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THE ROLE OF MAGNETIC RESONANCE IMAGING IN DIAGNOSIS OF ARRHYTHMOGENIC RIGHT VENTRICLE DYSPLASIA

ARİTMOJENİK SAĞ VENTRİKÜL DİSPLAZİSİ TANISINDA MANYETİK REZONANS GÖRÜNTÜLEMENİN YERİ

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Abstract

Aim: Arrhythmogenic right ventricle dysplasia and cardiomyopathy (ARVD/C) occurs due to fibrofatty tissue infiltration in the ventricle myocardium.. Although the etiology has not been completely understood, it is responsible for the sudden death in early adolescents and athletics. We aimed to demonstrate the accuracy of ARVD/C findings in clinically confirmed ARVD/C cases by cardiac magnetic resonance imaging (MRI).

Materials and Methods: Cardiac MRI findings of clinically diagnosed 23 ARVD/C patients (18 male, 5 female; mean age: 38.2; SD: ± 13) were evaluated retrospectively. In four chamber cine images at both end-diastolic and end-systolic phases of cardiac cycle, the endocardial diameters of both atria and ventricles were measured.

Results: Findings were found as following: Right ventricular dilation in end-diastolic phase (>42 mm: 78.2%), right atrial dilation (> 41 mm: 78.2%), myocardial fatty replacement (74%) and left ventricular dilation (>42 mm: 74%), ventricle wall motion disorders on Cine MRI (43.5%), decrease in myocardial contractions (30.4%), postcontrast enhancement due to fibrosis (21.7%), right ventricle outflow tract dilation (21.7%), trabeculation (17.4%). Tricuspid insufficiency was 4.3% and Ebstain's anomaly was found as 4.3%.

Conclusion: MRI is useful for the diagnosis of ARVD/C. In the current study, the most frequent MRI findings were right ventricular and atrial dilation, myocardial fatty infiltration and left ventricular dilation respectively. The presence of fibrosis is significant for the diagnosis of ARVD/C. The left ventricle involvement is rare and may occur in ARVD/C.

Keywords: Arrhythmogenic right ventricle dysplasia, Myocardium, Magnetic resonance imaging

Öz

Amaç: Aritmojenik sağ ventrikül displazisi ve kardiyomiyopati (ARVD/C) ventrikül miyokardındaki fibröz ve yağ dokusunun infiltrasyonuna bağlı olarak ortaya çıkar. Etiyolojisi tam olarak anlaşılmamış olmasına rağmen, erken ergenlik döneminde ve atletlerdeki ani ölümlerden sorumludur. Klinik olarak onaylanmış ARVD / C vakalarında ARVD / C bulgularının kardiyak manyetik rezonans görüntüleme (MRG) ile hassasiyetini incelemeyi amaçladık

Materyal ve Metot: Bu çalışmada, 23 (18 Erkek, 5 Kadın; yaş ortalaması 38.2 SD: ± 13) ARVD tanılı hastanın kardiyak MRG bulguları retrospektif olarak değerlendirildi. Kardiyak siklusun diyastol sonu ve sistol sonu fazlarda dört boşluk (4CH) görüntülerde, atrium ve ventriküllerin endokardiyal çapları ölçüldü.

Bulgular: Elde edilen sonuçlar sırasıyla end-diyastolik fazda sağ ventrikül dilatasyonu (42 mm< %78.2) ve sağ atrium dilatasyonu (41 mm< %78.2) olup, bunu takiben miyokardda yağ infiltrasyonu (%74), sol ventrikül dilatasyonu (42 mm< %74), Cine MRG incelemede paradoks hareket ve duvar hareket bozuklukları (%43.5), miyokard kontraksiyonunda azlama (%30.4), postkontrast fibrozis lehine tutulum (%21.7), RVOT dilatasyonu (%21.7), trabekülasyon artışı (%17.4) ve eşlik eden diğer anomaliler (triküspit kapak yetersizliği: %4.3 ve Ebstain anomalisi: %4.3) şeklinde bulundu.

