Heart failure in pregnancy – another possibility beyond peripartum cardiomyopathy

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Abstract

Non-compacted myocardium is a rare primary genetic cardiomyopathy described as an abnormality in endomyocardial morphogenesis where there is interruption of myocardial fiber compaction, generating a loose trabecular tissue. It is manifested with heart failure, embolism and ventricular arrhythmia. This is the case of systemic non-compacted cardiomyopathy in a 22-year-old woman, who had recently given birth and has type 1 diabetes mellitus.

Introduction

non-compacted cardiomyopathy (NCC) is a rare heart disease based on genetic mutations, prevailing in 0.014–1.3% of the general population according to different studies.[1,2] NCM is characterized by a thin epicardial layer and an extensive non-compacted endocardial layer. It is an abnormality of endomyocardial morphogenesis with interruption of myocardial fiber compaction, generating a loose trabecular tissue and it is more commonly found in men.[3]

Differential diagnosis of NCC must be made with peripartum cardiomyopathy (PCM) in pregnant patients with onset of heart failure (HF). Women who develop NCC present inconstant clinical expression, ranging from absence of symptoms to classic manifestations of HF, systemic embolism and ventricular arrhythmia, present between the last month of gestation or up to the fifth month of the postpartum period, including left ventricular systolic dysfunction, defined only in the absence of another identifiable cause.[4]

This is a case of a woman who had recently given birth, with type I Diabetes Mellitus (type I DM) and clinical picture of HF.

Case report

a 22-year-old African-descendant woman with G1P0A0 pregnancy, type I DM and a history of diabetic nephropathy. She seeks the emergency department at the beginning of the third trimester of pregnancy presenting lower limb edema, orthopnea, nocturnal paroxysmal dyspnea and dry cough. The patient was initially treated for nephrotic syndrome due to previous glomerular lesion by type I DM.

At the 37th week of pregnancy, the patient started labor and was submitted to cesarean section as prescribed by the medical team. In the immediate postoperative period, she had warm profile HF and congestion. On examination, she presented jugular vein turgescence and hepatojugular reflux; bilateral crackling rales and vesicular murmur abolished at the bases; regular heart rhythm, painful hepatomegaly, ascites and 3+/4 lower limb edema to the root of the thigh.

Furosemide, spironolactone, bisoprolol and enalapril were initiated. Laboratory tests consisted of alkaline phosphatase 257 U/L, gamma GT 199 U/L, AST 18 U/L, ALT 20 U/L, total bilirubin 0.3 mg/dL, urea 60 mg/dL and creatinine 1.04 mg/dL. Electrocardiogram with no ventricular overload, with ventricular bigeminism.

Transthoracic echocardiogram (TTECHO) using iE33 (Philips) after clinical compensation presented severe global systolic dysfunction, intertrabecular recesses filled with blood, enlarged left ventricular systolic-diastolic diameters with LV diastolic diameter of 3.55 cm/m^2, LV systolic diameter 2.51 cm/m^2, pulmonary artery systolic pressure (PASP) 49 mmHg, mean E/e' ratio 24.15 and moderate mitral regurgitation. Mean global circumferential systolic strain (GCSS) was -15% and mean global systolic longitudinal strain (GLSS), -13%. Presence of ventricular trabeculations in the inferior and anterolateral middle walls and lateral apical walls. Non-compacted LV/compacted LV ratio of 2.07. Analysis of myocardial mechanism revealed Bull's-eye map with the presence of apical sparing (Figure 1).

Cardiac magnetic resonance imaging (CMR) showed preserved-size right ventricle (RV) with global LV systolic dysfunction with LV ejection fraction estimated at 23%, with marked LV trabeculation on the lateral, anterior and inferior walls, meeting non-compaction criteria, with an average non-compacted/compacted myocardial ratio of about 3.1 and severe systolic dysfunction. Enlarged pulmonary artery trunk. No late enhancement with pleural effusion on the right, and pericardial effusion (Figure 2).

The patient was discharged with outpatient monitoring in NYHA functional class II after stabilization.

Discussion

non-compacted LV myocardium is defined as congenital myocardiopathy[5] resulting from modifications in genes G4.5 and alpha-dystrobrevin[6] and it is more common in males. Many patients have minimal symptoms or are asymptomatic,

Keywords

Pregnancy; Myocardium; Diagnosis.

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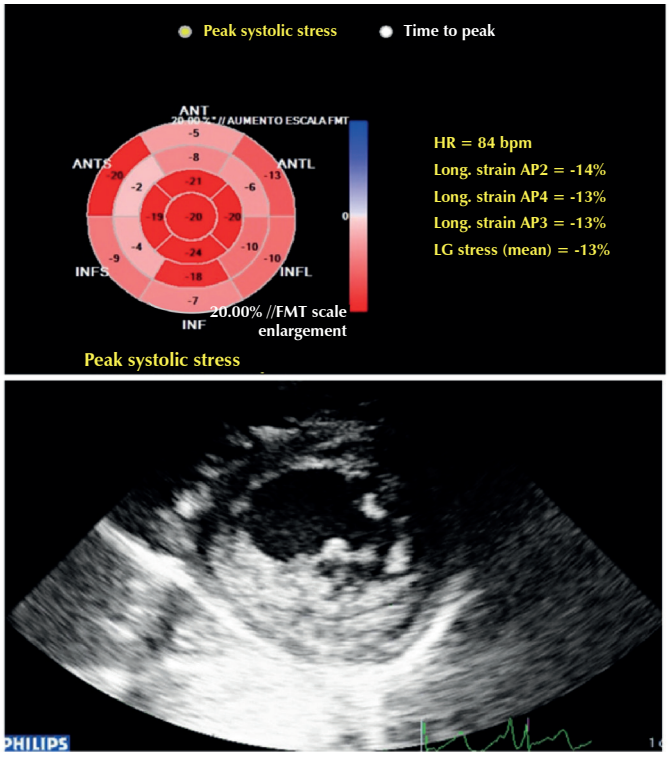

