An adolescent with laminopathy presenting as ventricular tachycardia and left ventricular apical aneurysm

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A 17-year-old girl was admitted because of recurrent ventricular tachycardia after sudden cardiac arrest (Panel A). Two-dimensional and strain echocardiography revealed a dilated left ventricle (LV) with apical aneurysm and dyskinesia (Panels B and C, and see Supplementary data online, Video S1). The normal coronary artery was confirmed under coronary angiography (Panel D, and see Supplementary data online, Videos S2 and S3). Cardiac magnetic resonance imaging revealed apical aneurysmal dilatation and wall thinning with thrombus in the LV (Panel E and see Supplementary data online, Video S4). Subendocardial and transmural late gadolinium enhancement in the LV apical and lateral walls was evident (Panel F, arrow heads). An implantable cardioverter-defibrillator was applied in addition to amiodarone, enalapril, carvedilol, and warfarin therapies.

The patient complained of a slowly progressive lower extremity weakness over the past 10 years. She showed a waddling gait, Gower’s sign, and Achilles tendon contracture. Under the impression of cardiomyopathy associated with muscular dystrophy, we performed genetic testing, which revealed a mutation (c.1621C>T [p.Arg541Ser]) in the LMNA gene.

This is an unusual case of laminopathy presenting as ventricular tachycardia associated with localized myocardial fibrosis and aneurysm.

Supplementary data are available at European Heart Journal — Cardiovascular Imaging online.