Sonuç: Kardiyak MRG, ARVD/C tanı için vazgeçilmez bir görüntüleme modalitesidir. Bu çalışmada en sık MRG bulguları sırasıyla sağ ventrikül ve atriyal dilatasyon, miyokard yağ infiltrasyonu ve sol ventrikül dilatasyonu olarak bulundu. Fibrozis varlığı ARVD/C tanısı için önemlidir. Sol ventrikül tutulumu nadir olup, detaylı bir şekilde değerlendirilmelidir.

Anahtar Kelimeler: Aritmojenik sağ ventrikül displazisi, Miyokard, Manyetik rezonans görüntüleme.

INTRODUCTION

Arrhythmogenic right ventricle dysplasia or cardiomyopathy (ARVD/C) is a disease characterized by the fibroadipose tissue infiltration of the right ventricle (RV) myocardium with unknown etiology which may lead to electrical disturbances and even sudden death[1-3]. Undiagnosed syncope,

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repetitive ventricular tachiarrhytmia, heart failure and sudden death can be seen[4]. ARVD/C is responsible for 3-4% deaths in athletic people and 5% of the sudden cardiac deaths below 65 years[2,5].

The estimated prevalence of ARVD/C in the population is about 1/1000-5000. Familial transition is seen in up to 30% to 50% of cases in different series[6]. Etiologic factors of the disease are mutations of genes encoding for desmosomal and non-desmosomal proteins [7-11].

Symptoms are frequently seen between ages of 15-40 [1]. The mean age is about 30 years occur during exercises and efforts with premature ventricular beats and non-sustained or sustained ventricular tachycardia of left bundle branch block morphology. Males are most commonly affected with the ratio of three male per one female[12,13].

There are minor and major Task Force diagnostic criterias (TFC) for ARVD/C including structural, histological, electrocardiographic, arrhythmic and genetic factors. Regarding to this classification, diagnosis of ARVD/C can be made by the presence of two major criteria, one major plus two minor criteria, or four minor criteria from different groups[14].

The diagnosis of ARVD/C in cardiac MRI can be made by the presence of intramyocardial fibrofatty infiltration, morphologically thickening or thining of the myocardial wall, motion abnormalities, myocardial inflammation and fibrosis. Angiography and echocardiography can only depict the focal or diffuse ventricular bulging[2,15].

In this study, we aimed aimed to demonstrate and reveal the accuracy of ARVD/C findings of 23 clinically confirmed ARVD/C patients on cardiac MRI.

MATERIAL AND METHODS

Patients:

40 patients with clinical diagnosis of ARVD/C, treated at cardiology department, were included in the study. Ten patients who had not cardiac MRI and 8 patients without follow-up, were excluded from the study. Of a total 23 patients, the age was varing between 20- 69 years (Mean age: 38.2, SD: ± 13), were included. Local ethical committee approved the study protocol (Istanbul university, Istanbul faculty of medicine, Date: 03 November 2011, Approval number:1771).

Magnetic Resonance Imaging (MRI):

MRI was performed on a 1.5 Tesla scanner (Symphony, Siemens Medical Systems, Erlangen, Germany) with electrocardiographic triggering. The MRI sequences and parameters were as following: four chamber (4CH), two chamber (2CH) and short-axis (SA) echo-planar cine true fast imaging with steady-state precession (FISP) (TR/TE, 50/1.70 ms; matrix, 256×256; slice thickness: 6 mm); 4CH and SA darkblood fast spin echo T1-weighted (TR/ TE, 700/26 ms; matrix, 133×256; slice thickness, 5 mm), 4CH and SA T2-weighted (TR/ TE, 1600/81 ms; matrix, 133×256; slice thickness, 5 mm), axial dark-blood half-Fourier single shot turbo spin echo (HASTE) (TR/TE, 800/26 ms; matrix, 256×256; slice thickness, 6 mm), SA and 4CH delayed contrast-enhanced images were obtained using three-dimensional (3D) inversion recovery FLASH (TR/ TE, 750/1.58 ms; time to inversion, 260 ms; matrix, 256×256; slice thickness, 4 mm) after 15±5 minutes delaying from initial time of injection.

