Acute episode of an arrhythmogenic right ventricular cardiomyopathy with vast necroses exclusively in right ventricular myocardium

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We describe the case of an athlete aged 18 years who died of sudden cardiac arrest without previously having been diagnosed with heart disease. The autopsy did not reveal signs of intoxication or endo-/myocarditis, including negative results for cardiotropic virus, and coronary heart disease/vasculitis was excluded by coronary angiography and histology.

In contrast, cardiac morphology showed classical signs of arrhythmogenic right ventricular cardiomyopathy (ARVC) with diffuse replacement of RV myocardium by fibro-fatty tissue (Panel A), detection of abnormal long desmosomes (Panel A, inset) and immunohistochemical lack of plakoglobin1 (Panel B) but strong expression of N-cadherin (Panel B, inset). More interestingly and exclusively in RV myocardium but not in left ventricle nor both atrias, large areas of acute/subacute cardiomyocyte necroses were detected (Panels C and D) with mild inflammatory infiltrates (Panel C), myocytolysis with loss of myofibrils2 (Panel C, inset) and myocardial contraction bands (Panel D) with hypercontracted sarcomeres (Panel D, inset).

Although death of single myocytes has been reported in ARVC,3 the vast necroses in RV myocardium shown here with an increase of the MB isoform of creatine kinase (CK-MB 85U/l; CK 530 U/l) detected in a blood sample collected immediately after start of reanimation have so far not been described and might be the morphological correlate of an acute episode of ARVC. In our opinion, these data confirm the ‘degenerative hypothesis’4 suggesting that the replacement of the RV myocardium is progressive with time starting from the epicardium and expanding transmurally to the endocardium3 and underline the relevance of markers as plakoglobin in preventing such fatal courses of ARVC.

Funding
Funding to pay open access publication charges for this article was provided by the Wilhelm-Sander-Stiftung (grant 2007.068.01) and the Deutsche Forschungs-Gesellschaft (DFG-SFB-Transregio 52, TPA8).

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