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PERIPARTUM MYOCARDITIS: CASE REPORT
An underdiagnosed pathology

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Abstract: Peripartum myocarditis, or pregnancy-associated cardiomyopathy, is a rare type of heart failure secondary to left ventricular systolic dysfunction that affects previously healthy women, at the end of pregnancy (last 30 days of pregnancy) or in the puerperium, and can extend to about five months after childbirth, with no identifiable cause, with a higher incidence in black women, aged over 30 years and multiparous. We report a case of a 20-year-old patient with a history of dyspnea and vomiting for 5 days. She was on the 60th day of puerperium, with normal delivery without complications, with an initial diagnostic hypothesis of massive pulmonary thromboembolism, which was later discarded due to the presence of clinical criteria for perinatal myocarditis. A bedside Doppler echocardiogram showed significant dilation of the right chamber, with reduced ejection fraction.
Keywords: Peripartum myocarditis; acute heart failure; puerperium.

INTRODUCTION

Peripartum cardiomyopathy syndrome (PPCM), despite being a rare form of heart failure, has an incidence of 1 to 100 in 10,000 live births7, depending on the geographic region, being more prevalent in the African continent3. This clinical entity is associated with high maternal mortality (between 10% and 32%)6, and has an idiopathic etiology. However, there are factors correlated with a higher incidence such as black race/color, multiparity, age over 30 years, preeclampsia and gestational hypertension2, in addition to the consumption of narcotics (such as cocaine), use of beta-agonists for more than 4 weeks, cesarean delivery and history of myocarditis.

This pathology was first described in 1849 by Ritchie et al.5, who defined it as idiopathic heart failure with onset between the last month of pregnancy and the sixth month after delivery. In addition, it characterized the presence of 4 criteria, which were confirmed and defined only in 1995 by the American Society of Cardiology: (1) Congestive Heart Failure developed in the last month of pregnancy or up to 5 months postpartum; (2) Decreased left ventricular systolic function; (3) Absence of previous heart disease; (4) Exclusion of other causes of CHF.

Diagnostic criteria include complementary tests: chest X-ray, electrocardiogram and echocardiogram/doppler echocardiogram, which is the gold standard for diagnosing heart failure. Thus, they include left ventricular ejection fraction less than 45% or M-mode fractional shortening less than 30% (or both) and left ventricular end-diastolic dimension greater than 2.7cm/m2. Electrocardiogram, endomyocardial biopsy, magnetic resonance imaging and myocardial catheterization help in the diagnosis and treatment of PPCM[3].

The clinical presentation of peripartum cardiomyopathy consists of symptoms, dyspnea, orthopnea, dry cough, palpitations, hemoptysis, nocturia, chest pain and abdominal pain. Signs include cardiomegaly, tachycardia, polypnea, hypotension, arrhythmic pulse, systolic murmur, mitral/ tricuspid insufficiency, jugular engorgement, peripheral edema/ascites, hepatomegaly. In this context, the differential diagnosis of the syndrome is pre-eclampsia; pulmonary thromboembolism (PTE)/ LA embolism; infectious myocarditis (by cocksachie B); dilated and hypertrophic and restrictive cardiomyopathy.

The treatment of such pathology has the objective of recovering the left ventricular function, reducing the cardiac pre and afterload and increasing the cardiac inotropism, since about 46% of the cases return to the left ventricular ejection fraction equal to or greater than 50% within six months, as described in

the study "Clinical presentation, management and 6-month outcomes in women with peripartum cardiomyopathy" by Karen Sliwa and colleagues, published in the European Journal of Cardiology[1]. Thus, the drugs that can be used are the same as for CHF, that is, inotropes (such as dobutamine), diuretics (such as spironolactone) and vasodilators. Thus, the prognosis of the disease depends on the degree of myocardial dysfunction, the diameter of the cardiac cavities and the complications that manifest themselves. Therefore, the mortality rate varies between 7 to 50% depending on the geographic region.

