INTRODUCTION

Arrhythmogenic cardiomyopathy (AC) is an uncommon disease characterized by progressive myocardial dystrophy with fibro-fatty replacement [1]. An under-recognized form of AC that predominantly presents with left ventricle (LV) involvement is frequently misdiagnosed with other cardiomyopathies with an enlarged LV and symptoms of heart failure. Late gadolinium enhancement (LGE) acquired by cardiovascular magnetic resonance (CMR) imaging can detect myocardial fibrosis and provide useful information for differential diagnosis, especially in cases with similar imaging manifestations such as dilated cardiomyopathy (DCM) [1,2].

CASE REPORT

A 35-year-old male was admitted with a 7-year history of exertional dyspnea and abdominal distention. His older brother, male cousin, and his mother's younger brother died of cardiomegaly and heart failure when they were young. Electrocardiogram showed right ventricular high voltage. Chest X-ray showed cardiomegaly with pulmonary venous congestion, and the cardiothoracic ratio was 59%. Holter monitoring revealed frequent premature ventricular contractions (PVCs) originating in the right ventricle (RV) and there were 2195 PVCs/24 h. Coronary arterial disease was excluded with CT coronary angiography. CMR imaging demonstrated a significantly dilated LV with severely reduced global systolic function (the normalized LV end-diastolic volume was 206 mL/m², and the LV ejection fraction was 13%). The RV was mildly dilated with moderate systolic dysfunction. The LV lateral wall appeared thin and akinetic (2 mm thickness, Fig. 1A and B). Transmural LGE was detected in the entire LV lateral wall and diffusely distributed focal LGE was demonstrated in the interventricular septum (Fig. 1C and D).

After the failure of guideline-based heart failure therapy, the patient received cardiac transplantation. Masson staining showed myocardial fibro-fatty replacement in the entire lateral wall and focally in the subepicardium of other LV walls as well as the RV free wall (Fig. 2). A diagnosis of left-dominant arrhythmogenic right ventricular cardiomyopathy was made.

Key words: Cardiomyopathy · MRI · Fibrosis · Arrhythmia.
cardiomyopathy (LDAC) was made. The patient was on regular anti-rejection treatment and lived well at the 4-year follow-up.

Publication of the present case and images has been approved by the Institutional Review Board at our hospital and the informed consent has been obtained from the patient.

**DISCUSSION**

AC is one of the most common forms of arrhythmogenic-inherited cardiomyopathy and is characterized pathologically by myocardial fibro-fatty replacement, which predisposes patients to life-threatening ventricular arrhythmias [3]. Although AC has a low incidence, it is a common cause of sudden death in the young and in athletes [3]. In the current case, the patient had a family history of sudden death, akinetic wall motion, and enlarged ventricular volume. These manifestations are considered as major criteria in the task force criteria (TFC) modified in 2010. However, TFC only apply to the right ventricular form of AC and provide limited information for the diagnosis of LDAC. Sen-Chowdhry et al. [1] established the clinical diagnostic features of LDAC as lateral or inferolateral T-wave inversion on electrocardiogram; right bundle branch block/polymorphic ventricular arrhythmias; LV aneurysms on imaging study; LV dilation with or without systolic impairment; and LGE in the LV free wall. Based on these criteria, most of the clinical presentation and imaging manifestations in the present case suggested the diagnosis of LDAC; however, it still needs to be differentiated from DCM because these two diseases share similar clinical features. In this regard, enhanced CMR can be a promising tool. In DCM, LGE is more likely to present with longitudinal striae located in the septum as midwall enhancement [4], while in LDAC, LGE is typically found in the lateral and inferolateral wall.

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**Fig. 1.** CMR images of the patient. CMR images showing that the LV was severely dilated and the LV lateral wall appeared thin on dark-blood sequence in transverse view (A) and in 4-chamber view cardiac cine imaging (B). LGE was detected in the entire LV lateral wall and diffusely distributed focal LGE was demonstrated in the interventricular septum in short axis and 4-chamber views (C and D). LV: left ventricle, LGE: late gadolinium enhancement, CMR: cardiovascular magnetic resonance.
in a subepicardial pattern [1,5]. In addition, LGE more frequently presents in patients with LDAC. In Sen-Choudhry’s study with 42 LDAC cases, LGE was detected in 40 patients [1], whereas in DCM patients, only 30–40% of the patients had LGE [6,7]. It should be noted that LDAC needs to be differentiated from other cardiomyopathies with similar magnification as well. In cardiac amyloidosis, patients usually have a thickened ventricular wall and the extracellular volume calculated by T1 mapping can be elevated [8]. In addition, LGE in cardiac sarcoidosis patients is prone to be located in the septum in a non-ischemic pattern [9]. Evidence of extracardiac involvement in these two diseases can be helpful in the differential diagnosis with LDAC.

This is a typical case of LDAC confirmed by pathology, which presented with predominant heart failure and LV impairments that led to a misdiagnosis with DCM. As contrast-enhanced CMR is highly specific for evaluation of myocardial tissue characteristics, LDAC should be differentiated in patients with cardiomegaly and an unusual scar distribution [1,10]. Because there are few randomized trials that compare the performance of LDAC treatment options, management of LDAC is largely based on clinical judgment. In the advanced stage, heart transplantation may be the ideal and only option.

Conflicts of Interest
The authors have no potential conflicts of interest to disclose.

Acknowledgments
None.
Left-Dominant Arrhythmogenic Cardiomyopathy

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