Cardiac Magnetic Resonance Imaging of Burned-out Hypertrophic Cardiomyopathy

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A 52 YEAR-OLD MAN WITH CHRONIC obstructive pulmonary disease and no prior cardiac history presented with new onset **congestive heart failure** (**CHF**). Echocardiography revealed severely depressed global **left ventricular** (**LV**) function and mildly reduced **right ventricular** (**RV**) function. His estimated pulmonary artery pressure was 80 mmHg, consistent with severe pulmonary hypertension. EKG revealed normal sinus rhythm with right bundle branch block. The patient was referred for **cardiac magnetic resonance imaging** (**CMR**) to evaluate the etiology of the **cardiomyopathy** (**CM**).

CMR confirmed severe global LV dysfunction with an ejection fraction of 15% and moderate RV dysfunction. While both the LV and RV free walls were mildly thickened, the interventricular septum was markedly thickened measuring 2.7 cm (normal ≤ 1.2 cm). (Figure 1a) Post-contrast imaging demonstrated patchy predominantly subepicardial enhancement throughout the LV, septum and apical RV consistent with fibrosis. (Figure 1b) This pattern of enhancement is non-specific, but excluded an ischemic CM where scar is sub*endo*cardial. The pattern is not typical for cardiac amyloid.[1] Though other non-ischemic etiologies such as cardiac sarcoid were considered, in light of the myocardial thickening, **hypertrophic CM** (**HCM**) was favored. Subsequent endomyocardial biopsy demonstrated myocyte disarray and hypertrophy, as well as areas of fibrosis, confirming HCM. (Figure 1c)

Discussion

HCM is a genetic disease of the cardiac sarcomere with an autosomal dominant inheritance pattern typically showing a high degree of penetrance. It is a relatively common disease occurring in up to 0.29% of individuals[2] and is among the most common cardiac lesions found in athletes with sudden cardiac death. Morphologically, several variants have been described: septal hypertrophy with or without LV outflow tract obstruction, concentric hypertrophy, apical hypertrophy, LV free wall hypertrophy, and RV hypertrophy. At histology myocardial disarray may be seen. Additionally, myocardial fibrosis is considered a hallmark of HCM and a potential substrate for arrhythmias and heart failure.[3]

CMR has emerged as a powerful tool for evaluating a host of non-ischemic cardiomyopathies, including HCM.[4] In addition to providing accurate quantification of wall thickness, chamber volumes and ejection fraction, assessment of regional and global wall motion, and quantification of valve lesion severity, CMR has the ability to characterize tissue pathology. It permits detection of edema, infarction, inflammation, and fibrosis. In our case, substantial fibrosis was detected in a non-infarct distribution confirming a non-ischemic etiology. It is proposed that the extensive fibrosis contributed to the marked LV dysfunction and eventual CHF observed in this patient.

References

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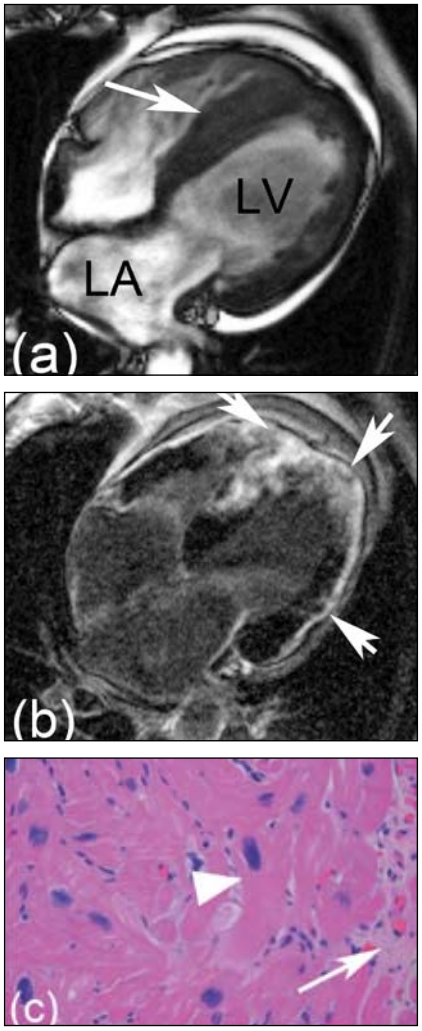

Figure 1. (a) Four-chamber bright-blood CMR image shows abnormal ventricular thickening, chiefly involving the interventricular septum (arrow). (b) Post-contrast imaging demonstrates extensive enhancement throughout the septum, distal right ventricle, and subepicardial left ventricular free wall (arrows). (c) Histologic specimen with H&E stain shows myocyte disarray, fibrosis (arrow), and hypertrophy (arrowhead). LA: Left atrium; LV: Left ventricle.

Disclosure of Financial Interests

The authors and/or their spouses/significant others have no financial interests to disclose.

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