The objective of this work is to report the case of a young woman who presented signs and symptoms of PPCM 60 days after an uneventful normal postpartum period, in order to elucidate the clinical pathology, since its definitive diagnosis is based on the exclusion of other comorbidities. This way, it is intended, through this case report, to carry out an early diagnosis of PPCM as well as therapeutic intervention, in an attempt to reduce mortality statistics, which are high, in addition to encouraging Brazil to carry out studies cohort these patients to get a true incidence of MCPP and the severity of cardiac anomalies in pregnancy.

CASE DESCRIPTION

E.C.A.S, 20 years old, female, black, primigravidae, primiparous, without previous abortion, was admitted to the first care service, complaining of dyspnea, nausea, vomiting and diarrhea for 5 days, and denies fever, blood or mucus in the stool, smoking, underlying disease or current pregnancy. The patient was on the 60th postpartum day without intercurrences, as well as the prenatal follow-up within the normal range, as reported by her companions, mother and aunt. On physical examination, she was in regular general condition (REG), anicteric, acyanotic, pale and dehydrated without measurement, flat and flaccid abdomen, not painful, bowel sounds present Glasgow Coma Scale scoring 15, RR 19 bpm, Temperature 35, 9ºC, SatO2 100% in room air and BP not checked and as a conduct, medication was started in the unit with 1000ml 0.9% SF + Complex B 1 ampoule + metoclopramide 10mg and laboratory tests were requested. However, approximately 7 hours after entering the service, a sepsis protocol was opened and she was referred to the emergency room due to hypotension (BP 82x60 mmHg), tachycardia (HR 125 bpm) with murmur, dyspnea (RR 28 bpm), evolving in poor general condition (MEG), with hypothermia (34.5ºC), dehydrated +++/4+, vesicular murmur present without adventitious sounds, painful globose abdomen in the right hypochondrium, sudden negative decompression, extremities with cyanosis +/4+, and started ceftriaxone 1g/vial/ ampoule 2000mg IV 24/24h + metronidazole injectable solution 5mg/ml vial 100ml 500 mg EV 8/8h, and approximately after 1h, patient evolved with agitation and lowered level of consciousness, orotracheal intubation (OTI) was performed, noradrenaline was started, a central venous catheter was inserted in the left internal jugular and CROSS was requested. Laboratory tests resulted in Hemoglobin 12.3, Leukocytes 11570, Neutrophils 71%, Lymphocytes 19%, Platelets 177,000, D-Dimer 26,779, Urea 72.6, Creatinine 0.7, Na 142, K 5.8, Ca 7,9, Mg 2.1, BT 0.8, TGO 753, TGP 689, GGT 11, BNP 22,958, Dyspnoea to be clarified, pulmonary thromboembolism (PTE) questioned and Abdominal pain to be clarified was proposed as a diagnosis.

Noradrenaline 20ml/h and sedation were maintained, thrombolysis was performed with tenecteplase 30mg (according to the criteria of hemodynamic instability, right

ventricular failure), dobutamine and the possibility of transfer to NIR-HSMI was verified.

However, after approximately another 8 hours, the patient was admitted to a shock room with a diagnostic hypothesis of massive PE, hemodynamically unstable, intubated, sedoanalgesia, using vasoactive drugs, noradrenaline, dobutamine and vasopressin, with a Wells Score of 4.5 medium probability.

When an echocardiogram was performed at the bedside, pulmonary artery hypertension and cardiac chamber dilatation were identified. Physical examination showed MEG, sedated, RASS-3, vesicular murmur present without adventitious sounds, tachycardia, semiglobose abdomen, present bowel sounds, extremities poorly perfused. Urgency was requested in an ICU vacancy and intensive support was given with the use of vasoactive drugs.

However, approximately 26 hours after her admission to the service, the patient suffered a cardiorespiratory arrest in asystole, cardiopulmonary resuscitation was immediately initiated according to the ACLS protocol, administered bolus of IV adrenaline every 2 minutes, reassessing pulse and rhythm on a monitor every 2 minutes. After 12 minutes of maneuvers, the patient returns to spontaneous body circulation.