Statistical Analysis: The statistical analysis (descriptive frequencies) was performed by using SPSS 18.0 (SPSS Inc., Chicago, Illinois. USA).

RESULTS:

Cardiac Chambers Evaluation: In four chamber cine images at both end-diastolic and end-systolic phases of cardiac cycle, the diameters (from endocard to endocard) of both atria and ventricles were measured (Table 1). The shortest diameter of the left ventricle in the end-diastolic phase, the longest diameter and the mean diameter of left ventricle in end-diastolic phase were 36 mm, 58 mm and 49.1 mm, respectively. The shortest diameter of the right ventricle in the end-diastolic phase, the longest diameter and the mean diameter of left ventricle in end-diastolic phase were 33 mm, 62 mm and 49 mm, respectively.

The shortest diameter of the left atrium in the end-diastolic phase was 20 mm, the longest diameter was 52, and the mean diameter of left atrium in end-diastolic phase was measured as 49.1 mm. The shortest diameter of the right atrium in the end-diastolic phase, the longest diameter, the mean diameter of right atrium in end-diastolic phase were 21mm, 65 mm and 46 mm, respectively (Table 1).

The shortest diameter of the left atrium in the end- systolic phase, the longest diameter and the mean diameter of left atrium in end-systolic phase were 24 mm, 58 mm and 40 mm, respectively. The shortest diameter of the right atrium in the end- systolic phase, the longest diameter and the mean diameter of right atrium in end- systolic phase were 27 mm, 64 mm and 46 mm, respectively(Table 2).

Motion Abnormalities: In 10 patients (43.5%) motion abnormalities detected in cine images as following: the dyskinetic wall in the right ventricle apical region of 3 patients (13%), at the right ventricle lateral wall in 5 patients (21.7%) and at bilateral ventricle lateral walls in 2 patients (8.7%).

Fatty Infiltration: On T1-weighted 4CH and SA images, myocardial fatty infiltration was detected in 17 patients (74%). Fifteen (88.2%) cases were male and 2 (11.2%) were female. In patients with myocardial fatty infiltration, 7 (30.4%) patients had right ventricle free wall, 6 (%26.1) had right ventricle apical, 3 (13%) had biventricular and 1 (4.3%) had interventricular septum involvement (Table 3, Figure 1-5).

Fibrosis: On postcontrast delayed enhanced MRI, there were contrast enhacement in 5 cases (21.7%) due to fibrosis. Of the five, 2 (8.7%) were detected at right ventricle free wall, 2 (8.7%) were in both right and left ventricle and 1 (4.3%) was at right ventricle myocardium and the apical region (Figure 3).

Right ventricle outflow tract (RVOT) dilation: On cine RVOT images, the minimum, maximum and mean diameter was measured as 21 mm, 36 mm and 28.6 mm, respectively. Of the total 23 patients, RVOT diameter was higher than 30 mm in 5 (21.7%) patients and this finding was assessed as RVOT dilation.

Accompanying anomalies

In one (4.3%) patient, tricuspidal regurgitation and in one (4.3%) patient Ebstain's anomaly was detected. Cine and T1-weighted SA images showed hypertrabeculation in 17.4% (n=4) patients, of which, 4.3% (n=1) was in right ventricle, 8.7% (n=2) were in mid-ventricular segments of both ventricles and

4.3% (n=1) was in apical segment of both ventricles. The ratio of trabeculated/non-trabeculated segments was higher than 2.3 and were diagnosed as non-compaction cardiomyopathy.

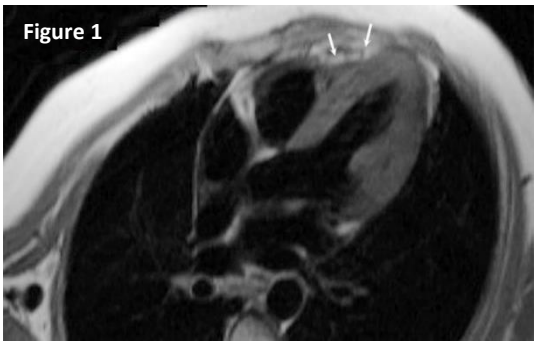

Figure 1: Cardiac MR imaging of 22-year-old female patient with ARVD/C shows the high signal intensity at right ventricle free wall due to fatty infiltration on T1 weighted images (arrows).

