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MEADOWS SYNDROM, A Poorly Understood Entity: Case Report

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Abstract

*Peripartum cardiomyopathy (PPCM) is a heart disease that occurs in previously healthy women towards the end of pregnancy or in the first months after childbirth (with two-thirds of all cases occurring during childbed) **[1]**. In Germany, around one in 1000 to 1500 pregnant women will develop PPCM. As the symptoms of PPCM (dyspnea, leg edema, orthopnea, fatigue) are unspecific and often difficult to distinguish from normal peripartum symptoms, there is often a considerable delay in diagnosis and consequently in therapy, which is associated with a poorer prognosis. This is precisely why we feel it is important to draw the attention to this dangerous peripartum disease, which can present differently, from acute heart failure to cardiogenic shock. The mortality rate from PPCM is 2% to 30%, and a significant proportion of women develops chronic heart failure. Therefore, early diagnosis and therapy are essential.*

***Keywords**: pregnancy, dyspnea, orthopnea, peripartum, echocardiography, heart failure.*

Introduction

Peripartum cardiomyopathy or MEADOWS syndrom is a form of systolic heart failure affecting young women toward the end of pregnancy or in the months following delivery. Incidence is higher in African-American women and in women with older maternal age, hypertensive disorders of pregnancy, and multiple gestation pregnancies. Symptoms of heart failure mimic those of normal pregnancy, often resulting in a delay in diagnosis and preventable complications. Echocardiography showing decreased myocardial function is essential for the diagnosis. Medical management is similar to heart failure with reduced ejection fraction of other etiologies, but adjustments during pregnancy are necessary to ensure fetal safety. Variable outcomes include complete recovery, persistent heart failure, arrhythmias, thromboembolic events, and death. Subsequent pregnancy confers substantial risk of relapse and even death if there is incomplete myocardial recovery.

Case Report

We report the case of a young 28-years-old parturient without any notable pathological medical history, G3P3, 2 children delivered by vaginal delivery the 3rd pregnancy was well followed and carried to term with spontaneous labor, the delivery was by cesarean section for suspicion of acute fetal distress.

The immediate post-partum follow up was simple, twelve hours later she suddenly presented a dyspnea with orthopnea. The examination found a normotensive, tachycardic, apyretic, dyspneic patient, with desaturation on room air up to 60%. We found a good safety globe, physiological lochia, and soft calves.

The patient was put in condition, hemodynamic and respiratory monitoring, half sitting position and oxygen therapy.

A pulmonary embolism was suspected given the trombo-embolic context, a D-Dimer dosage was performed, coming back slightly elevated, completed by a thoracic angioscanner which did not reveal signs in favor of pulmonary embolism, the echodoppler of the lower limbs was normal.

This led to an ECG, a troponin assay which was also elevated, and then an echocardiography which showed a myocardial contractility failure with a systolic function of 25%, thus concluding that the patient had acute cardiac insufficiency linked to cardiomyopathy, without any previous history of cardiac disease, revealed in the context of peripartum.

This clinical profile reminds us of peripartum cardiomyopathy or MEADOWS syndrome, which is a rare entity but with a high risk of morbidity and mortality.

Our patient was put on treatment for heart failure, with a combination of beta-blocker, ACE inhibitor and diuretics. The monitoring shows a good clinical evolution but also a quick improvement of the cardiac function after initiation of the heart failure treatment with a control systolic function at 60%.

This is a rare entity that should be considered when faced with such a clinical condition in order to guarantee adequate and, above all, early management in order to improve the prognosis, which may be poor in the case of delayed diagnosis or therapy.

Discussion

Peripartum cardiomyopathy (PPCM) is a diagnosis of exclusion in women presenting with HF due to left ventricular (LV) systolic dysfunction and should be considered when no other cause is evident. PPCM was previously defined as symptomatic HF presenting in the last month of pregnancy and up to 5 months postpartum **[**2-3**]**. The diagnostic criteria indicate that LVEF is <45% and there may or may not be ventricular dilatation.

The incidence of PPCM differs widely depending on the ethnic/racial and regional background of women. Africans and African, Americans are at a higher risk for developing PPCM, with an estimated incidence of 1:100 pregnancies in Nigeria and 1:299 in Haiti, whereas incidences in Caucasian populations range from 1:1500 pregnancies in Germany to 1:10 000 in Denmark. In a large United States cohort of well-phenotyped patients, African American women were diagnosed with PPCM at a younger age and later in the postpartum period, and were more likely to present with a LVEF < 30% compared with non – African American women. In the USA, an increasing incidence was described over the past years. In a Japanese cohort the incidence was as low as 1:20 000**[**4**]**.