Peripartum myocarditis was suggested as a diagnostic hypothesis with a score of 4/4 criteria (1- Development of HF in the last month, 2- Absence of another cause justifying ADHF, 3- Absence of known heart disease before the last month, 4- Dysfunction systolic with reduced ejection fraction), PTE and mixed cardiogenic/obstructive shock.

An electrocardiogram was performed (Figure 2) showing sinus rhythm, heart rate of 154 bpm, sinus tachycardia, no change in ventricular repolarization, presence of low amplitude in other leads. A new echocardiogram was performed at the bedside, which showed significant cardiomegaly with dilation of the right ventricle, also presenting significant hepatomegaly, in addition to the presence of upstream B lines.

As a conduct, it was decided to increase the noradrenaline flow to 60ml/h with a calculation of 3mcg/kg/min, increase the dobutamine flow to the maximum dose with 21 mcg/min with a flow of 21ml/h, increase the vasopressin flow to maximum dose of 0.04 IU/min with a flow rate of 12ml/h, intensive support, due to the need to be transferred to the ICU, negative fluid balance was tried, and gasometry was requested for metabolic control and correction of possible acid-base disorders.

Four hours after his first arrest, the patient had bradycardia on the monitor and no pulse, so when he had pulseless electrical activity (PEA), CPR maneuvers were started according to the ACLS protocol, 2 cycles were performed with the administration of 1 ampoule of adrenaline, obtaining reversal for spontaneous rhythm in the sequence. at the same time, bicarbonate was started due to metabolic acidosis (pH 7, BIC 9, lactate 14), and 1 hour after the incident, the patient had no central pulse and CPR was started for the third time with 2 cycles and 1 ampoule of adrenaline, and again returning to spontaneous circulation, but in this condition, during the maneuvers, the patient presented a large amount of yellowish, slightly bloody secretion coming out of the orotracheal tube, and the amount was aspirated. After 5 minutes there was the fourth cardiac arrest, and given the numerous cardiorespiratory arrests and irreversible condition with no therapeutic response, it was decided to cease efforts in order to avoid dysthanasia, requesting the presence of family members to report the death.