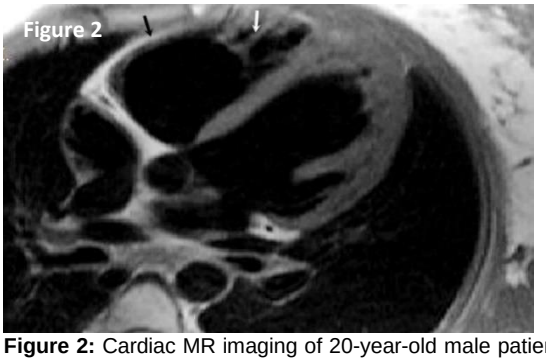

Figure 2: Cardiac MR imaging of 20-year-old male patient with ARVD/C shows high signal intensity due to fatty tissue infiltration (black arrow) and trabeculations (white arrow) on T1-WI and There was motion dyskinesia at the fatty tissue involved area on Cine 4CH images.

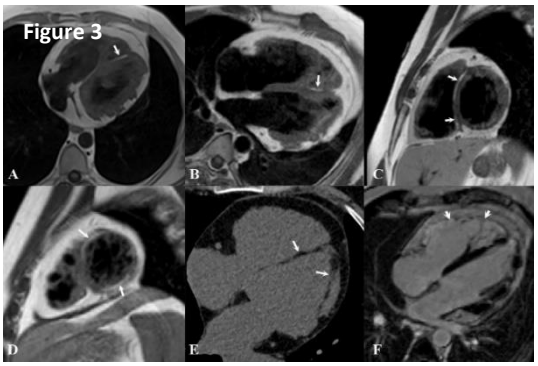

Figure 3. Cardiac MR and computed tomography (CT) studies of 35-year-old male patient with diagnosis of ARVD/C. On 4CH T1-WI (A,B) and SA (C,D) studies there was high signal intensity representing fatty tissue infiltration at the right portion of interventricular septum and in the subepicardial region of the left ventricle wall (white arrows). Long axis cardiac CT images (E) show the low density fatty tissue infiltrations at left ventricle lateral wall and the septum (white arrows). Post-contrast imaging (F) depicts contrast material enhancement which represents fibrosis at right ventricle wall (short arrows).

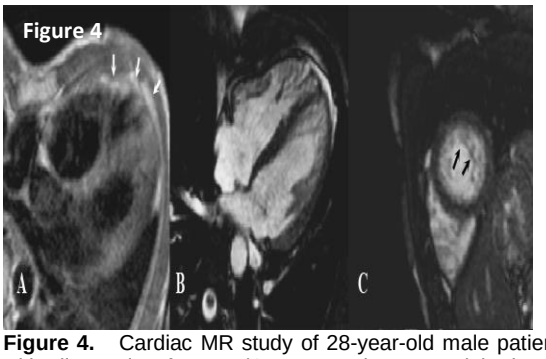

Figure 4. Cardiac MR study of 28-year-old male patient with diagnosis of ARVD/C. Long axis T1-WI (A) shows high signal intensity of fatty tissue involvement at apical region (white arrows). Long axis (B) and short axis (C) studies show the marked trabeculation which is compatible with non-compaction and the thinning in the lateral wall (black arrow).