Figure 1 – *Bull's eye map showing the presence of apical sparing. Echocardiogram with ventricular trabeculations in inferior, mid anterolateral and lateral apical walls.*

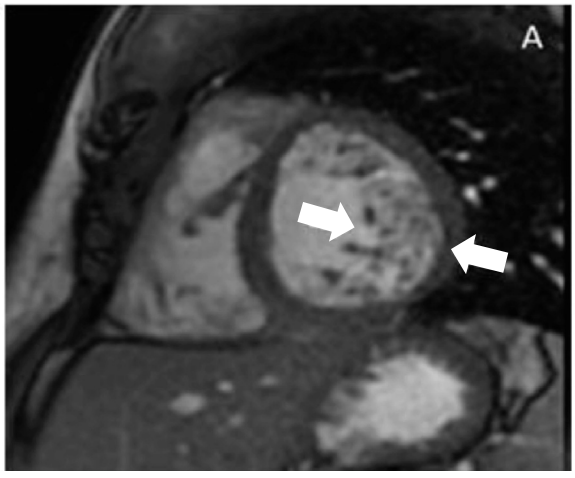

Figure 2 – *Cardiac magnetic resonance imaging — short axis view showing increased ratio between compacted and non-compacted myocardium in the left ventricular lateral wall.*

and disease progression in these patients is not well established, although there is evidence that long-term prognosis is not good.[1,5,7] In symptomatic patients, the course of the disease is usually progressive deterioration of cardiac function.

The patient had an onset of this condition with acute HF at the end of pregnancy but, in general, the disease has an insidious onset presenting compensatory ventricular hypertrophy and asymptomatic ventricular dilation as observed in other reports.[2] Predisposition to onset of acute HF was probably due to the high cardiac output during pregnancy.

The patient had no previous history of pulmonary thromboembolism, although it is common for patients with NCC to have blood stasis in the chambers with thrombus

formation in the intertrabecular recess leading to pulmonary or systemic complications due to thromboembolism.[8,9]

TTECHO is useful in diagnosis, by combining the established criteria described by three authors[7,10-12] (Table 1) while offering low sensitivity and greater specificity. The echocardiographic abnormalities observed in the speckle tracking raised the suspicion of NCC and the patient was referred to CMR, which confirmed the diagnosis.[12,13] When CMR is not available, NCC can be misdiagnosed as hypertrophic cardiomyopathy, dilated cardiomyopathy (DCM), endocardial fibroelastosis, restrictive cardiomyopathy and myocarditis and, in these cases, apical sparing longitudinal strain may be used in dilated phenotype cardiomyopathies to differentiate compaction and non-compaction. Tarando F et al.[14] compared 48 patients with NCC and 45 with DCM and performed LV longitudinal strain analysis. The results were compared to define the best tool to differentiate NCC and DCM. Longitudinal shortening was greater in individuals with NCC than in those with DCM. Mean base-to-apex LV wall strain gradient had 88.4% sensitivity and 66.7% specificity. In a multivariate model, Mean base-to-apex LV wall strain gradient was the only independent echocardiographic criterion (OR=0.76, 95% CI [0.66; 0.90], p=0.0010) that allowed distinguishing between NCC and DCM. In this case, longitudinal strain showed a non-specific DCM pattern and gradient calculation was not performed; diagnosis was confirmed by CMR.

Conclusion

NCC is a rare congenital disease and the patient was asymptomatic, but the high cardiac output due to pregnancy was the trigger for acute HF. TTECHO may raise suspicions of uncompressed myocardium, and differential diagnosis for DCM is difficult, but cardiac magnetic resonance imaging is the most appropriate test to confirm diagnosis. The disease has no definitive treatment and, in some cases, heart transplantation may be necessary.

Quadro 1 - Critério de diagnostico para miocárdio não compactado isolado.

1. No coexisting cardiac abnormalities (other than topic 2 to 4) by definition.
2. Typical two-layer myocardial structure: a thin compacted outer layer (epicardial) and a thicker non-compacted inner layer. Consisting of a trabecular meshwork with deep endocardial spaces, characterized by final systolic ratio of the non-compacted versus compacted endocardial layer greater than 2.
3. Predominant segmental location of the abnormality, i.e., non-compressed myocardium is predominantly (>80%) found in the apical and mid ventricular areas of the lower and lateral walls.
4. Color Doppler echocardiography shows deeply perfused intertrabecular recesses (in contrast to myocardial sinusoids, and intertrabecular spaces do not communicate with the coronary circulation).

Adapted from Jenni R et al.15

Authors' contribution:

Data acquisition: Martins DLD**, Martino OS,** Ribeiro ML, Neves DG, Ávila DX. Manuscript writing: Jorge AJL, Martins DLD**,** Ribeiro ML, Martins WA, Ávila DX. Critical revision of the manuscript for important intellectual content: Jorge **AJL,** Martins WA.

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