Pak Heart J

CLINICAL SPECTRUM AND OUTCOME OF PATIENTS WITH PERI-PARTUM CARDIOMYOPATHY

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Date Received: June 28, 2019

Date Revised: Aug 15, 2019

Date Accepted: Sep 02, 2019

Contribution

FATS conceived the idea, designed the study and did the final review. AKS contributed in data collection and analysis. Both authors contributed equally to the submitted manuscript.

All authors declare no conflict of interest.

This article may be cited as: Sultan FAT, Shadmani AK. Clinical spectrum and outcome of patients with peri-partum cardiomyopathy. Pak Heart J 2019; 52 (04):302-6

ABSTRACT

Objective: To study the clinical spectrum and outcome of the patients with peri-partum cardiomyopathy (PPCM) at a tertiary care hospital of Pakistan.

Methodology: In this cross-sectional study, all consecutive patients with the diagnosis of Peripartum Cardiomyopathy, admitted at Agha Khan University Hospital Karachi, Pakistan, during the year 2008 to 2015 were included. Data was collected on a pre - designed questionnaire by reviewing the files. Follow up was done by reviewing files and by telephone calls, where required.

Results: A total of 32 patients were included in the study. Mean age was 27.4 ± 5.8 years. Only 12% were primi-gravida. Half of them (51.5%) had no associated Co-morbidity while 18.7% were diabetics and 28% were diagnosed to have pregnancy-induced hypertension. In 31.2% of patients symptoms were started during the last month of pregnancy while 68.8% had their symptom onset after the delivery. Main symptom was shortness of breath, present in almost all the patients, with the signs of heart failure in 96.9% at the time of presentation). About 25% patients had no ECG changes while 71.8% showed sinus tachycardia, 74.9% had severely reduced left ventricular systolic function on echocardiogram with enlarged left ventricle in 56.2%. In the mean follow up of 24 months 1 patient died because of heart failure with an overall mortality of 12.5%. Symptoms improved in 59.4% and worsened in 12.5%.

Conclusion: Peripartum Cardiomyopathy usually affects young and multigravida women. About one third of patients have their symptom onset in the last month of pregnancy while two thirds after the delivery. Peripartum Cardiomyopathy has a high maternal and fetal mortality, however more than half of the patients improve.

Key Words: Peripartum cardiomyopathy, Heart failure, Left ventricular systolic dysfunction.

INTRODUCTION

PPCM is a disease of unknown cause. It has been recognized as a serious complication of pregnancy since the 18th century. Ritchie, described heart failure in late pregnancy and the puerperium associated with a primary heart muscle disease.[1] Virchow and Porak found myocardial degeneration to be a cause of death in women dying in the puerperium.[2] In 1930s descriptions of women with heart failure in the post-partum period and autopsy findings of enlarged hearts with focal areas of necrosis, were came out in the United States.[3,4]

PPCM is now characterized by symptoms and signs of heart failure associated with decreased left ventricular systolic function, that develops either in the last month of pregnancy or in the first five months after delivery, in women without pre-existing symptoms, signs or history of heart disease.[5,6] It is established by exclusion of other causes of left ventricular dilatation and systolic dysfunction.

Peripartum cardiomyopathy occurs in one out of every 3000 to 15,000 pregnancies, with a higher incidence in Africa.[7-11] In spite of the higher incidence in Africans, all races can be affected.

Not much data is available from Pakistan, which has assessed the clinical spectrum and outcome of patients with PPCM. We evaluated the data of patients with PPCM, from a tertiary care center of Pakistan. The main aim of this study is to get better knowledge about the disease, which can help us in making the strategies to reduce the mortality associated with PPCM.

METHODOLOGY

In this cross-sectional study, all consecutive patients with the diagnosis of PPCM, admitted at AKUH Karachi Pakistan, during the year 2008-2015 were reviewed. Patients with development of cardiac failure in the last month of pregnancy or within 5 months after delivery, absence of a demonstrable cause for the cardiac failure, absence of demonstrable heart disease before the last month of pregnancy, and documented systolic dysfunction on echocardiogram were included. Data was collected by reviewing the files and by telephone calls, where required. SPSS version 18 was used for data analysis. Frequency and percentages ware calculated .

RESULTS

A total of 33 patients were found fulfilling the criteria for diagnosis of PPCM, however 32 patients were included in the study due to incomplete data in one patient . The baseline characteristics of the patients are shown in table 1.

Mean age was 27.4 ± 5.8 years. Majority of them were multi-gravida (87.5%) and multi-para (81.2%). Pregnancy induced hypertension (PIH) was the most common co-morbid present in 28% of the patients followed by diabetes mellitus in 18.7% while only 3% developed pre-eclampsia.

Majority (68.8%) had their symptom onset after the delivery. Main symptom was shortness of breath, present in almost all the patients, with the signs of heart failure present in 96.9% of patients at the time of presentation. ECG was done in all patients with a normal ECG in 25% of patients. About 71.8% had sinus tachycardia.

The echocardiographic features. Patients were divided into three categories on the basis of ejection fraction (EF) on echocardiogram are shown in table 2. Majority (74.9%) had severely reduced left ventricular systolic function, with enlarged left ventricle in 56.2% on echocardiogram.

The medications received and the outcome are shown in table 3. All of the patients received diuretics. Angiotensin converting enzyme inhibitors (ACEI) were used in 90.6%, digoxin in 81.2% and beta blockers (BB) in 71.9%.

Four patients died with an overall mortality of 12.5% in a mean follow up of 24 months. Follow up was not available in 4 patients. About 3 patients died early during their initial presentation while 1 died later. Follow up echocardiogram was not available in all the patients. Symptoms improved in 59.4% and worsened in 12.5%.

