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Multimodality imaging in a case of arrhythmogenic left ventricular cardiomyopathy: a case report

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Case description

A 17-year-old girl with no prior medical or family history presented with palpitations. There was no abnormality on clinical examination. Trans-thoracic echocardiography was normal. Continuous electrocardiogram Holter monitoring showed frequent ventricular ectopic beats. A 12-lead electrocardiogram showed small R waves in V1 and V2 suggested the origin was the LVOT rather than the right (*Figure 1*).

She went on to have cardiac magnetic resonance imaging (CMR). This showed mildly impaired left ventricular function (left ventricular ejection fraction 48%, EDV 71 mL/m^2, ESV 37 mL/m^2). There was

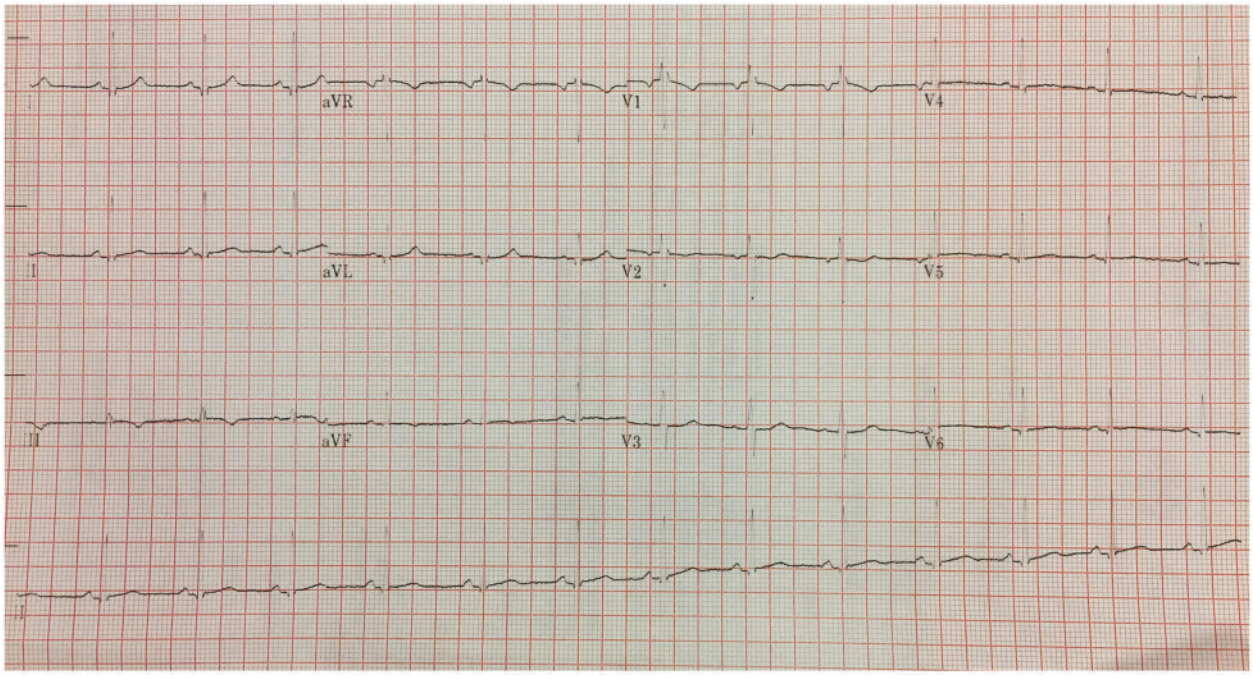

Figure 1 A 12 lead electrocardiogram showing small R waves in V1 and V2.

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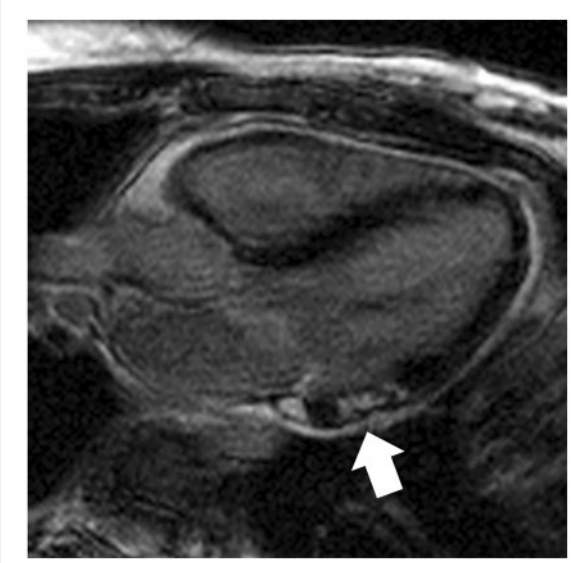

Figure 2 Three-chamber late gadolinium enhancement magnitude image demonstrating mid to epicardial increased signal return from the basal inferolateral segment.

