An Atypical Presentation of Endomyocardial Fibrosis Diagnosed by Cardiac MRI

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Cardiovascular MRI (CMR) is complementary to transthoracic echocardiography in the characterization of cardiac masses by offering a more complete view through its unlimited imaging planes and tissue characterization with the use of multiple imaging sequences. In this instance, CMR was able to noninvasively diagnose an atypical presentation of endomyocardial fibrosis (EMF) in a 44-year-old woman who presented with new-onset heart failure and a cardiac mass. Transthoracic echocardiography showed a 9-cm² mass arising from the inferoposterior wall of the left ventricle (Figure 1, Supplementary Videos 1 and 2) resulting in a left ventricular outflow tract obstruction with a dynamic 30-mm Hg gradient. The posterior leaflet was tented, producing severe mitral regurgitation (Figure 1, Supplementary Videos 3 and 4). Left ventricular contractility appeared normal. Cardiac catheterization revealed moderate pulmonary hypertension with an elevated pulmonary capillary wedge.

CMR further defined the mass as hypointense (on cine images), heterogeneous, and sessile arising from the left ventricular basal lateral wall (Figure 2A, Supplementary Video 5). The papillary muscles were surrounded by the mass resulting in severe mitral regurgitation with a posterolateral jet. The mass did not appear to be perfused (Figures 2B and 2C, Supplementary Video 6). There was diffuse delayed enhancement of the left ventricular endocardium and focal enhancement of the right ventricular apical endocardium (Figure 2D). No enhancement of the myocardium was visualized. The mass itself was hyperintense with short inversion times and hypointense with long inversion times consistent with overlying thrombus.

Intraoperatively, a soft, yellow-green mass measuring 7×5 cm was debulked from the left ventricle. No mass was visualized in either apex. The friable mass, which was adherent to the posterior mitral valve leaflet and encased the papillary muscles, was removed. A firmly adherent fibrotic base was peeled away from the endocardium (Figure 3). The mitral valve annulus was debulked, and a bioprosthetic valve was placed. Histopathology revealed organizing and disorganized thrombus involving the mitral valve leaflets, papillary muscle, and ventricular endocardium, consistent with EMF.

EMF is characterized by deposition of fibrous tissue on the endocardium of either ventricle, leading to obstruction of the inflow tract. It commonly involves the ventricular apices and papillary muscles, which can lead to mitral or tricuspid regurgitation and heart failure.

Echocardiography is a useful diagnostic tool for EMF. Typical findings include dilated atria, obliterated ventricular cavities, and normal systolic function. Thrombus is commonly in the apex with extension to the posterior papillary muscles, with concomitant mitral or tricuspid regurgitation. Mitral and tricuspid inflow velocities by Doppler demonstrate a restrictive pattern with short deceleration times. In this patient, the thrombus was located in the posterolateral base, and except for mitral regurgitation, she lacked these typical echocardiographic features.

Although the use of CMR in the diagnosis of EMF is limited in the literature, this imaging modality led to the diagnosis preoperatively. The delayed enhancement of the endocardium in the absence of significant myocardial delayed enhancement and in the setting of normal systolic function is found in few other diseases. One exception is cardiac amyloidosis, in which a pattern of circumferential subendocardial enhancement occurs in most patients, albeit with varying extension into the neighboring myocardium. Although characteristics of restrictive physiology may also be present on imaging modalities in this disease entity, the presence of a mass extending into the ventricular cavity is inconsistent with cardiac amyloidosis. Thus, the ability to identify thrombus, observe isolated delayed enhancement of the endocardium, and visualize other structural and valvular abnormalities made CMR the ideal imaging modality for diagnosing EMF, even with atypical findings on echocardiography.

In this patient, the pulmonary artery pressures decreased postoperatively. Two weeks later, the patient was ambulatory with assistance and discharged to a rehabilitation facility.

Disclosures
None.

References

From the Zena and Michael A. Wiener Cardiovascular Institute, Marie-Josée and Henry R. Kravis Center for Cardiovascular Health, Mount Sinai School of Medicine, New York, NY. The online-only Data Supplement is available at http://circ失败.ahajournals.org/cgi/content/full/CIRCEP.108.811109/DC1.
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Figure 1. Transthoracic echocardiogram of intracardiac mass. A 9-cm² mass (asterisk) from the inferoposterior wall of the left ventricle encompassing the posterolateral papillary muscle in the parasternal long (A) and apical 4-chamber (B) views. The mass led to severe mitral regurgitation as seen in the parasternal long (C) and apical 3-chamber (D) views.
Figure 2. CMR of intracardiac mass. A, CMR confirmed the large, hypointense, heterogeneous sessile mass (asterisk) arising from the left ventricular basal lateral wall leading to severe mitral regurgitation and outflow obstruction. Serial perfusion images reveal uptake in the right and left ventricles (B) and myocardium (C), without any uptake in the mass. D, Delayed enhancement images showed uptake in the left ventricular endocardium (arrows) as well as focal enhancement of the right ventricular apical endocardium. No delayed enhancement of the myocardium was visualized.
Figure 3. Intraoperative images of cardiac mass. A, Mass through the mitral valve from the left atrium. A gelatinous mass almost completely obstructs the mitral valve orifice from the anterior mitral leaflet (retracted), and the posterior mitral leaflet and is partly adherent to the interventricular septum. All of the chordal apparatus and papillary muscles are encased in this mass. B, Left ventricular outflow tract through the aortic valve. The interventricular septum is the muscle prominence at the top of the image, and the ventricular aspect of the anterior mitral leaflet can be seen at the bottom of the screen. Again, a gelatinous mass is seen, which almost totally occludes the left ventricular outflow tract. The mass encases the papillary muscles and is also partly adherent to the septum.
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