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CARDIAC AMYLOIDOSIS DIAGNOSED ON MRI

P. Lu[1], H. Van Acker[2], P. Waer[1]

The frequency of cardiac involvement varies among other types of amyloidosis. Cardiac amyloidosis leads to systolic and diastolic dysfunction with symptoms of heart failure. Cardiac magnetic resonance (CMR) findings are helpful in supporting the diagnosis of amyloid cardiomyopathy. We report a case of a 73-year-old man who presented with shortness of breath. Echocardiography showed a hypertrophic, diffusely hypocontractile left ventricle with a restrictive filling pattern. The diagnosis of an isolated amyloidosis was made on CMR.

Key-words: Amyloidosis – Heart, MR.

Case report

We report the case of a 73-year-old man with history of hypertension and paroxysmal atrial fibrillation, who presented with recent dyspnea. The patient was admitted to the hospital for evaluation and treatment. Echocardiography showed a hypertrophied left ventricle with a systolic and diastolic dysfunction, no significant valve disease was found. Coronarography did not show vascular impairment.

CMR was performed for assessment of the heart. It revealed a biventricular increase of wall thickness and dilated atria. The functional studies showed a decrease of systolic function with an ejection fraction of 37%. Late gadolinium enhancement images showed global subendocardial hypersignal of both ventricles and atria. Phase-contrast sequences acquired on the mitral valve and the pulmonary vein showed a type II diastolic dysfunction (pseudonormalisation). Based on MRI findings, the diagnosis of cardiac amyloidosis was highly suggested. A subsequent rectal biopsy confirmed the amyloidosis.

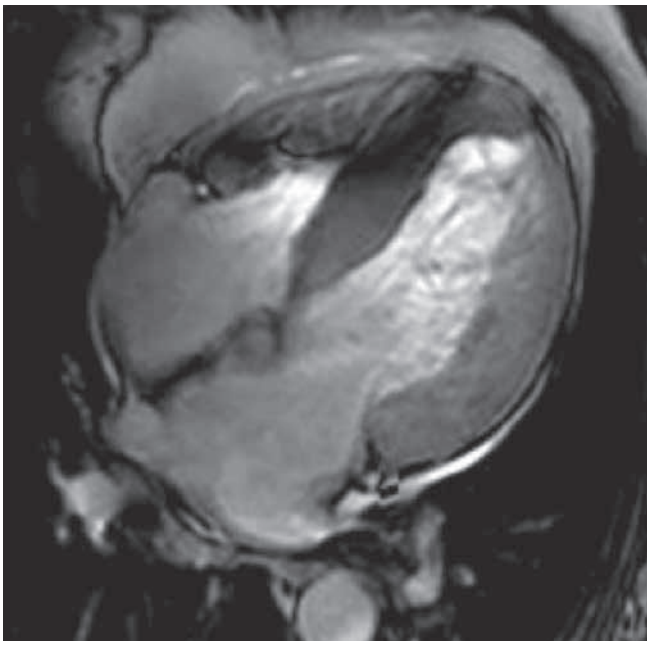

Fig. 1. — bSSFP image in four chamber orientation show a biventricular increase of wall thickness and dilated atria.

Discussion

Amyloidosis is defined by an extracellular deposit of fibrils which are composed of low molecular weight subunits. These deposits of fibrils lead to a histologic change of the affected tissues. The most common type is systemic light-chain amyloidosis (AL) and is due to deposition of protein derived from immunoglobulin light chain fragments, often associated with multiple myeloma or other monoclonal gammopathies. Cardiac involvement occurs in up to 50% or more of cases in AL.

Cardiac involvement is also found in almost all cases of senile type amyloidosis (Wild-Type ATTR) and is frequent in some types of hereditary amyloidosis (variant ATTR) while it is rather uncommon in secondary amyloidosis (AA) complicating chronic inflammatory diseases (1).

The cardiac involvement in the amyloidosis is, in most cases, secondary to a systemic disease. However an isolated cardiac involvement can occur. Cardiac amyloidosis presents as a restrictive cardiomyopathy with progressive diastolic and systolic dysfunction. Dyspnea and oedema are the most common clinical manifestations of heart failure due to cardiac amyloidosis.

The gold standard for the diagnosis of a cardiac amyloidosis remains the endocardial biopsy, but this is an invasive procedure and is related to severe complications. A strong diagnostic suspicion can be made by identifying several specific bio-clinical and imaging findings (1, 2).

The differential diagnosis of amyloid heart disease includes hypertrophic cardiomyopathy, various causes of restrictive cardiomyopathies and hypertensive heart disease.

Echocardiography is a simple non invasive technique for the morphological study and evaluation of myocardial function. However these indicators aren't specific of cardiac amyloidosis.

MRI is a non-invasive and non-operator dependent technique. It allows a three-dimensional assessment of the heart and evaluates more accurately changes in cardiac function.

