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Case Report

Peripartum Cardiomyopathy: About Two Cases and Review of Literature

Fatnassi Ridha*, Turki Elyes, Marouen Nedia and Zgaya Rym

Department of Gynecology and Obstetrics, Ibn El Jazzar Hospital, Kairouan, Tunisia

***Corresponding Author:** Fatnassi Ridha, Department of Gynecology and Obstetrics, Ibn El Jazzar Hospital, Kairouan, Tunisia.

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Abstract

Peripartum cardiomyopathy (PPCM) is a dangerous and a rare pathology; with an inaccurate etiology. Some risk factors have been identified such as advanced maternal age, multiparity, pregnancy and induced hypertension.

The diagnosis is based on the association of clinical presentation of an acute heart failure, and echographic signs of systolic dysfunction. The main differential diagnoses are myocardial infarction, myocarditis, and familial cardiomyopathy. The treatment is based on Digitalis and Diuretics, however a specific treatment based on Bromocriptine and immunosuppressive agents could be indicated. The evolution of this pathology remains unforeseen ranging from complete recovery to maternal death.

We are reporting two cases of PPCM which occurred in two parturient of 31 years old and 35 years old. Symptomatic treatments have been initiated for both patients, and the evolution was marked by intrauterine fetal death with a mother recovery for the first case, and a maternal death for the second case.

Keywords: *Cardiomyopathy; Pregnancy; Peripartum; Heart Failure*

Introduction

Peripartum cardiomyopathy is a serious heart failure that occurs during or following pregnancy. It has been initially described in 1849, but the first anatomoclincal description was made by Meadows in 1957, for this reason the PPCM is often referred to as Meadows syndrome [1].

It is about a rare and primary dilated cardiomyopathy [2,3], and etiopathogenesis of which is still a mystery. Its diagnosis should be mentioned in case of heart failure clinical signs occurring in late pregnancy or postpartum in absence of cardiomyopathy history. The confirmation is made essentially by echocardiography [4]. The evolution of this affection remains unpredictable [5]. The prognosis mainly depends on the precocity of the management.

We are presenting two new cases in the third trimester and postpartum, and we are discussing the clinical and therapeutic sides of PPCM.

Case Report

Case no 1

It's about M Z, a 31 year-patient with no particular pathological history, gravida 2, para 0, caucasian, from an average socio-economic level. she consulted our emergency at 36 WA, complaining of chest pain, dyspnea and dry cough dating back to three days ago. The clinical examination showed;

Blood pressure at 150/100 mmHg, tachycardia, polypnea with orthopnea, vesicular murmurings were perceived, but markedly diminished at the level of the right pulmonary base. Proteinuria with Labstix at +++. On the obstetrical examination the uterine height was at 34 cm with a normally relaxed uterus, and pelvic examination was without abnormalities with closed and posterior cervix.

The electrocardiogram showed sinus tachycardia without repolarization disorders (Figure 1). Chest x-ray revealed pleural effusion and increased cardiac chest index. Cardiac echocardiography revealed an enlarged left ventricle with hypokinesia of the anterior wall with a left ventricular ejection fraction (LVEF) at 28%, low to moderate abundance pericardial detachment opposite the right ventricle and a right moderate pleural effusion (Figure 2).

