Acta Scientific Gastrointestinal Disorders(ISSN: 2582-1091)

Volume 3 Issue 2 February 2020

Case Report

Amyloidosis and Cardiac Magnetic Resonance

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Received: November 14, 2019

Published: January 03, 2020

Abstract

Advances in cardiac imaging have resulted in greater recognition of light-chain (AL) cardiac amyloidosis in everyday clinical practice.

AL amyloidosis is a rare and multisystem disease characterized by deposition of misfolded immunoglobulin light chain (LC) in the heart that causes a restrictive cardiomyopathy.

We describe a 50-year-old melanodermic male that is followed at our Hospital with AL amyloidosis diagnosed since 2016, with renal and cardiac involvement. At the time of diagnosis, the echo showed left ventricular hypertrophy with moderately impaired function and typical apical sparing. A CMR demonstrated pleural and pericardial effusions; diffuse subendocardial staining pattern and remarkable difficult "nulling" of the myocardium of the left ventricle after gadolinium administration. An abdominal biopsy was positive for amyloidosis. In the last consultation, June/2018, the echo showed a moderately decreased of the left and the right ventricular function and blood and urine tests with NT-proBNPb of 4048 ng/l, hsTnTc 30 ng/L and albuminuria 0.58 g/day with MDRDd 64ml/min.

Improved medical recognition is necessary for earlier diagnosis and treatment and prompt outcomes4. Ventricular hypertrophy or heart failure without a known case maybe the first red flag for cardiac amyloid fibril infiltration and CMR should be considered.

The CMR is useful in distinguishing ventricular wall thickening as a result of amyloid infiltration. Late gadolinium enhancement adds a unique advantage in evaluating myocardial tissue characterization. Although, CMR be a non-invasive exam that is similar to cardiac biopsy, delayed hyper-enhancement imaging may lead to the development of new classification and prognostic systems.

Keywords: Cardiac Magnetic Resonance; Cardiac Amyloidosis; Late Gadolinium Enhancement and Extracellular Volume

Introduction

Advances in cardiac imaging, especially with cardiac magnetic resonance (CMR) may provide a method for evaluation of cardiac involvement and have resulted in greater recognition of light-chain (AL) cardiac amyloidosis in everyday clinical practice [1,2].

Cardiac amyloidosis is a restrictive and rare cardiomyopathy marked by extracellular accumulation of misfolded protein fragments [4,7].

This case report should be an alert for choosing the best imaging tests.

Case Report

We describe a 50-year-old melanodermic male that is followed at our Hospital with AL amyloidosis diagnosed since 2016, with renal and cardiac involvement. He has also a medical history of heart failure (NYHA II-III) with chronic pleural and pericardial effusions, 1st-degree atrioventricular block; nephrotic syndrome; α-sickle cell carrier; monoclonal gammopathy IgG lambda and bilateral pulmonary embolism (2016), chronically medicated with bisoprolol, torasemide, ivabradine, spironolactone, and rivaroxaban. At the time of diagnosis, the echocardiogram trans-thoracic showed left ventricular hypertrophy with moderately impaired function and typical apical sparing. A cardiac magnetic resonance demonstrated pleural and pericardial effusions; diffuse subendocardial staining pattern and remarkable difficult "nulling" of the myocardium of the left ventricle after gadolinium administration. An abdominal biopsy was positive for amyloidosis. He underwent 4 CyBorD cycles[1a] and Bortezomib support therapy, with complete remission. In the last consultation, June of 2018, the echocardiogram showed a moderately decreased of the left and the right ventricular function

[1a] Therapeutic with Cyclophosphamide, Bortezomib e Dexamethasone

and blood and urine tests with NT-proBNP[b] of 4048 ng/l, hsTnT[c] 30 ng/L and albuminuria 0.58 g/day with MDRD[d] 64ml/min.

