Peripartum Cardiomyopathy: Experience at a Tertiary Care Center in Puerto Rico

Anthony D. Osterman-Pla, MD*; Ronald López-Cepero, MD†; Luis Jiménez, MD*;
Josefina Romaguera, MD, MPH†; Juan Aranda, MD*

Objective: Peripartum cardiomyopathy is a rare type of heart failure to which only pregnant women are susceptible. In Puerto Rico there is a paucity of information regarding this condition. In this report we describe our experience with peripartum cardiomyopathy.

Methods: This was a descriptive retrospective study. We conducted a chart review of all the patients with peripartum cardiomyopathy managed at our institution from January 2006 through December 2012. A total of 12 patients qualified for our analysis.

Results: The mean age of our population was 27 (± 8) years. Eight of the twelve (67%) patients were multigravid women, with a mean parity of 2.6 (± 1.6). Most patients (75%) showed clinical evidence of anemia while pregnant, and more than half (58%) delivered prematurely. Six patients (50%) had a prior history of preeclampsia. Cardiac imaging detected a mean ejection fraction of 35% (± 8%) at the time of diagnosis. Only 1 maternal death was documented.

Conclusion: Our study cohort was very similar to those in most case series in which the same condition has been studied. In order to better understand the pathophysiology of this entity and, thus, improve treatment for these patients, prospective studies are needed. [P R Health Sci J 2016;35:224-227]

Key words: Peripartum cardiomyopathy, Pregnancy, Heart failure

Peripartum cardiomyopathy (PPCM) is an unusual cause of heart failure that is unique to pregnant women of all reproductive ages. It is a separate entity from other known existing cardiomyopathies, and it has a poorly understood pathophysiology. PPCM occurs during late pregnancy or early in the first months postpartum and is characterized by a reduced ejection fraction and the absence of any other identifiable cause of heart failure (1). Patients with PPCM have left ventricular systolic dysfunction with an ejection fraction below 45% (1).

Wide variations exist regarding the true incidence of PPCM, with several reports placing it between 1 in 300 pregnancies in places of high incidence, such as Haiti, and 1 in 4025 in areas in which the incidence is lower, such as the United States and other western societies (2–6). These discrepancies may reflect the differences in the incidence of PPCM between ethnic groups and geographical regions, as well as differences in study methodology. In the United States, there has been an overall increasing trend in the incidence of PPCM, which may be due to a heightened awareness of the condition, increases in the risk-factor burden of PPCM among pregnant women, or the expanded timeframe for diagnosis in the newer definition of PPCM (4).

Although the natural history of this condition has not been entirely elucidated, a dysfunction involving the autoimmune system (as well as abnormalities of hormonal function) is likely to be involved (7). In this report, we review our experience at the University District Hospital of Puerto Rico in treating patients diagnosed with PPCM.

Methods

This was a descriptive retrospective study that was completed after having been approved by the Institutional Review Board of the Medical Sciences Campus of the University of Puerto Rico. We conducted a chart review of all the patients who had been admitted with a diagnosis of peripartum cardiomyopathy (ICD-9 code 674.5) to the University District Hospital of Puerto Rico from January 2006 to December 2012. The inclusion criteria were as follows: 1) systolic dysfunction, as documented with an appropriate imaging study, 2) no previous history of heart failure, and 3) onset in the last month of gestation or the first 5 months of puerperium.

*Department of Medicine; †Obstetrics and Gynecology of the University of Puerto Rico School of Medicine, San Juan, PR

Address correspondence to: Anthony D. Osterman-Pla, MD, University of Puerto Rico Medical Sciences Campus, Department of Medicine, PO Box 365067, San Juan, PR 00936. Email: anthony.osterman@upr.edu

Quantitative and qualitative data were collected from medical charts. The following variables were recorded for each patient: age, the moment of clinical onset, medical history, gravidity and parity, the mode of delivery, perinatal complications, laboratory test parameters, the findings of a chest x-ray, the ejection fraction according to the echocardiographic report, the ejection fraction according to the multi-gated acquisition scan (if performed), demographic characteristics, and the levels of B-type natriuretic peptide. A statistical analysis was performed using the SPSS software program (SPSS Inc. Released 2009. PASW Statistics for Windows, Version 18.0. Chicago: SPSS Inc.). Descriptive analysis was performed to describe frequencies, proportions, minimum, maximum, means, and medians (± standard deviations).

Results

Twelve cases that met the criteria for PPCM were identified. The mean age at diagnosis was 27 (± 8) years (range 15–39), with 7 patients being 30 or older at the time of delivery. The majority of patients were multigravid, with a mean parity of 2.6 (± 1.6). There were 2 cases involving multiple gestations.

