

# Noncompaction myocardium as a differential diagnosis of peripartum cardiomyopathy. Study of a case

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## Summary

Peripartum cardiomyopathy (PPCM) is a common cause of secondary cardiomyopathy of unknown etiology. It is characterized by the presence of congestive heart failure (CHF) in the mother in the last month of pregnancy or until five months after birth with left ventricular systolic dysfunction in the absence of other causes of heart failure in previously healthy women. Medical treatment consists of neurohormonal blockade, inotropic support, reduced pre- and post-cardiac load and anticoagulation. Heart transplantation is reserved for severe cases refractory to medical therapy. The prognosis is variable: approximately 50-60% of patients recover full cardiac function, in most cases in the first six months.

We report the case of a black woman of 37 years, multiparous, prenatal without complications, comorbidities or previous drug use that developed CHF, sudden onset, demonstrated ventricular dysfunction on echocardiography 15 days after childbirth normal. There was no clinical suspicion of non-compaction cardiomyopathy (NCC) which gave rise to doubt about the diagnosis of PPCM, since this is a diagnosis of exclusion. The presence of NCC was ruled out by cardiac magnetic resonance imaging.

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**Keywords:** Peripartum cardiomyopathy - Heart failure - Pregnancy - Non-compaction cardiomyopathy - Cardiac magnetic resonance imaging

## Introduction

Peripartum cardiomyopathy (PPCM) is a disease of undefined etiology, characterized by the presence of four criteria established in the age of 1995, by the American Society of Cardiology (AHA/ACC): 1) development of congestive heart failure (CHF) in the last month of pregnancy or up to five months after delivery, 2) left ventricular systolic dysfunction, 3) absence of previous heart disease, and 4) exclusion of other causes of CHF.

Non-compaction cardiomyopathy (NCC) is a congenital heart defect that results from an embryological malformation, with deep grooves and persistence of sinusoids in the left ventricular wall, typical of the embryonic heart. In most literature reports, the NCC is associated with other congenital heart disease, predominantly pulmonary atresia and ventricular outflow obstruction combined with a left ventricular intact septum<sup>1-5</sup>. It was first described in 1932, after the autopsy and has facilitated the diagnosis with the use of cardiac magnetic resonance imaging (MRI)<sup>1,6</sup>. This

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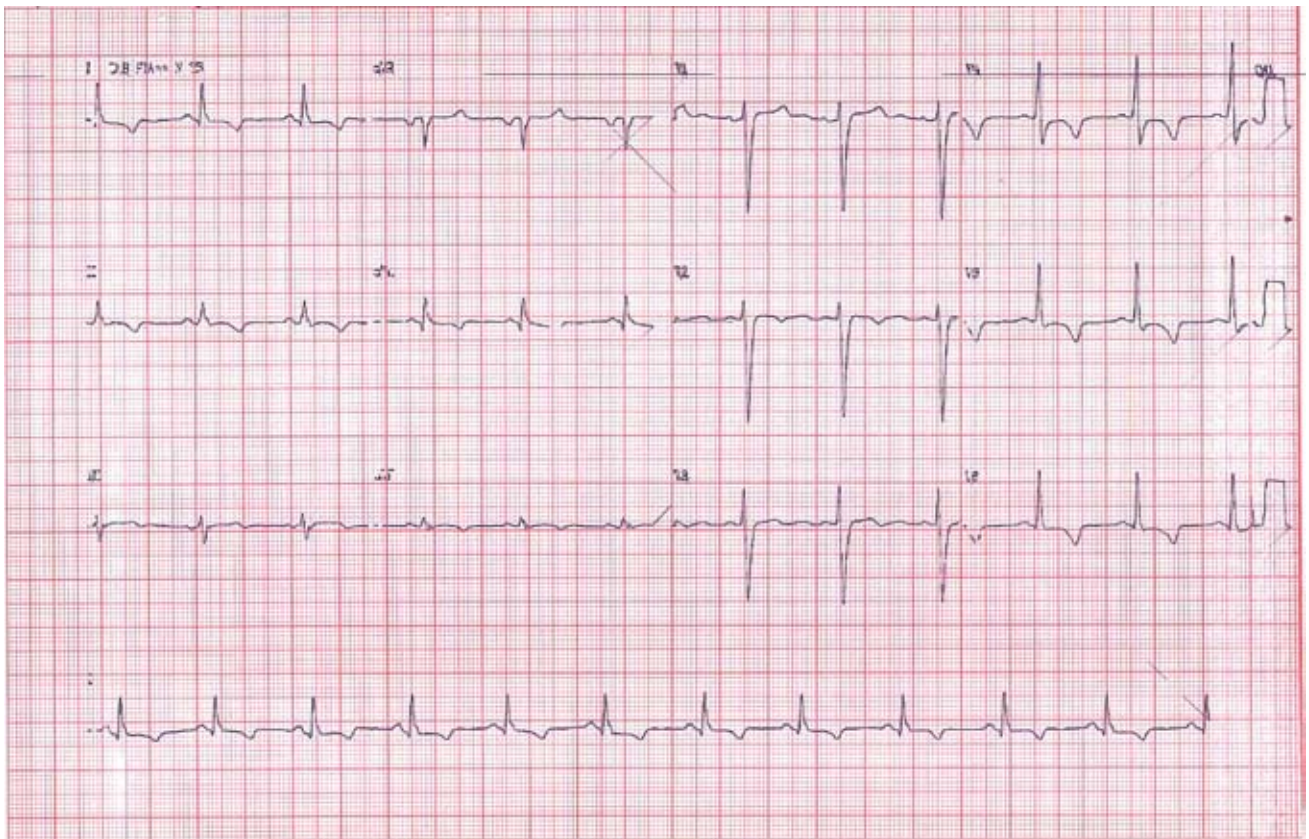
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**Figure 1.** Electrocardiogram with pattern of ventricular overload.