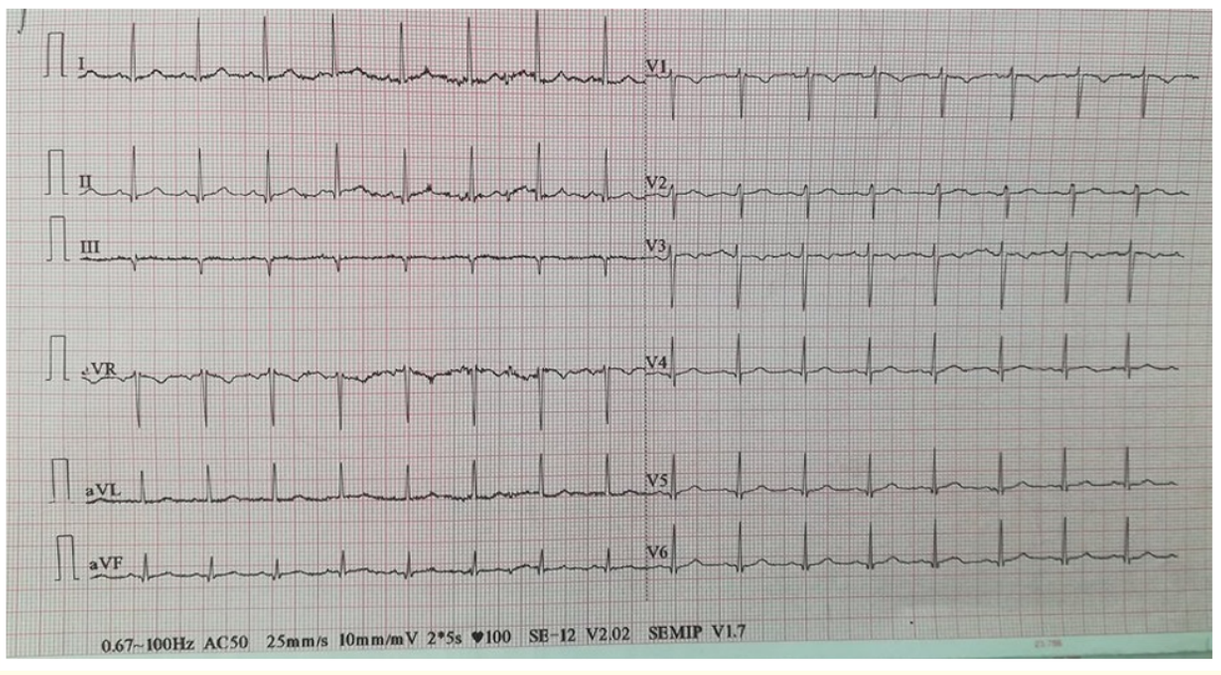
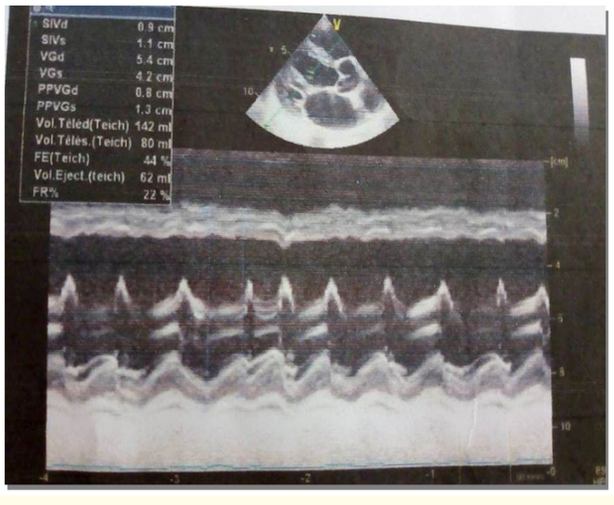


Figure 1: *The electrocardiogram showing sinus tachycardia without repolarization disorders.*



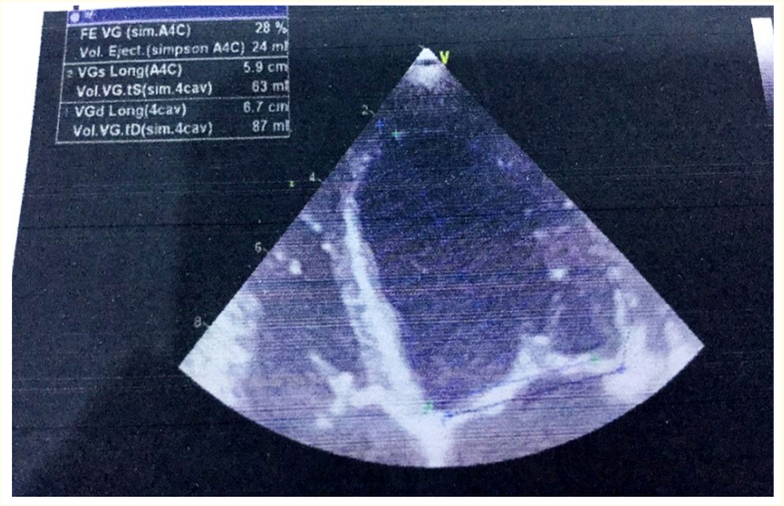


Figure 2: *Cardiac echocardiography: Enlarged left ventricle with hypokinesia of the anterior wall with a left ventricular ejection fraction (LVEF) at 28%.*

The diagnosis of peripartum cardiomyopathy was retained and the patient was hospitalized in the cardiology department for management of pleuro-pericarditis and was put under diuretic and nitrates. A right thoracentesis was also made as well as a careful fetal and maternal monitoring. The evolution was marked by the occurrence of fetal death in utero a day after hospitalization, complicating the clinical picture and indicating the delivery by caesarean section.

