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CASE REPORT

INTERMEDIATE

CLINICAL CASE SERIES

Right Ventricular Fatty Infiltration With an Abnormal ECG

Natalija Odanovic, MD,[a] Hamid R. Mojibian, MD,[b] Lawrence H. Young, MD[a]

ABSTRACT

Two middle-aged women had evidence suggesting right ventricular hypertrophy on routine electrocardiograms. Their echocardiograms showed right ventricular thickening and cardiac magnetic resonance imaging revealed right ventricular fatty infiltration. Neither patient fulfilled the criteria for arrhythmogenic right ventricular cardiomyopathy, and both had a benign clinical course. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2021;3:314–8)

CASE 1

A 53-year-old woman had a routine electrocardiogram (ECG) showing tall R waves in V_1 and V_2 suggesting possible right ventricular hypertrophy (RVH), and nonspecific ST-segment depression and T-wave inversions in V_1 to V_5 (**Figure 1**). Echocardiogram showed increased right ventricular (RV) free wall thickness (14 mm in the parasternal long-axis view), with normal RV dimension (20 mm in the apical 4-chamber view) (**Figure 2**, Videos 1 to 5) and no valvular disease.

MEDICAL HISTORY. She had hypertension and a body mass index (BMI) of 24.4 kg/m^2. Her sister died of congenital heart disease and a paternal uncle died suddenly in his forties while running.

LEARNING OBJECTIVES

- To recognize the presentation of fatty infiltration of the right ventricle associated with an abnormal ECG, suggesting RV hypertrophy, and a thickened RV on echocardiogram.
- To understand the diagnostic approach and criteria that distinguish fatty infiltration of the right ventricle from normal and ARVC.

CASE 2

A 59-year-old woman complained of atypical chest pain to her primary care physician. An ECG showed a tall R-wave in V_1, and S waves throughout the precordial leads with S>R in V_6 with nonspecific T-wave inversions in V_1 to V_3, suggesting RVH (**Figure 3**). Echocardiogram revealed increased RV free wall thickness (8 mm in the parasternal long-axis view), normal RV midcavity dimension (22 mm in the apical 4-chamber view), and no valvular heart disease (**Figure 4**, Videos 6 to 10).

MEDICAL HISTORY. She had musculoskeletal discomfort and a BMI of 28.0 kg/m^2. There was no family history of cardiovascular disease or sudden death.

DIFFERENTIAL DIAGNOSIS

The differential diagnoses for both patients included RVH, infiltrative cardiomyopathy, epicardial RV fat,

From the [a]Department of Internal Medicine, Section of Cardiovascular Medicine, Yale School of Medicine, New Haven, Connecticut, USA; and the [b]Department of Diagnostic Imaging, Yale School of Medicine, New Haven, Connecticut, USA.
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fatty infiltration of the RV, RV neoplasm, and arrhythmogenic right ventricular cardiomyopathy (ARVC).

INVESTIGATIONS

Cardiac magnetic resonance imaging (MRI) was performed in both patients, demonstrating lipomatous infiltration of the RV free wall, normal RV function and wall motion, and no late gadolinium enhancement (**Figures 5A, 5B, 6A, and 6B**, Videos 11 and 12). In Patient #1 where the lipomatous infiltration was marked, we considered a diagnosis of liposarcoma; however, because of the lack of late gadolinium enhancement and imaging stability over the years, we ruled liposarcoma out. We also considered a diagnosis of lipoma; however, the lipomatous infiltration was not encapsulated but interspersed with the myocardium and there was separate minor lipomatous infiltration of the apical septum, so we thought lipoma was less likely. The lipomatous infiltration of the RV in Patient #2 was more subtle and there was no left ventricular involvement. Additionally, both patients had cardiac computed tomography, the first to confirm the diagnosis and the second for attenuation correction during a nuclear stress test, both of which showed RV fatty infiltration (**Figures 5C and 6C**). Holter monitor in Patient #1 showed frequent premature ventricular contractions, whereas Patient #2 had 1 premature ventricular contraction and a single ventricular triplet. Because of the family history of sudden death Patient 1 underwent genetic testing for 5 ARVC genes that was negative.

MANAGEMENT

Patient #1 was treated with beta-blockers for hypertension, which lessened her premature ventricular contractions. Patient #2 was monitored without additional interventions.

DISCUSSION

These cases illustrate RV fatty infiltration associated with abnormal ECGs suggesting RVH. They then underwent echocardiography, including RV-focused imaging from the apical 4-chamber and RV inflow views. Both patients had increased RV free wall thickness, which also confused the diagnosis, before additional imaging (cardiac computed tomography and MRI) that revealed fatty infiltration. The degree of RV wall thickening and intramyocardial location of the fat also distinguished the images from normal. Neither patient had MRI evidence of ARVC, such as RV fibrosis, dilatation, or regional wall motion abnormality.

