Short Communication

Longitudinal Strain and Cardiac Amyloidosis: Case Reports

Strain Longitudinal Bidimensional e Amiloidose Cardíaca: Série de Casos

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Abstract

Introduction: Cardiac involvement in patients with systemic amyloidosis indicates a worse prognosis. Moreover, the diagnosis is delayed, since the signs and symptoms of this disease are nonspecific. Global longitudinal strain has been shown to be an important predictor of cardiac events. We report four cases of patients with cardiac amyloidosis submitted to ECHO Strain and magnetic resonance for comparison and diagnostic confirmation.

Case reports: The medical records of four patients with cardiac amyloidosis admitted in 2017 at a tertiary hospital were evaluated. The preservation of the apical segments of the ventricle was the pattern found in all global longitudinal strain. Magnetic resonance image showed predominantly subendocardial, diffuse and circumferential mesocardial late enhancement in all patients reported.

Discussion: On the spectrum of clinical manifestations of cardiac amyloidosis, heart failure was the most prevalent among the patients reported. Magnetic resonance image showed patterns of myocardial thickening and late subendocardial enhancement. In global longitudinal strain, we observed the preservation of the deformity in the apical segments, associated with a significant reduction of the deformity in the basal and middle segments of the left ventricle. The global longitudinal strain also allows differential diagnosis with hypertrophic cardiomyopathy and its findings were consistent with those of the magnetic resonance image, which reinforces its value in the early detection of the condition.

Keywords: Echocardiography; Amyloidosis; Magnetic Resonance Imaging; Hypertrophic Cardiomyopathy.

Resumo

Introdução: *O acometimento cardíaco em pacientes com amiloidose sistêmica indica pior prognóstico. Além disso, o diagnóstico é tardio, pois seus sinais e sintomas são inespecíficos. O strain longitudinal bidimensional tem se mostrado importante preditor de eventos cardíacos. Relatamos quatro casos de pacientes com amiloidose cardíaca submetidos ao strain longitudinal bidimensional e à ressonância magnética cardíaca, para comparação e confirmação diagnóstica.*

Relatos dos casos: *Foram avaliados os prontuários de quatro pacientes com amiloidose cardíaca admitidos em 2017 em um hospital terciário. A preservação da deformidade miocárdica nos segmentos apicais (apical sparing) do ventrículo esquerdo no strain longitudinal bidimensional foi encontrada em todos os pacientes estudados. A ressonância magnética cardíaca evidenciou realce tardio predominantemente subendocárdico, mesocárdico difuso e circunferencial em todos pacientes relatados.*

Discussão: *Diante do espectro de manifestações clínicas da amiloidose cardíaca, a insuficiência cardíaca foi a mais prevalente dentre os pacientes relatados. A ressonância magnética cardíaca mostra padrões de espessamento miocárdico e realce tardio subendocárdico. Ao strain longitudinal bidimensional, é possível observar a preservação da deformidade nos segmentos apicais, associada à redução significativa da deformidade nos segmentos basais e médios do ventrículo esquerdo. O exame também possibilita o diagnóstico diferencial com a miocardiopatia hipertrófica. Os achados ao strain longitudinal bidimensional foram condizentes com os da ressonância magnética cardíaca, o que reforça seu valor na detecção precoce da condição.*

Descritores: *Ecocardiografia; Amiloidose; Ressonância Magnética Nuclear; Cardiomiopatia Hipertrófica.*

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Introduction

Amyloidosis is a disease characterized by extracellular deposition of amyloid substance, a process that can cause structural and functional disorders in the affected tissue. At the molecular level, deposits arise from inadequate protein folding processes, generating toxic, insoluble aggregates with the capacity to settle in tissue B protein chains.[1]

Patients with amyloidosis and cardiac involvement have a worse prognosis, and their diagnosis is delayed due to nonspecific signs and symptoms of the disease.[1] Although the gold standard is endomyocardial biopsy, this new technique — two-dimensional longitudinal Strain (Strain 2D) — is another helpful early detection tool and is also considered a strong prognostic marker.[2]

We report four cases of patients with cardiac amyloidosis who underwent strain 2D echocardiography and Cardiac Magnetic Resonance Imaging (CMRI) for comparison and diagnostic confirmation.