Predisposing factors for PPCM seem to be multiparity and multiple pregnancies, family history, ethnicity, smoking, diabetes, hypertension, pre-eclampsia, malnutrition, age of mother (with older mothers being at greater risk), and prolonged use of tocolytic **[**5**]**.

The aetiology of PPCM is uncertain. Possible factors leading to PPCM include genetic predisposition, low selenium levels, viral infections, stress-activated cytokines, inflammation, autoimmune reaction, pathological response to haemodynamic stress, unbalanced oxidative stress and induction of antiangiogenic factor. Particularly, the oxidative stress-mediated cleavage of the hormone prolactin into a smaller antiangiogenic sub fragment, 16-kDa prolactin, may drive PPCM by inducing endothelial damage. Release of endothelial microparticles loaded with active compounds such as microRNAs, whose release into the circulation is also induced by 16-kDa prolactin, may subsequently impair cardiomyocyte metabolism and further contribute to the manifestation of PPCM **[**1**]**.

The link between vascular pregnancy complications (e.g. pre-eclampsia) and PPCM was strengthened by the observation that women with PPCM had high levels of soluble fms-like tyrosine kinase 1 (sFlt-1), a potent vascular endothelial growth factor inhibitor, which has been implicated in the pathogenesis of pre-eclampsia, suggesting an overlap between these conditions. Indeed, pro-angiogenic therapies could rescue the PPCM phenotype in experimental models. In conclusion, PPCM is a complex disease with a quite heterogeneous and incompletely understood pathophysiology involving angiogenic, metabolic, hormonal and oxidative stress factors **[**1**]**.

PPCM is a diagnosis of exclusion, the clinical features and diagnosis include Symptoms of shortness of breath on exertion and orthopnoea are common in the third trimester. Diagnosis of PPCM can be delayed if these symptoms are erroneously attributed to the normal physiological changes of pregnancy. Symptoms of PPCM

include dyspnoea, fatigue, orthopnoea, paroxysmal nocturnal dyspnoea, palpitations, haemoptysis and peripheral oedema. These may present as acute or subacute episodes of left ventricular failure **[**6**]**.

Tachyarrhythmias can occur in PPCM, including supraventricular tachycardia (SVT), atrial fibrillation (AF) and, rarely, ventricular tachycardia (VT). There is an increased risk of thromboembolism due to the potential development of mural thrombus. Diagnosis of PPCM requires four criteria to be met **[**1**]**:

1. Heart failure developing towards the end of pregnancy or up to 5 months post-partum
2. Absence of another identifiable cause of cardiac failure
3. Absence of cardiac symptoms or disease prior to late pregnancy
4. Left ventricular dysfunction – defined as an ejection fraction less than 45% or reduced fractional shortening of less than 30%.

Echocardiography should be performed in any suspected case of PPCM as the LVEF is typically <45%. In addition to systolic dysfunction, the echocardiogram may demonstrate LV and right ventricular dilatation and/or dysfunction, functional mitral and/or tricuspid regurgitation, pulmonary hypertension, and left atrial or biatrial enlargement. Intracardiac thrombus may occur, and the LV apex should be clearly visualized particularly when the LVEF is severely reduced **[**7**]**.

Levels of brain natriuretic peptide (BNP) and N-terminal pro-BNP, which do not change significantly during normal pregnancy and may be mildly elevated in the setting of pre-eclampsia, are usually markedly elevated in PPCM. The electrocardiogram may show nonspecific abnormalities, but a normal electrocardiogram does not rule out PPCM **[**7**]**.

PPCM is a diagnosis of exclusion. To avoid overdiagnosis, careful attention to possible pre-existing heart disease including cardiomyopathies and valvular disease is important. Severe pre-eclampsia can cause Heart Failure (HF) related to diastolic dysfunction, but PPCM is only diagnosed in the presence of systolic dysfunction.

Early identification of PPCM enables optimal management. Multidisciplinary management by senior staff is essential, and should include involvement from an obstetrician, a cardiologist with a special interest in obstetric disorders, and an anaesthetist. Should PPCM present during pregnancy, maternal optimisation is key, which may necessitation expedited delivery. A neonatologist should be involved in discussions regarding timing of delivery and to address the potential impact of maternal disease on neonatal outcome. If critical care admission is likely, early involvement of a critical care specialist is important PPCM **[**8**]**.

