

An Autopsy Case of Arrhythmogenic Right Ventricular Cardiomyopathy – Radiological/Pathological Comparison –

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A 40-year-old female patient was admitted to the Tokyo Women's Medical University Hospital due to heart failure (HF) and life-threatening arrhythmia. Cardiovascular magnetic resonance (CMR) findings revealed biventricular dilatation; furthermore, late gadolinium enhancement (LGE) imaging in CMR showed diffuse pericardial LGE superior to the left ventricular (LV) wall (**Figure A**). In contrast, electrocardiogram-gated computed tomography (CT) images revealed fatty tissue in the right ventricular (RV) wall (**Figure B**). Arrhythmogenic RV cardiomyopathy (ARVC) was diagnosed based on the criteria.¹ Despite outpatient care and placement of an implantable cardioverter defibrillator, the patient died from HF 6 years later. The postmortem evaluation revealed epicardial fatty tissue and biventricular enlargement (**Figure D**). Transmural myocardial loss and fibro-fatty replacement in the RV wall, diffuse interstitial fibrosis within the LV wall, and minor LV pericardial-side fatty infiltration were

detected histopathologically (**Figure E–H**). Archived CMR and CT imaging data revealed that an RV lesion appearing as an area of low CT density or positive LGE-CMR was identified as fatty tissue (**Figure A–C**); however, a positive LGE-CMR LV lesion was characterized as interstitial fibrosis. Based on the pathological findings, ARVC was suspected, but dilated cardiomyopathy could not be ruled out. This case demonstrates the value of CT-based imaging, and highlights the need for plausible diagnosis applying radiologic/pathologic relationships in ARVC.

Disclosures

N.H. is a member of *Circulation Journal* Editorial Team.

Reference

1. Marcus FI, McKenna WJ, Sherrill D, Basso C, Bauce B, Bluemke DA, et al. Diagnosis of arrhythmogenic right ventricular cardiomyopathy/dysplasia. *Circulation* 2010; **121**: 1533–1541.

Received March 11, 2020; accepted March 13, 2020; J-STAGE Advance Publication released online April 9, 2020 Time for primary review: 2 days

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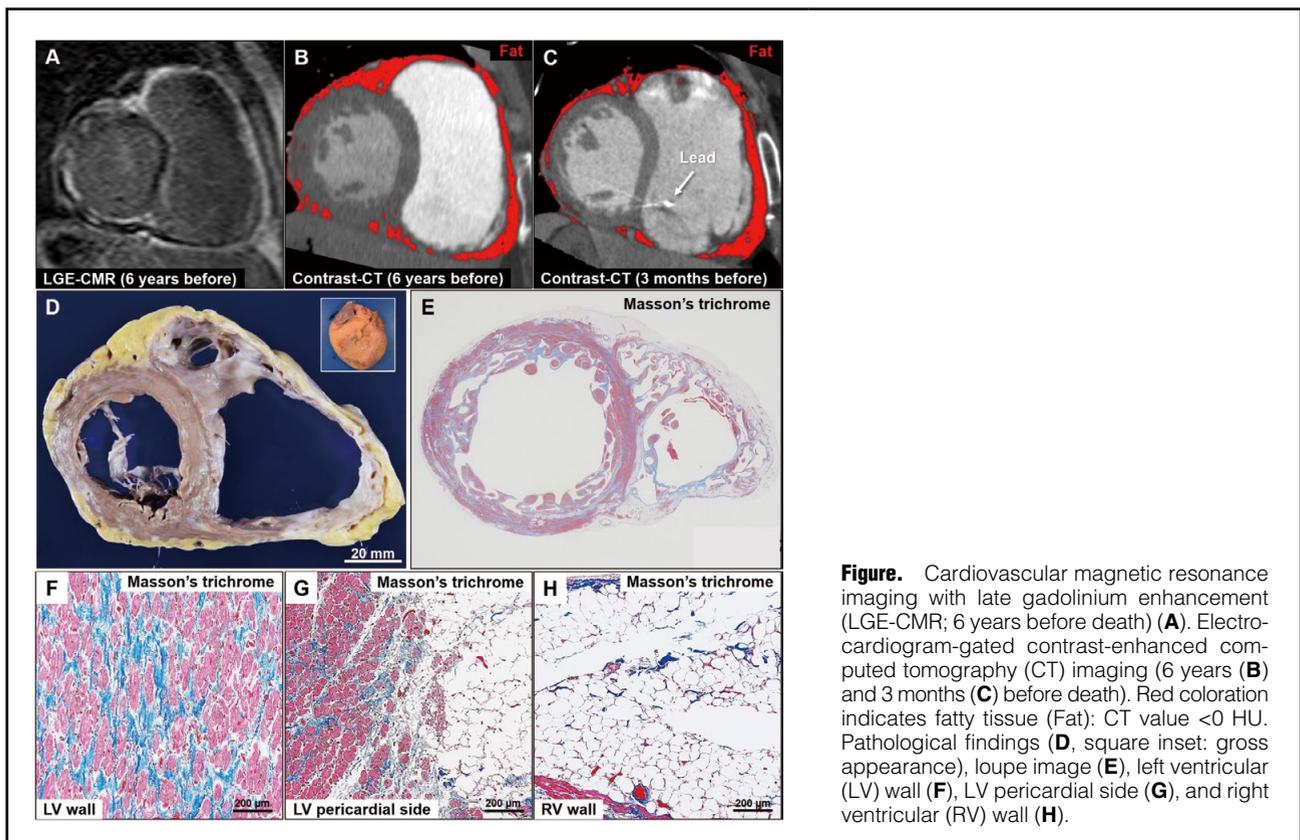


Figure. Cardiovascular magnetic resonance imaging with late gadolinium enhancement (LGE-CMR; 6 years before death) (**A**). Electrocardiogram-gated contrast-enhanced computed tomography (CT) imaging (6 years (**B**) and 3 months (**C**) before death). Red coloration indicates fatty tissue (Fat); CT value <0 HU. Pathological findings (**D**, square inset: gross appearance), loupe image (**E**), left ventricular (LV) wall (**F**), LV pericardial side (**G**), and right ventricular (RV) wall (**H**).