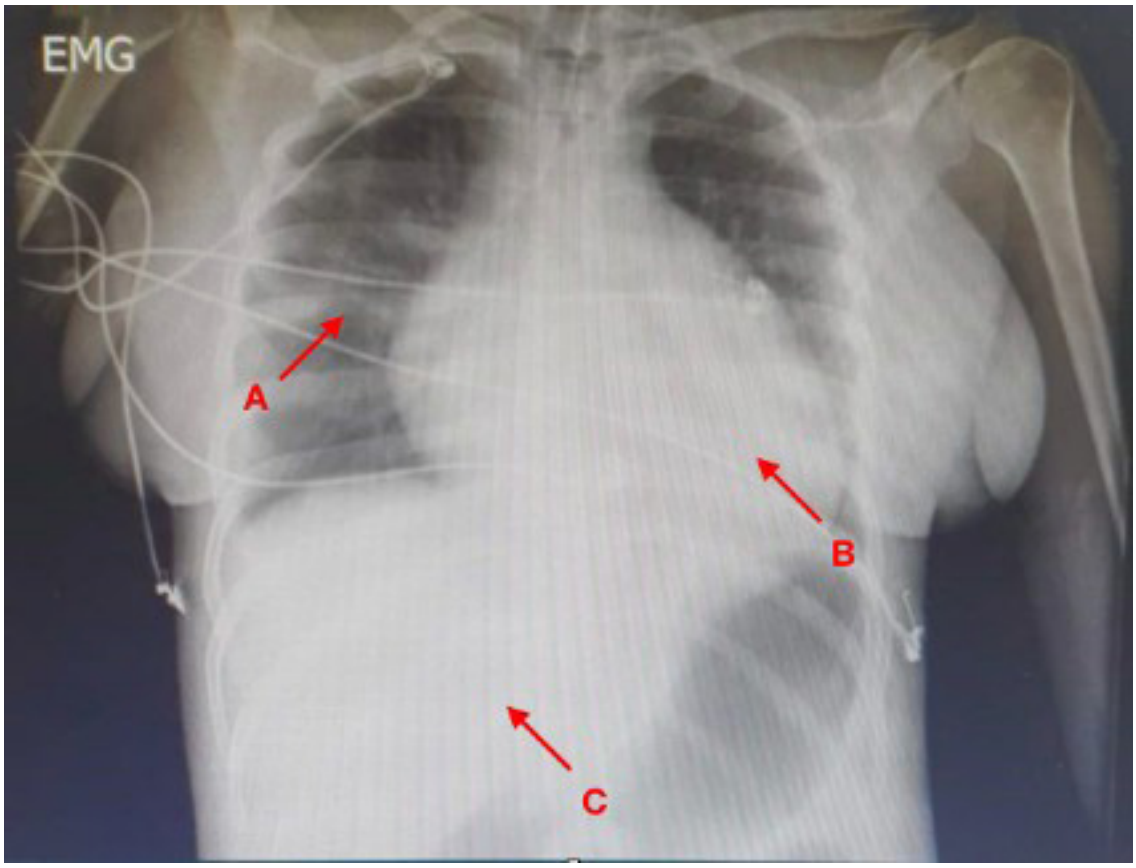

Figure 1. Posteroanterior chest X-ray showing straightening of the costal arches (A), cardiomegaly (B) and hepatomegaly (C).

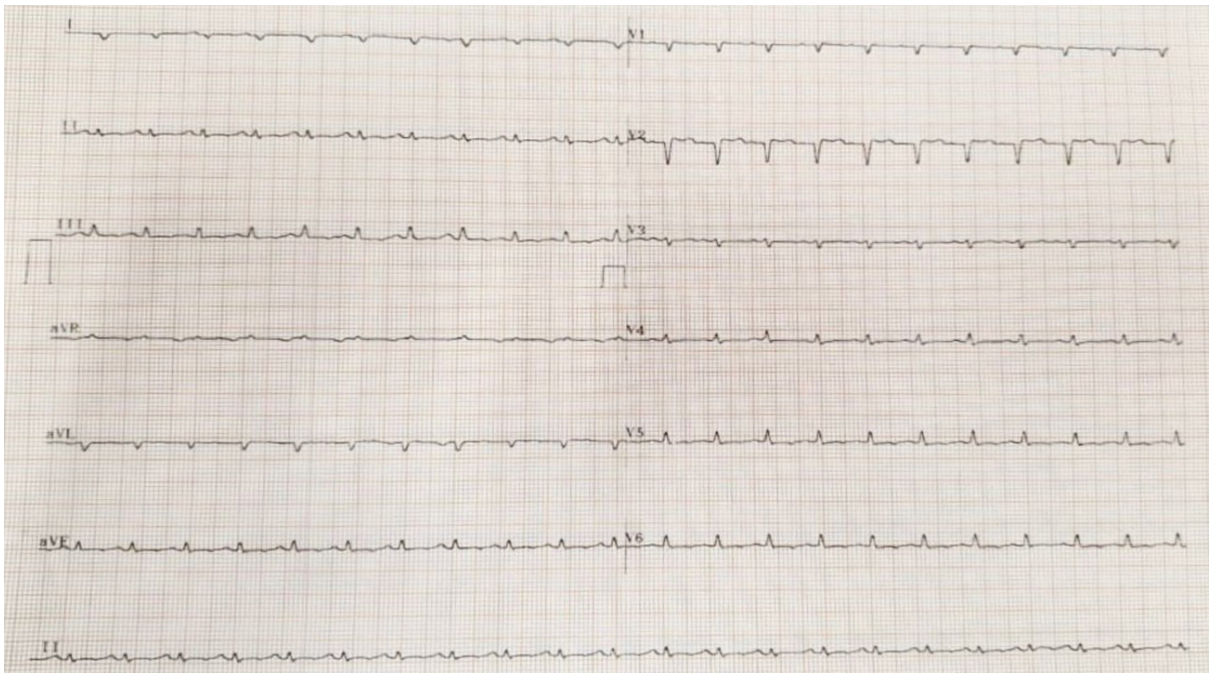

Figure 2. Electrocardiogram showing sinus rhythm, sinus tachycardia, presence of low amplitude in other leads.

