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(CASE REPORT)

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Peripartum cardiomyopathy: How to be sure of the diagnosis?

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Abstract

Peripartum cardiomyopathy is an entity within the spectrum of heart failure that is related to pregnancy, which can debut with a clinical variety whose diagnosis in many cases is late.

Because the diagnosis is usually of exclusion, it is of interest to the physician, a high clinical suspicion is required to initiate management and avoid irreversible sequelae.

Keywords: Peripartum Period; Cardiomyopathies; Heart Failure; Pregnancy; Cardiac lesions.

1. Introduction

Peripartum cardiomyopathy or also known as pregnancy-associated cardiomyopathy is a cardiac degeneration that leads to heart failure during pregnancy or early postpartum, leading to the development of heart failure in the last month of pregnancy and up to the fifth month after the end of pregnancy (2).

It is known as a cause of heart failure since the XIX century where it was recognized and until now the hypotheses that revolve around the presentation of this pathology are currently related to genetic components and factors dependent on functional alterations triggered by hormonal imbalance dependent on the placenta and the pituitary gland that induce damage in the cardiac vasculature and myocardiocytes (2,13).

Its clinical course is highly variable and may range from progression to terminal heart failure or to spontaneous and complete recovery (13).

2. Case presentation

A 22-year-old female who is in the distant puerperium of 45 days of a second pregnancy by cesarean section, which was motivated by previous cesarean section, with no known pathological history and family history of a twin sister who died at 18 years of age in the immediate puerperium, related to severe preeclampsia and pneumonia. She attended a level IV clinic in the city of Barranquilla with clinical symptoms of 5 days of evolution at admission characterized by dyspnea of medium effort with progression to rest, associated with orthopnea of recent onset, and dry cough unrelated to fever. On physical examination patient with orthopnea, tachycardia with rhythmic heart sounds with pansystolic murmur in mitral focus grade 3, tachypneic with decreased breath sounds in right hemithorax, no pulmonary aggregates, abdomen without visceromegaly, limbs with acropacity without cyanosis, no evidence of tissue congestion, in paraclinical tests

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performed in initial care is evidenced: 12-lead electrocardiogram with sinus tachycardia, normal axis, regular R-R, T wave inversion in V5-V6 and flattening in V2-V4, no ischemic data, no blockages. Chest X-ray in AP projection with findings showing an increase in the cardiac silhouette grade II, with flow cephalization predominantly right pulmonary and right alveolar occupation in the middle lobe, hemogram leukocytosis predominantly polymorphonuclear.

Given the clinical context of the patient, risk factors and dyspnea of acute onset, complementary studies were requested for diagnostic possibilities of infectious and/or peripartum cardiac etiology, antimicrobial coverage with amino-penicillin and beta-lactamase inhibitor was started and a non-contrast chest CT scan was performed, showing multilobar cottony infiltrates in the pulmonary window, predominantly in the lower, middle, right apical and left basal lobe, with right pleural effusion, and cardiomegaly without pericardial effusion. Maintaining antimicrobial therapy, initiation of diuretic therapy and continuous hemodynamic monitoring due to the high risk of cardiogenic shock, during his stay in the unit he presented marked dyspnea on decubitus with increased ventilatory effort requiring oxygen by high flow cannula due to hypoxemia in arterial blood gases.

Studies were extended to determine the autoimmune cause or in relation to the puerperium; taking into account the family history of a sister who died in the puerperium, a transthoracic echocardiogram was requested with the result of severe eccentric hypertrophy of the left ventricle with LVEF severely depressed to 18%, with severe functional mitral insufficiency and data of mild pulmonary hypertension, so studies were extended to determine the autoimmune cause.

Taking into account the family history of a sister who died in the puerperium, a transesophageal echocardiogram was requested for better documentation of cardiac anatomy and valvular planes, which concluded: left ventricle slightly dilated with severely depressed systolic function and alterations of LVEF contractility: 24%. Spontaneous echo-contrast in left ventricle, left atrium and left atrial appendage, right ventricle with mild systolic dysfunction and moderate to severe functional mitral insufficiency. With negative quantitative troponins and highly positive natriuretic peptides for acutely decompensated heart failure, autoimmune pathology is ruled out and complete antimicrobial scheme, pharmacological management is optimized with foundational therapy (ARNI, Beta-blocker, IGLST2 and MRA) for management of heart failure with favorable clinical response given the weaning of supplemental oxygen, resolution of cottony infiltrates in chest X-ray control so it is indicated transfer to general ward and is submitted to cardiovascular board where cardio-resonance is requested by high risk of being a candidate for cardiac transplantation if there is no improvement of LVEF. (Figure 1).

