Giant Cell Myocarditis Depicted by Cardiac Magnetic Resonance Imaging

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Summary: Giant cell myocarditis is a rare condition, the cardiac magnetic resonance imaging findings of which have not been previously described. The disease usually occurs in young previously healthy people and is typified by rapidly progressive cardiac dysfunction, often requiring cardiac transplantation. A case of giant cell myocarditis is presented, with associated pathologic and imaging findings.

Key Words: cardiac magnetic resonance imaging, heart failure, myocarditis, giant cell myocarditis

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CASE REPORT

A 33-year-old woman of Chinese descent presented to the emergency department with palpitations and syncope, and was found to be in ventricular tachycardia. Transthoracic echocardiography and left heart catheterization demonstrated normal left ventricular systolic function and normal coronary arteries. Cine cardiac magnetic resonance imaging (MRI) demonstrated right ventricular dilatation and systolic dysfunction with a right ventricular ejection fraction of 25% (Fig. 1). The left ventricular ejection fraction was minimally depressed at 49%. Delayed postcontrast images demonstrated abnormal subendocardial enhancement involving a large segment of the right ventricular free wall and several smaller foci of abnormal subendocardial enhancement in the left ventricle (Fig. 2).

An initial endomyocardial biopsy was nonspecific but demonstrated an inflammatory infiltrate, predominantly mononuclear. Cardiac function progressively worsened, with the left ventricular ejection fraction falling to 10% by transthoracic echocardiography performed approximately 4 weeks after the MRI. Left and right ventricular assist devices were placed as a bridge to heart transplantation. Surgical pathology specimens demonstrated exuberant mononuclear-macrophage infiltrate with intermixed multinucleated giant cells (Fig. 3). There was also extensive myocardial destruction with myocyte dropout as well as granulation tissue and early fibrosis. There was no evidence of granuloma formation or viral inclusions. Findings were consistent with giant cell myocarditis.

DISCUSSION

Giant cell myocarditis is a rare condition, the cardiac MRI findings of which have not been previously described. Delayed enhancement MRI is most commonly used for identification of fibrosis associated with myocardial infarction, but the same technique is useful to depict the extent and location of inflammatory cardiomyopathy. In this case, the biventricular nonanatomic distribution excluded coronary artery disease. Another diagnostic consideration was arrhythmogenic right ventricular dysplasia attributable to ventricular arrhythmia and symptoms of right ventricular failure. Arrhythmogenic right ventricular dysplasia presents primarily with right-sided enhancement, however. Viral myocarditis or infiltrative disease such as sarcoidosis was the primary diagnostic consideration based on the imaging findings.

Whereas giant cell myocarditis has been reported in association with various autoimmune conditions, it usually occurs in young previously healthy people. The disease is typified by rapidly progressive cardiac dysfunction, most commonly with biventricular failure, and ventricular tachycardia is also a common presentation. Giant cell myocarditis is treated with immunosuppression but often requires cardiac transplantation because of progressive cardiac failure. Median transplant-free survival is approximately 3 months without treatment and is reported to be longer with immunosuppressive therapy. Less typically, giant cell myocarditis may present with a chronic course of dilated cardiomyopathy. Classic pathologic features of giant cell myocarditis include prominent serpiginous regions of myocyte destruction and necrosis as well as multinucleated giant cells at the margins of necrosis.

Like other forms of myocarditis, the diagnosis of giant cell myocarditis is often made only in retrospect, on autopsy or transplant specimens. Cardiac MRI may assist in the diagnosis by depicting areas of involvement, directing biopsy, excluding other types of cardiomyopathy, and providing quantitative measures of right ventricular and left ventricular function for assessing disease progression and prognosis.
FIGURE 1. Horizontal long-axis, cine, steady-state free precession images at end-diastole (A) and end-systole (B) demonstrate moderately depressed right ventricular systolic function and minimally depressed left ventricular systolic function.

FIGURE 2. Myocardium-suppressed, delayed, postcontrast images obtained in the short axis (A, B) and axial plane (C, D) demonstrate abnormal subendocardial delayed enhancement in the right ventricular free wall (black arrowheads) as well as patchy abnormal enhancement in the left ventricle (white arrows).
FIGURE 3. A, Pathology specimen obtained at the time of ventricular assist device placement demonstrates exuberant inflammatory infiltrate consisting predominantly of macrophages and mononuclear cells with occasional eosinophils (hematoxylin–eosin, 200×). B, Intermixed in the infiltrate are collections of multinucleated giant cells; under polarization, they show no evidence of foreign material and are not members of discrete granulomata (CD68 stain, 400×). The destructive nature of this infiltrate is consistent with active giant cell myocarditis.

REFERENCES


