Cardiac Magnetic Resonance Imaging in Amyloidosis

To the Editor:

We present a case that illustrates the usefulness of cardiac magnetic resonance (CMR) in amyloid heart disease. A 71-year-old woman with diabetes went to the emergency room for syncope of 2 months’ evolution, with 2-3 daily episodes but no focal neurological deficits. The examination revealed pallor of the skin and mucosa and a grade II/VI apical systolic murmur. Blood pressure and heart rate in decubitus and standing position were 130/70 mm Hg and 68 bpm, and 80/40 mm Hg and 74 bpm, respectively. The rest of the examination was normal.

The analyses indicated normochromic, normocytic anemia; elevated urea, creatinine, and sedimentation rate; hypalbuminemia, hypergammaglobulinemia with monoclonal peak; elevated immunoglobulin G, and enlarged lambda chains in serum and urine.

Figure 1. Inversion-recovery sequence after administration of a gadolinium-DTPA bolus, 4-chamber view. Diffuse myocardial thickening and diffuse subendocardial gadolinium enhancement are observed, mainly in the lateral wall of the left ventricle.
and loss of longitudinal systolic function; and
gadolinium kinetics in blood and myocardium, with subendocardial
the lateral LV wall (Figures 1 and 2), accelerated gadolin-
ing of the right ventricular free wall and interatrial septum; c)
s) of the right ventricular free wall and interatrial septum; b) slight thicke-
ning of the right ventricular free wall and interatrial septum; c) aortic valve thickening, mild mitral and aortic regurgita-
tion; d) diffuse, subendocardial late enhancement, mainly in
the lateral LV wall (Figures 1 and 2), accelerated gadolinium
kinetics in blood and myocardium, with subendocardial T1 of 510 ms 4 min following gadolinium administration; and e) mild pericardial and pleural effusion.

Based on these findings, a sural nerve biopsy was per-
formed and primary amyloidosis was diagnosed. Because of the
patient’s age and the fact that she was not eligible for
bone marrow transplantation, melphalan and prednisone
therapy was initiated. The patient died some months later of
progressive heart failure.

Amyloidosis is a complex systemic disease characterized
by extracellular deposits of amyloid fibrillary protein in the
body.1 The diagnosis of this condition is usually established
in advanced stages; structural heart disease is the most fre-
quent cause of death.17 Treatment for primary amyloidosis is
unsatisfactory, although some improvement in survival and
functional status has been attained with alkylating agents.4

The patient did not respond well in this case, probably due
to the advanced state of the disease. Doppler echocardiography, the initial imaging technique,
can detect wall thickening with small chambers, dilated atria, valvular and interatrial septum thickening, and diastolic and systolic dysfunction.12 On occasions, myocardial thickening is not uniform and the features resemble hyper-
trrophic cardiomyopathy, although in amyloidosis the elec-
tric cardiogram usually exhibits low voltage. The granular sparkling appearance caused by the deposits may be diffi-
cult to visualize. In the present case, the echocardiogram ac-
tually did not detect the granular texture, or the severe valve
regurgitation or pericardial effusion, possibly because they
were mild or had not yet appeared at the time.

Cardiac magnetic resonance can provide more complete
information for the diagnosis of this entity. In addition to the
morphological findings described on echocardiography, amyloid deposits specifically affect the gadolinium kinetics
in the blood and myocardium.12 Following gadolinium ad-
ministration, there is greater shortening of the subendocar-
dial T1 and a larger difference between subendocardial T1 and blood; hence a cut-off point can be established with
good diagnostic accuracy. In addition, there is a typical pat-
tern of diffuse late subendocardial enhancement, which re-
ffects the greater infiltration of amyloidosis in the subendo-
cardium with expansion of the extracelllar space.1

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179

Figure 2. Identical sequence as in the previous figure, basal, mid, and apical short-axis views. More intense diffuse subendocardial gadolinium enhance-
ment in the side wall of the left ventri-
cle is evident.