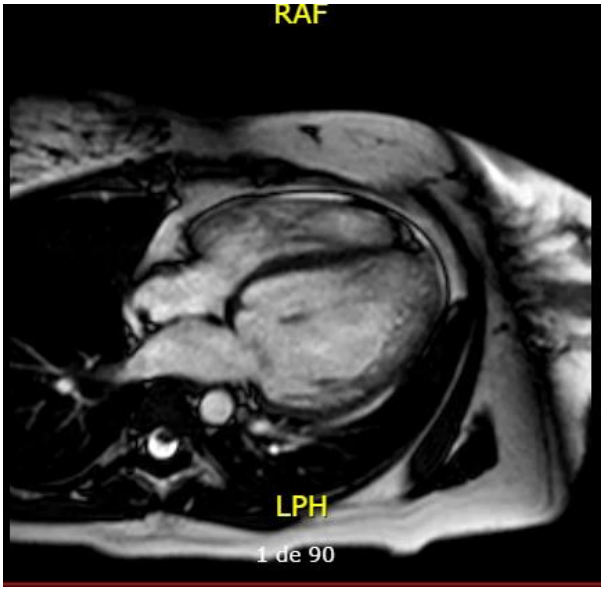

Figure 1 Cardiac magnetic resonance in axial view: 1. dilatation of left cavities, without presence of intracavitary thrombi. 2. myocardial enhancement pattern of non-ischemic and non-specific type. 3. no myocardial perfusion defects at rest. 4. moderate mitral insufficiency (Carpentier 1) and mild tricuspid insufficiency. 5. severe biventricular systolic dysfunction LVEF: 16%. Left ventricular ejection fraction (LVEF).

Findings that reinforce the diagnosis of peripartum cardiomyopathy. Given the good response to treatment, the patient was discharged for outpatient follow-up with cardiology and cardiac imaging studies for control and to define additional behaviors.

3. Discussion

La Heart failure (HF) in pregnancy occupies an etiological range that includes arrhythmias, congenital heart disease, valvular disease, pulmonary hypertension, ischemic heart disease and the group of cardiomyopathies (1).

Sixty percent of HF cases usually present in the postpartum period, followed by 27% during labor and 13% during gestation. But there is a bimodal peak between 23 to 30 weeks and after delivery (1).

The timing of onset of heart failure is relevant for the diagnostic algorithm. It should be noted that in patients with pre-existing cardiac lesions, HF is likely to present throughout gestation, whereas in the case of cardiomyopathy it usually appears weeks before or after delivery (1).

Worldwide, HF during pregnancy has a prevalence of 13% and up to 33% in patients with pre-existing cardiomyopathy (1).

Heart disease is the leading cause of maternal morbidity and mortality in developed countries, including the United States, and is attributed to almost 27% of gestation-related deaths (1). In fact, 60% of maternal cardiogenic shock cases are caused by peripartum cardiomyopathy (2).

The epidemiology of peripartum cardiomyopathy varies according to the geographical location and can complicate births with a rate ranging from 1 case per 2000 births in the world (2).

In Colombia, the main cause of maternal death is pregnancy-associated hypertension. In the first semester of the year 2022 there were a total of 122 maternal deaths, of these, 2 deaths were attributed to cardiac disorders without specifying type; however, this value could be underestimated since it is unknown whether when grouping the causes (such as other direct causes and other non-indirect causes, for a total of 29 deaths) they could be cardiac affectations related to peripartum cardiomyopathy (3).

In general, pregnant women with cardiomyopathy (any type) are at increased risk of adverse events such as maternal death and increased maternal morbidity, and, in the case of peripartum cardiomyopathy, it is associated with an increased risk of decompensation, obstetric complications and hospital readmission rates (4).

Not only maternal involvement, but there is evidence of adverse neonatal effects such as increased perinatal mortality, preterm birth, low Apgar scores, low birth weight and respiratory distress syndrome (1).

Peripartum cardiomyopathy is defined as maternal heart failure with systolic dysfunction with idiopathic left ventricular ejection fraction <45% that develops in the last month of pregnancy, delivery or in the first 5 months of puerperium (the term of which could be extended), in the absence of known pre-existing cardiac dysfunction (2,5).

Among the risk factors for this entity, African ancestry, preeclampsia, systemic arterial hypertension, multiple gestation, advanced maternal age (over 30 years), anemia, thyroid dysfunction, prolonged tocolysis and obesity have been documented (5,6).

At present, the pathophysiology is not clear, but different hypotheses have been described that include an underlying predisposition, as could happen with variants of genes encoding the sarcomeric proteins titin, myosin and troponin. And, subsequently, a vascular aggression that could originate in different ways; among these are (6).

