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Short Report

Atrial tachycardia in a patient with arrhythmogenic right ventricular cardiomyopathy/dysplasia

Hiroaki Mano, MD, Ichiro Watanabe, MD*, Yasuo Okumura, MD, Kazumasa Sonoda, MD, Koichi Nagashima, MD, Toshiko Nakai, MD, Kimie Ohkubo, MD, Tatsuya Kofune, MD, Hironori Haruta, MD, Masayoshi Kofune, MD, Satoshi Kunimoto, MD, Atsushi Hirayama, MD

Division of Cardiology, Department of Medicine, Nihon University School of Medicine, 30-1 Ohyaguchi-kami machi, Tokyo 173-8610, Japan

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ABSTRACT

We describe a 49-year-old woman with atrial tachycardia (AT) and arrhythmogenic right ventricular cardiomyopathy/dysplasia. Cardiac magnetic resonance images showed a markedly dilated right atrium and right ventricle. Electroanatomical mapping showed that the AT originated from the right atrial appendage.

1. Introduction

Arrhythmogenic right ventricular cardiomyopathy/dysplasia (ARVC/D) is a myocardial disease that primarily affects the right ventricle (RV) and causes replacement of the myocardium with fatty and fibrous tissue. The predominant presenting symptoms – palpitations, syncope, or uncommonly, sudden cardiac death – result from ventricular tachyarrhythmias. This report describes a case of atrial tachycardia originating from the right atrium (RA) in a patient with ARVC/D.

2. Case report

A 49-year-old woman was admitted to our hospital because of palpitations. She had no family history of cardiac arrhythmias or sudden cardiac death. A cardioverter-defibrillator had been implanted 7 years earlier because of nonsustained ventricular tachycardia associated with ARVC/D. This case fulfills the 2 major criteria of ARVC/D [1]: (a) global or regional dysfunction and structural alterations detected by magnetic resonance imaging (MRI) (regional RV akinesia, RV contraction, or RV ejection fraction $\leq 40\%$), and (b) arrhythmias (nonsustained ventricular tachycardia of left bundle-branch morphology with a superior axis). The patient's chest radiograph showed an increased cardiothoracic ratio of 59% without pulmonary congestion. A 12-lead electrocardiogram (ECG) showed tiny P waves preceded by narrow QRS complexes at a cycle length of 300 ms with variable atrioventricular conduction (Fig. 1A). The signal-averaged ECG showed positive late potentials. Transthoracic echocardiography showed a dilated RA and RV and a left ventricular end-diastolic diameter of 52 mm with an LV ejection fraction (EF) of 35%. MRI angiography showed an enlarged RV cavity with an RVEF of 6% (Fig. 2). A cardiac biopsy of the RV endocardium showed fibrofatty replacement of the myocardium. During the electrophysiological study, narrow QRS tachycardia was noted. The tachycardia was sustained, although transient termination was observed by pacing from the high RA. A standard mapping and ablation catheter with a 4-mm tip (Navistar; Biosense Webster, Diamond Bar, CA, USA) was introduced into the RA from the right femoral vein. Activation mapping, guided by a CARTO XP system (Biosense Webster), showed that the earliest atrial endocardial activation site of the narrow QRS tachycardia (36 ms before the onset of the P wave) was located at the orifice of the RA appendage (RAA) (Fig. 3A) with a cycle length (CL) of 365 ms (AT1). The patient was therefore diagnosed with atrial tachycardia (AT) originating from the RAA. Radiofrequency ablation (RFA) (55 °C, 25 W, 30 s per cycle) was delivered, and after the fourth application to the same site, the AT–CL changed to 252 ms (AT2). Activation mapping of AT2 showed that the earliest endocardial activation (42 ms before the P wave) was localized to the tricuspid annulus (Fig. 3B). RFA at that site transiently terminated the AT. Atrial burst pacing induced a different AT (AT3; CL=430 ms), for which activation mapping showed that the earliest endocardial activation (46 ms before the

* Corresponding author. Tel.: +81 3 3972 8111; fax: +81 3 3972 1098.
E-mail address: iwatanab1953@gmail.com (I. Watanabe).

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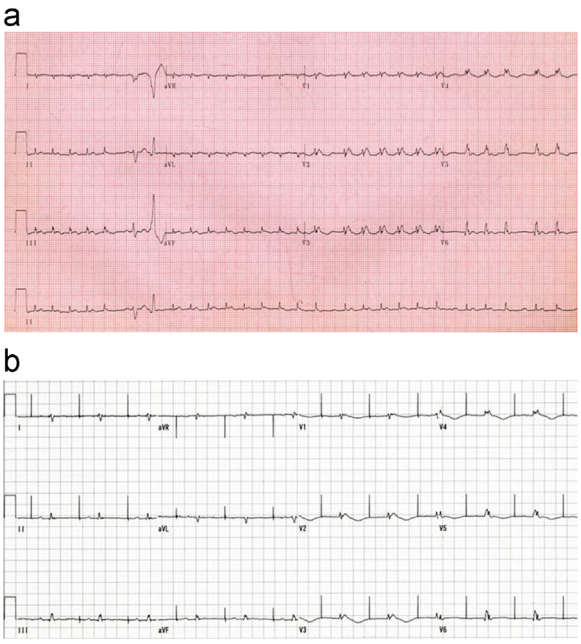