Management of PPCM is similar to that of other causes of cardiac failure and includes maintenance of adequate oxygenation, fluid and salt restriction and ventricular off-loading by vasodilation and diuresis. Hydralazine and nitrate therapy, such as isosorbide dinitrate, can be used safely during pregnancy. These drugs reduce afterload, preload and intracardiac filling pressures. Beta-blockers have been shown to improve survival and may be protective against tachyarrhythmias. Digoxin is considered safe in pregnancy and may be used for its positive

inotropic effect and for the treatment of atrial fibrillation should it occur. Digoxin plasma levels should be carefully monitored **[**7**]**.

Owing to the risk of venous thromboembolism and mural thrombus associated with PPCM, prophylactic low-molecular-weight heparin (LMWH) therapy is indicated. If thromboembolic sequelae or mural thrombus have been identified, full anticoagulation is indicated **[**9**]**.

Patients in the acute setting may need supportive therapy with non- invasive ventilation or intubation, ventilation and inotropic support. In severe cases an intra-aortic balloon pump, left ventricular assist device (LVAD) or extracorporeal membrane oxygenation (ECMO) may be required. Heart transplantation may be indicated for patients with severe disease who do not respond to pharmacological treatments. Implantable defibrillators and cardiac resynchronisation are possible interventions in those women who survive the acute phase but experience on-going significant functional impairment **[**9**]**.

Vaginal delivery is always preferable if the patient is haemodynamically stable and there are no absolute obstetric indications for caesarean delivery. Close haemodynamic monitoring is required. Epidural analgesia is preferred. Urgent delivery irrespective of gestation duration should be considered in women with advanced heart failure and haemodynamic instability despite optimal heart failure treatment, in these cases, caesarean section is recommended with central neuraxial anaesthesia **[**10**]**. Uterotonic drugs should be used cautiously because of their associated side-effects.

Prognosis is poor and PPCM is one of the leading causes of maternal death. A prognostic indicator is the degree of dysfunction at presentation, defined either by the New York Heart Association (NYHA) functional classification or by the findings on transthoracic echocardiography. The mortality rate is between 15% and 50%, while 30–50% will improve and recover a left ventricular ejection fraction of 50% or more. Death results from intractable heart failure, arrhythmias and thromboembolism **[**11**]**.

Breastfeeding in patients with heart failure is controversial. The observation that breastfeeding seems to be safe in PPCM suggests that continued stimulation of prolactin secretion may not be harmful. Most HF medications can be given safely with breastfeeding and should not be a reason to advise women against lactation **[**12**]**.

The importance of contraception should be emphasized by the cardiologist, as well as the obstetrician/ gynecologist. In the early postpartum setting with severe LV (left ventricul) dysfunction, the increased risk of thromboembolism should dissuade the use of estrogen-containing contraceptives. Progesterone-releasing subcutaneous implants or the Mirena intrauterine device are safe and effective choices. Injectable depot medroxyprogesterone acetate is less effective and is considered a second-line option. Nonhormonal barrier methods are less effective. Tubal ligation and vasectomy are other options. Therefore, women should be encouraged to select the method they will use most consistently.

The safety of a subsequent pregnancy is a frequent concern for patients and their families. Appropriate and accurate counseling is essential. The risks associated with a subsequent pregnancy depend primarily upon

whether the myocardial function has fully recovered, and the pre-pregnancy LVEF is the strongest predictor of outcomes. Detection of subclinical LV dysfunction by stress testing and strain imaging has been proposed, but further research about the predictive utility of these tests is needed.

Conclusion

The diagnosis of PPCM should be considered in any pregnant or postpartum woman with symptoms concerning for heart failure. An elevated BNP level should always be followed by an echocardiogram to assess for systolic dysfunction. Prompt treatment with medications tailored for pregnancy and lactation may prevent adverse outcomes. Limited studies suggest breastfeeding is safe. Acutely ill women should be managed by specialized multidisciplinary teams, and may require advanced heart failure therapies. Women considering a subsequent pregnancy should be counseled and monitored by physicians familiar with PPCM. Long-term follow-up is important, but the optimal duration of medications following recovery is unknown. Given the overall rarity of PPCM, collabo- ration among multiple.

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