Case Reports

Cardiac MRI in a Patient with Amyloidosis

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Abstract

A 42-year-old man presented with orthopnea, paroxysmal nocturnal dyspnea, and ascites, which had progressed for the previous two months. Electrocardiogram was low voltage. Transthoracic echocardiography showed concentric left ventricular hypertrophy and increased brightness and speckling pattern in the ventricular septum, consistent with amyloidosis. Cardiac magnetic resonance imaging confirmed the echocardiographic findings, and gingival biopsy was positive for amyloidosis ***(Iranian Heart Journal 2010; 11 (2):59-61).***

Key words: magnetic resonance imaging ■ cardiac MRI ■ amyloidosis

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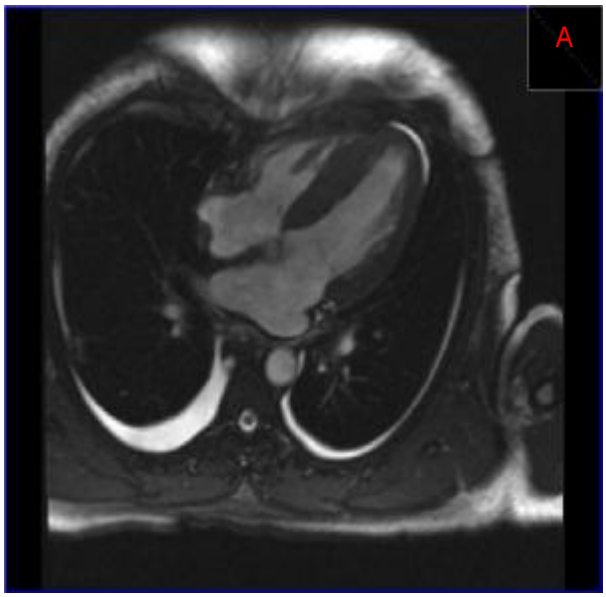
Case report

A 42-year-old man presented with orthopnea, paroxysmal nocturnal dyspnea, and ascites, which had progressed for the previous two months.

On physical examination, jugular venous pulse was prominent and there was evidence of abdominal ascites. Electrocardiogram was low voltage with left anterior hemiblock pattern. Transthoracic echocardiography was done in our center and revealed the following data: bi-atrial enlargement, left ventricular ejection fraction (LVEF) of 45-50%, concentric LV hypertrophy, severe grade III diastolic dysfunction, pulmonary arterial hypertension, SPAP 60mmHg without any intracardiac shunt, increased brightness, and speckling pattern in the ventricular septum consistent with amyloidosis.

Cardiac magnetic resonance imaging was performed with a 1.5 T Siemens Avanto using protocols for functional and flow study with T1, retro IPAT, magnitude and phase-sensitive techniques (different planes, two-chamber, three-chamber, four-chamber, short-axis views, and MRA with Gadolinium) and revealed: normal LV size with mildly reduced systolic function, LVEF 45%, LV mass (ED: 177.58g), average LV mass (avg 167.25g), normal right ventricle (RV) size with mildly reduced systolic function (RVEF 50%), RV pressure overload, mildly increased interatrial septal thickness (5.3 mm), and dilated inferior vena cava (Fig.1).



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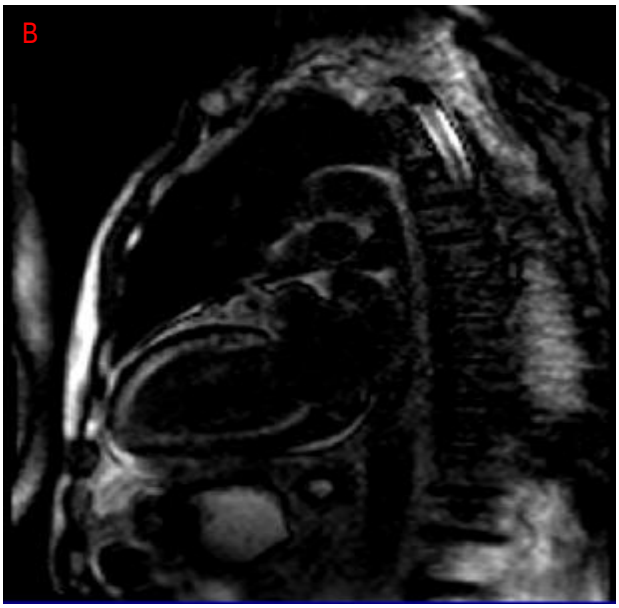

Fig. 1, A, B. View of four chambers demonstrated increased thickness of left ventricle walls and interatrial septum, bilateral pleural effusion, and biatrial enlargement.

In the first pass imaging, there was diffuse global subendocardial hypoperfusion.
In the late enhancement imaging, achievement of myocardial nulling with multiple TI was difficult and diffuse global subendocardial enhancement with extension to intramural LV myocardium together with deposition of Gd in the interatrial septum and RV inferior wall was visible. Given these findings, restrictive cardiomyopathy secondary to cardiac amyloidosis deposition was considered (Fig. 2). On coronary angiography, the patient had normal epicardial coronary arteries. Endomyocardial, fat pad, and gingival biopsies were done and ultimately gingival biopsy was positive for amyloidosis.

