Case Report

Typical Cardiac Magnetic Resonance Imaging Findings of Cardiac Amyloidosis

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We report the case of a 62-year-old man who presented with shortness of breath, lower extremity edema and clinical signs of congestive heart failure. Transthoracic echocardiography demonstrated concentric left ventricular hypertrophy with severe diastolic dysfunction and biatrial enlargement. After aggressive diuresis and clinical improvement, a cardiac magnetic resonance imaging (CMR) examination was performed. The study confirmed the presence of concentric left ventricular hypertrophy with borderline systolic function and impaired diastolic function. Delayed contrast-enhanced imaging indicated diffuse enhancement and lack of adequate signal suppression of the left ventricular myocardium, suggesting the diagnosis of infiltrative heart disease. Rectal biopsy confirmed the diagnosis of amyloidosis. This report presents the typical noninvasive imaging findings of cardiac amyloidosis.

Cardiac amyloidosis is a rare condition with a wide range of clinical manifestations. Cardiac magnetic resonance imaging (CMR) provides valuable information that can be of assistance in establishing the diagnosis of this disease.

Case presentation

We report the case of a 62-year-old man with hypertension and family history of coronary artery disease, who presented with progressive shortness of breath, lower extremity edema and clinical signs of congestive heart failure. The patient was admitted to the hospital for evaluation and treatment. An acute coronary event was ruled out by cardiac enzymes and serial electrocardiograms. Transthoracic echocardiography demonstrated concentric left ventricular hypertrophy with severe diastolic dysfunction and biatrial enlargement. Intravenous diuretics were administered with prompt improvement of

the clinical status and a CMR examination was ordered.

Black blood transverse T1-weighted images of the heart were acquired before and after contrast administration (gadodiamide 0.2 mmol/kg). T2-weighted images and delayed-enhanced images using an inversion-recovery technique with heavy T1-weighting were also obtained. Functional assessment was performed with a bright-blood breath-hold steady-state free-precession cine technique. Lastly, real-time images were obtained in the short-axis orientation during deep inspiration and expiration.

The CMR study demonstrated left ventricular hypertrophy (117 g/m^2, normal <95 g/m^2) involving predominantly the basal segments of the ventricle (diastolic diameter ~15-16 mm) (Figure 1). Left ventricular systolic function was marginally decreased (ejection fraction 60%, normal >61%). Right ventricular systolic function was qualitatively normal. There were no findings of constrictive physiol-
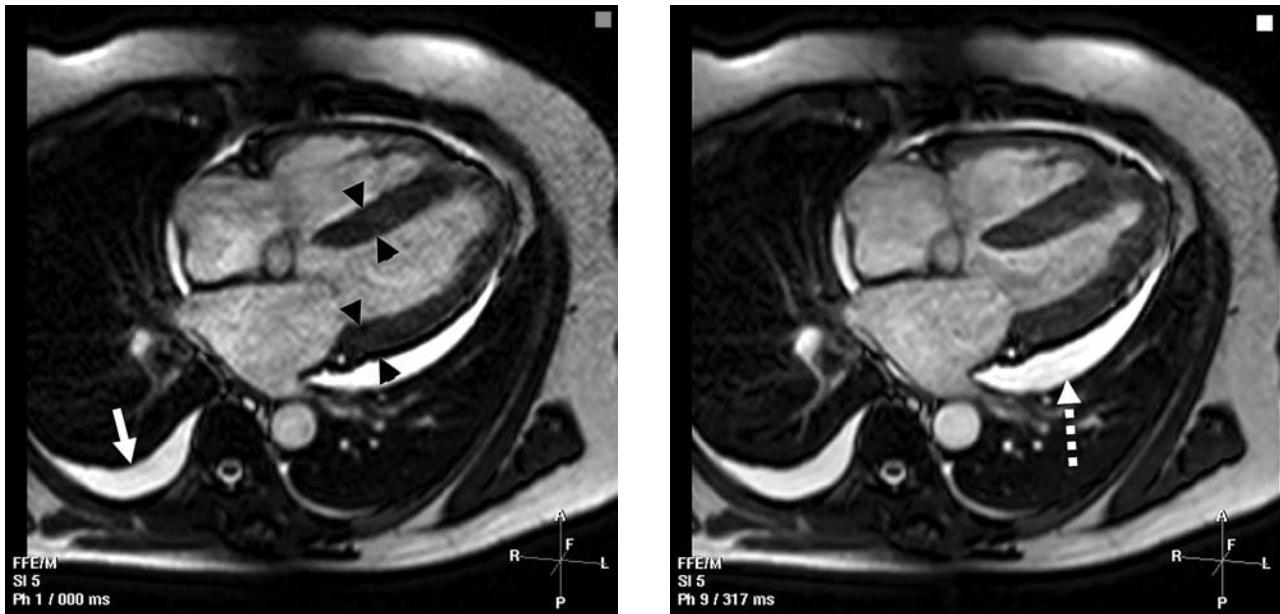
Figure 1. Diastolic (left) and systolic (right) images from the functional cardiac magnetic resonance study in the four-chamber orientation, demonstrating increased thickness of the myocardial walls (arrowheads) and borderline left ventricular function. A small right pleural effusion (solid arrow) and a small pericardial effusion (dotted arrow) are present.