Fatty infiltration of the RV is an unusual entity, but has been well-described in autopsy series (1–4). Intramyocardial fat in the RV has a higher prevalence in older patients and women (1,2). The average percentage of fat has been reported from 0.3% to 17% (5), with a predilection for the anterior and lateral apex (3). In one autopsy series, including people with

ABBREVIATIONS AND ACRONYMS

ARVC = arrhythmogenic right ventricular cardiomyopathy

BMI = body mass index

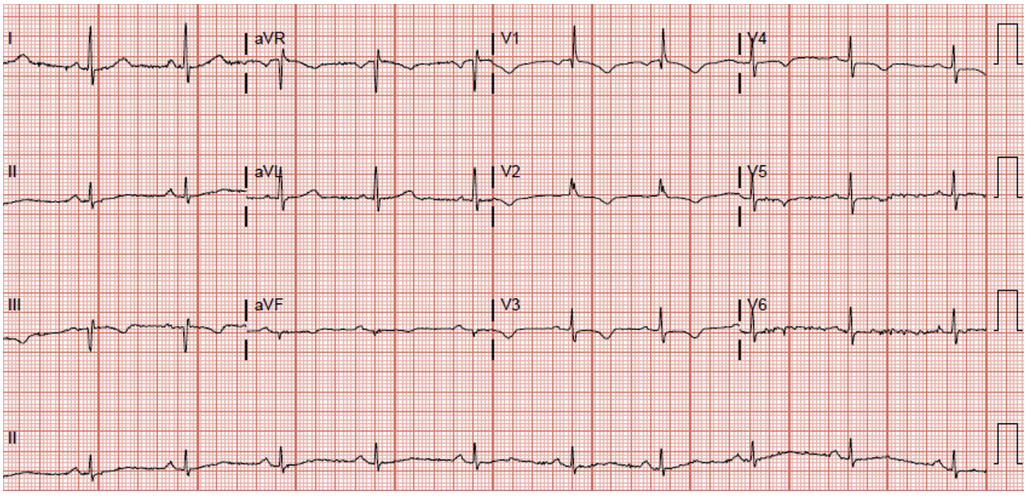
ECG = electrocardiogram

MRI = magnetic resonance imaging

RV = right ventricle

RVH = right ventricular hypertrophy

FIGURE 1 Electrocardiogram in Case 1



Tall R waves in V_1 to V_2 suggestive of right ventricular hypertrophy.

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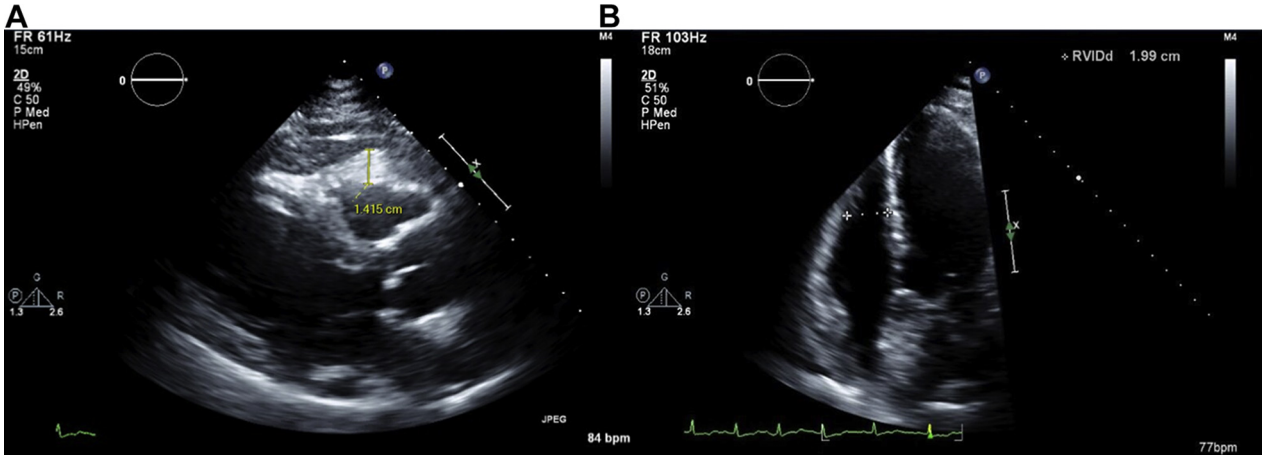
FIGURE 2 Echocardiogram in Case 1