Case report

We reviewed the medical records of four patients with cardiac amyloidosis admitted to a tertiary hospital in the city of Brasilia, in 2017, with diagnostic suspicion by strain 2D and characteristic pattern by CMRI.

Case 1 involves an 84-year-old female patient, with mild controlled hypertension, clinical presentation of heart failure (HF), who had clinical improvement after treatment, maintaining dyspnea on exertion. On echocardiogram (ECHO) (Figure 1): EF = 64%, with LV mass index = 150 g/m², septum = 18 mm; PW = 22 mm, E/e' ratio = 26, LAV index = 54 ml/m², Strain 2D = -8.8%, less reduced in the LV apical portions (Figure 2a). Cardiac catheterization showed no significant aortic valve injury. CMRI showed predominantly subendocardial and diffuse mesocardial circumferential late enhancement without respecting vascular territories.

Case 2 involves a 79-year-old female patient with HF despite optimal treatment and borderline renal failure. On ECHO: Severe functional mitral regurgitation due to annular dilation — Carpentier type IIIb (Figure 3), EF = 48%, LV mass index = 140 mg/m², septum = 15 mm, PW = 13 mm, E/e' = 18.5, LAV index = 78 ml/m², Strain 2D = -14.4%, preserved in the apical portions of the LV (Figure 2b). CMRI revealed abnormal findings similar to case 1, besides late atrial enhancement (Figure 4).

Case 3 involves a 60-year-old female patient with multiple myeloma and HF. On ECHO: EF = 63%, LV mass index = 123 mg/m², GLS = -6.5% (Figure 2c), preserved in the apical portions of the LV. CMRI revealed abnormal findings that are similar to previous cases.

Case 4 involves a 70-year-old male patient with HF and prostate cancer with previous surgeries. On ECHO: EF = 27%, LV mass index = 135 mg/m², septum = 15 mm, PP = 14 mm, E/e' = 23.7, LAV index = 47 ml/m², Strain 2D = -12.4% (Figure 2d), preserved in the apical portions of the LV. CMRI revealed abnormal findings that are similar to previous cases.

Table 1 shows the electrocardiographic findings. Reported patients were not submitted to endomyocardial biopsy until data collection.

Discussion

Amyloid infiltration in the cardiac form is usually due to light chain (AL) immunoglobulin deposits. The second form of amyloidosis with the highest prevalence of cardiac involvement is associated with transthyretin mutations (ATTR).[3] The differentiation between the two types may be suggested by some findings, such as wall thickness greater than 18 mm and septal involvement, both more frequent in the ATTR form.[4]

Clinical manifestations of amyloidosis begin nonspecifically; however, as the disease progresses and the degree of infiltration of amyloid deposits increases, the condition becomes more characteristic.[1] HF was the main clinical manifestation in all reported patients, in whom the signs of increased ventricular filling pressures were detected on echocardiography by high E/e' ratio in all patients. Myocardial infiltration in cardiac