The patient showed a clinical and echographic improvement on the second day after delivery having allowed her to be discharged on 10th day of hospitalization (Figure 3).

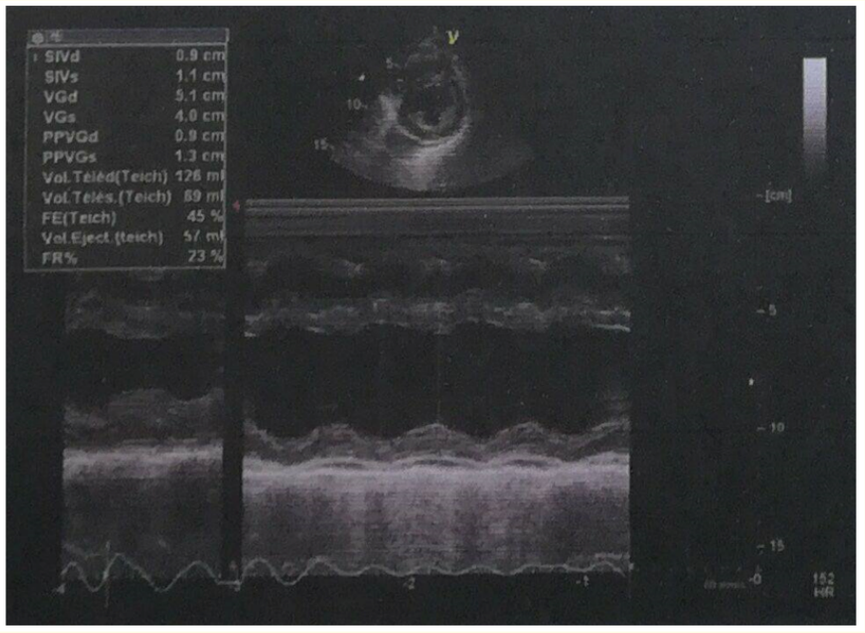


Figure 3: *Cardiac echocardiography after 10 days of hospitalization: the left ventricular ejection fraction (LVEF) at 45%.*

The subsequent evolution was marked by a restitution of cardiac function as evidenced by the ultrasound performed six months later.

Case No 2

It is about O S., a 35-year-old patient with no history of disease, brunette, with an average body build, gravida 3; para 3, with an average socio-economic level, ended with cesarean delivery for acute fetal distress with normal postoperative course and the parturient left the hospital on the second postoperative day.

The subsequent development was marked by the installation of productive cough and gradual increasing exertional dyspnea. The patient was re-hospitalized on the fourth postoperative day and the clinical examination revealed an orthopnea, a blood pressure at 110/70 cm Hg, a heart rate at 100 bat/mn, a temperature of 37°C, crackles at the pulmonary base. Gynecologic examination revealed the presence of lochia made of blood tissue, and mucous with an increased and uterus volume (postpartum) that was painless. A chest x-ray was performed and objectified an alveolar-interstitial syndrome at both lung fields and the borders of the heart was of a normal appearance (Figure 4). The electrocardiogram showed sinus tachycardia without repolarization disorders. Echocardiography revealed a hypokinesia with a left ventricular ejection fraction (LVEF) at 30%, evoking a perinatal cardiomyopathy.