**Figure 2.** Chest X-ray posterior-anterior view: middle arch rectified; cardiomegaly; double contour and increased left ventricular.

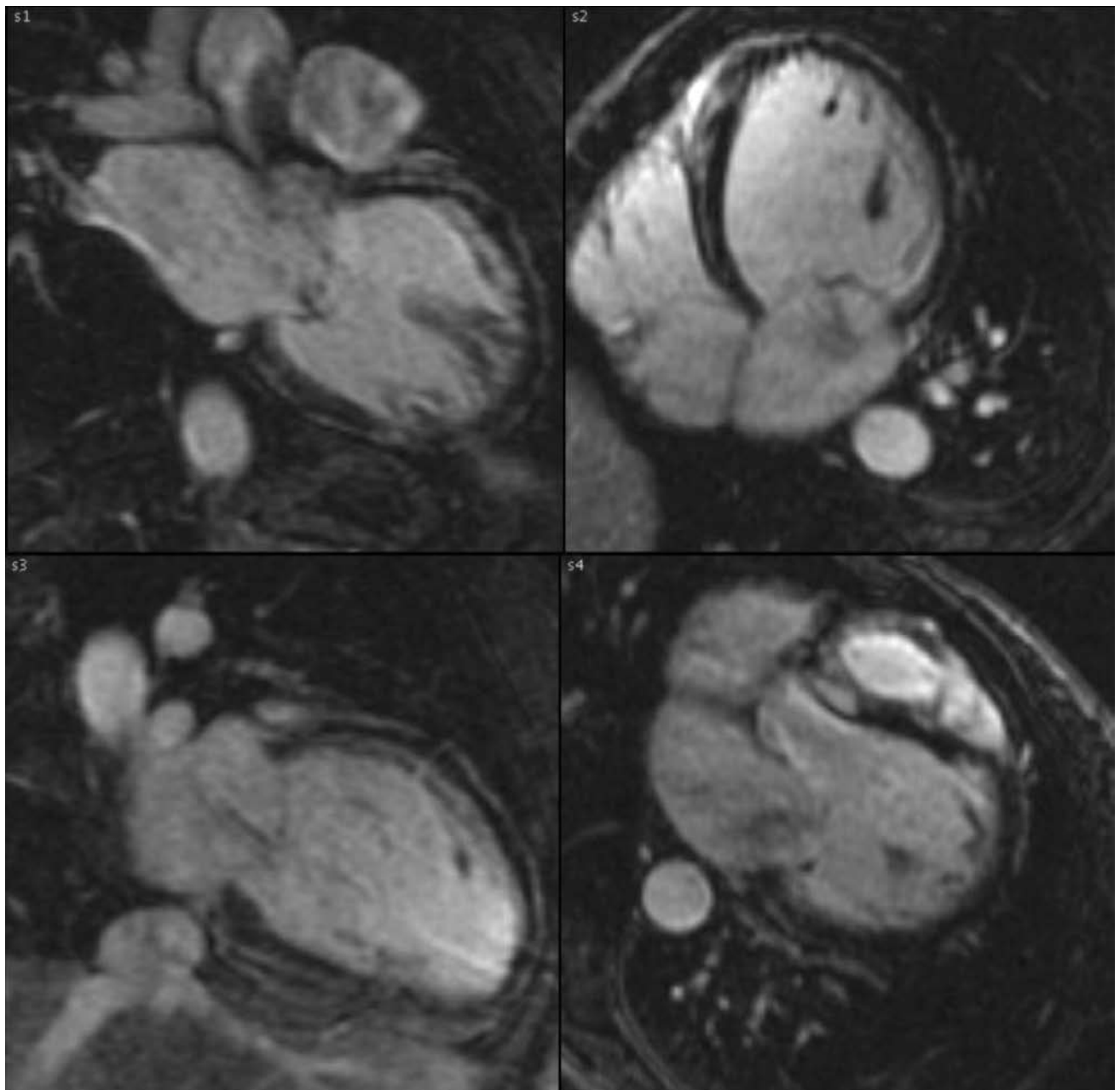
also is associated with heart failure, severe arrhythmias and embolic events. The objective is to report the case of PPCM where the diagnosis of NCC was suggested as a differential diagnosis.

