Sudden cardiac death due to acute right heart failure in a patient with arrhythmogenic right ventricular dysplasia and acute myocardial infarction

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A 40-year-old patient with toothache and shoulder pain for 10 days died without former known history of cardiac arrhythmias or myocardial infarction (MI) on sleep-related sudden cardiac death (SCD). The autopsy did not reveal signs for traumatic violence or intoxications nor for acute/chronic infections particularly of the upper aerodigestive tract. Furthermore, no aspects of endo-/myocarditis including negative results for cardiotropic virus were evident.

In contrast, the right ventricular (RV) myocardium showed a diffuse replacement by fatty tissue (>70%) and dystrophy of cardiomycocytes (Panel A, arrows indicate endocardium), corresponding to an arrhythmogenic RV dysplasia (ARVD) with immunohistochemical up-regulation of the cytoskeleton protein desmin (Panel A, inset) and electronmicroscopic detection of abnormal desmosomes (Panel B, arrowheads and inset). Moreover, the proximal right coronary arteria showed an isolated high-grade stenosis (~80%) with acute subtotal thrombotic obliteration (Panel C, inset histological image) resulting in acute MIs of subendocardial RV myocardium (Panel D, hyper eosinophilic cardiomyocytes without nuclei). Since any signs of backward failure of the left ventricle such as lung oedema were lacking, we diagnosed an SCD due to acute RV failure in a patient with ARVD and acute MI.

ARVD is behind hypertrophic concentric cardiomyopathy the major non-ischaemic cause for SCD in association with sports activity in young patients (age <40 years). Here, we describe, to our knowledge, the first case of an ARVD in combination with coronary heart disease/MIs, which might have triggered acute ventricular arrhythmias/failure independently from physical exertion and should be therefore considered for in vivo/autopsy investigations in family members of ARVD patients.

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