A 48-year-old man presented with syncope due to tachycardia of left ventricular origin and was electrically cardioverted to sinus rhythm. There was no medical history of cardiac disease or family history of sudden cardiac death. Resting electrocardiography displayed epsilon waves (Figure, A). Coronary angiography revealed unobstructed arteries. Transthoracic echocardiography (Movies I and II) showed normal left ventricular dimensions with mild systolic dysfunction. The right ventricle was mildly dilated, with reduced systolic function but no regional wall-motion abnormalities or aneurysms. The right ventricular outflow tract measured 41 mm.

Cardiac magnetic resonance (CMR) imaging demonstrated a dilated right ventricle and biventricular systolic dysfunction (Table; Movies III and IV). Short-axis imaging at the midventricular level after injection of gadolinium at the midventricular level (Figure, B) identified patchy late enhancement of the right ventricular border of the septum, which extended into the subepicardium of the anterior and inferior walls. A 2-chamber view of the left ventricle (Figure, C) demonstrated extensive midmyocardial and subepicardial enhancement. In addition, short TI inversion recovery sequences (Figure, D and E) revealed fatty infiltration of the right ventricle in the absence of myocardial edema. The CMR differential diagnosis included chronic myocarditis and arrhythmogenic right ventricular cardiomyopathy (ARVC). Further investigation confirmed a mutation in the desmoplakin gene, which supports a diagnosis of this variant of ARVC.

The features presented fulfill the recently modified Task Force Criteria for the diagnosis of ARVC, produced with the aim of increasing the sensitivity of ARVC diagnosis. However, diagnostic doubt was raised because the CMR findings shown are typical of chronic myocarditis, with severe biventricular impairment and extensive diffuse midmyocardial and subepicardial late gadolinium enhancement, particularly of the left ventricle. Of note, CMR findings of late gadolinium enhancement are not included in the recently modified diagnostic criteria for ARVC, perhaps reflecting the difficulty in reliably determining pathological right ventricular enhancement patterns and underrepresentation of the left ventricular dominant subtype in clinical case series.

An explanation for these CMR appearances is offered by the suggestion that diffuse myocardial inflammation is part of the ARVC disease process. An association between myocardial inflammation and lipomatous infiltration in ARVC has been previously described.

Figure. A. Epsilon waves. B. Short-axis image at midventricular level. Late enhancement of the right ventricular border of the septum is seen, extending into epicardium of the anterior and inferior walls. Further late enhancement is seen throughout the right ventricular epicardium. C. Two-chamber view of the left ventricle. Extensive patchy enhancement was seen throughout the left ventricular mid wall and epicardium. D-E. Four-chamber and midventricular short-axis short TI inversion recovery sequences demonstrating fat suppression in the right ventricular myocardium.
ditis and ARVC has been postulated previously, supported by the frequent demonstration of myocardial inflammation in postmortem histological analysis together with the presence of cardiotropic viruses in diseased hearts. However, the precise link between the 2 conditions has yet to be determined; these findings may indicate an inflammatory process related to progression of the disease itself or an increased susceptibility to myocardial infection due to apoptosis and cell debris within the diseased myocardium.

In the present case, the demonstration of CMR features typical of chronic myocarditis in a patient with ARVC confirms the close relationship between these 2 conditions, with extensive involvement of the left ventricle characteristic of the desmoplakin variant of ARVC. Because images were taken during a period of intermittent arrhythmia, the CMR appearance may illustrate an “active myocarditic” phase of the ARVC disease process.

Disclosures
None.

References

Table. Ventricular Wall Dimensions From CMR Imaging

<table>
<thead>
<tr>
<th></th>
<th>EDV, mL</th>
<th>ESV, mL</th>
<th>SV, mL</th>
<th>EF, %</th>
<th>Mass Index, g/m²</th>
</tr>
</thead>
<tbody>
<tr>
<td>LV</td>
<td>158</td>
<td>83</td>
<td>75</td>
<td>49</td>
<td>60</td>
</tr>
<tr>
<td>RV</td>
<td>195 (88–227)</td>
<td>118 (23–103)</td>
<td>77 (52–138)</td>
<td>39 (47–74)</td>
<td>. . .</td>
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</table>

EDV indicates end-diastolic volume; ESV, end-systolic volume; SV, stroke volume; EF, ejection fraction; LV, left ventricle; and RV, right ventricle.

Normal ranges for right ventricular volumes are given in parentheses.
Myocarditic Appearance of Arrhythmogenic Right Ventricular Cardiomyopathy
Andrew N. Jordan, Jonathan Lyne, Ranil De Silva and Tom Wong

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