Table 1. Clinical data on patients with peripartum cardiomyopathy

Mean age at diagnosis (years)	27 ± 8
Mean ejection fraction (%)	35% ± 8
Mean frequency of chest X-ray abnormalities	1.7 ± 0.8
Mean hemoglobin level (g/dL)	10.5 ± 1.3

Cesarean delivery (n = 9), preterm birth (n = 7), and preeclampsia (n = 6) were the most common complications observed in this population. Maternal comorbidities included pregestational diabetes (n = 2), gestational diabetes (n = 1), and systemic lupus erythematosus (n = 1). All the individuals were diagnosed (with PPCM) during the puerperium and for the first time in their lives. One maternal death (a consequence of complications related to cardiomyopathy) was documented. No neonatal deaths were recorded in our population.

A mean ejection fraction of 35% (± 8) was noted overall. For 1 patient, the ejection fraction was obtained from a multi-gated acquisition scan because she had a poor echocardiographic window. Frequent echocardiographic findings included left atrial enlargement (n = 4), moderate–severe mitral regurgitation (n = 3), and pericardial effusion (n = 3). Electrocardiographic data revealed that sinus tachycardia was the most common (n = 5) abnormal finding. Four patients exhibited the electrocardiographic criteria for left ventricular hypertrophy. Six patients had evidence of pulmonary congestion on their chest radiographs. The most frequent hematological laboratory abnormality was anemia, which was present in 9 patients. Anemia was defined as a hemoglobin concentration of less than 10 g/dL (8). There was no evidence of renal failure in any of the patients evaluated. Brain natriuretic peptide (BNP) or N-terminal pro-brain natriuretic peptide (NT-proBNP) was elevated in all the patients in whom one or the other was measured (n = 10).
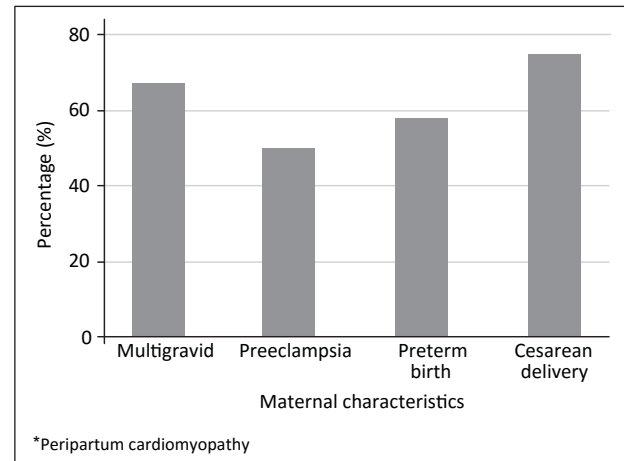
Figure 1. Maternal characteristics of patients with PPCM* (n = 12)

Discussion

This retrospective review accounts for 12 cases of Puerto Rican women who developed PPCM during pregnancy. To date, this is the only study describing pregnant Hispanic women in Puerto Rico with this condition.

Several clinical characteristics have been identified as risk factors for developing PPCM. It has been shown that a maternal age of 30 years or older is a risk factor for PPCM (9). Even though the mean age (27 ± 8 years) of our sample was below this threshold, it is interesting to note that 58% of the patients were older than 30 at the time of delivery. The majority of the patients (67%) were in their second or subsequent pregnancy, implying that having been pregnant at least once is an important risk factor for the development of this condition (mean parity: 2.6 ± 1.6). Similar findings have been well documented by Elkayam, et al (9). Although multiple-gestation pregnancies have been noted to be a risk factor for PPCM (9), only 2 of our patients had twin pregnancies. We noted that preeclampsia is strongly associated with the development of PPCM in our population, as the former was present in 50% of the investigated pregnancies. This correlates with previous accounts that report a strong clinical and pathophysiological association between these 2 disease entities (10).

All the patients included in this study met the definition of PPCM, including their having evidence of a left ventricular systolic dysfunction. In none of the patients examined was there evidence (from the clinical presentation, past medical history, family history, physical examination, laboratory data, or echocardiographic findings) to suggest that there were other possible causes of heart failure, including congenital heart disease, familial cardiomyopathy, severe valvular heart