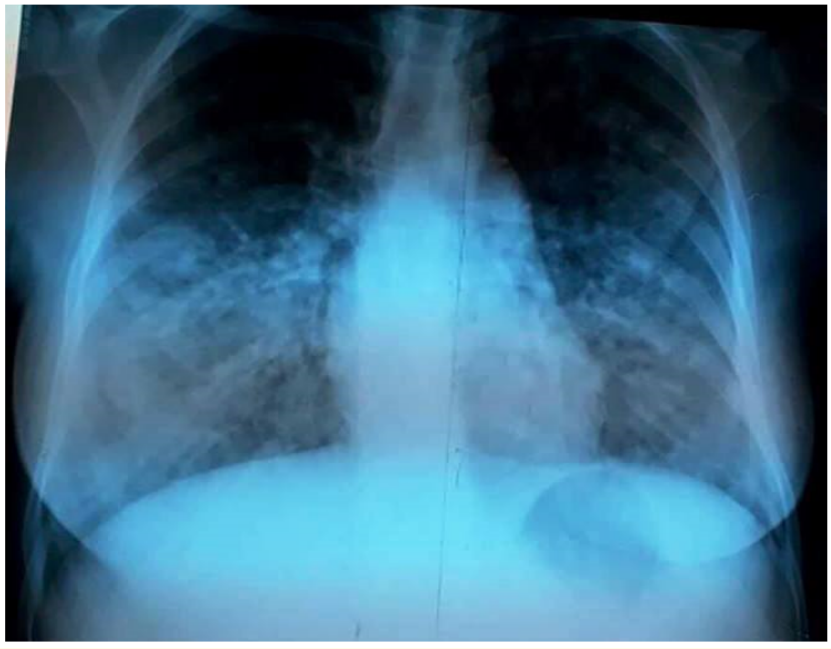


Figure 4: *Chest x-ray: alveolar-interstitial syndrome at both lung fields. Normal appearance of the borders of the heart.*

The patient was monitored and maintained in a semi-sitting position with oxygen therapy by face mask at a rate of 9 liters per minute and inotropic dobutamine therapy initiated by electric syringe shot at the dose of 5 gamma/Kg/mn. However, the course was rapidly fatal within 2 hours to acute pulmonary edema with respiratory distress, sudden desaturation, severe hypoxia, and neurological distress requiring orotracheal intubation and ventilatory support and fatally the occurrence of a cardio-circulatory distress and a cardio-circulatory stop with failure of resuscitation.

A medico-legal autopsy was performed and allowed the diagnosis of peripartum cardiomyopathy by showing significant pulmonary congestion, an enlarged heart with dilated left ventricle thinned wall.

Discussion

PPCM is a dilated cardiomyopathy manifesting in the peripartum period in a patient previously free from pre-existing cardiomyopathy [6]. Diagnosis combines four criteria:

- The occurrence of peripartum congestive heart failure, classically in the month before the delivery or the five months following it.
- The absence of a recognized etiology despite an exhaustive review
- The absence of a history of heart disease before the onset of the first symptoms.
- Echocardiography demonstrating left ventricular dysfunction with a LVEF of less than 45% and/or a shortening fraction (SF) of less than 30% and/or cavitary dilatation with a ventricular end-diastolic diameter greater than 2.7 cm/ m^2 of corporeal area [6].

The incidence of PPCM is difficult to assess, because of the difficulties encountered in differentiating a PPCM from a pre-existing dilated cardiopathy with decompensation, during pregnancy. However, its incidence seems very low, very variable from one country to another. It is estimated at 1/100 births in Nigeria while it is only 1/4000 in the United States. In Tunisia, the PPCM is rare and limited to sporadic cases [7].

The etiopathogenesis of the PPCM is mysterious. However, some risk factors have been identified with maternal age beyond 30 years, multiparity, multiple pregnancies, obesity, high blood pressure, pre-eclampsia and prolonged tocolysis. Other factors seem to influence the occurrence of the PPCM such as African ethnic groups, the low socio-economic level, some vitamin deficiencies as well as zinc, copper, magnesium and selenium [8]. With the exception of maternal age, no other risk factors were identified for the two parturients of our study.

The physiopathology of the PPCM has remained unclear and hypothetical until now. The inflammatory hypothesis is based on the release of mediators of inflammation during pregnancy that may contribute to the formation of myocardial lesions. This release may be exaggerated by stressful situations and infectious conditions [9]. The autoimmune theory is the modification of the immune profile of the parturient at the end of pregnancy and the release during delivery of cross-affinity antibodies with cardiac myocytes [6,9]. For other authors, the PPCM may be the consequence of a hormonal imbalance with a prolactin cleavage under the effect of cathepsin-D protease activated during pregnancy. This cleavage results in the production of 16 Kda-prolactin, the protein responsible for myocardial dysfunction [10]. The hormonal theory also involves the abrupt postpartum drop of estrogen, a cardio-protective hormone [11].