Fig. 1. Twelve-lead electrocardiograms obtained during atrial tachycardia (A) and after the ablation (B) a 12-lead electrocardiogram shows a tiny P wave preceded by a narrow QRS rhythm at a cycle length of 300 ms (A). Baseline rhythm during atrial pacing at a cycle length of 1000 ms (B).

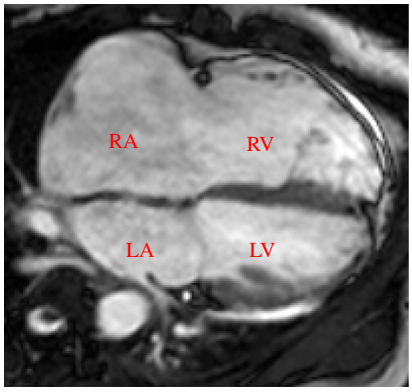

Fig. 2. Magnetic resonance imaging scan shows right atrial and ventricular dilatation.

P wave) was at the base of the RAA (Fig. 3C). RFA at that site changed the AT–CL to 360 ms (AT4). The earliest endocardial activation of AT4 (54 ms before the P wave) was located adjacent to AT1, and RFA at that site transiently terminated AT4. The P-wave morphology did not differ among each AT (Fig. 3A–C). In the voltage map during sinus rhythm, a low voltage area was widely observed throughout the RA, except in the RA appendage (Fig. 4). Thereafter, AT was not induced further by atrial extrastimulation or burst atrial pacing. The patient has been free of any atrial or ventricular tachyarrhythmias for 8 months.

3. Discussion

In this study, AT originated in the orifice of the RAA in a patient with ARVC/D. The origins of these ATs were quite close, which is supported by similar P-wave morphology of each AT. The ATs could be ablated by multiple endocardial RF applications. In this case, the exact mechanism underlying the ATs was unclear because of the failure of detailed electrophysiologic testing. However, the activations of each AT spread from a single earliest point of activation, and each AT activation time did not cover $>90\%$ of the tachycardia cycle length. The termination by pacing and these activation features suggest that the mechanism of AT is micro-reentry or triggered activity. To the best of our knowledge, there have been no reports of AT combined with ARVC/D, and only a few reports have described atrial involvement in ARVC/D. In a single case, right atrial biopsies verified the presence of fibrotic replacement of the atrial myocardium in a patient with ARVC/D, sick sinus syndrome, and an atrioventricular conduction disturbance [2]. A previous report showed that patients with ARVC/D had not only prolonged interatrial conduction expressed as a longer P-wave duration but also significant abnormalities in the P-wave morphology [3]. Atrial involvement in ARVC/D was also noted in several animal models of the disease, that is, the loss of the atrial myocardium and the presence of bilateral, focal, fibrofatty atrial lesions in up to one-third of affected boxer dogs and cats [4,5]. Takemura et al. [6] found that the RA was electrically silent with no capture during electrical stimulation at all sites, except for small areas in the lower RA in a patient with ARVC/D. In fact, this patient had an extensive low voltage area throughout the RA, and the P-wave duration during atrial pacing was substantially prolonged after ablation (Fig. 1B). Furthermore, the AT in our patient originated from the low voltage zone (LVZ) or the border zone adjacent to the LVZ (Fig. 4). The LVZ contained low-amplitude and fractionated electrograms (pink dots in Fig. 3), suggesting slowed or anisotropic conduction through the diseased RA. These anatomic changes could be a potential source for the development of AT and may be related to the fibrotic replacement of the atrial myocardium in a patient with ARVC/D. The occurrence of AT in patients with ARVC is rare; however, care is warranted because AT may cause inappropriate ICD discharges in patients with ARVC/D.

Disclosures and funding sources

None.

