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CARDIOMIOPATIA PERIPARTO: A PROPÓSITO DE UM CASO CLÍNICO

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RESUMO

A cardiomiopatia periparto é uma forma rara de cardiomiopatia dilatada definida pelo desenvolvimento de insuficiência cardíaca materna no último mês de gestação ou nos cinco meses pós-parto. O diagnóstico baseia-se na documentação de disfunção sistólica ventricular esquerda, na ausência de outras causas, numa mulher previamente saudável. A mortalidade pode atingir os 50%. Apresenta-se o caso de uma mulher de 36 anos, IV gesta IV para, obesa, fumadora, com quadro clínico de insuficiência cardíaca de novo nas primeiras horas após o parto. Apesar da instituição de terapêutica adequada com melhoria clínica significativa, não se verificou normalização da função ventricular sistólica ao fim de 1 ano de vigilância, estabelecendo o diagnóstico de cardiomiopatia periparto sem recuperação da função ventricular sistólica.

PALAVRAS-CHAVE: CARDIOMIOPATIA PERIPARTO, INSUFICIÊNCIA CARDÍACA, GRAVIDEZ

PERIPARTUM CARDIOMYOPATHY: A CASE REPORT

ABSTRACT

The peripartum cardiomyopathy is a rare form of heart disease, defined as the development of maternal congestive heart failure, in the last month of pregnancy or within five months after delivery, with documented left ventricular systolic dysfunction, in the absence of a demonstrable cause in a previously healthy woman. Mortality rates could reach 50%. The authors report a case of 36-year-old woman, gravida IV para IV, obese and smoker, with the diagnosis of peripartum cardiomyopathy established based on clinical status after delivery. Although heart failure treatment optimization was performed with significant clinical improvement, normalization of left ventricle systolic function did not occur at one-year follow-up, establishing the diagnosis of peripartum cardiomyopathy without left ventricular systolic function recovery.

KEY-WORDS: PERIPARTUM CARDIOMYOPATHY, HEART FAILURE, PREGNANCY

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BACKGROUND

Peripartum Cardiomyopathy (PPCM) is a rare but serious form of heart failure (HF) affecting women in the last month of pregnancy or early postpartum, associated with high morbimortality.[1,2] Prompt diagnosis with institution of intensive therapy by a multidisciplinary team is a prerequisite for improved outcome. Prognosis is highly related with reversal of ventricular dysfunction.[2]

CASE REPORT

A 36 year old caucasian pregnant, gravida IV para III, obese (BMI 36,5 Kg/m^2), smoker (10 cigars/day), was admitted at 38 weeks with erysipela of the left leg.

No personal or family history of cardiovascular disease was present.

Obstetric surveillance was irregular. Fasting blood glucose at 9 weeks suggested previous diabetes (146g/dl).

Two days after admission, due to suspected macrossomia and fetal distress (hydramnios, umbilical artery pulsatility index >P_{95}), elective cesarean section with tubal ligation was performed. Birth weight was 4950g and Apgar score was 2-7-10.

Twelve hours after surgery, the patient initiated dyspnea, cough and orthopnea. Physical examination revealed sweating, normal blood pressure, polypnea and respiratory distress signs. Cardiac auscultation revealed tachycardia, with no murmur or gallop sounds. Pulmonary auscultation showed bilateral rales and wheezing. Arterial gasometry revealed chronic respiratory alkalosis and oxygen saturation of 95%. Oxygen therapy by high-flow mask, hydrocortisone, aminophylline and furosemide were immediately started with clinical improvement. Cardiac enzyme levels were normal. Chest x-ray showed moderate cardiomegaly and bibasilar interstitial infiltrate. Electrocardiography (ECG) registered sinus tachycardia. Pulmonary multi-detector computed tomography angiography excluded acute pulmonary embolism but revealed bilateral pulmonary infiltration and pleural effusion suggesting acute pulmonary edema. Transthoracic echocardiography (TTE) revealed four-chamber dilatation with a left ventricle (LV) end-diastolic dimension of 67mm and a left atrium of 50mm, severe mitral insufficiency secondary to mitral annulus dilatation, global hypokinesia of the LV with severe LV systolic dysfunction and ejection fraction (EF) of 26%.

Diagnosis of congestive HF [New York Heart Association (NYHA) class IV] secondary to PPCM was established and the patient was admitted to coronary intensive care unit. Bi-level positive pressure airway (BiPAP) ventilation, nitrates, diuretics, ACE inhibitor and enoxaparin were started with slow symptomatic improvement. Once clinical and hemodynamic stability was ascertained β-blocker was initiated.