disease, tachycardia-induced cardiomyopathy, hypertrophic cardiomyopathy, and ischemic heart disease. The mean ejection fraction was 35 (± 8). This is slightly above that of reports from populations of other ethnic backgrounds, which reports detail a left ventricular ejection fraction (LVEF) at diagnosis in the range of 20% to 31% (9, 11, 12). Although the majority of patients with PPCM experience a recovery of their cardiac function (9), it has been suggested that a low LVEF at presentation is associated with a poor prognosis and a lower probability of recovery of cardiac function (13). Regrettably, our patient data is limited to that contained in the hospital medical charts, which document only inpatient encounters. Other echocardiographic findings included mitral regurgitation, left atrial enlargement, and pericardial effusion, which have been previously described as possible echocardiographic findings in this patient population (14). The significance of these echocardiographic findings is still unknown. In addition, all the studied patients had normal sinus rhythms on their electrocardiograms, with only a minority of those patients having abnormalities such as sinus tachycardia (42%) or left ventricular hypertrophy (33%). None of the patients evaluated had evidence of arrhythmias, ST-T wave changes, or conduction abnormalities, which have been previously described (12). With regards to the laboratory test variables, most patients were noted to have significant abnormalities in terms of their hemoglobin levels and BNP or NT-proBNP levels. When using the WHO definition of anemia for pregnant patients (8), 9 patients (75%) were classified as having anemia (mean hemoglobin level of 10.5 ± 1.3). This is consistent with several studies that have identified an association between anemia and PPCM (15, 16). Furthermore, increased BNP or NT-proBNP levels were noted in all the patients in our study in whom those levels were measured (n = 10). This is consistent with the clinical presentation of the studied patients, who had clinical evidence of heart failure. Nevertheless, obtaining BNP or NT-proBNP levels is not required to make the diagnosis.

When evaluating obstetrical and neonatal outcomes, we noted a significant rate of preterm deliveries among the included cases (58%). Even though this rate is higher than what others have reported for their populations (25%) (9), in our population, it (the rate) could be secondary to a higher number of patients presenting with severe preeclampsia in whom early delivery was indicated. Accordingly, the cesarean delivery rate was substantial in our population (75%) and higher than what has been presented in other reports (40%) (9). This is expected in a group of pregnant women with multiple comorbidities, most of whom have already had a cesarean delivery in the past. Nevertheless, despite the complexity of managing these patients, only 1 patient died during hospitalization, and it was due to congestive heart failure secondary to PPCM. Thus, our mortality rate (8%) was noted to be lower than what has been reported in other populations (15%) (5).

Our study had several limitations. As noted previously, the data were obtained from a tertiary care center in Puerto Rico. Thus, our patient population was limited to cases that were referred from other institutions, which patients tended to be those with higher numbers of complications and poorer prognoses. This selection bias had a profound effect on obstetric outcome variables, such as preterm delivery rate and cesarean section rate. Additionally, this system limited the availability of long-term follow-up data because we did not have access to patient data beyond the hospitalization of that patient. As such, a future study evaluating long-term outcomes is needed in order to evaluate other possible clinical correlations in our population. Moreover, we recognize that the small sample size was an important limitation of this study, specifically, a retrospective study, which in turn severely limited the statistical analysis that could be performed and the strength of the associations. In addition, our study had several strengths, the primary being that it is the first study describing the clinical characteristics of Hispanic women in Puerto Rico with peripartum cardiomyopathy. Furthermore, given that the University District Hospital is the largest medical facility providing high-risk obstetrical services on the island and the only one that offers treatment for patients with PPCM, it is probably reasonable to conclude that our patient population represents the general population with PPCM on the island.

Resumen

Objetivos La Cardiomiopatía Periparto es una condición rara que causa fallo cardiaco en mujeres embarazadas. En Puerto Rico hay muy poca información relacionada a esta condición. En este reporte, describimos la experiencia con esta condición en nuestro centro de cuidado terciario. Métodos: Esto fue un estudio retrospectivo descriptivo. Se revisaron los expedientes médicos de las pacientes con Cardiomiopatía Periparto que fueron atendidas en nuestra institución entre enero 2006 y diciembre 2012. Doce pacientes fueron incluidas en nuestro análisis. Resultados: La edad promedio de nuestra población fue 27 ± 8 años. Ocho de las doce (67%) pacientes eran mujeres multíparas con un promedio de 2.6 ± 1.6 nacimientos vivos. La mayoría de las pacientes (75%) mostraron evidencia clínica de anemia durante el embarazo y más de la mitad (58%) tuvieron un parto prematuro. Seis pacientes (50%) tenían un historial previo de preeclampsia. Imágenes cardiacas detectaron una fracción de expulsión promedio de 35% ± 8 al momento del diagnóstico. Una muerte materna fue documentada. Conclusión: Nuestro cohorte de pacientes era similar a las demás series de casos de Cardiomiopatía Periparto que han sido reportadas en la literatura. Se necesitan estudios prospectivos para poder entender mejor la fisiopatología de la enfermedad y mejorar el tratamiento de estas pacientes.

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