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Multidetector Computed Tomography Findings of Arrhythmogenic Right Ventricular Dysplasia: A Case Report

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ABSTRACT

Arrhythmogenic right ventricular dysplasia (ARVD) is a heart muscle disorder characterized pathologically by fatty or fibrofatty replacement and electrical instability of the right ventricular myocardium. This cardiac entity leads to sudden cardiac death, syncope, recurrent ventricular tachycardia, and in some cases, heart failure in a younger population. Contrast angiography, echocardiography, radionuclide angiography, ultrafast computed tomography (CT), and cardiovascular magnetic resonance imaging are techniques used to diagnose functional and morphologic characteristics of the disease. CT is sensitive in detecting intramyocardial fat because of its low attenuation. Recently the advances in multislice CT (MDCT) have improved temporal resolution, which has increased effectiveness in providing morphologic and functional information. We present a case with ARVD evaluated through 16-row MDCT. Fatty infiltration was clearly demonstrated by 16-slice CT; thus, multislice CT may have a significant role in the assessment and follow-up of patients with ARVD.

INTRODUCTION

Arrhythmogenic right ventricular dysplasia (ARVD) is a rare disorder characterized by structural and functional abnormalities of the right ventricle and a propensity for ventricular arrhythmias and sudden death. The diagnosis of ARVD is based on the presence of major and minor criteria encompassing structural, histologic, electrocardiographic, arrhythmic, and genetic factors proposed by the ARVD Task Force in 1994 [Hermida 1997]. ARVD occurs in young adults with a male-to-female ratio of 2.7/1.0, but the true incidence is unknown. This cardiac entity should be suspected in all young patients presenting with syncope, ventricular tachycardia, or cardiac arrest [Kayser 2002]. Eighty percent of reported incidences are diagnosed in patients younger than 40 years [Hermida 1997]. The newest 16-row multidetector computed tomography (MDCT) has provided high temporal resolution and information about morphology. In our case, fatty infiltration of the right ventricular wall was clearly shown by MDCT.

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CASE REPORT

A 35-year-old male patient presented with dyspnea and recurrent syncope. Electrocardiography (ECG) revealed nonsustained ventricular tachycardia with frequent, multifocal premature ventricular beats and inversion of T waves (Figure 1). The patient denied any known family history of cardiomyopathy or sudden death.

Sixteen-slice CT was performed (Aquilion 16; Toshiba Medical Systems, Tokyo, Japan) with ECG gating after an intravenous injection of contrast medium. Axial and sagittal CT images demonstrated wall thinning and low-density areas (–120 HU), indicative of focal fatty infiltration along the right ventricular free wall (Figure 2). However, we could not perform biopsy evaluation of this abnormal ventricular segment to verify the diagnosis because of technical limitations and substantial risk of perforation. ARVD is known to be the only disease showing fatty infiltration along the right ventricular wall; thus, indication of focal fatty infiltration by MDCT provided the diagnosis.

Ventricular tachycardia was induced in electrophysiological study, and in the treatment of the patient, implantable cardioverter-defibrillator therapy was performed. The patient is being followed-up asymptomatically.

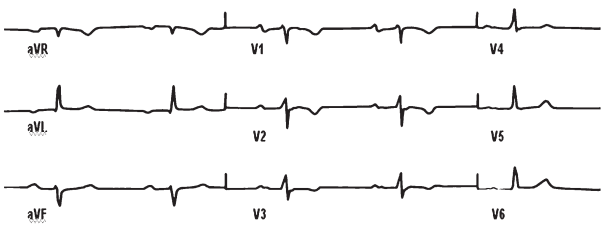
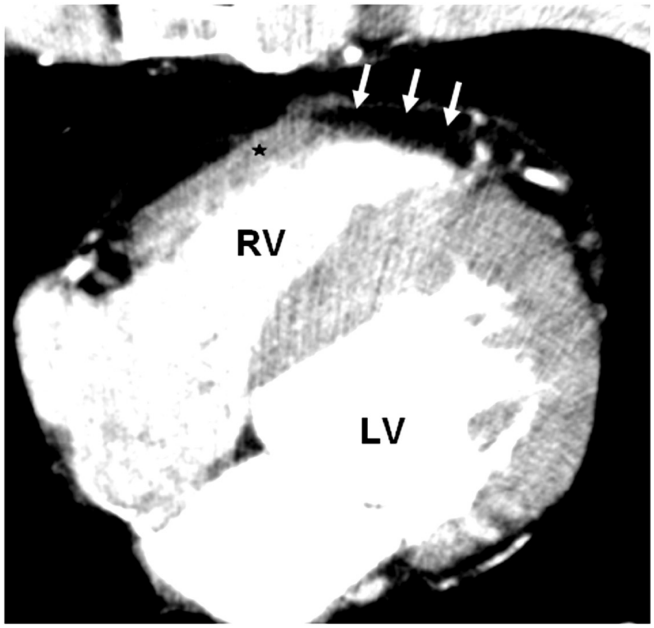


Figure 1. Electrocardiography in sinus rhythm showing inverted T waves in the precordial leads.



