A 78-year-old man experienced a first-time syncpe with falling resulting in raccoon eyes and a single episode of non-sustained ventricular tachycardia (>10 beats) was documented. Creatine kinase and troponin T were only weakly abnormal. Echocardiography and invasive coronary angiography were unremarkable.

Cardiac magnetic resonance (CMR) imaging revealed the presence of intramyocardial fatty spots and streaks of the left ventricular (LV) inferolateral wall and the interventricular septum (arrowheads in Panel A, steady-state free precession (SSFP) imaging; Panels B and C, T2-weighted black blood imaging without and with fat suppression; white arrows in Panel D, delayed enhancement imaging with diffuse and streaky intramyocardial signal enhancement; see Supplementary material online, Movie 1). Right ventricular morphology and regional and global function were normal. Lipomatous metaplasia of chronic myocardial infarction was ruled out by missing regional subendocardial delayed enhancement. Hence, CMR findings were suggestive of isolated arrhythmogenic dysplasia of the LV.

Histopathological work-up including transmission electron microscopy of a deep LV biopsy demonstrated disrupted myocardial architecture with extensive fibrofatty replacement [Panels E and F, haematoxylin and eosin staining; Panels G and H, transmission electron microscopy, white arrows indicate cardiomyocytes with several cytoplasmatic vacuoles separated by a collagen rich matrix (asterisk); arrowheads, nuclei of fibrocytes; black-lined white arrows, lipid droplets; rectangle in G delineates the magnified image area in Panel H].

A ‘left-dominant’ subtype of arrhythmogenic right ventricular cardiomyopathy (ARVC) has been recognized with multicentre studies reporting LV involvement in ARVC to occur with disease progression in >75% of cases. However, primary LV involvement has been acknowledged as a phenotype expression of ARVC only recently.

Supplementary material: Supplementary material is available at European Heart Journal online.