ogy. Biatrial enlargement was noted. A small right pleural effusion (~175 ml) and a medium-size pericardial effusion (~110 ml) were noted. Myocardial signal intensity was homogeneous in the pre- and early post-contrast images. Delayed-enhanced imaging indicated diffuse myocardial signal enhancement and lack of signal suppression of the left ventricular myocardium with several different inversion times and at several time points, even quite late after contrast agent administration (Figure 2). The above findings were characteristic of infiltrative heart disease and particularly cardiac amyloidosis, with diffuse deposition of amyloid in the extracellular matrix.

Rectal biopsy confirmed the diagnosis of amyloidosis in this patient. An extensive workup, including serum and protein electrophoresis and bone marrow biopsy, revealed multiple myeloma as the underlying disease.

Discussion

Amyloidosis is an uncommon condition, characterized by interstitial deposition of insoluble abnormal fibrous protein aggregates. In patients with amyloidosis, a familial type with frequent cardiac involvement has been described. In non-familial cases, also known as "AA systemic" or "secondary" amyloidosis, the disease is often associated with chronic inflammation or malignancies[1,2] and has a lower incidence of cardiac involvement. In cases where no underlying disease can be identified, the term "primary" or "AL systemic" amyloidosis is used. In these patients, involvement of the heart (infiltration of the extracellular space by glycoproteins) is very frequent and has been reported in up to 90% of patients with AL systemic amyloidosis.[3]

Clinically, cardiac amyloidosis is almost always symptomatic.[4] Dyspnea during exertion or at rest is common. The presence of clinical congestive heart failure suggests a poor prognosis.[5,6] Arrhythmias, including atrial fibrillation and atrioventricular conduction disturbances, are common. Electrocardiographic abnormalities include low voltage and a pseudo-infarction pattern.[7]