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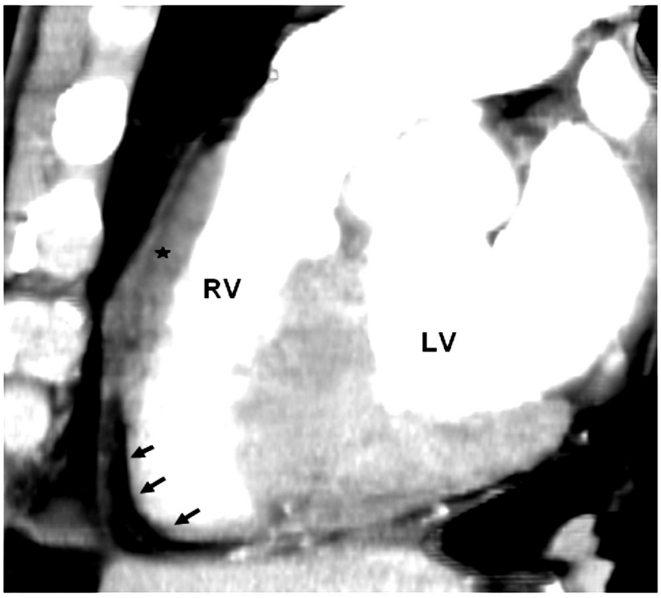

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Figure 2. Axial (A) and sagittal (B) MIP images demonstrate marked wall thinning and fat infiltration along right ventricular free side. Asterisk indicates right ventricular muscular wall; arrows, fat infiltration; LV, left ventricle; RV, right ventricle.

DISCUSSION

ARVD is an uncommon inheritable cardiomyopathy involving predominantly the right ventricle with progressive fibrofatty tissue replacement [Kies 2006]. Electrical, functional, and anatomic abnormalities and family history are used for diagnosis [McKenna 1996]. A family history of ARVD is present in 30% to 50% of cases [Nova 1988].

The most evident morphologic feature of the disease is the diffuse or segmental loss of right ventricular myocytes with replacement by fibrofatty tissue and thinning of the right ventricular wall. Fibrofatty replacement usually begins in the subepicardium or mediomural layers and progresses to the subendocardium. Only the endocardium and the myocardium of the trabeculae may be spared [Peters 2006]. Anatomic malformations of the right ventricle consist of mild-to-severe global right ventricular dilatation, right ventricular aneurysms, and segmental right ventricular hypokinesia [Rampazzo 2002].

One of the main diagnostic features of ARVD is ECG findings in standard and alternative recording techniques. Localized right precordial QRS prolongation, prolongation of right precordial S wave, complete or incomplete right bundle branch block, and inversion of T waves are the ECG findings [Tada 1996].

Antiarrhythmogenic agents, radiofrequency ablation, implantable cardioverter-defibrillator therapy, heart failure treatment, and surgical treatment are therapeutic options in patients with ARVD [Tandri 2004]. In our case, ventricular tachycardia was induced in electrophysiological study, and in the treatment of the patient, implantable cardioverter-defibrillator therapy was performed.

Imaging modalities such as contrast angiography, echocardiography, radioisotope techniques, CT, and cardiovascular magnetic resonance imaging are used to establish diagnosis of ARVD [van der Wall 2000]. Right ventricular contrast angiography is usually regarded as the reference standard for the diagnosis [Wu 2007]. Akinesis/dyskinetic bulging and the presence of hypertrophic trabeculae are findings of angiography. Its invasive nature, x-ray exposure, and ventricular extrasystoles, which may occur during contrast injection, are the limitations of the technique [Yamamuro 2005].

Echocardiography is used in assessing patients with widespread ARVD. Right ventricular dilatation, enlargement of the right atrium, dilatation of right ventricular outflow tract, and localized aneurysms are the findings detected by this technique [Yamamuro 2005].

A radioisotope technique such as myocardial perfusion scintigraphy provides noninvasive assessment of right ventricular damage in patients with ARVD. Limitations of this technique are suboptimal spatial resolution and inherent radiation burden [Yamamuro 2005].

Cardiovascular magnetic resonance imaging allows visualization of the right ventricle anatomically, morphologically, and functionally [Tandri 2004]. Intramyocardial fat deposits, focal wall thinning, wall hypertrophy, trabecular disarray, right ventricular outflow tract enlargement, right ventricular aneurysms, and impaired right ventricular function are the findings of cardiovascular magnetic resonance [van der Wall 2000].

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Although cardiovascular magnetic resonance is suggested to be a noninvasive diagnostic standard, typical finding of fat infiltration in the right ventricle is detected in up to 40% of cases [Wu 2007].

MDCT has demonstrated its excellent spatial and temporal resolution, which provides the anatomical and functional changes in ARVD. MDCT may be considered a reliable imaging technique in the assessment and follow-up of patients with ARVD.

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