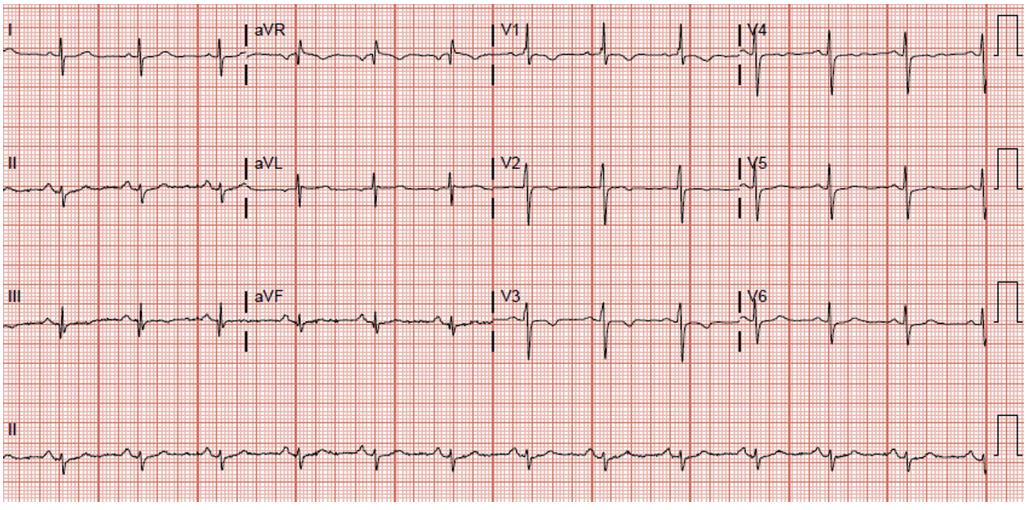


Parasternal long-axis view **(A)** and apical 4-chamber view **(B)**.

noncardiac deaths at >40 years of age, almost two-thirds of women and 30% of men had >25% of fat in the RV lateral wall, and one-third of women and 10% of men had >50% fat (2). In another series of 7 patients with an average BMI of 26.4 kg/m^2, isolated RV fat content averaged 32%, which was more than in comparative ARVC or control groups (3). These patients had thickening of the epicardial fat layer and the myocardium itself.

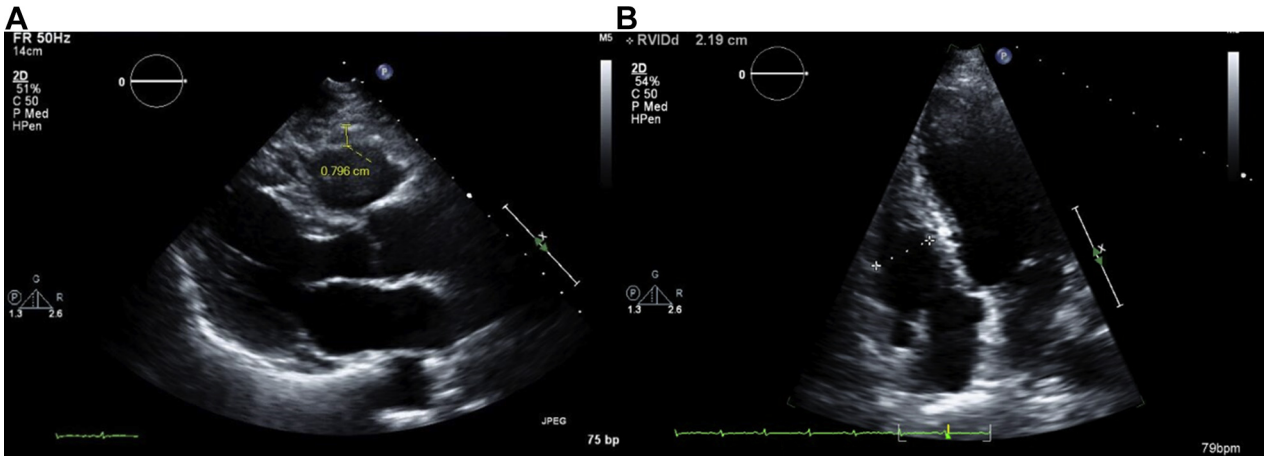
An MRI series described 4 men and 9 women with marked lipomatous infiltration of the RV on cardiac imaging and no features of ARVC. Like our second patient, they were overweight with a BMI averaging 28 kg/m^2. RV wall thickness averaged 9 mm, compared with 4 mm in the control group. However, their RV size, function, and wall motion were not different from control subjects (6). These imaging features are very similar to our patients, but ECG results were not reported. Thus, our cases are unique in demonstrating an abnormal ECG in patients with fatty infiltration of the RV.

FIGURE 3 Electrocardiogram in Case 2



Tall R-wave in V_1 and anterior S waves with S>R in V_5 and V_6, suggestive of right ventricular hypertrophy.