A small left ventricular cavity size with biventricular and atrial septal thickening, increased echogenicity and a granular or "sparkling" appearance of the myocardium, biatrial enlargement and ventricular diastolic dysfunction with a restrictive left ventricular filling pattern at late stages, are the most characteristic echocardiographic findings for cardiac involvement in amyloidosis.[7,8] Echocardiography may be highly suggestive of the disease in many cases, but several patients may not have all the above findings, or have limited acoustic windows that preclude accurate assessment. CMR can offer additional diagnostic information, as it provides precise anatomic and
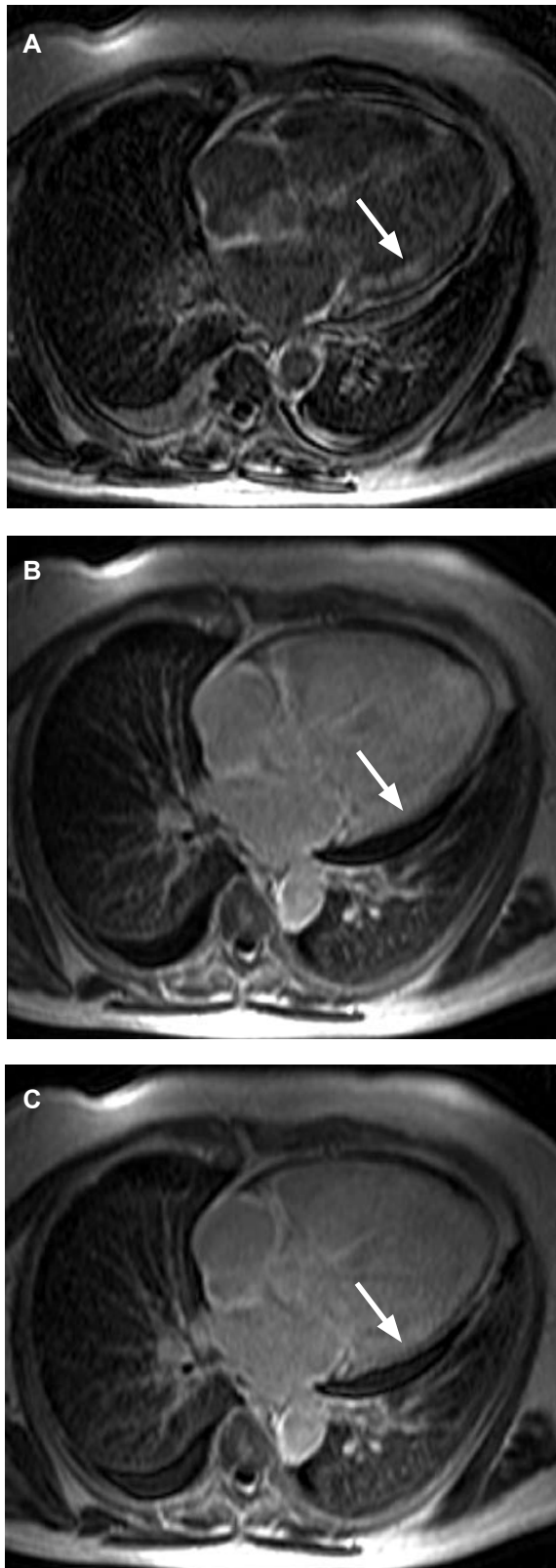
Figure 2. Delayed gadolinium-enhanced images at the four-chamber orientation. Diffuse myocardial signal enhancement is present (arrows) despite the use of multiple (A, B, C) inversion times (TI), that all fail to suppress the myocardial signal.

functional assessment, and, importantly, noninvasive myocardial tissue characterization, particularly with paramagnetic contrast enhancement. The expansion of interstitial space increases the volume of distribution of gadolinium-based agents. Therefore, there is usually a homogeneous pattern of enhancement, such that the signal from the myocardium cannot be adequately suppressed and differentiated from the adjacent blood pool with delayed-enhanced inversion recovery sequences.[9,10] A heterogeneous pattern of late enhancement has also been reported, presumably due to variable deposition of amyloid in the myocardium.[11,12] Early gadolinium kinetics may also have value for the diagnosis of cardiac amyloidosis. The subendocardial longitudinal relaxation time (T1) in patients with cardiac amyloidosis is shorter than in normal individuals.[9] This decrease is inversely correlated with markers of myocardial amyloid load, including left ventricular mass, wall thickness, interatrial septal thickness and diastolic function.[9] Perugini et al[12] reported an atypically dark appearance of the blood pool, which reflects the similar myocardial and blood T1 values attributable to high myocardial uptake and fast blood pool washout.

In conclusion, CMR is a valuable noninvasive imaging method in the study of infiltrative cardiomyopathies and particularly amyloidosis. CMR provides data on anatomy, function and tissue characterization and can help establish the diagnosis, as exemplified in the case discussed here.

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