Prolactin secretion: through cathepsin D, secreted by cardiomyocytes, is cleaved into a 16kDa prolactin fragment that would promote injury especially when there are low levels or deficiency of signal transducer and activator of transcription 3 (STAT 3) (6). Positive regulation of endothelial microRNA-146a (miR-146a): stimulated by the 16 kDa protein whose levels have been normalized with the administration of bromocriptine and antisense oligonucleotides, this miR-146-a is released by exosomes and blocks pathways such as Erbb4, Nras and Notch1, leading to cardiomyocyte apoptosis (6).

- Placental secretion of soluble fms-like tyrosine kinase receptor 1 (sFlt-1), secreted to a greater extent in late gestation, acts as an anti-angiogenic protein by sequestering vascular endothelial growth factor (VEGF) and placental growth factor (PIGF) leading to endothelial dysfunction and hypertension; furthermore, in mice with cardiac deletion of the

PGC-1α (proliferator-activated receptor gamma-1α coactivator) gene, the action of exogenous sFlt-1 led to cardiomyopathy and heart failure, the effects of which were mitigated by bromocriptine and VEGF (2,6).

All these mechanisms triggered by pituitary and placental hormones would lead to endothelial dysfunction and cardiomyocyte death and may explain, in part, the findings in peripartum cardiomyopathy (6).

The clinical presentation does not vary according to the signs and symptoms found in patients with heart failure, they can include from signs of vascular and tissue congestion to hypoperfusion, on the other hand, according to the cardiac compromise they can reach cardiogenic shock (2,6,7).

The diagnosis of peripartum cardiomyopathy is of exclusion, as shown in figure 2, sinus tachycardia can be found in the electrocardiogram, signs of pulmonary venous congestion in the chest x-ray (2,6,8).

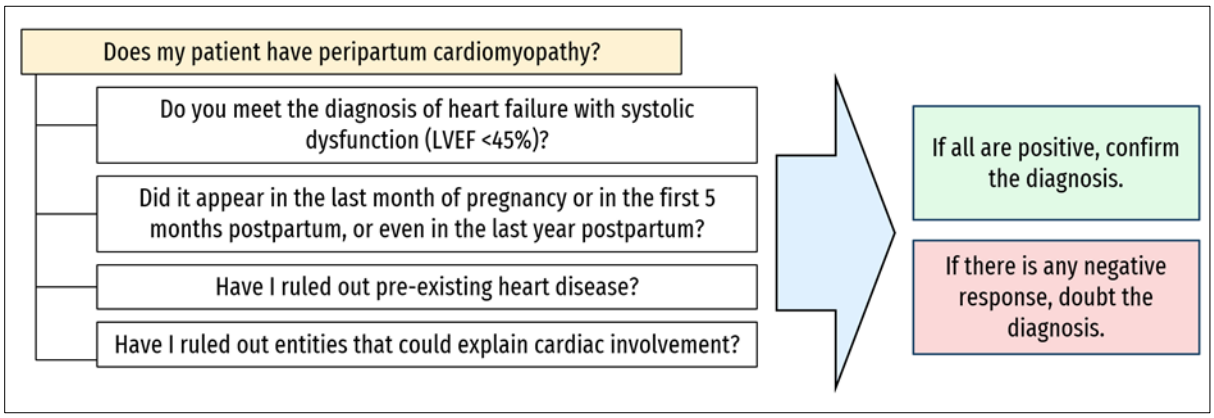

Figure 2 Diagnostic algorithm for peripartum cardiomyopathy. Own elaboration.

The diagnosis is generally made by echocardiography where systolic dysfunction is documented in the absence of other structural heart diseases that may explain this finding, MRI has a diagnostic yield of up to 80%, it is useful to evaluate the volumes of the chambers and the global and segmental myocardial function, it identifies the presence of thrombi in the left ventricle. (2,6,8) A useful tool to rule out other entities that have a characteristic pattern, T1 and T1 edema, small fibrosis areas and late gadolinium enhancement have been documented in a large proportion of patients (9,10). Endomyocardial biopsy is the last resort to be used and sometimes it is necessary to rule out acute myocarditis whose diagnosis is made with the Dallas histological criteria, to exclude significant viral presence, autoimmune etiologies, deposit diseases or of metabolic origin (2,6,8,9).

It is important to perform an adequate diagnostic algorithm where entities such as pulmonary embolism, amniotic fluid embolism, pre-existing cardiomyopathy induced by toxins or drugs, valvular diseases, congenital heart disease, pulmonary hypertension, MINOCA, myocarditis, sepsis, thyrotoxicosis, acute aortic syndrome, among others, must be ruled out (Figure 3) (4,6,8,11).

