Multimodal imaging-guided endomyocardial delivery


Isolated left ventricular arrhythmogenic dysplasia

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A 37-year-old man with history of tobacco use presented to a hospital reporting several episodes of syncope preceded by palpitations. At admission, he was hypotensive (80/50 mmHg), normocardic (80 bpm), and diaphoretic. The 12-lead electrocardiogram showed sinus rhythm with epsilon waves in II, III, and aVF (arrows, Panel A) and T-wave inversion in inferolateral leads. Blood analyses revealed discrete troponin I elevation (0.29 ng/mL) and normal D-dimer. Echocardiogram evidenced mild dilatation of left cardiac chambers and moderate left ventricular (LV) systolic dysfunction with diffuse hyperechogenic regions. Myocardial perfusion scan was negative for myocardial ischaemia. Cardiac MRI showed LV systolic dysfunction (ejection fraction of 41%) with subepicardial fat deposits in LV lateral wall apical segment (arrows in Panels B and C, which represent, respectively, T1-weighted images without and with fat suppression in four-chamber plane) and extensive LV intramural and subepicardial fibrosis on late-enhancement images (arrows, Panel D), suggesting isolated LV arrhythmogenic dysplasia. Pharmacological therapy with β-blocker and ACE-inhibitor was initiated. To document the aetiology of syncope, an electrophysiology study was performed with induction of monomorphic ventricular tachycardia with LV apex origin (corresponding to the area of dysplasia). An implantable cardioverter-defibrillator was implanted and during follow-up (6 months) the patient had some episodes of ventricular tachycardia successfully treated with anti-tachycardia pacing.

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