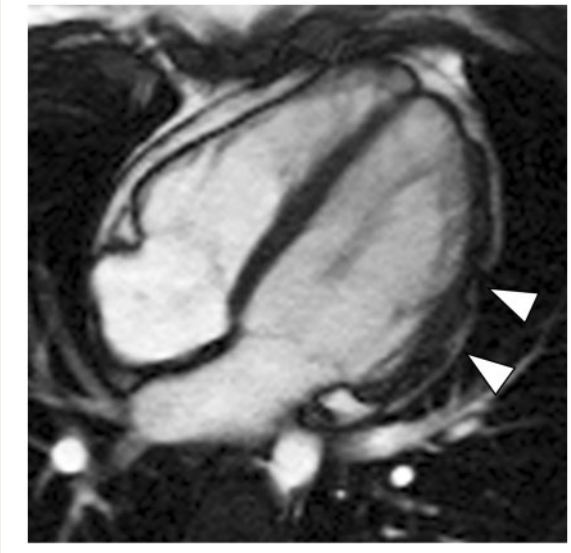

Figure 3 Four-chamber steady state free precession cine image at end-diastole demonstrating lobulated contour of the anterolateral segments.

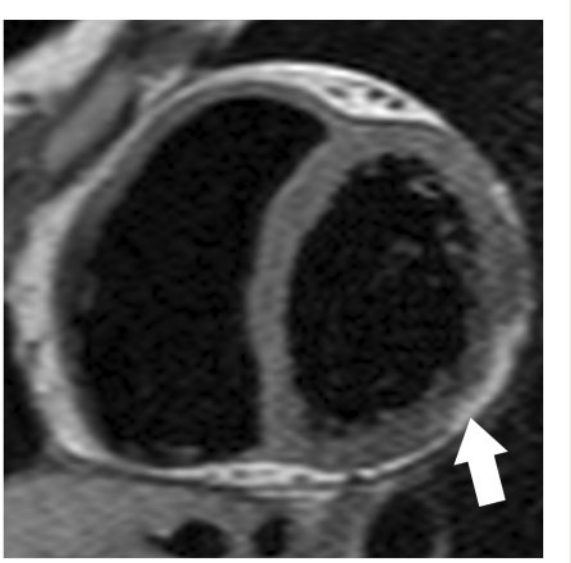

Figure 4 Mid left ventricular short-axis turbo-spin echo T1 weighted image at end-diastole demonstrating high signal in the anterolateral and inferolateral segments.

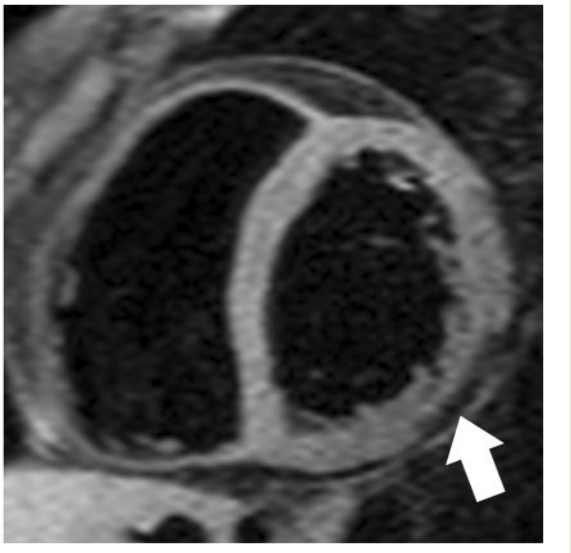

Figure 5 Mid left ventricular short-axis turbo-spin echo T1 weighted image with fat saturation at end-diastole at the same slice position as *Figure 3* demonstrating homogenous signal drop out in the epicardial region of the anterolateral and inferolateral segment consistent with foci of fat.