CMR imaging can provide evidence strongly suggestive of amyloid cardiomyopathy. One of the characteristic features of cardiac am-

From: Department of 1. Radiology, 2. Cardiology, Europe Hospitals, Site S^te^ Elisabeth, Brussels, Belgium.
Address for correspondence: Dr P. Lu, M.D., Radiology Department, Europe Hospitals, Site S^te^ Elisabeth, Av. De Fré 206, B-1180, Brussels, Belgium.
E-mail: luanhtai2@gmail.com

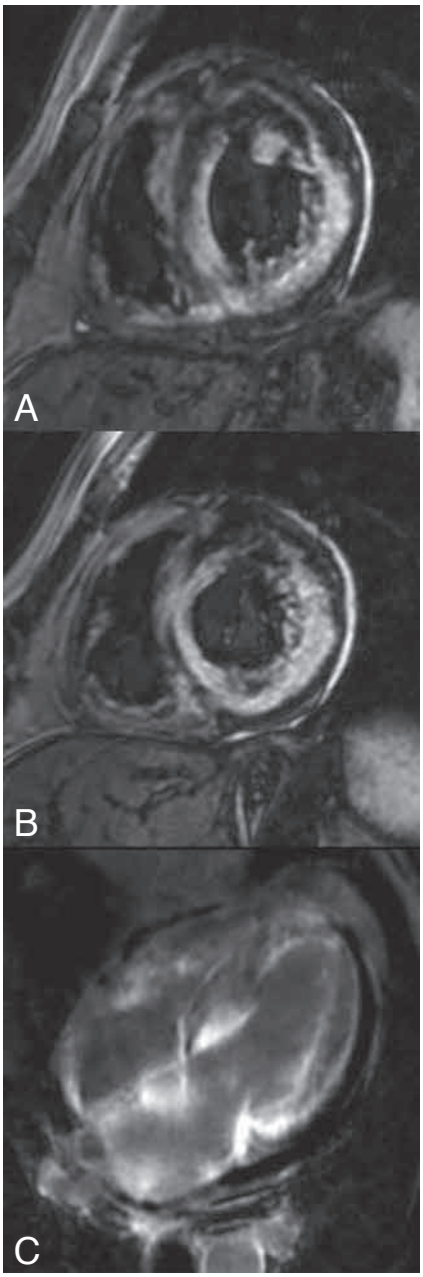

Fig. 2. — Late gadolinium enhancement images with inversion recovery saturation of the myocardium in short axis (A, B) and four chamber orientation (C) show classic amyloid global, subendocardial LGE pattern in both ventricle and atria.

yloidosis by CMR imaging is an increased ventricular wall thickness. A thickening of the interatrial septum and right atrial free wall is highly specific (3). Pericardial effusion is often present (4). CMR can also confirm the presence of myocardial dysfunction.

Finally, the "late gadolinium enhancement" (LGE) technique has an established role in diagnosis. It shows particularly a distinctive pattern of global late gadolinium enhancement (LGE) rarely seen in other cardiomyopathies. A global subendocardial LGE is the dominant pattern that corresponds to the myocardial distribution of amyloid protein in the extracellular space that is expanded. A patchy focal LGE pattern can also occur, but is rare (5).

Among other there is a positive correlation between degree of LGE and markers of disease severity (NYHA, cardiac biomarkers) and prognosis (6).

Treatment of the different types of amyloidosis varies with the cause of fibril production. Cardiac amyloidosis is generally diagnosed when there are already morphologic changes apparent in echocardiography. LGE-CMR represents a good way for screening with an adequate sensitivity and specificity of a subclinical amyloid infiltration. It allows to detect a cardiac involvement earlier than could otherwise be possible by morphologic assessment and therefore could change the clinical course and prognosis.

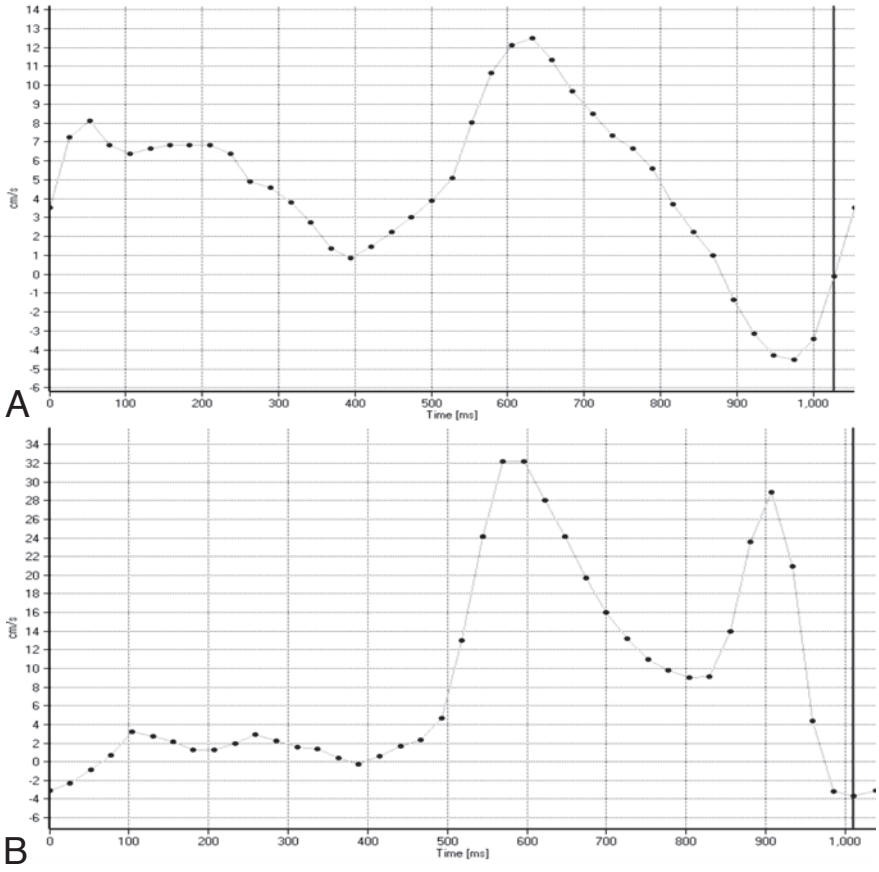

Fig. 3. — Phase contrast sequences acquired on pulmonary vein S < D (A) and mitral valve 0.75 < E/A < 1.5 (B) show a moderate diastolic dysfunction (type II).

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