On 16th day she was discharged, medicated with furosemide, carvedilol, lisinopril and aspirin. TTE showed slight improvement of LV systolic function, with an EF of 38%. N-terminal pro-B-type natriuretic peptide (NT-Pro-BNP) was 4897pg/ml.

One year later, she was in NYHA class I, medicated with metolazone, carvedilol, furosemide, lisinopril, spironolactone and aspirin. NT-Pro-BNP was 416pg/ml. TTE showed persistent heart dysfunction, with EF of 36%.

DISCUSSION

PPCM is a rare form of dilated cardiomyopathy associated with a high maternal morbidity and mortality. Its definition is not consensual (**Table 1**).[2]

Reported incidences range from 1/299 live births in Haiti to 1/1000 in South Africa or 1/4000 in the USA.[1]

Although the precise mechanisms that lead to PPCM remain unknown, some possible etiological processes have been suggested, including viral myocarditis, abnormal immune response to pregnancy, maladaptive response to hemodynamic stresses of pregnancy, stress-activated cytokines, prolonged tocolysis, malnutrition and selenium deficiency and familial predisposition.[1,2,3] Several risk factors predispose to PPCM including general risk factors for cardiovascular disease (hypertension, diabetes, obesity, smoking, African descent) and pregnancy related factors (maternal age>30 years, multiparity, multiple pregnancy, preeclampsia and gestational hypertension).[1,3]

TABLE 1 DEFINITIONS OF PERIPARTUM CARDIOMYOPATHY[2]

	Definition of PPCM
European Society of Cardiology on the classification of cardiomyopathies	A non-familial, non-genetic form of dilated cardiomyopathy associated with pregnancy.
American Heart Association Scientific Statement on contemporary definitions and classification of the cardiomyopathies	A rare and dilated acquired primary cardiomyopathy associated with left ventricular dysfunction and HF.
The National Heart Lung And Blood Institute and The Office Of Rare Diseases	The development of HF in the last month of pregnancy or within 5 months post-partum. The absence of an identifiable cause of HF. The absence of recognizable heart disease prior to the last month of pregnancy. Left ventricular systolic dysfunction demonstrated by classical echocardiographic criteria. The latter may be characterized as a LVEF < 45%, fractional shortening <30%, or both, with or without a left ventricle end-diastolic dimension >2.7cm/m2 body surface area.
Heart Failure Association of the European Society of Cardiology Working Group on PPCM 2010	PPCM is an idiopathic cardiomyopathy presenting with HF secondary to left ventricular systolic dysfunction towards the end of pregnancy or in the months following delivery, where no other cause of HF is found. It is a diagnosis of exclusion. The left ventricle may not be dilated but the ejection fraction is nearly always <45%.

HF- HEART FAILURE; **LVEF-** LEFT VENTRICLE EJECTION FRACTION.

PPCM presents with signs and symptoms of HF, with NYHA functional class III/IV in most cases.[2] In the majority of patients, symptoms develop in the first four months postpartum.[2] Features of a normal pregnancy and early puerperium can mimic those of mild HF, including peripheral edema, nonspecific fatigue and exertion dyspnea.[2,3] Thus, a high index of suspicion is necessary for diagnosis. Thromboembolic events are more common in PPCM than in other types of cardiomyopathy, probably associated with pregnancy induced hypercoagulable state.[4] PPCM is a diagnosis of exclusion and other causes of HF (myocardial infarction, myocarditis, sepsis, severe preeclampsia, pulmonary embolism, valvular diseases, pre-existing idiopathic dilated cardiomyopathy unmasked by pregnancy) should be excluded.[2] ECG, chest radiographs, laboratory tests and cardiac imaging should be performed. As the definitive diagnosis of PPCM requires identification of LV systolic dysfunction, any peripartum woman with HF symptoms should perform a cardiac imaging technique.[2,3] Although echocardiography is the most widely available imaging modality, magnetic resonance may be helpful allowing more accurate measurement of chamber volumes and ventricular function and in the differential diagnosis of myocarditis.[5]

