Arrhythmogenic cardiomyopathy suspected by electrocardiogram: confirmed by angiography

Ardan M. Saguner¹, Laurent M. Haegeli¹, Christian Templin¹, Thomas Wolber¹, Ulf Landmesser¹,², and Firat Duru¹,²*

¹Clinic for Cardiology, Cardiovascular Center, University Hospital Zurich, Ramistrasse 100, CH-8091 Zurich, Switzerland; and ²Center for Integrative Human Physiology, University of Zurich, Zurich, Switzerland

* Corresponding author. Tel: +41 44 2553565, Fax: +41 44 2554401, Email: firat.duru@usz.ch

This paper was guest edited by Prof. Frank Rademakers, University Hospital Gasthuisberg, Leuven, Belgium

A 58-year-old woman was hospitalized for presyncope. The 12-lead surface electrocardiogram (ECG) in the emergency room showed a sustained ventricular tachycardia (VT) at a rate of 143 b.p.m. with a left bundle branch block (LBBB) morphology and inferior axis (Panel A). The ECG was remarkable for pronounced notching of the QRS complexes across multiple leads. Idiopathic right ventricular (RV) outflow tract tachycardia (RVOT-VT) was initially suspected. Intravenous metoprolol, verapamil, and amiodarone were administered without success. Electrical cardioversion (150 J biphasic) converted the arrhythmia into sinus rhythm at a rate of 57 b.p.m. with first-degree atrioventricular block and late potentials suggesting epsilon waves as well as ST-T wave changes in various leads (Panel B). Transthoracic echocardiography revealed regional wall motion abnormalities, a dilated RVOT (22.7 mm/m² at parasternal long axis), and a reduced RV fractional area change (18%) with extensive left ventricular (LV) involvement (ejection fraction biplane 26%) (Panel C; see Supplementary material online, Movie S1). Angiography showed a reduced global RV function, RV dilatation, and regional wall motion abnormalities (dyskinesis and aneurysm of the inferior RV wall and the apex) (Panel D, arrows; Supplementary material online, Movie S2). The pile d’assiettes sign—a pathognomonic angiographic sign for advanced RV involvement—was visible (Panel D, arrowhead). The diagnosis of arrhythmogenic RV cardiomyopathy (ARVC) was made on the basis of two major (epsilon wave and RV aneurysm) and one minor criterion (LBBB-VT with inferior axis) according to the Revised Task Force Criteria. On surface ECG, the presence of multiple QRS notches across several leads during VT with LBBB morphology and inferior axis represents a conduction delay due to fibro-fatty replacement and is highly suggestive for the presence of ARVC when compared with idiopathic RVOT-VT.

Supplementary material is available at European Heart Journal online.

Published on behalf of the European Society of Cardiology. All rights reserved. © The Author 2012. For permissions please email: journals.permissions@oup.com