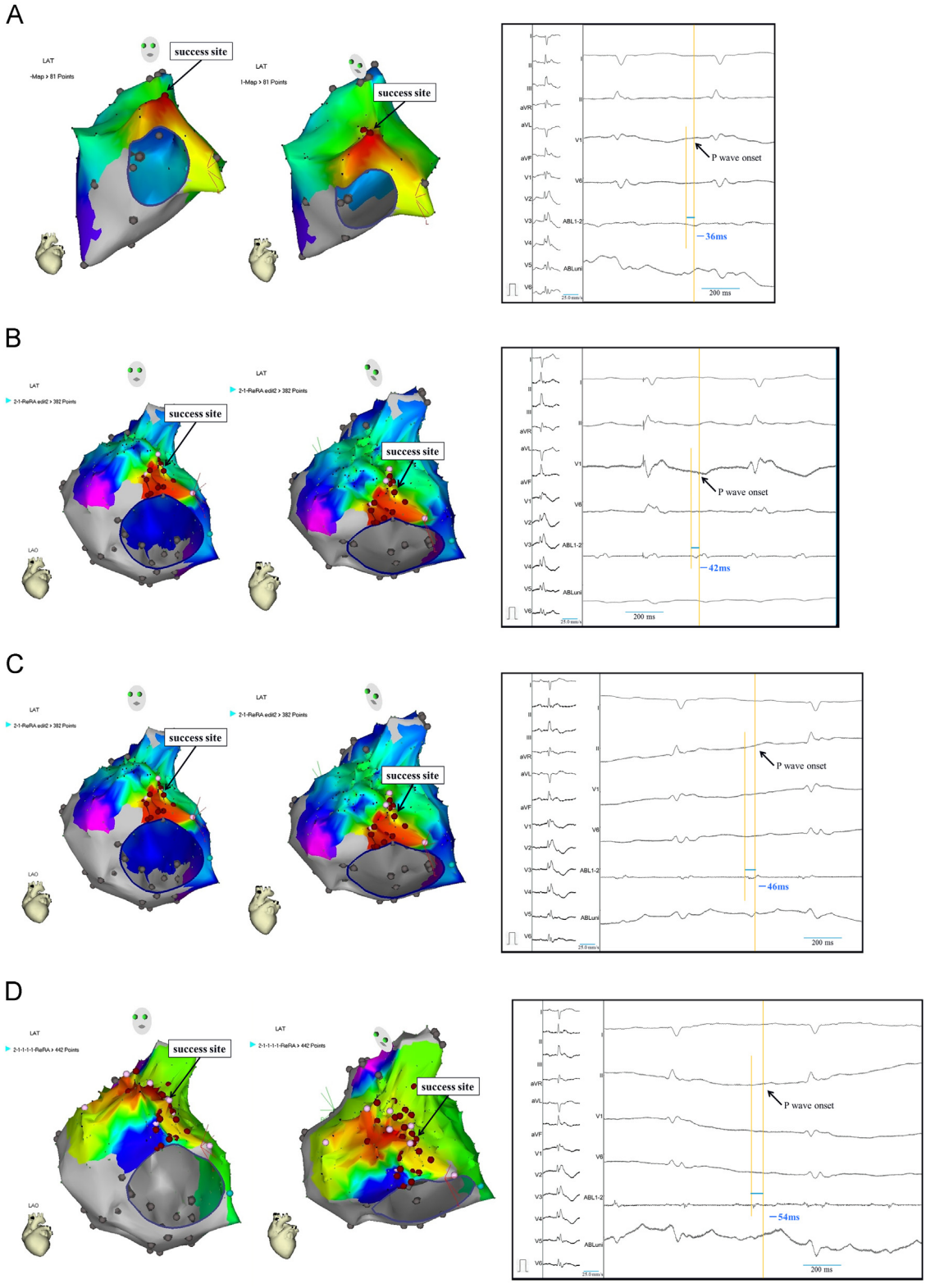

Fig. 3. Activation maps of the right atrium in the left anterior oblique (LAO) and in the cranial LAO. The earliest activation site (red) of atrial tachycardia (AT1) is located at the orifice of the right appendage (A), the tricuspid annulus for AT2 (B), the base of the RAA for AT3 (C), and the site adjacent to the AT1 for AT4 (D). The local intracardiac electrograms at each successful ablation site are shown in the right panels. The red dots represent the points of ablation. The pink dots represent the points of fragment potential. The blue dots represent the points of double potential.

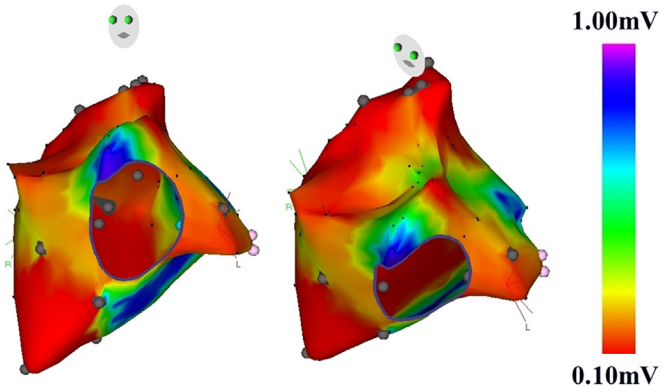

Fig. 4. Voltage map of the right atrium in the left anterior oblique (LAO) and in the cranial LAO. Red indicates areas of amplitude=0.1 mV; purple, amplitude >1.00 mV.

Conflict of interest statement

None.

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