DISCUSSION

Peripartum myocarditis is an infrequent comorbidity, but with a high rate of maternal mortality, with heart failure, arrhythmia or embolic event being the main causes of death. Risk factors associated with the development of the disease are advanced maternal age (> 35 years), multiparity (> 3 years), multifetal pregnancy, pre-eclampsia/ eclampsia or gestational hypertension, black skin color and use of tocolytics (over 4 weeks). The most considered causes are viral myocarditis, genetic or immunological causes, inadequate response to the hemodynamic stress generated by pregnancy, activation of inflammatory cytokines, prolonged tocolysis or the deleterious effect of prolactin, which induces cell apoptosis. Regarding the patient in the case report, it can be said with certainty that the present risk factor is ethnicity.

Clinical symptoms are the classic ones of heart failure, such as dyspnea, fatigue and edema of the lower limbs and are often associated with those present in advanced pregnancy and in the postpartum period, making the diagnosis more difficult. In addition to the clinical diagnosis, whose criteria are (obligatorily four) development of heart failure in the last month of pregnancy or within the last five/six months postpartum; no other identifiable cause of heart failure; absence of known heart disease before the last month of pregnancy and left ventricular systolic dysfunction (ejection fraction below 45% or reduced shortening fraction), the patient met the four criteria, as there was no previous history of heart failure and had a of reduced ejection as seen on Doppler echocardiography.

With regard to suspected perinatal myocarditis, it is essential to request an electrocardiogram, chest X-ray and echocardiogram. The findings of the patient's complementary exams reveal, on the electrocardiogram, sinus rhythm, heart rate of 154 sinus tachycardia, no changes in ventricular repolarization, presence of low amplitude in other leads. In the first echocardiogram performed, arterial hypertension of the pulmonary artery was evidenced, with dilation of the heart chambers, and in the second, significant cardiomegaly with dilation of the right ventricle, also presenting important hepatomegaly, in addition to the presence of upstream B lines. The chest X-ray showed an enlarged cardiac silhouette, with no pleural effusion and infiltrates. In addition to imaging tests, laboratories can be requested, such as brain natriuretic peptide, which has high values (the patient had BNP 22,958 (reference value: up to 70 pg/ml).

What the treatment consists of is based on the treatment of heart failure, using angiotensin-converting enzyme inhibitors or angiotensin receptor blockers, which are contraindicated during pregnancy. The use of diuretics is for symptomatic relief of pulmonary congestion. Anticoagulant therapy with unfractionated heparin or low molecular weight heparin due to the high incidence of thromboembolism, especially when ejection fraction is < 30%. In more severe cases, the use of dopamine or dobutamine is indicated. In the case of the patient, thrombolysis was performed with tenecteplase due to the high probability of pulmonary thromboembolism according to the Wells score, in addition to the high value of d-dimer 26,776, in use of vasoactive drugs due to mixed cardiogenic/distributive shock.

There are few studies in the national literature that describe the clinical evolution of these patients. Moreira and cols.[4] evaluated 12 patients diagnosed with peripartum cardiomyopathy, divided into two groups: 6 without changes and 6 with persistent cardiomegaly and ventricular dysfunction.

The mean age of patients (8 white, 2 black and 2 brown) was 15 to 36 years; 10 patients were primigravidae or secundiparous and 8 had resolution of the pregnancy by cesarean section. One of these patients died suddenly during one of the admissions for treatment of congestive heart failure; she was white, had a singleton pregnancy, was 17 years old and died 2 months after the diagnosis, carried out 5 months postpartum.