After delivery, PPCM should be treated in accordance with the current guidelines for HF.[2] However, modifications to standard therapy are often necessary regarding the safety of the pregnant and the fetus or breastfeeding child. Recently, excessive production of prolactin has been implicated in the pathogenic mechanisms of PPCM and several reports have suggested that the addition of bromocriptine may be beneficial.[2,6,7] Anticoagulation therapy should be considered in patients with EF<35%.[1,2] Mechanical cardiovascular support may be required.[2,3] Published data show that up to 11% of patients with PPCM undergo heart transplantation, with similar results to idiopathic dilated cardiomyopathy.[2,8]

When PPCM presents during pregnancy, collaboration between cardiologists and obstetricians is essential to define the therapeutic strategy and deli-

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very time and mode. Unless there's maternal or fetal deterioration early delivery isn't necessary.[2] Cesarean section is only indicated if obstetric indication is present and in critically ill woman needing inotropic support.[2,4] Epidural analgesia and shortening of expulsive period are indicated.[2]

Based on the possible negative effects of prolactin and the high metabolic demands of lactation, breastfeeding is not advised.[2]

Prognosis is variable and appears to be positively related to LV size and function recovery.[1,2] Fractional shortening <20%, LV diastolic dimension ≥6cm at the time of diagnosis, LVEF≤30% and elevated troponin I are associated with higher risk of persistent cardiac dysfunction.[3,9,10,11] Aproximately 30-50% of women with PPCM have complete recovery of cardiac function.[2,3] Reported mortality rates for PPCM vary widely, up to 50%.[2,12] Two large studies in USA reported mortality rates of 1.36%-2.5%, suggesting that PPCM's current mortality may be less than reported in older series.[13,14]

Currently, there is no consensus regarding recommendations for future pregnancy. LV recovery and function are considered the most reliable prognostic factors and predictors of survival in subsequent pregnancies.[15] Women with LVEF<25% at diagnosis or where LV function hasn't normalized should be advised against subsequent pregnancy.[2] Women whose cardiomyopathy apparently resolved completely are a more difficult group to counsel. As multiparity is considered a risk factor for PPCM, subsequent pregnancies may increase the risk of recurrence, irreversible cardiac damage and decreased LV function.[1] In addition, even when LV size and function returns to normal, there is no evidence that contractile reserve isn't impaired1. All patients should be informed of the possible negative effects of pregnancy on cardiac function and that development of HF and death may occur.[2] Subsequent pregnancies, if not avoided, should be managed in collaboration with a high-risk perinatal center.[1] Careful counseling about contraception should be provided (**Table 2**). In the presented case, if tubal ligation was not performed, a subsequent pregnancy should be discouraged as LVEF remained low at one-year follow-up, establishing the diagnosis of peripartum cardiomyopathy without left ventricular systolic function recovery. In this condition, follow-up must be kept, with clinical evaluation and echocardiogram performed every 6-12 months.

TABLE 2 CONTRACEPTION IN PERIPARTUM CARDIOMYOPATHY[2,16,17]

METHOD	PEARL INDEX	WHO CLASSIFICATION[a]		CONSIDERATIONS
		NYHA Class I or II	NYHA Class III or IV	
COMBINED HORMONAL CONTRACEPTION (Oral /Vaginal /Transdermal)	8	3/4	4	Estrogen related risks of thromboembolic events
PROGESTOGEN-ONLY CONTRACEPTION		1	2	
Oral	8			
Intramuscular	3			
Subdermal	0,5			
INTRAUTERINE DEVICES		2	2	
Copper	0,5			Risk of menorrhagia in hipocoagulated women
Progestogen-releasing	0,1			Specially indicated in controlling menorrhagia associated with hipocoagulation
BARRIER METHODS	15	1	1	High failure rate
FEMALE STERILIZATION				
Tubal ligation	0,1-0,5	2	2	Anesthetic risk
Intratubal stents	0,1	1	1	Anesthesia not required
MALE STERILIZATION				
Vasectomy	0,15			

PEARL INDEX – number of unwanted pregnancies using a contraceptive method per year. **a WHO** (World Health Organization) Classification: 1- No restriction to use; 2- Advantages outweigh possible risks; use requires adequate surveillance; 3- Risks outweigh advantages; use not recommended; 4- Unacceptable risks; contraindicated use.

CONCLUSION

PPCM is a rare disease that affects previously healthy young women. The rarity of this condition makes it particularly difficult to study and may lead to late diagnosis and treatment. Although some women experience clinical and echocardiographic recovery, it's still associated with high mortality.

Close collaboration between obstetricians and cardiologists is essential for appropriate management. Subsequent pregnancies remain controversial, as recurrence may occur.

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