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Arrhythmogenic Cardiomyopathy

Suspected by ECG - Confirmed by Angiography

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Conflict of interest: none declared.
A 58-year-old woman was hospitalized for presyncope. The 12-lead surface ECG in the emergency room showed a sustained ventricular tachycardia (VT) at a rate of 143 bpm 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 Joules biphasic) converted the arrhythmia into sinus rhythm at a rate of 57 bpm 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 (22.7mm/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; Supplementary material online, Movie 1). 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 2). The pile d’assiettes sign - a pathognomonic angiographic sign for advanced RV involvement- was visible (Panel D, arrowhead). The diagnosis of arrhythmogenic right ventricular 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 conduction delay due to fibro-fatty replacement and is highly suggestive for the presence of ARVC as compared to idiopathic RVOT-VT.