The management of this entity can revert cardiac function to normal in 50% of the cases within the first 3 to 6 months, although they will have a higher risk of recurrence in future pregnancies. The mortality rate is usually between 11 to 32% of cases. Medical treatment is usually the same as for other causes of systolic dysfunction, however, it should be noted that if the event occurs during pregnancy, angiotensin-converting enzyme inhibitors and angiotensin receptor blockers are contraindicated. Diuretics, metoprolol and spironolactone, hydralazine, isosorbide dinitrate, nitroglycerin, digoxin and low molecular weight heparin can be used (5, 7,12).

In stable patients the route of delivery should be vaginal, except if there is an obstetric indication, and breastfeeding is still controversial (5).

According to the described pathophysiology, the action of dopamine agonists and prolactin release inhibitors could have a beneficial effect in this type of HF; however, more studies are required to make a recommendation (5).

In cases of non-improvement, ventricular assist devices and cardiac transplantation may be used if compliant with indications (7). The threshold for anticoagulation is usually lower and consider if LVEF <35% (5).

Complications of this entity include: brain injury, cardiopulmonary arrest, pulmonary edema, thromboembolic disease and death (7).

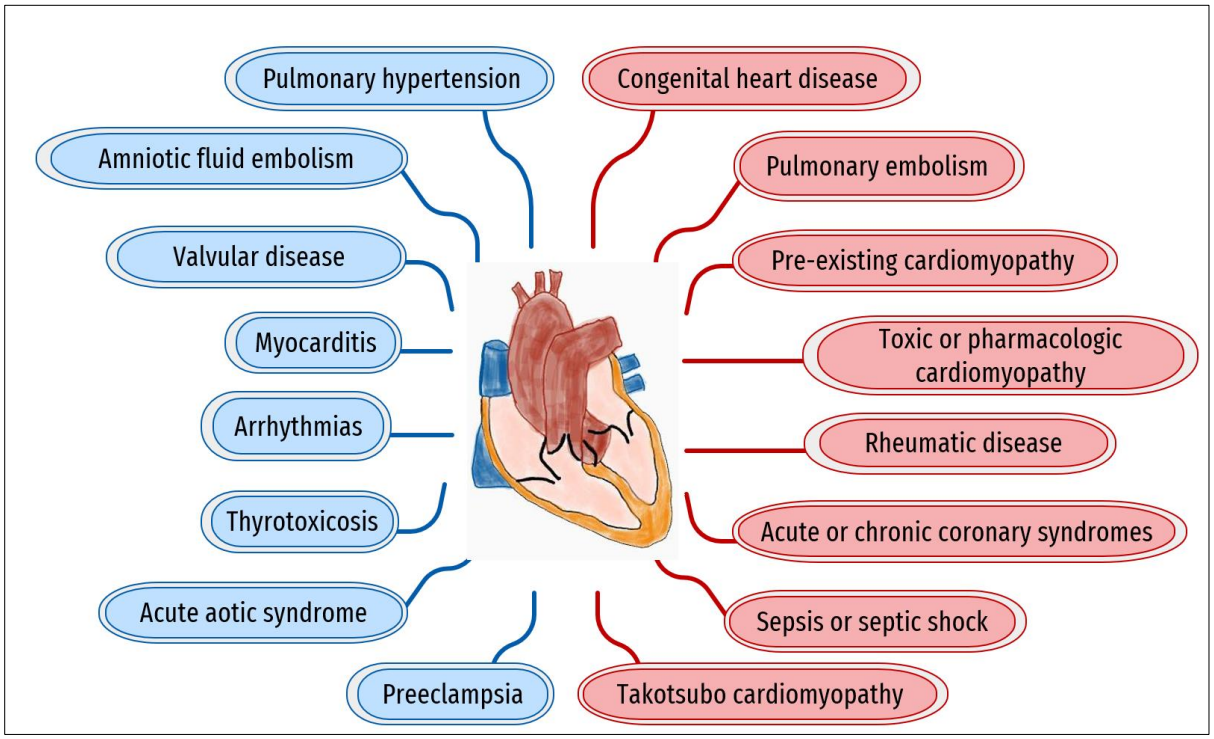

Figure 3 Differential diagnoses. Own elaboration.

4. Conclusion

A high clinical suspicion is required for the diagnosis of peripartum cardiomyopathy, it is important to take a clinical history of the patient and to use the appropriate diagnostic tools according to the clinical characteristics of the patient in order to separate other entities that could explain the clinical picture of these patients.

Compliance with ethical standards

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Disclosure of conflict of interest

The authors declare no conflicts of interest.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study

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