Discussion and Conclusion

In AL amyloidosis, light-chain fibrils infiltrate the myocardium, interfere with cell–cell coupling, disrupt cellular integrity and may contribute to cell injury and death [7]. AL monomers elicit oxidative damage by interacting with proteins involved in cell viability and metabolism. AL also promotes cardiac stress hormone brain natriuretic peptide (BNP) expression through p38 mitogen-activated protein kinase (MAPK), reason why we had done sequencial pro-BNP measurements [8].

Cardiac amyloidosis results in increased biventricular wall thickness and ventricular stiffness. The myocardial deposition of amyloid fibrils increases extracellular volume (ECV) and results in the accumulation of gadolinium contrast. For this reason, late gadolinium enhancement (LGE) imaging has proven effective in identifying cardiac amyloidosis [8].

The cardiac amyloidosis is typified by a characteristic pattern of diffuse sub-endocardial LGE that has been associated with clinical heart failure and survival. Additionally, LGE by CMR is an independent predictor of mortality in patients with AL cardiac amyloidosis as prognostic value [8].

LGE is very useful for differentiating abnormal from normal myocardium, but this presupposes regions of normal are present. Amyloidosis is a diffuse disease, and in many instances, there is no "normal" myocardium to contrast. Quantitative imaging techniques including non-contrast (native) T1 mapping and direct extracellular volume fraction (ECV) [7] determination have been recently explored in cardiomyopathic diseases including amyloidosis [8].

Improved medical recognition is necessary for earlier diagnosis, treatment and prompt outcomes [4]. Clues to diagnosis include ventricular hypertrophy, heart failure associated with neuropathy, nephrotic syndrome, hepatomegaly, periorbital bleeding and macroglossia [3,4] and CMR should be considered.

To summarize, the CMR analyses heart's structure, ventricular volumes and function and is useful in distinguishing ventricular wall thickening as a result of amyloid infiltration. Late gadolinium enhancement adds a unique advantage in evaluating myocardial tissue characterization [6]. T1 mapping can also measure the amyloid burden and the myocyte response to infiltration with advantages for tracking change during therapy. Although, delayed hyper-enhancement imaging may lead to the development of new classification and prognostic systems [5], it is a non-invasive exam that is similar to cardiac biopsy.

This case report also illustrated the need of new studies to amyloidosis early diagnosis.

Bibliography

1. Singh A and Falk R. "A new staging system for cardiac transthyretin amyloidosis: is it already on the verge of obsolescence?". *European Heart Journal* 39 (2018): 2807-2809.

2. Maurer MS., *et al*. "Addressing Common Questions Encountered in the Diagnosis and Management of Cardiac Amyloidosis". *Circulation* 135 (2017): 1357-1377.

3. Khalid U., *et al*. "Case Report: Isolated Cardiac Amyloidosis: An Enigma Unravelled". *Methodist Debakey Cardiovascular Journal* 11 (2015): 53-58.

4. Grogan M., *et al*. "Light-chain cardiac amyloidosis: strategies to promote early diagnosis and cardiac response". *Heart* 103 (2017): 1065-1072.

5. White J., *et al*. "Prognostic utility of delayed enhancement magnetic resonance imaging in patients with systemic amyloidosis and suspected cardiac involvement". *Circulation* 114 (2006): 679.

6. Fontana M., *et al*. "Prognostic value of late gadolinium enhancement cardiovascular magnetic resonance in cardiac amyloidosis". *Circulation* 132 (2015): 1570-1579.

7. Siddigi OK and Ruberg FL. "Cardiac Amyloidosis: An Update on Pathophysiology, Diagnosis, and Treatment". *Trends in Cardiovascular Medicine* 28 (2018): 10-21.

8. Palladini G., *et al*. "The combination of high-sensitivity cardiac troponin T (hs-cTnT) at presentation and changes in N-terminal natriuretic peptide type B (NT-proBNP) after chemotherapy best predicts survival in AL amyloidosis". *Blood* 116 (2010): 3426-3430.

[1a] Therapeutic with Cyclophosphamide, Bortezomib e Dexamethasone
[b] Peptide natriuretic type B
[c] Hypersensitivity Troponin
[d] Creatinine clearance with Cockroft-Gault