hypokinesia and thinning of the lateral left ventricular wall. There was evidence of patchy epicardial late gadolinium enhancement in the lateral left ventricular wall (*Figure 2*). The right ventricle was normal in size and function (right ventricular ejection fraction 40%, EDV 70 mL/m^2, ESV 42 mL/m^2). Follow-up CMR performed 1 year later showed signal abnormality in the left ventricular lateral wall, which was hyper-intense to myocardium on steady-state free precession images, iso-intense to fat on T1-weighted sequences and showed homogenous signal drop out with fat suppression techniques (*Figures 3–5*). This favoured fibro-fatty replacement secondary to arrhythmogenic left ventricular cardiomyopathy (ALVC) over prior myocarditis.[1,2] Fibrous metaplasia was also considered unlikely in a

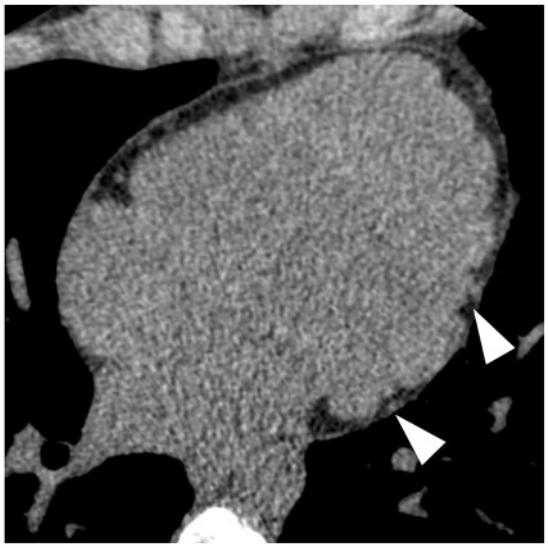

Figure 6 End-diastolic unenhanced electrocardiogram gated cardiac computed tomography image reconstructed in the four-chamber plane demonstrating foci of fat attenuation in the left anterolateral wall (Hounsfield units ranging from -76 to -132).

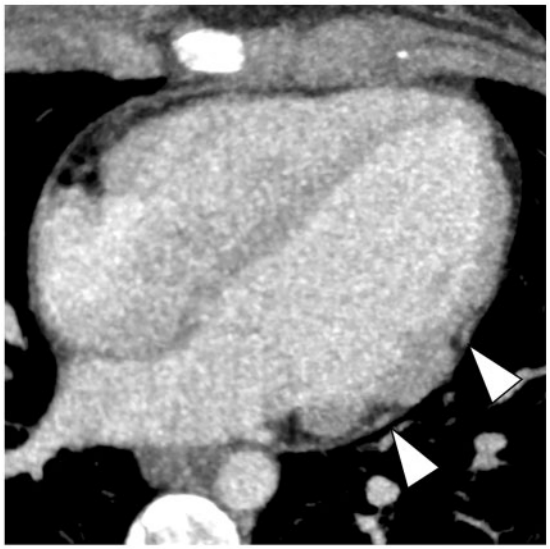

Figure 7 End-diastolic electrocardiogram gated cardiac computed tomography image reconstructed in the four-chamber plane post-contrast with 50-s delay demonstrating foci of fat attenuation that is discrete to the enhancing myocardium in the left ventricular anterolateral wall.

young patient without evidence of prior myocardial infarction and without ischaemic heart disease risk factors.

Unenhanced and delayed phase contrast cardiac computed tomography imaging confirmed an abnormal lobular contour of the left ventricular wall with multiple areas of focal epicardial thinning. The thin walled sections were invaginated by epicardial fat (*Figures 6* and *7*). The right ventricle again was normal.

Arrhythmogenic left ventricular cardiomyopathy without right ventricular involvement is a rare, but increasingly recognised condition, characterised by fibro-fatty replacement in the left ventricle.[3,4] It is closely related to the more widely recognized arrhythmogenic right ventricular cardiomyopathy (ARVC). The two disease patterns can co-exist within the same family and genes implicated in ARVC have also been identified in patients with ALVC.[3] The major complication of arrhythmogenic cardiomyopathies is sudden cardiac death due to malignant arrhythmia. Clinical heart failure is rarely seen.[3]

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

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Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

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