A study published by Carvalho and cols.[1] concluded that some echocardiographic characteristics, such as an increase in the left ventricular end-diastolic diameter, in the acute phase of the disease, and the late onset of symptoms, in the postpartum period, are associated with a poor prognosis. Patients with previous peripartum cardiomyopathy have an unfavorable evolution associated with black race and more pronounced initial cardiac alterations, and a favorable evolution is associated with a reduction in myocardial mass and an increase in the relative thickness of the ventricular wall.

The only risk factor that our patient had was black race, as it did not fit the parameter of multiparity, age over 30 years, gestational hypertension, pre-eclampsia. However, as reported by Moreira et cols.[6], all the bad prognoses were in black/brown women and all the good prognoses were only in white women, given that, despite a single factor, it corroborates to a bad prognosis and death.

Myocardial recovery of left ventricular function within 6 months occurred less frequently in Africa than in other countries, such as the USA and Germany, according to Sliwa et al7 in the article published in the European Heart Journal.

There must be a high prevalence of occult, underdiagnosed cases, of women who died due to a late diagnosis of PPCM or because they suggested massive PTE as a diagnosis, knowing that the gestational state is a risk factor for thrombotic events. The existence of a massive PE is a fact, but it does not determine the end of the diagnosis, since PPCM is a diagnosis of exclusion, thus embodying the fact that the immediate diagnosis must be a priority to allow the timely initiation of therapies for cardiac insufficiency.

FINAL CONSIDERATIONS

Perinatal cardiomyopathy is a disease with high maternal morbidity and mortality, but it is often not diagnosed early. Most reported cases suggest that patients have severe heart failure at the time of presentation, suggesting that the condition was not diagnosed during its initial phase, perhaps because symptoms and signs such as dyspnoea, fatigue and edema are attributed to pregnancy or the puerperium in instead of heart failure, or because these findings are common to several other pathologies. As seen in the article *"Internationally, the median time from symptom onset to diagnosis was 10 days, but ranged from 6 days in Europe to 23 days in Africa"*[7].

The immediate diagnosis has a direct impact on the patient's prognosis and, therefore, the patient's complaints must be taken into account, and the existence of this pathology must be known. The diagnosis of PPCM requires heightened awareness of multidisciplinary patient care teams and a high degree of suspicion. Management of peripartum cardiomyopathy must aim first to improve symptoms of heart failure through conventional therapies and then to administer targeted therapies.

However, we would like to emphasize the contribution of this case report to greater knowledge about the presentation of the disease in order to reduce the waiting time for the diagnosis and thus improve the prognosis of patients.

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ANNEXES

Entry labs	Values
Erythrocytes	4,53 10ˆ6/mm3
Hemoglobin	12.3g/dl
Hematocrit	39%
VCM	86.1fl
HCM	27.2pg
CHCM	31.5g/dl
RDW	15.7%
Platelets	177.000/mm3
Leukocytes	11.57 mil/mm3
Serum urea dosage	72,6 mg/dl
Creatinine	0,7 mg/dl
Serum sodium dosage	142 mmol/L
Serum potassium dosage	5,8 mmol/L
Serum magnesium dosage	2,1mg/dl
Serum calcium dosage	7,9 mg/dl

Annex 1 - Table of entrance exams.

Laboratories Emergency Room	Values
Serum D-dimer dosage	26.779,0ng/ml
Arterial blood gases	pH 7,23 PO2 166 mmHg SatO2 99,7% PCO2 13 mmHg HCO3 5,4 mmol/L ctCO2 5,8 mmol/L ctO2 16,5 mmol/L BE -22.2 mmol/L Na 134 mmol/L K 4,8 mmol/L Hb 11,9 g/dlHt * Lactato 4.9 mmol/L

Annex 2 - Table of laboratory tests in the Emergency Room.