The clinical diagnosis of PPCM is an acute heart failure that is often global, sometimes left heart failure. It is usually severe with quick installation [12]. The main functional signs are exertional dyspnea, orthopnea, asthenia, tachycardia, edema of the lower limbs and sometimes even chest pain. As for the physical signs, they are not specific and include signs of left or global heart failure [13]. In our two parturient, dyspnea was the main symptom and clinical examination revealed signs of heart failure.

The observation of clinical signs of acute global heart failure during the peripartum period should lead to the suspicion of a diagnosis of PPCM and to perform transthoracic Doppler echocardiography (ETT) which constitutes the confirmation examination. Ultrasonographic signs associate left ventricular (LV) dilation and impaired contractile function (FE < 45% et/ou FR < 30%) on the absence of underlying heart disease [14]. In addition, other complementary tests may be useful such as ECG and chest X-ray. The ECG often shows sinus tachycardia. Other electrocardiographic abnormalities can be visualized such as atrial arrhythmia, subepicardial ischemia, necrosis Q wave, ST segment changes and signs of left ventricular hypertrophy. As for the chest X-ray, it is looking for cardiomegaly with or without signs of pulmonary congestion [14]. In our work, the diagnosis of PPCM focused on a bundle of clinical, radiographic and ultrasound elements. In the second case the autopsy validated the diagnosis.

The symptomatology of PPCM can be misleading and confusing with many other conditions. In fact, dyspnea, asthenia and edema of the lower limbs are often attributed to the increase of the uterine volume at the end of pregnancy.

Moreover, other pathologies can be evoked in front of these symptoms of PPCM mainly involving the pre-existent cardiopathy which constitute an acute cause of left ventricular systolic dysfunction, but also the pulmonary embolisms and the attacks of asthma [1,12].

The treatment of this condition is similar to that of congestive left heart failure. It aims to decrease pre-load and post-load and increase myocardial contractility. Because of its contraindication during pregnancy, angiotensin-converting enzyme (ACE) inhibitors are the treatment of choice in postpartum.

The other therapeutic alternatives are based on the limitation of the physical activity, a restriction of fluid and sodium, the prescription of diuretics, digitalis and sometimes even cardiotropic drugs [15]. The treatment initiated for our two patients included nitrates and diuretics for the first case and a cardiotonic agent for the second.

Thromboembolic events constitute a serious complication of the CMPP. They are seen in 50% of the cases [16]. This complication is the consequence of hypercoagulability secondary to pregnancy and blood stasis in right heart chambers [18]. As a result, curative anticoagulation with heparin therapy is mandatory in the event of a proven thromboembolic event and strongly recommended as a preventive measure [17].

Immunosuppressive agents are a therapeutic alternative indicated in cases there is no improvement by conventional treatment with biopsy confirmation of myocarditis [18]. As for bromocriptine, and because of its anti-prolactin effect, it has proved its effectiveness by reducing the rate of morbidity and mortality. It is considered in the case of LVEF < 35% or in case of persistent symptomatology under well-conducted conventional treatment [18].

The obstetric management of these PPCs is based on rapid extraction of the fetus to be able to implement better maternal care. Thus, depending on the gestational age, hemodynamic status of the mother and obstetric conditions, the vaginal approach may be allowed under epidural anesthesia. However, in cases where obstetric conditions are unfavorable and / or heart failure is advanced, a cesarean section is indicated [12].

The evolution of the PPCM is variable. It can lead to complete recovery in 40 to 75% of cases. However, the persistence of stable chronic heart failure under medical treatment is reported in one third of cases. In addition, severe myocardial failure followed by death from terminal heart failure or thromboembolic complications is described in 25 to 50% of cases [4,12].

Several factors of poor prognosis have been linked to this high mortality. These factors included the black race, advanced age (> 30 years), multiparity, persistence of symptomatology beyond two weeks postpartum, onset of symptoms after delivery, persistence of cardiomegaly beyond six weeks and ECG conduction disorders [19].

Conclusion

PPCM is a rare pathology, of an unpredictable evolution. Diagnosis is based on clinical signs of heart failure and ultrasonographic confirmation. Rapid diagnosis and treatment determine the maternal and fetal prognosis. This management involves the usual treatment of heart failure. However, specific treatment with bromocriptine and immunosuppressive agents seems useful.

Urgent fetal extraction conditions adequate maternal management through the introduction of angiotensin converting enzyme inhibitors.

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