Case Report: Abnormal Myocardial Enhancement

Alan C. Legasto, M.D., 1 Stephen Waite, M.D. 2

1 Department of Radiology, Weill Cornell Medical Center, New York, NY
2 Department of Radiology, SUNY Downstate Medical Center, New York, NY

Case Presentation

A 21-year-old man presented to the emergency department with fever and chest pain. Chest radiographs were normal. Electrocardiogram showed elevation of the ST-segments in leads II, III, and a VF, as well as reciprocal depression in leads I and a VL. Serum tests showed elevation of troponin T, creatine kinase (CK), and CK-MB. Echocardiography performed in the emergency department measured the left ventricular ejection fraction at 48%. Cardiac MRI with and without contrast was obtained to evaluate for myocarditis (Figure 1).

**FIGURE 1.** Delayed contrast-enhanced MRI image of the heart in short axis (A) demonstrates significant left ventricular epicardial hyperenhancement (arrow), particularly within the inferolateral walls. Three-chamber contrast-enhanced view of the heart (B) demonstrates additional foci of scattered midmyocardial hyperenhancement, particularly within the septum (arrow).
Key Imaging Finding(s)
Abnormal delayed epicardial and midmyocardial hyperenhancement

Differential Diagnoses
Infectious/inflammatory cardiomyopathy
  Ischemic cardiomyopathy
  Hypertrophic cardiomyopathy
  Sarcoidosis
  Amyloidosis

Discussion
Delayed contrast-enhanced MRI images of the heart demonstrate significant left ventricular epicardial hyperenhancement, particularly involving the inferolateral walls along with foci of scattered midmyocardial hyperenhancement, particularly involving the septum. Given the patient’s age, EKG changes and clinical symptoms of fever and chest pain, nonischemic cardiomyopathy is the favored diagnosis. Examples of nonischemic cardiomyopathy include infectious cardiomyopathy, hypertrophic cardiomyopathy, sarcoidosis and amyloidosis. While ischemic cardiomyopathy is not favored, it is to be excluded whenever delayed contrast-enhanced cardiac MRI images are obtained.

Infectious Cardiomyopathy
Inflammation of the myocardium, or myocarditis, can result from several infectious, systemic or toxic etiologies. The majority of infectious processes are viral with the Coxsackie B virus responsible for about 50% of cases. The most common systemic processes to result in myocarditis include systemic lupus erythematosus (SLE), inflammatory bowel disease, and mixed connective tissue diseases. Toxic exposures from hydralazine, procainamide, hep- arin, warfarin, and anthracyclines are another source of myocarditis. The clinical picture of myocarditis is often vague; therefore, MR imaging may help establish a diagnosis or occasionally guide endomyocardial biopsy, when necessary. When there is a clear diagnostic picture with self-limited disease, however, cross-sectional imaging is usually not required. The typical MRI appearance of myocarditis is delayed ventricular wall enhancement that does not conform to a vascular distribution. Myocarditis typically affects the epicardial region of the myocardium with the lateral wall most often involved. Enhancement becomes more diffuse and faint over time, with or without associated wall motion abnormalities. As expected, there is often associated T2 hyperintense signal from inflammation. Pericardial effusions can occur but are often small.

In the chronic setting, fibrous tissue can replace inflammatory cells leading to an enhancing scar, similar to ischemic cardiomyopathy. The distinction from ischemic cardiomyopathy may be made if the scar does not conform to a vascular distribution or if there is preferential subepicardial involvement, as opposed to subendocardial involvement seen with ischemic disease. Involvement of the cardiac apex can be particularly difficult to distinguish from ischemic disease.

Ischemic Cardiomyopathy
Myocardial scarring and wall motion abnormality in a coronary artery distribution are the hallmarks of ischemic disease. The degree of wall involvement determines the likelihood of recovering contractility after reperfusion. Subendocardial infarction and scar involving <50% of the wall thickness is associated with reversible ischemia of the remaining myocardium, which can regain or improve contractility when corrected (often with reperfusion procedures, such as stenting or revascularization). Transmural enhancement, on the other hand, is associated with decreased likelihood of recovery. In chronic cases, transmural infarctions will typically show myocardial thinning, while subendocardial infarctions frequently have normal wall thickness.

Hypertrophic Cardiomyopathy
Hypertrophic cardiomyopathy (HCM) is the most common inherited cardiac disorder and is autosomal dominant in most cases. HCM is characterized by diffuse or focal left ventricular wall thickening, >1.5 cm at end diastole but typically closer to 2.5 cm. Asymmetric interventricular septal involvement is the most common pattern; however, symmetric, apical (first described in Asian populations), mass-like, and end-stage or burned out (similar in appearance to dilated cardiomyopathy) forms also occur. Right ventricular involvement occurs in around 20% of patients. In addition to myocardial wall thickening, important MR imaging findings include midmyocardial patchy delayed enhancement – in particular at the right ventricular insertion points in the septum – in about 80% of patients, regional wall dyskinesia, and systolic anterior motion (SAM) of the mitral valve contributing to subaortic outflow tract obstruction.3,5,6,7

Sarcoidosis
Sarcoidosis is a multisystem non-necrotizing (noncaseating) granulomatous disease that most commonly involves the lungs and lymph nodes. It presents in young to middle-aged adults with a higher incidence in African-Americans and slight increased incidence in women. Approximately 5% of sarcoid patients will develop symptomatic cardiac disease, although up to 50% of patients have myocardial involvement on autopsy. Symptomatic patients often present with heart block, restrictive cardiomyopathy, and/or dysrhythmias and are at increased risk for sudden death. Delayed enhancement is typically patchy with involvement of the midmyocardium or subepicardium. The basal septum and lateral left ventricular free wall are often involved with sparing of the papillary muscles and right ventricular wall. Both chronic sarcoid and severe myocarditis can have transmural, whole-heart hyperenhancement, which may be distinguished from chronic ischemic disease by the lack of myocardial thinning. Sarcoidosis can also mimic myocarditis with subepicardial enhancement.8,9
**Amyloidosis**

The heart is affected pathologically in up to 90% of systemic light chain (AL) amyloidosis patients. Systemic amyloid AL amyloidosis is associated with monoclonal plasma cell dyscrasias, including multiple myeloma. Asymptomatic ECG abnormalities, followed by restrictive cardiomyopathy and congestive heart failure, are common clinical manifestations. The characteristic MR imaging appearance of amyloidosis consists of diffuse left ventricular subendocardial delayed enhancement with biventricular myocardial thickening. Diffuse biventricular delayed enhancement is another common appearance. A classic pitfall in amyloid imaging can occur during selection of inversion time to null the myocardial signal due to the intrinsic T1 prolongation caused by amyloid protein. With diffuse amyloid involvement of the myocardium, significant enhancement may be missed and mistakenly imaged as normal due to inappropriate nulling of this signal.

**Diagnosis**

Infectious cardiomyopathy

**Summary**

The pattern of delayed enhancement on cardiac MRI, when correlated with the appropriate clinical context and additional MRI findings, can typically differentiate nonischemic from ischemic cardiomyopathies. Delayed enhancement affecting the midmyocardial and epicardial regions in a nonvascular distribution is characteristic of nonischemic cardiomyopathies, while ischemic cardiomyopathies tend to have subendocardial or transmural delayed enhancement in a vascular distribution with associated wall motion abnormalities.

**References**