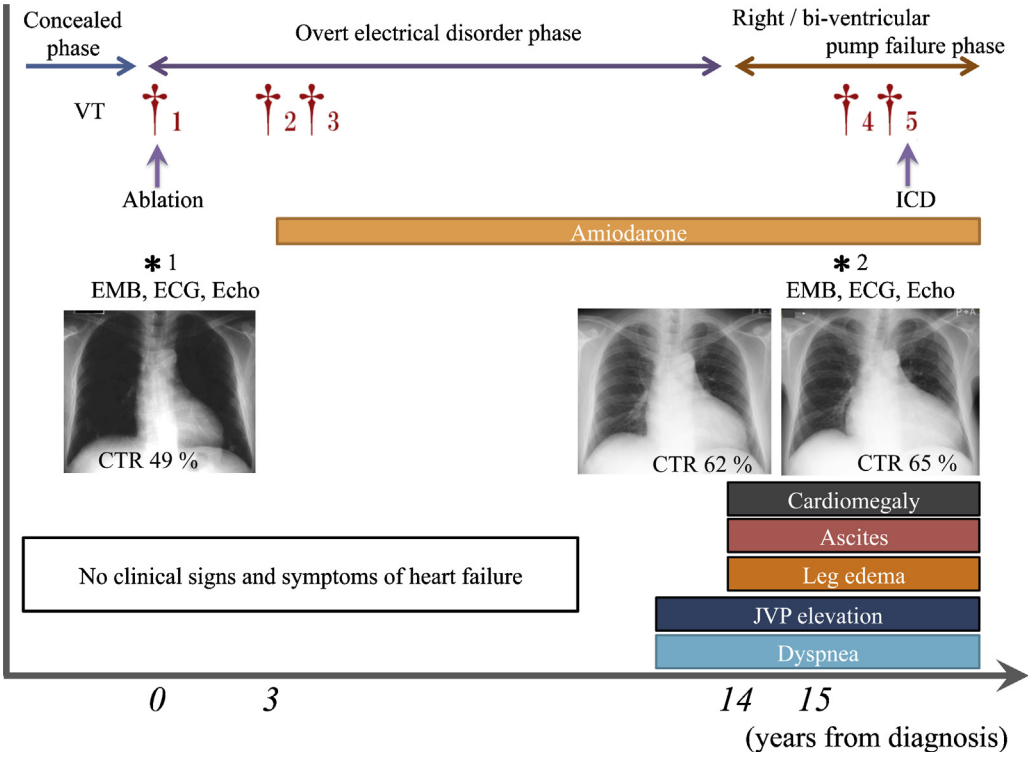

Fig. 1. Clinical course during the 15 years of follow-up. Initially, the patient presented with a ventricular tachycardia (VT) but did not have the signs and symptoms of mechanical heart failure except for right ventricular dilatation. As the disease progressed, the signs and symptoms of right heart failure gradually became overt as manifested by leg edema, ascites, elevated jugular venous pulse (JVP) and increased cardiothoracic ratio (CTR). *1 represents the initial presentation and *2 represents the current presentation. †1–5 represent episodes of VT. EMB, endomyocardial biopsy; ECG, electrocardiogram; Echo, echocardiogram.

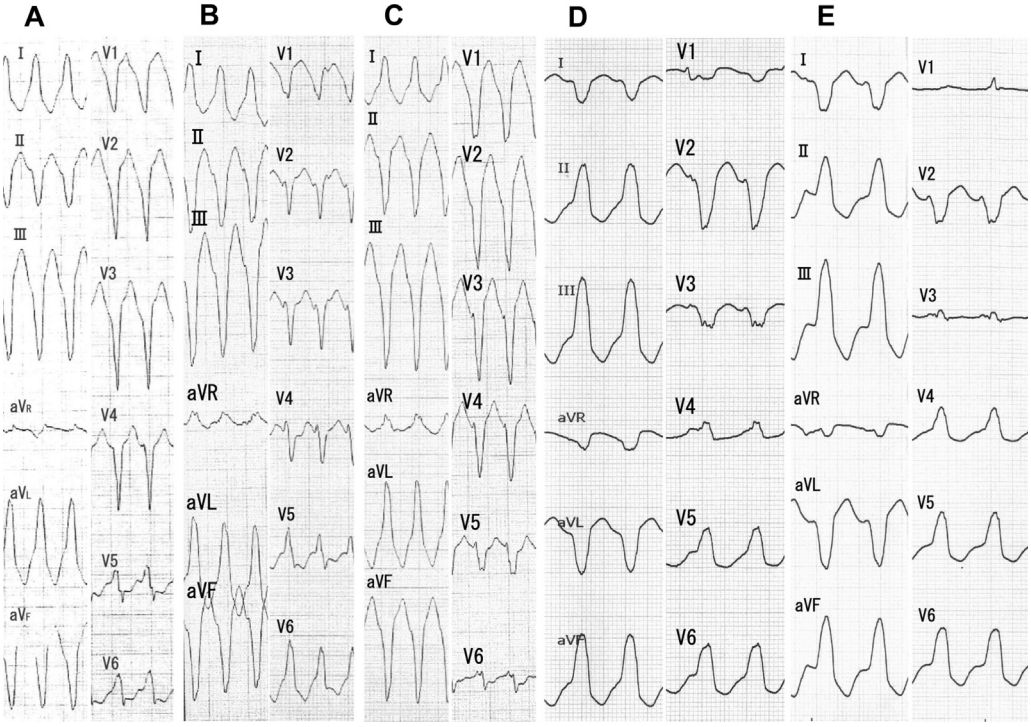

Fig. 2. Ventricular tachycardia (VT) morphology. (A) The first VT recorded at the time of diagnosis. (B–E) Four different forms of VT occurred during the 15 years of follow-up. (A–E) Respectively represent †1–5 in Fig. 1. (D) and (E) show VT morphology of the inferior axis.