Isolated Primary Cardiac Sarcoidosis
MRI Diagnosis and Monitoring of Treatment Response With Cardiac Enzymes

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A 48-year-old woman without known cardiovascular disease presented with progressive dyspnea. The ECG showed first-degree heart block. Plasma Troponin I and T were persistently elevated. Chest x-ray was normal. Echocardiography demonstrated asymmetrical left ventricular (LV) hypertrophy with septal predominance and moderately impaired LV systolic function. Coronary angiography demonstrated angiographically normal coronary arteries.

Although the echocardiographic features were consistent with a diagnosis of hypertrophic cardiomyopathy, the ECG evidence of conduction abnormality and persistent elevation of cardiac enzymes were more suggestive of a myocarditic process. A cardiac MRI (CMR) was requested. This demonstrated asymmetrical septal hypertrophy with thickening of the basal-mid septum extending to the apex (Figure 1). On delayed-enhancement imaging, multiple discrete areas of hyperenhancement were noted throughout the LV myocardium that deformed the endocardial and epicardial borders (Figure 2). The LV ejection fraction was 31%. The clinical presentation and CMR appearances were suggestive of sarcoidosis; however, high-resolution CT of the thorax to look for confirmatory evidence of pulmonary sarcoidosis showed no evidence of lymphadenopathy or other pulmonary involvement. She therefore proceeded to cardiac biopsy to establish a diagnosis. This showed focal granulomas composed of multinucleated giant cells with central necrosis and associated lymphocytes. Tuberculous and fungal infections were excluded by negative Ziehl-Neelsen and periodic acid-Schiff staining. A diagnosis of isolated primary cardiac sarcoidosis (PCS) was made.

The clinical course was complicated by complete heart block. She received an implantable cardiac defibrillator. Prednisone was commenced at 40 mg per day for treatment of PCS, with a rapid response of dyspnea and normalization of the Troponin I within 1 week, remaining suppressed over 6 months of follow-up. Repeat echocardiography at 1 month showed no change in LV function.

Cardiac involvement has been reported in up to 30% of patients with sarcoidosis, but the prevalence of isolated PCS is not well defined.1,2 Nonspecific CMR features of sarcoidosis may include segmental wall motion abnormalities in nonvascular distributions and focal wall thickening,3,4 changes that may mimic hypertrophic cardiomyopathy. CMR has been shown to be of greater utility than echocardiography and nuclear imaging modalities (other than PET) in the diagnosis of PCS.4 Although not absolutely contraindicated, the utility of CMR may be limited in patients with pacemakers.4 Cardiac enzymes may be helpful in guiding treatment in patients such as this woman in whom serial CMR evaluations are contraindicated by insertion of pacing devices.

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Disclosures
None.

References

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Figure 1. Asymmetrical LV hypertrophy shown in CMR steady-state-free precession images in LV short-axis and 3-chamber views.

Figure 2. Late gadolinium enhancement (arrows) shown in CMR delayed-enhancement images in 2-, 4-, and 3-chamber projections.
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