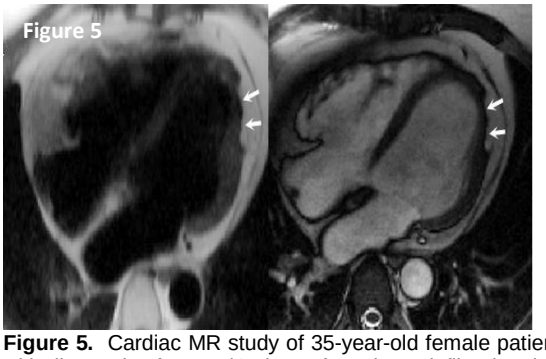

Figure 5. Cardiac MR study of 35-year-old female patient with diagnosis of ARVD/C shows fatty tissue infiltration, left ventricle involvement and on SE and GE sequence images wall thinning beginning from subepicardial region can also be seen (arrows).

DISCUSSION

In diagnosis of ARVD/C, according to TFC, the diagnosis can be made by the presence of two major criteria, one major plus two minor criteria, or four minor criteria from different groups[14].

Myocardial wall motion abnormalities, electropotential (depolarization and repolarization) changes in electrocardiographic study, structural changes such as silent aneurysmatic dilation and fatty tissue infiltration of right venrtricular wall are among the main findings of ARVD/C. The fibrofatty replacement in ARVD/C frequently involves the anterolateral side of right ventricle or the right portion of the septum, however, it may rarely involve the left ventricle myocardium.

In the presence of the intramyocardial fibrofatty infiltration, morphologically thickening or thinning of the myocardial wall, motion abnormalities, myocardial inflammation and the fibrosis, it is possible to make ARVD/C diagnosis in a cardiac MRI study[2,15]. The sensitivity and the specifity of cardiac MRI in the diagnosis of ARVD/C are variable between 22-100%[2,13,16].

Owing to the adipose tissue that has high signal intensity on T1-weighted imaging (T1-WI) [2,17], the minimal and focal intramyocardial fatty deposition can be detected on T1-WI and it is associated with ARVD/C[2,18,19]. However, intramyocardial adipose tissue is not a pathognomonic finding for the diagnosis of ARVD/C because the presence of fatty tissue particularly in the anterio-apical region may be a condition that is also observed in a normal heart[20].

Bluemke et al. published a similar study; 6 of the 7 patients who have received ARVD/C diagnosis according to TFC were studied and RV enlargement, RV abnormal morphology, left ventricle dilation, fatty tissue infiltration of high signal in myocardium on T1-WI and the location were evaluated. In ARVD/C patients, right ventricle dilation was more than in control group (definite diagnosis of ARVD/C group 58%, control group 14%; $p<0.0001$). The results of this study show that detecting T1 high signal intensity of fatty tissue is a less reliable finding than RV enlargement and morphological changes[21]. Similarly, in our clinically diagnosed 23 ARVD/C patients; the most common MR imaging finding of ARVD/C was right ventricular dilation (78%). In our retrospective study, myocardial fatty infiltration was found in 74% of ARVD/C patients (88.2% male, 11.8% female), the most common sites were right ventricle lateral wall (30.4%) and apical regions (26.1%), respectively.

Cardiac MR study is able to detect the regional and diastolic (right and left) ventricular disfunctions and the earlier findings of ARVD/C uniquely[16,22]. Disfunctional regions are typically seen at right ventricular free wall, the apex cordis and RV input/output tract location. The involvement may be variable from patchy infiltration to uniform or diffuse infiltration pattern [23].

The contrast material uptake pattern in cardiac MR study is essential to detect the myocardial fibrofatty changes of biventricular involvement in ARVD/C patients[24-26]. The fibrofatty areas, detected in cardiac MR, depict the closed correlation with histologic and electro-physiologic studies. It has been shown that RV disfunction is associated with the stimulation of ventricular tachycardia in electro-physiologic studies[25].

It is believed that the presence of fibrous tissue in the myocardium is more arrhitmogenic than the fatty tissue [20]. In non-ischemic condition like ARVD/C, the contrast material uptake pattern is not compatible with the distribution of coronary arteries, it is frequently more seen in the middle portion of the myocardial wall rather than the subendocardial or transmural area.[26] Tandri et al. pointed to the relationship of histologically confirmed fibrosis and the presence of late contrast enhancement in MR images of ARVD/C patients [27]. In none of the patients there was contrast uptake without diagnosis of ARVD/C. Similar results have been reported by other study groups[20,24].

