

Peripartum cardiomyopathy: A case report

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ABSTRACT: Peripartum cardiomyopathy (PPCM) is a rare cardiac disorder of unknown origin associated with a high risk of mortality (50%). Echocardiography confirms the diagnosis by showing a left ventricular dilatation and a decreased ejection fraction. Up to date, the treatment remains symptomatic. The authors report the case of a woman of 36 years old who presented a peripartum cardiomyopathy.

KEYWORDS: peripartum, cardiomyopathy, pregnancy, heart failure, left ventricular dysfunction.

1 INTRODUCTION

Peripartum cardiomyopathy (PPCM) is a dilated and hypokinetic cardiomyopathie occurring during pregnancy or after delivery with an estimated between 1/1000 and 1/4000 births. It has been defined as a new onset of heart failure in the month preceding or following delivery, without demonstrated aetiology nor previously known heart disease and without echocardiographic evidence of left ventricular (LV) dysfunction (LV ejection fraction <0,45). It's a multifactorial disease, immunologic, hormonal, and possibly viral mechanisms playing a determinant pathophysiological role. The classical clinical presentation is a rapid and unexpected onset of heart failure in a previously healthy woman, echocardiography being the key examination for positive and differential diagnosis, prognostication, therapeutic decision making, and follow-up. The potential severity of PPCM, and its unpredictable evolution in the first days following diagnosis, require that patients be referred to a tertiary care centre with a high skill in intensive cardiology care. Therapeutic management of PPCM does not offer any specificity when compared to other causes of acute or chronic heart failure (from diuretics to extracorporeal life support), except for ACE inhibitors, that are contraindicated before delivery. The high incidence of thromboembolic complications observed in the disease requires however rapid and curative anticoagulation and immunosuppressive treatment has been proposed in fulminant and highly inflammatory presentation, but its efficacy remains controversial. Very recently, promising results have been reported with bromocriptin-a prolactin secretion inhibitor for reducing 6 month morbidity and mortality, but these findings have to be confirmed in larger scale randomised trials. As for the long-term evolution, approximately half of the patients will heal, while half of the women will keep some degree of LV dysfunction, 25% of them developing moderate to severe chronic heart failure. We report a case of an acute lung edema secondary to a left heart failure at 37 weeks of gestation in a woman of 36 years old.

2 CASE HISTORY

Our case is about a 36 years old woman, low socioeconomic status, without medical history, Gravida 3 Para 3. The first and second pregnancies were natural deliveries at home without abnormality. In fact, the patient did not report a high blood pressure after her deliveries. The current pregnancy was poorly monitored. It was estimated at 37 weeks of gestation according to the last menstrual period. The patient was admitted to obstetric emergencies for acute dyspnea NYHA stage III of the New-York Heart Association (NYHA) associated to orthopnea, cough and frothy sputum. The overall examination showed an obese patient, conscious with profuse sweat, heart rate at 100 beats / minute, breath rate at 30 cycles / min with high blood pressure ranging from 180 to 160 mmHg for systolic and 120-110 for diastolic associated to neurosensory signs

and important edema of the lower limbs. Besides, the patient reported a quick weight gain since a month. Urinary Dipstick looking for proteinuria was positive with 4 cross albumin which it was in favor of severe preeclampsia. The chest auscultation showed crackles at the base and summits. The obstetric examination revealed a fundal height at 35 cm with blubber, fetal heart sounds were well perceived. On palpation, the uterus was soft with cephalic presentation back to the left. There was no bleeding in the speculum examination. In the vaginal touch, the cervix was posterior, long, admitting the pulp of the finger with a mobile cephalic presentation and a flexible lower segment of the uterus. Ultrasound showed a cephalic presentation, the estimation of fetal weight was 3400 g, an anterior placenta, the quantity of amniotic fluid was normal. Umbilical and cerebral Doppler were normal. The electrocardiogram showed sinus tachycardia at 110 beats / min with some atrial extrasystoles. Chest-X-rays radiography with abdominal protection was in favor of an acute pulmonary edema. Transthoracic echocardiography showed a dilated left ventricle, global hypokinesia and left ventricular dysfunction. Left ventricular ejection fraction was at 31% (Figure 1). The left atrium was dilated, the right cavities were non-dilated and the right ventricle systolic function was normal. On the Doppler, there was a mitral regurgitation grade I-II and a tricuspid insufficiency grade I with significant pulmonary hypertension. The pericardium was dry. These results were in favor of dilated cardiomyopathy. The patient was quickly put in condition (half sitting position, oxygen, urinary catheterization). She received loop diuretic (Furosemide) 40 mg intravenous and an antihypertensive parenterally (calcium inhibitor). A biological assessment with: blood count with platelets, complete blood ionogram with liver function and renal function, C-reactive protein (CRP), lipid profile, thyroid balance and cardiac enzymes was made. The blood count was unremarkable, the electrolytes, the liver and renal function tests were normal, CRP was elevated at 10.4 mg/l. The thyroid function was normal and the troponin IC was negative. A C-section was carried out under general anesthesia allowing the extraction of a newborn baby girl weighing 3200g Apgar 10/10 at 1,5 and 10 minutes of life. Examination of the newborn was unremarkable. After the C-section, the patient was transferred to the intensive care unit (ICU). Diuretic therapy was continued. The blood pressure remained high and calcium inhibitor continued parenterally. Prophylactic heparin therapy was instituted for the prevention of thromboembolic complication. An inhibitor of lactation (cabergoline) and a pressure bandage was in place to prevent breast engorgement. The blood pressure returned to normal in the third day of the postpartum period. After five days of hospitalization in ICU and a good clinical course, the patient was transferred to the cardiology department. She received Furosemide, beta-blockers and angiotensin converting enzyme inhibitor given orally as well as Proton-pump inhibitors, iron (curative dose) and prophylactic heparin. Clinically, improvement was observed. The last transthoracic echocardiography before discharge found an ejection fraction of 32%. The patient was the sixth outgoing days within inhibitors converting enzyme, beta-blockers. Microprogestative pill was prescribed as a contraceptive.