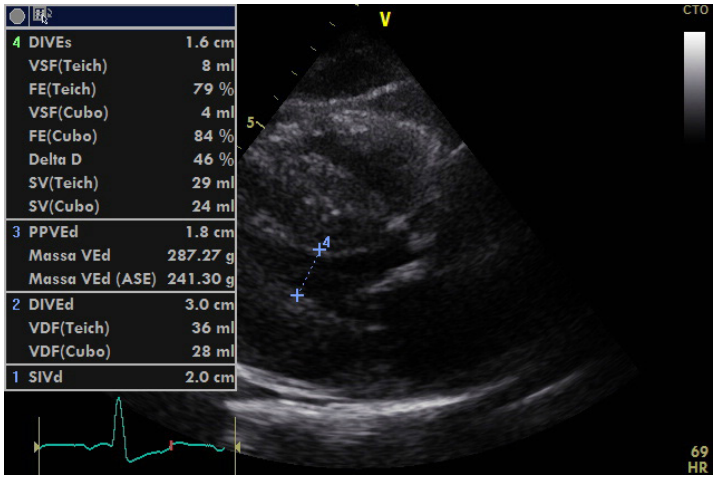

Figure 1 – *Parasternal longitudinal section on two-dimensional echocardiogram revealing significant increase in myocardial thickness in Case 1 patient.*

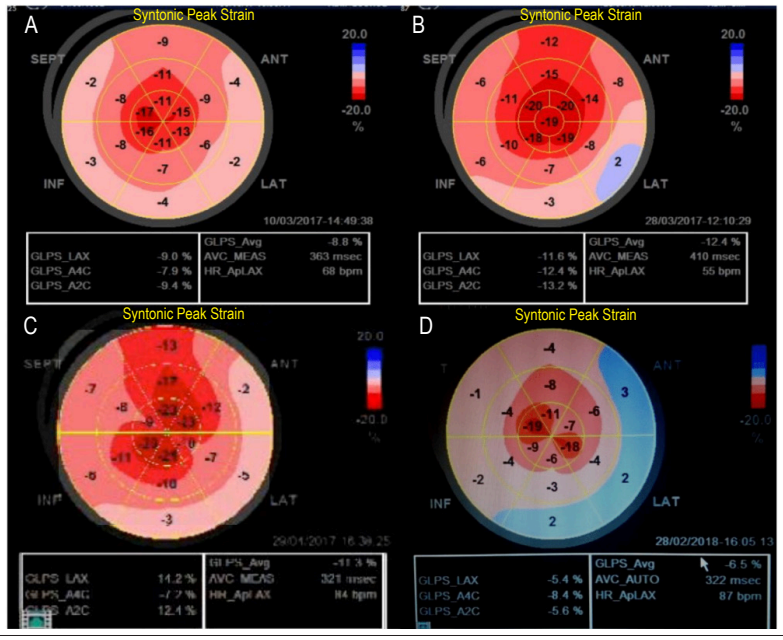

Figure 2 – *Bull's eye plot of two-dimensional longitudinal strain in cases 1 to 4 (fig 1A to 1D, respectively). Note that, in all cases, the longitudinal strain is more preserved in the apical region, unlike the middle and basal left ventricular segments.*

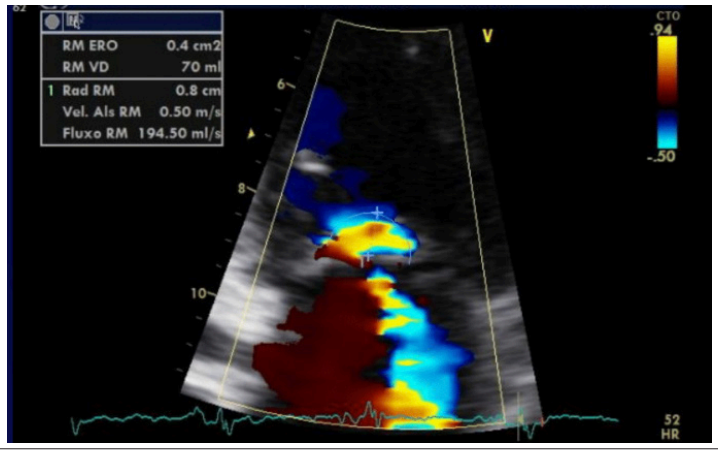

Figure 3 – *Calculation by the PISA (proximal isovelocity surface area) method demonstrating the quantification of the degree of mitral regurgitation in case 2 patient.*