FIGURE 4 Echocardiogram in Case 2



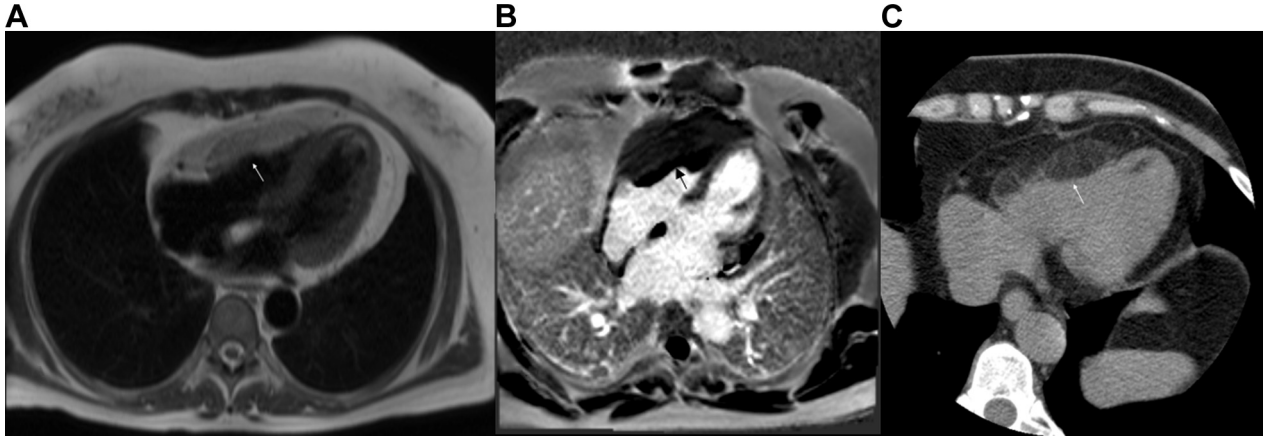
Parasternal long-axis view **(A)** and apical 4-chamber view **(B)**.

It is clinically imperative to distinguish fatty infiltration of the RV from ARVC. Fat can comprise between 13% and 77% of the RV wall in ARVC (3,5), but does not define ARVC, which is also characterized by myocardial fibrosis and contractile dysfunction (4). In contrast to our patients, the fat infiltration in ARVC is located predominantly in the RV outflow tract and is associated with myocyte atrophy and RV wall thinning. Clinical criteria for the diagnosis of ARVC include a family history, arrhythmias (left bundle branch block with an inferior axis), epsilon waves, depolarization abnormalities and T-wave inversions, and evidence of RV enlargement and/or dysfunction (7). In contrast, our patients had a prominent R-wave with RS complexes in the anterolateral leads, ECG findings that are not characteristic of ARVC.

FOLLOW-UP

Cardiac MRI was repeated at 2- to 3-year intervals in Patient #1 and showed stable RV function and fatty

FIGURE 5 Cardiac Magnetic Resonance Imaging and Chest Computed Tomography Images in Case 1



Contrast magnetic resonance imaging T1-weighted black blood image **(A)** and myocardial delayed enhancement image **(B)**, and noncontrast computed tomography **(C)** show thick right ventricle free wall with diffuse fatty infiltration **(arrows)**.

FIGURE 6 Cardiac Magnetic Resonance Imaging and Chest Computed Tomography Images in Case 2

Magnetic resonance imaging cine image **(A)** and post-contrast image **(B)**, and noncontrast computed tomography **(C)** show thick right ventricle free wall with diffuse fatty infiltration **(arrows)**.

infiltration and no cardiac problems over 10 years. Patient #2 has had a benign 2-year follow-up.

CONCLUSIONS

We present 2 cases of isolated RV fatty infiltration that came to our attention because of the incidental finding of an abnormal ECG suggestive of RVH. Both cases initially represented a diagnostic dilemma, which was ultimately clarified by cardiac MRI imaging. Our patients have done well, and most authors agree that RV fatty infiltration does not have significant potential for malignant arrhythmias or sudden cardiac death. However, rare instances of sudden death have been reported (8,9), including a highly unusual case of pancreatitis-mediated fat necrosis of a lipomatous RV (10). The clinical implications of isolated RV fatty infiltration with an RVH pattern on ECG remain unclear, and further investigation is warranted to further understand its prevalence and significance.

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ADDRESS FOR CORRESPONDENCE: Dr. Lawrence H. Young, Yale University School of Medicine, 333 Cedar Street, P.O. Box 208017, New Haven, Connecticut 06520-8017, USA. E-mail: Lawrence.young@yale.edu.

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KEY WORDS electrocardiogram, magnetic resonance imaging, right ventricle

APPENDIX For supplemental videos, please see the online version of this paper.