Unusual Clinical Presentation of a Patient With an Extreme Form of Right Ventricular Dysplasia

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A previously healthy 42-year-old woman presented with recurrent reversible neurological deficits. On admission, the surface ECG showed a very low voltage QRS pattern along with prominent P-waves (Figure 1). Computerized tomography of the brain was normal. Transthoracic echocardiography showed massive dilatation and extensive dyskinesia of the right ventricle and evidence of fatty deposits in the myocardial structure, suggesting a diagnosis of right ventricular dysplasia. The patient had a patent foramen ovale, which could have caused paradoxical emboli to the brain. A coronary angiogram was normal. Right ventriculography showed sacculations that are typically observed in patients with right ventricular dysplasia. Left ventriculography showed normal myocardial function. In electrophysiological testing, no arrhythmias could be induced by programmed ventricular stimulation. MRI showed fatty replacement of right ventricular myocardium (Figure 2). In addition, there was massive dilatation of the right heart chambers, which essentially encircled the left heart chambers (Figures 2 and 3). Because the left ventricle makes the most significant contribution to QRS forces, these findings would explain the discrepancy between the markedly attenuated QRS complexes and the prominent P-waves, a rarely observed ECG pattern in clinical practice.

Figure 1. A 12-lead ECG showing low-voltage QRS complexes and prominent P-waves.

Figure 2. Short-axis gradient-echo MRI demonstrating a normal-sized left ventricle (arrowhead) covered by an extremely dilated right ventricle with fatty infiltration (arrow).
Figure 3. An axial T1-weighted spin-echo sequence of the heart showing massive dilatation of the right atrium (arrow) and the right ventricle (arrowhead).
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