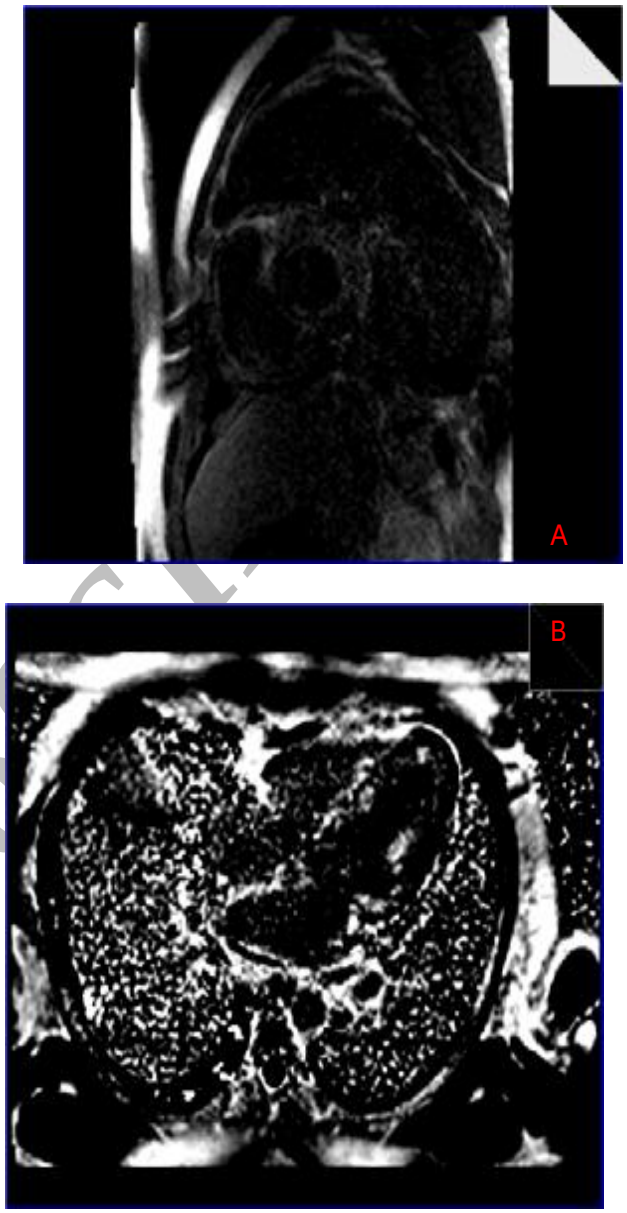

Fig. 2, A, B. Views of PSIR sequence for delayed Gd enhancement. Myocardial nulling with multiple TIs was difficult. Diffuse global subendocardial enhancement with extension to mural left ventricle myocardial wall (thin white arrows) and also deposition of Gd in interatrial septum (thick white arrow) are demonstrated.

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Discussion

Amyloidosis represents a diverse group of diseases characterized by the common factor of deposition of twisted ß-pleated sheet fibrils (amyloid), formed as a result of the misfolding of various proteins produced in several different pathological states.[1] Cardiac involvement is the most common cause of death in systemic amyloidosis, and cardiac amyloidosis is the most common identifiable cause of restrictive cardiomyopathy.[2] Cine-MRI imaging typically demonstrates global hypertrophy, often involving the right ventricle as well. Additionally, thickening of the interatrial septum and of the atrial walls is a finding strongly suggestive of the diagnosis.[4,5] Delayed-enhancement imaging typically demonstrates diffuse LV hyper-enhancement in these patients. Although often subendocardial, the pattern is clearly distinct from the usual coronary artery disease pattern in that the hyper-enhancement does not follow a coronary artery distribution.[3]

From an imaging point-of-view, the presence of diffuse myocardial involvement may make setting the parameters for delayed-enhancement imaging problematic. Specifically, it may be difficult to determine the optimal inversion time that will null "normal" myocardium, as there may be few areas that are completely normal. Additionally, unlike the situation in normal patients, in whom the LV blood pool (post-contrast administration) has far shorter T1 relaxation than myocardium, in patients with cardiac amyloidosis, the myocardium may have similar T1 to that of the blood pool and image intensities may be similar on delayed-enhancement imaging. Reports indicate that this finding is likely due to the profound expansion of the extracellular volume of the myocardium, as well as the more rapid clearance of gadolinium contrast from the blood pool. This results in the near equilibration of the blood pool and myocardial gadolinium concentrations, and partially explains the difficulty in nulling.

Conclusion

The potential role of cardiac MRI in the assessment of cardiac involvement in amyloidosis is well known. In this patient, this imaging modality was key in establishing the correct etiology of LV hypertrophy.

References

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