3 DISCUSSION

Peripartum cardiomyopathy has been defined by the Working Group on Heart Failure of the European Society of Cardiology as an idiopathic cardiomyopathy manifested by signs of heart failure secondary to left systolic dysfunction between the last month of pregnancy and the five months following the delivery disease when no other etiology was found. Dilatation of the left ventricle is not compulsory but the left ventricular ejection fraction (LVEF) is almost always less than 45% [1]. Several risk factors for PPCM were identified: maternal age > 30 years, multiparity, multiple pregnancy, obesity, high blood pressure, pre-eclampsia [2]. In fact, in 1/3 of cases, no risk factor was found. Our patient had four of these risk factors: She was 36 years old, she was Para 3, obese with preeclampsia in her admission. The clinical picture of PPCM is systolic heart failure. Functional signs are dyspnea, cough, edema of the lower limbs. The cardiopulmonary auscultation found crackles or wheezes related to pulmonary congestion [3]. Our patient presented the same clinical picture. The differential diagnosis includes pulmonary embolism, pneumonia, infarction or aortic dissection. The electrocardiogram (ECG) usually shows sinus tachycardia, but atrial arrhythmias is not uncommon [4,5]. Besides, Left ventricular hypertrophy, ischemia in epicardial Q waves, ST segment changes may also see [6]. ECG our patient showed sinus tachycardia at 110 beats / min with some atrial extrasystoles. Chest-X-rays is not specific showing signs of heart failure and inconstant cardiomegaly. Successive radiography will allow to follow the evolution of congestive signs. In our case, chest-X-rays (made with protection of the abdomen) objectified an acute pulmonary edema. Echocardiography is the key examination for positive and differential diagnosis, prognostication, therapeutic decision making, and follow-up. She often found a left ventricular dilatation, decreased less than 45% ejection fraction (the left ventricular ejection fraction in our patient was 35%) and possible right ventricular disease associated [1]. Finally, it is often used to confirm the diagnosis of PPCM by eliminating pre-existing heart disease (hypertrophic, rheumatic valvular, ischemic). Biology is not specific. It assesses the severity of cardiac disease and follow the evolution: Troponin Ic or T related to myocyte necrosis, C-reactive protein (CRP) reflecting the scalability of inflammatory processes, transaminases, creatinine and lactate, to assess the visceral impact of heart failure. The evolution of the PPCM is often extremely fast and unpredictable with possibility of sudden onset of refractory cardiogenic shock in the first 24-48 hours. The treatment of PPCM in acute phase is symptomatic: it is that of all heart failure. It is to reduce preload and afterload and increase myocardial contractility. Several drugs are usable depending on the clinical and the period of

