A 54-year-old female was evaluated for worsening dyspnea on exertion and palpitations. As a young adult, a murmur had been detected during a routine physical examination of the patient, and a 2-dimensional echocardiogram revealed increased left ventricular (LV) wall thickness, which resulted in a presumed diagnosis of hypertrophic cardiomyopathy (HCM). Her medical history was significant for hypothyroidism and breast cancer treated with chemotherapy and radiation 4 years previously. The patient had no family history of heart disease or unexplained sudden death. She was treated with β-blockers and calcium channel blockers for heart failure symptoms.

Physical examination was notable for a III/VI systolic ejection murmur at the left upper sternal border and a body weight of 160 lb (body mass index = 30 kg/m²). A 12-lead ECG demonstrated normal sinus rhythm with right-axis deviation and marked LV hypertrophy with nonspecific ST/T-wave changes; a 24-hour ambulatory Holter ECG recorded 1733 premature ventricular contractions with 7 runs of nonsustained ventricular tachycardia. Although the LV wall thickness appeared qualitatively increased. In all echocardiographic imaging planes bright echodensities were present within the myocardium, which produced substantial acoustic shadowing that resulted in poor image quality.

To further characterize cardiac morphology, the patient underwent cardiovascular magnetic resonance imaging (CMR). Cine steady-state free precession imaging demonstrated normal systolic function (ejection fraction 83%) of both the LV and right ventricle, with normal regional wall motion. However, there was increased wall thickness of both ventricles with a maximal thickness in the LV lateral wall of 33 mm (ventricular septum 25 mm) and 23 mm in the right ventricle, with a significantly increased LV mass index of 162 g/m² (normal < 75 g/m²) and normal left and right atrial...
sizes (Figure 1; Movies IA and IIA). In addition, there were large areas of increased intramyocardial signal intensity that involved the entire ventricular myocardium and septum (Figures 1 and 2A).

On contrast-enhanced CMR imaging with late gadolinium enhancement without fat saturation, high signal intensity was present in nearly the identical regions of myocardium where areas of increased signal intensity were observed on the cine images (Figure 2B; Movie II). However, with the addition of a fat-saturation sequence to the same contrast-enhanced images, all of the previous intramyocardial high signal intensity regions were completely nulled, which confirmed that these areas were composed predominantly of adipose tissue (Figure 2C).

To confirm the diagnosis, the patient underwent a cardiac biopsy that demonstrated structurally normal myocytes diffusely infiltrated with adipose cells (Figure 3). In addition, there was no evidence of other abnormal histopathology including: myocyte disarray, amyloid (by Congo Red staining), glycogen or lysosomal products within myocytes, granulomas, or lymphocytic infiltration. Genetic testing of the 5 known genetic mutations for arrhythmogenic right ventricular cardiomyopathy revealed no mutations, nor did testing of the 9 common mutations for sarcomeric HCM. In addition, the extensive involvement of fat infiltration in the LV was not consistent with arrhythmogenic right ventricular cardiomyopathy.

Therefore, the totality of the clinical, imaging, and histological data confirmed a diagnosis of adipositas cordis. An extraordinarily rare cardiomyopathy adipositas cordis is characterized by myocardial fatty infiltration of both ventricles and has been diagnosed predominantly as an incidental finding at autopsy in male patients with generalized obesity.2 This disease has generally been considered to be a benign clinical entity in the obese, although there are reports of patients with adipositas cordis experiencing ventricular and supraventricular arrhythmias and restrictive cardiomyopathy.3,4 On the basis of the presence of a substantial burden of nonsustained ventricular tachycardia demonstrated on the ambulatory Holter monitor, an implantable cardioverter-defibrillator was placed for primary prevention of sudden death. Medical treatment with calcium channel blockers and β-blockers was continued for symptoms.

This patient had a long-standing diagnosis of HCM based on the presence of increased wall thickness on echocardiography. In the later part of her clinical course, the extensive myocardial fatty infiltration resulted in poor acoustic image quality that compromised the morphological information that could be reliably obtained with echocardiography. However, with its unique ability to identify adipose tissue using fat-saturation radiofrequency pulses and high spatial and temporal resolution imaging, cardiovascular magnetic reso-

Figure 2. Cine and contrast-enhanced cardiovascular magnetic resonance images in adipositas cordis. A, Basal end-diastolic short-axis cine image showing multiple areas of increased signal intensity in both ventricles, including 2 large areas in the inferior right ventricular wall extending into the posterior septum (thick arrow) and the inferior wall of the LV (thin arrow). B, Contrast-enhanced cardiovascular magnetic resonance image without fat saturation in the identical short-axis imaging plane as panel A, showing areas of hyperintensity in similar regions as those corresponding to increased signal intensity in the previous cine image, including the inferior wall of the right ventricle (thick arrow) and inferior wall of the LV (thin arrow). These areas of myocardial hyperintensity have similar signal intensity as that of subcutaneous fat (*). C, Contrast-enhanced basal short-axis image in the identical imaging plane as the previous 2 panels but now with the addition of a fat-saturation pulse sequence, demonstrating that all regions of myocardium are nulled (ie, black) and of similar signal intensity as subcutaneous fat (*). The nulling of regions of increased signal intensity with fat saturation confirmed that these areas of myocardium were predominantly composed of fatty infiltration and not fibrosis. RV indicates right ventricle free wall; VS, ventricular septum.

Figure 3. Histological specimen in this patient with adipositas cordis obtained during biopsy of the right ventricle. There are extensive areas of adipocyte cells (thin arrow) intermixed with small areas of myocytes of normal size and features (thick arrow).
nance ultimately raised consideration of the alternative diagnosis of adipositas cordis in this patient. This case also highlights the growing role of cardiovascular magnetic resonance in the evaluation of patients with structural heart disease, especially in those with the possibility of an unusual form of infiltrative cardiomyopathy.

Disclosures
Dr Maron has served as a consultant for PGxHealth. The remaining authors report no conflicts.

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