In 5 (% 21.7) patients at our study, there was contrast enhacement due to fibrosis in late phase of dynamic post-contrast series. In 2 (% 8.7) patients the right ventricle myocardium, in 2 (%8.7) both right and left ventricle myocardium and in 1 (% 4.3) patient the right ventricle myocardium and the apical region were affected.

RVOT tachycardia should be considered in the differential diagnosis of ARVD/C. White et al. reported wall thinning or wall motion disorders in 74% of 46 cases with RVOT tachycardia. These abnormalities are frequently located at right ventricle anterior wall and in 8 (25%) of cases fatty infiltration has been reported [28]. On the other hand, Grimm et al. declared that no abnormality detected in 14 cases with RVOT tachycardia[29].

In ARVD/C the serious fatty infiltration results in trabecular derangement associated with wall thickening (8mm <)[30]. In our study, trabecular derangement was detected in 4 (17.3%) of 23 ARVD/C patients, 1 (4.3%) at right ventricle, 2 (8.7%) at both right and left ventricles, 1 (4.3%) at biventricular and the apical region.

The prevalence of left ventricle involvement in ARVD/C has been reported as %16 [31] - %76 [32] according to different authors. Bauce et al. revealed left ventricular disorders in half of ARVD/C patients in electrocardiographic examination[33]. Lindstrom et al. demonstrated the presence of left ventricular disorders in 93% of ARVD/C patients by using scintigraphic myocardial perfusion and the echocardiography studies[34].

Although there is not an exact consensus in the literature, the left ventricular involvement in ARVD/C is not considered as a different disorder, also it may represent or be a component of a disease process called "arrhythmogenic cardiomyopathy" with appropriate subclassifications. LV involvement in ARVD/C may precede the begining of significant right ventricular dysfunction[35].

Table 1. The end-diastolic diameters of four chambers

End-diastolic phase	Longest Diameter (mm)	Shortest Diameter (mm)	Mean Diameter (mm)
Left Venticle	58	36	49.1
Right Venticle	62	33	49
Left Atrium	52	20	49.1
Right Atrium	65	21	46

Table 2. The end-systolic diameters of four chambers

End-systolic phase	Longest Diameter (mm)	Shortest Diameter (mm)	Mean Diameter (mm)
Left Venticle	53	19	37
Right Venticle	55	21	39
Left Atrium	58	24	40
Right Atrium	64	27	46

Table 3. The distribution of fatty infiltration in ARVD/C patients.

Fatty Infiltration	Patients (n=17)	%
Right ventricle free wall	7	30.4
Right ventricle apex	6	26.1
Right and left ventricle	3	13
Interventricular septum	1	4.3

The fibrofatty replacement of the left ventricle shows correlation with the significant perfusion defect in the anterioseptal and the posteriobasal segments of the left ventricle[34]. The reported qualitative and quantitative left ventricle disorders as well as in right ventricle in ARVD/C including increase in left ventricle end-diastolic pressure, decreased ejection fraction, atrioventricular block, regional wall motion abnormalities, aneurysms (frequently in apical regions), presence of asinergic regions (frequently the apex and inferior-posterior wall) and dilation[36]. Moreover, the left ventricle involvement has been detected in asymptomatic or short-time symptomatic patients[34,37].

Left ventricle involvement, rather than right ventricle, is more common and severe in

families with several affected members with ARVD/C [36]. In our study, in 4 (17%) of 23 ARVD/C patients T1 hiperintensity of fatty infiltration at the left ventricle and in 2 (%8.7) of cases, the postcontrast enhancement due to fibrosis at the left ventricle wall, was detected.

CONCLUSION

MR imaging is an indispensable and non-invasive imaging modality in the assessment of right ventricle size and morphology as well as motion disorders. Right ventricular and atrial dilation as well as the presence of fibrosis and the involvement of the left ventricle should be cared in ARVD/C. A careful assessment is required regarding to the possible accompanying anomalies.

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