### Case report

Patient female, black, 37 years, multiparous, prenatal without complications, comorbidities or previous drug use. After 15 days of a normal delivery at term, the clinical onset with fatigue, dyspnea on slight exertion, orthopnea and paroxysmal nocturnal dyspnea. She evolved with worsening of New York Heart Association (NYHA) functional class III-IV associated with cold, pitting, ascending and symmetrical in both lower limbs. The presence of persistent dry cough led to the suspicion of pulmonary tuberculosis. We performed bronchoscopy and bronchoalveolar lavage fluid was normal. After admission to the intensive care unit and initiate treatment for CHF, she evolved in good general condition, eupneic, breathing room air, without jugular venous distention at 45 degrees. Blood pressure of 80X60mmHg and a



**Figure 3.** Transthoracic echocardiography. Left atrium and left ventricle (LV) increased, significant LV systolic dysfunction and trabecular aspect of lateral and anterior wall.



**Figure 4.** Cardiac magnetic resonance imaging: dilated cardiomyopathy, significant left ventricular dysfunction and increased subendocardial trabeculations, no criteria for non-compacted cardiomyopathy.



heart rate of 62bpm. Apical impulse deviated to the left end on the 5th interspaces, regular heart rhythm, split second heart sound and no murmurs. Respiratory system, abdomen and lower limbs showed no alterations. The electrocardiogram (Figure 1) showed sinus rhythm with biventricular hypertrophy signs. The chest X-ray (Figure 2) showed cardiomegaly, medial arch contour grinding and double into the right atrium. Transthoracic echocardiography (Figure 1) showed a significant increase in atrial and ventricular diameters, severe mitral regurgitation, systolic dysfunction, left ventricular ejection fraction of 42% (Simpson method), diastolic function with a restrictive pattern III-b and trabecular appearance of myocardium, suggesting the diagnosis of NCC. Conducting a hypothesis the cardiac MRI excluded NCC (Figure 4). The patient recovered and was discharged in NYHA functional class I with carvedilol (12.5 mg/day), enalapril (10 mg/day), spironolactone (25 mg/day) and warfarin sodium (5 mg/day).

## Discussion

In this case presented we found three of the four criteria recommended by the AHA/ACC for the diagnosis of PPCM. However, the possibility of another diagnosis -NCC- could exclude the first. This patient also had demographic characteristics associated with PPCM: black, multiparous with pregnancy after age 40. Several inflammatory mechanisms are admitted as *etiopathogenic* factors of PPCM<sup>7-10</sup>. Ultrasensitive C-reactive protein is present in 50% of cases<sup>11</sup>. Studies have shown that the *16-kDa fragment of prolactin* was shown to have antiangiogenic activity in some women and, consequently, could result in a cardiomyopathy<sup>12</sup>. Since in severe cases of PPCM this mechanism has therapeutic implications, bromocriptine added to standard therapy improves the outcome of the disease. Pentoxifylline may also be added in severe cases, according to some authors<sup>13</sup>.

## Conclusion

The undefined criteria for the echocardiographic diagnosis of the NCC has brought a great number of clinical suspicion of this disease, but confirmation has not been found yet in the cardiac MRI, as in the case presented. The presence of trabeculations in the ventricular wall is normal and so only carefully to the possibility of diagnosis. The cardiac MRI is essential in cases of unexplained heart failure. The inflammatory nature of the PPCM could *predispose to intracardiac thrombus formation*. Thus anticoagulation is recommended in these patients. Although the diagnosis of NCC must be a hypothesis in patients with trabeculations visualized by echocardiography, the clinical picture suggested the diagnosis of PPCM. The cardiac MRI helped the exclusion of the NCC.

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## Conflict of interest

The authors have no conflicts of interest to declare.

## Linking university

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