Arrhythmogenic right ventricular cardiomyopathy with biventricular involvement and heart failure in a 9-year old girl

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Arrhythmogenic right ventricular cardiomyopathy (ARVC) is seldom recognized clinically in infancy or under the age of 10. We report a case of a 9-year-old girl with ARVC, who presented with signs and symptoms of heart failure and palpitations. Holter monitoring showed frequent premature ventricular beats and echocardiogram revealed dilated and dysfunctional right ventricle with normal tricuspid valve and no evidence of intracardiac shunt. Cardiac magnetic resonance showed classical features of ARVC with both ventricular involvements. After optimization of medical treatment the patient was referred for ICD implantation.

Keywords: ARVC, CMR, Dilated RV

Introduction

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is an inherited cardiomyopathy, characterized by fibro-fatty replacement of the right ventricular (RV) myocardium. It is associated with cardiac arrhythmias of right ventricular origin, right ventricular dysfunction, heart failure, and sudden cardiac death [1]. Clinically recognized ARVC is unusual in children, probably because the clinical expression of the disease is

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CASE REPORT

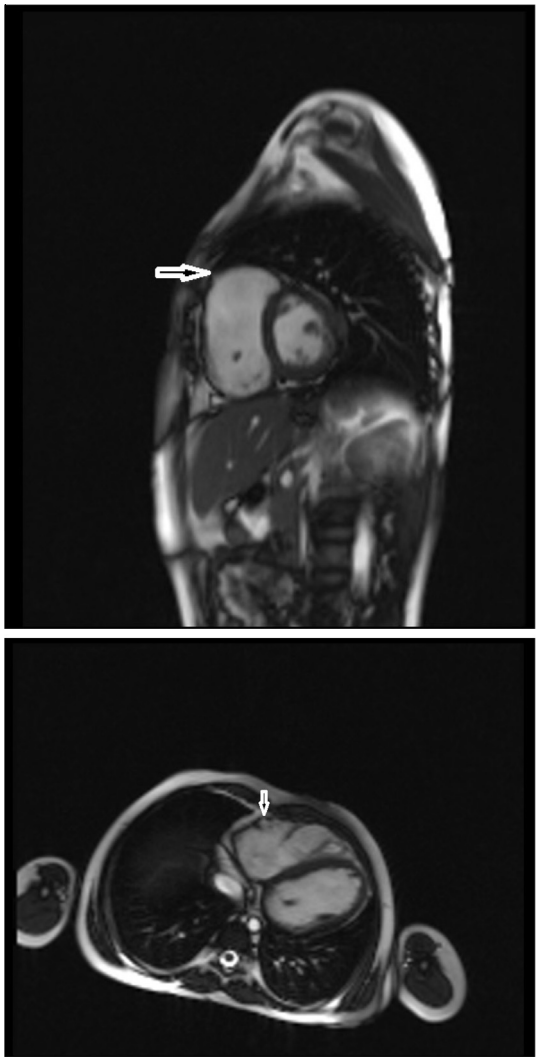

Figure 1. Steady-state free precession still frame, showing focal aneurysmal areas in the right ventricular free wall, indicated by the arrow.

normally postponed to youth and adulthood. There is now an increased awareness about nonclassical phenotypes of ARVC, including left dominant and biventricular types, and its presence in children.

Here, we report a case of a young girl with ARVC, biventricular involvement, and heart failure.

Case report

A 9-year-old girl presented with signs and symptoms of heart failure and palpitations. Electrocardiography showed sinus rhythm with T wave inversion in chest leads. Morphology and duration of QRS complexes in right precordial leads were normal. There was no delay in activation. Holter monitoring showed frequent ventricular premature beats of left bundle branch block morphology. Echocardiographic examination revealed dilated and dysfunctional right ventricle with normally placed tricuspid valve and no evidence of intracardiac shunt. Holter monitoring showed frequent isolated ventricular ectopies but no evidence of sustained ventricular tachycardia. No information was available about the family history, as the child was adopted by the family from an orphan house. Neither parent was alive and the cause of death was not known.

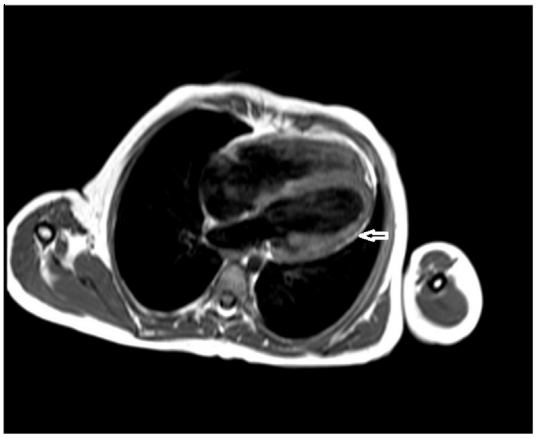

Figure 2. Turbo spin echo T1 weighted image, showing fatty infiltration of the myocardium, indicated by the arrow.

With the clinical and echocardiographic suspicion of ARVC, the child was referred for cardiac magnetic resonance (CMR), which was done on a Siemens Avanto 1.5 T machine (Siemens, Munich, Germany) with ARVC protocol. Image sequences acquired included: black blood T1-weighted turbo spin echo images with and without fat suppression; steady-state free precession or cine images; and late gadolinium images after gadolinium injection.

RV was dilated (end diastolic volume 132 mL) with dys-synchronous contractility and severely reduced systolic function (ejection fraction 26%). RV free wall was thin looking with evidence of regional/focal dyskinesis (Fig. 1). Left ventricular (LV) volumes were normal with wall motion abnormalities involving the inferolateral wall and apex and mildly reduced systolic function (ejection fraction 44%).

There was fatty infiltration on the epicardial surface of RV free wall and also fatty replacement of myocardium in the mid to distal lateral wall of the LV (Fig. 2). On late gadolinium images, diffuse hyperenhancement of the RV free wall and RV side of the interventricular septum was noted. There was also hyperenhancement involving the inferolateral segments and apex of the LV (Fig. 3).

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