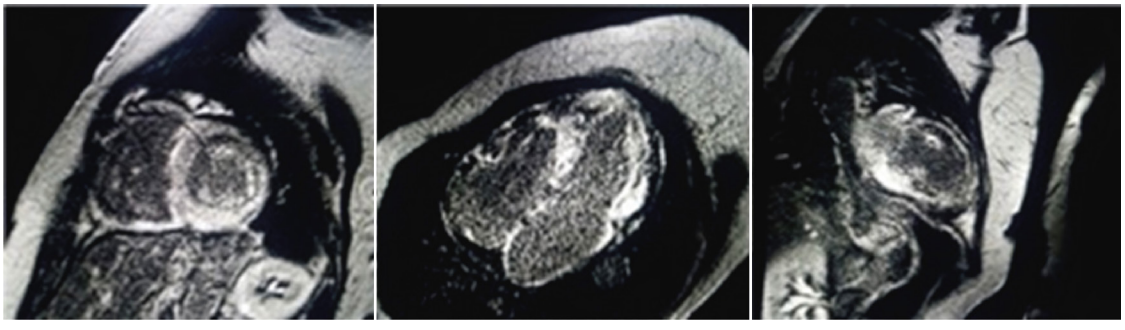

Figure 4 – *Cardiac magnetic resonance imaging showing mesocardial enhancement in the left ventricle, right atrium and left atrium, typical of amyloid infiltration.*

Table 1 - Ultrasound findings of reported cases.

Patient	LA (mm)	LAV index (ml/m²)	LVEDD (mm)	S (mm)	PW (mm)	EF (%)	LV mass (g/m²)	GLS (%)	E/e'
1	45	54	36	18	22	64	150	-8.8%	26
2	46	78	41	15	13	48	140	-14.4	18.5
3	33	48	43	19	18	63	123	-6.5	20
4	84	47	45	15	14	27	135	-12.4	23.7

LA: left atrium; LVA: left atrial volume; LVEDD: left ventricular end diastolic diameter; PW: posterior wall; EF: ejection fraction; GLS: global longitudinal strain.

amyloidosis may cause HF with preserved EF or reduced EF in later stages.

The diagnosis of cardiac amyloidosis is usually made late, mainly due to nonspecific signs and symptoms. New diagnostic techniques are proving useful in early detection.[5] They include strain 2D echocardiography, which is based on the average of the values resulting from evaluation of longitudinal myocardial fiber shortening during systole at different ventricular sites,[6] providing information that is useful in diagnosis and prognosis.[2]

Hypertrophic cardiomyopathy is an important differential diagnosis to be considered in patients with suspected cardiac amyloidosis because it has similar morphological structures. Clinically, amyloidosis is a systemic disease and may present pleural effusion, which is commonly observed in hypertrophic cardiomyopathy in later stages of the disease.[7] Strain 2D helps distinguish amyloidosis from pathologies that include left ventricular hypertrophy;[6] however, specificity is 82%.[8] In cases of cardiac amyloidosis, the most observed pattern includes apical sparing, associated with significantly reduced strain in the basal and middle ventricular segments, as evidenced in all reported cases. In hypertrophic cardiomyopathy, on the other hand, a significant reduction in strain in the hypertrophic regions is observed, with no pattern of apical sparing.[9] Patients with hypertrophic cardiomyopathy typically demonstrate a pattern of fall in longitudinal strain from the ventricular apex to the base, although this tendency may be reversed in apical hypertrophy patterns and may lead to confusion with the apical sparing most commonly observed in amyloidosis.[10]

CMRI is an alternative to the diagnosis of cardiac amyloidosis, with good sensitivity (87%) and specificity (96%) for the form associated with ATTR. CMRI identifies myocardial thickening, and interatrial septal thickening may also occur,[5] as observed in the patient of case 2. Subendocardial late enhancement pattern in the left ventricle was observed in all reported cases. These findings are consistent with the results found in strain 2D, which underscores their value in the early detection of the condition and in the differentiation with pathologies that include similar clinical conditions and myocardial hypertrophy features.

Conflict of interest

The authors declare that there is no conflict of interest regarding this manuscript.

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