pregnancy. Decreasing the precharge can be provided by nitro-derivatives and diuretics. Angiotensin converting enzyme inhibitor are the best treatment to decrease afterload, but they are contraindicated during pregnancy because of the side effects on the foetus (oligoamnios, prematurity, bone malformation....)[7]. Amlodipine or hydralazine-nitro-derivatives association are alternatives during pregnancy. Beta-blockers are prescribed during tachycardia or arrhythmia. Dobutamine increases myocardial contractility without risk during pregnancy [8,9]. Therapeutic abortion may sometimes be justified in case of worsening heart failure despite optimal medical therapy. The total duration of this medical long-term treatment is not well defined. It must be at least a year and then based on data of the ultrasound monitoring. Some authors advocate unconventional treatments etiological as immunosuppressive (azathioprine, prednisolone), plasmapheresis, immunoglobulin, interferon or immunomodulation for patients who remain symptomatic under conventional medical treatment and in whom myocarditis is proved by endomyocardial biopsy [10]. A small randomized study published in 2010 has opened up new perspectives. This open study comparing 20 patients placed under bromocriptine (dopamine agonist anti-prolactin activity: 2.5mg two times a day over 15 days followed by 2.5 mg / day for 6 weeks) versus conventional treatment isolated, showed a benefit effect in terms of improvement in LVEF to 4 months. Other complications associated to PPCM include thromboembolic events which are found in nearly 50% of cases. The hypercoagulability secondary to pregnancy and blood stasis in dilated cavities promote the formation of thrombi. In fact, anticoagulation is justified and should be pursued as ejection fraction is less than 35% [11]. The management of the patients with PPCM is multidisciplinary including obstetrician, cardiologist, anesthetist and pediatrician. Childbirth can be done vaginally and epidural analgesia is then medically indicated, as it enables the reduction of myocardial work during labor and an improvement in cardiac function by decreasing load conditions. An indication for c-section may be raised by the obstetrician or in case of severe cardiac decompensation. General or regional anesthesia is then used, the goal remains to maintain a good hemodynamic stability. Spinal anesthesia alone is strongly discouraged because of the sudden hemodynamic changes it engenders and which can be fatal in this context. Epidural anesthesia and combined epidural spinal anesthesia are preferred. The regional anesthesia is preferred except in cases of emergency obstetric and contraindicated. If the PPCM occurs in the last months of pregnancy, in order to benefit the mother of a better therapeutic management, rapid extraction of the fetus is often indicated. If the cervix "is ready" and if maternal hemodynamic status permits, vaginal delivery may be authorized. Otherwise (cervix "closed" and / or heart failure NYHA class > I), a C-section is preferred [12]. Recurrence of PPCM during a subsequent pregnancy occurs in 25-100% of cases depending on the study [12]. Although it has no reliable predictor, the failure to recover normal LVEF waning of a first episode predispose to recurrence. Finally, the European Society of Cardiology recommends to [13]:

- Discourage a future pregnancy in a patient with ventricular sequelae post PPCM.
- Against state-of pregnancy if LVEF <50%.
- Perform a therapeutic abortion in case of deterioration of LVEF <50% in the 1st or 2nd quarter, due to the risk of heart failure may jeopardize the mother's prognosis.
- Perform echocardiography screening and regular monitoring in patients with a history of PPCM with normalization of LVEF, as well as in patients with a family history of cardiomyopathy or PPCM ascendants and collaterals in the first degree.

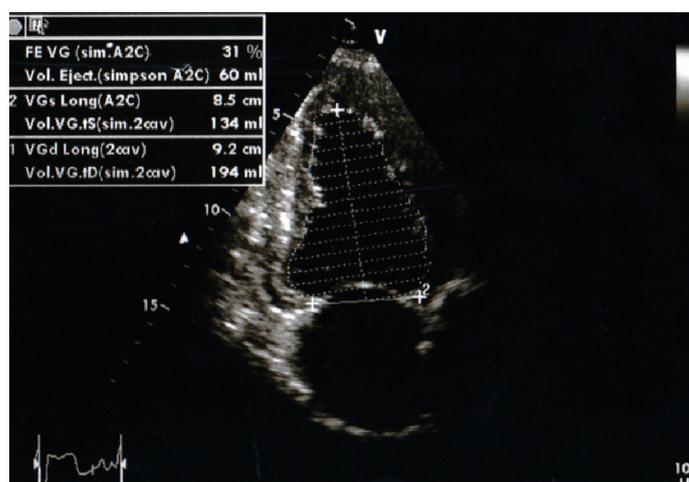


Figure 1: Echocardiography showing a dilated left ventricle, global hypokinesia and left ventricular dysfunction. Left ventricular ejection fraction is 31%.

4 CONCLUSION

Peripartum cardiomyopathy is a rare but serious disease in pregnant women. It is a hypokinetic dilated cardiomyopathy. Its pathophysiology is unknown and multifactorial. Its treatment is symptomatic, but must take into account the state of pregnancy. Angiotensin converting enzyme inhibitor are the treatment of choice but they are contraindicated during pregnancy. Other specific therapies are being evaluated based on etiological hypotheses. A Secondary normalization of ventricular function does not exclude a risk of recurrence in a subsequent pregnancy justifying a multidisciplinary long-term monitoring.

CONFLICTS OF INTEREST

The authors declare that they have no conflicts of interest related to this article.

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