Table 1: Baseline Characteristics of the Patients with PPCM (n = 32)

Variables	Frequency (n)	Percentage (%)
Age ± SD	27.4 ± 5.8	------
Multi –gravid a	28	87.5%
Multi -para	26	81.2%
PIH	9	28%
Diabetic	6	18.7%
Pre-eclampsia	1	3.1%
SOB	32	100%
Orthopnea & PND	20	62.4%
Cough	17	53%
Palpitation	6	18.7%
Syncope	2	6.2%
Signs of heart failure	31	96.9%
Edema	27	84.2%
Raised JVP	17	53.1%
Crepitations	30	93.5%
S3 gallop	19	59.3%
Normal ECG	8	25%
Sinus tachycardia	23	71.8%
Atrial fibrillation	3	9.3%
PVCs	4	12.4%
Bundle branch block	3	9.3%
Cardiomegaly on X-ray	15	46.7%
Pulmonary edema	19	59.1%
Pleural effusion	17	53%

Table 2: Findings on Baseline Echocardiogram (n = 32)

Echo features	Frequency (n)	Percentages (%)
Severe LV dysfunction	24	74.9%
Moderate LV dysfunction	7	21.8%
Mild LV dysfunction	1	3.1%
Enlarged LV	18	56.2%
Enlarged LA	13	40.6%

Table 3: Medications and Outcome (n = 32)

Medications and Outcome	Frequency (n)	Percentages (%)
Diuretics	32	100%
ACEI	29	90.6%
Digoxin	26	81.2%
B Blockers	23	71.9%
Inotropes	2	6.2%
Death	4	12.5%

DISCUSSION

This is a small case series of an uncommon condition, which documented the clinical profile and outcome of 32 patients diagnosed as PPCM at a tertiary care hospital of Pakistan. These are relatively younger patients with a mean age of 27.4+5.8 years, which is similar to 29+6 years noted in a study of 55 patients, published in Am Heart J, in 2006.[12] However, earlier series suggested the syndrome was more prevalent in older women.[7,8,13-16] In the original description by Demakis, where he proposed clinical criteria for diagnosing the condition, he included 27 patients with the diagnosis of PPCM. Fourteen of these patients were older than 30 years, while 13 patients were younger than 30 years of age.[17] More recently, Fett et al. published a series of women from Haiti diagnosed with PPCM, where the mean age was 31.8 years (SD+8.1years).[18] Elkayam et al., in a retrospective multi-center study, with a survey questionnaire, found the mean age of PPCM patients to be 30.7 years (SD+6.4years).[19]

Majority of patients were multi-gravida and multi-para in this study, which is similar to the study by Demakis et al., in which 71% of women diagnosed with PPCM had three or more prior pregnancies, while 8% were primigravidas.[17] It has been repeatedly demonstrated that PPCM occurs with much higher incidence and prevalence in women with high parity and gravidity.[18-20]

Twin pregnancy appears to cause a higher risk of developing PPCM.[21-23] While the majority of women with PPCM have a single pregnancy, the prevalence of the syndrome amongst women with twin pregnancies is much higher.[24,25] In one study of 100 women with PPCM, 13% had twin pregnancies.[19] However, no patient in this study had twin pregnancy.

Pregnancy induced hypertension was the most common co-morbid found in 28% patients in this study, while only 3% developed pre-eclampsia. Hypertension and pre-eclampsia have been associated with a significant number of PPCM cases.[11,14,15,16,19,26] In the original description of the condition by Demakis in 1971, 22% of the women diagnosed with the condition had pregnancy related hypertension.[17] In a different study of women diagnosed with PPCM, 43% had hypertensive disorders during pregnancy.[18] Amos et al. published a series of 55 patients with PPCM, in which hypertension was present in 56% and pre-eclampsia/eclampsia in 46%.[12] Many authors even report PPCM as a variety of hypertensive heart failure.[9,18,27] However, others argue for a stricter definition of PPCM that excludes pregnancy related hypertensive disorders.[28]

Majority of the patients in this study had their symptom onset after the delivery. This is in keeping with the studies performed by others, and in contrast with a study by Sliwa et al, in which all patients had their symptom onset in the post-partum period [11,29,30].

Malnutrition, low socioeconomic status and poor antenatal care are also mentioned as risk factors in the earlier reports.[7,8,11,22,23] However, this was not the case in this study as all the patients belonged to a good socioeconomic class, with adequate antenatal care. In fact substantial correlations of these factors have not been found in further studies.[13,14]

Signs and symptoms of heart failure were the main presenting features in this series, similar to what is reported in the literature.[7,8,11,14-16,21,22] But no embolic complication was observed in contrast to the literature.[7,8,11,14,15,21-23,26]

Diagnosis of PPCM is based on excluding common causes of cardiac failure. Early diagnosis of PPCM may be difficult because many of the similarities of its presenting features with that of advanced pregnancy. Echocardiography and other laboratory evaluations strengthen the clinical diagnosis. Baseline transthoracic echocardiogram was done in all the patients in this series and majority of the patients (74.9%) had severely reduced left ventricular systolic function, while left ventricle was dilated in more than half of the patients. This indicates the severity of the disease at presentation.

Medical management of PPCM is similar to that of heart failure. Fluid and salt restriction, diuretics, digoxin, vasodilators and beta blockers are the mainstays of treatment.[11,14,15,21-23,26] All the patients in this study received diuretics while use of ACE inhibitors (90.6%) and digoxin (81.2%) was also quite high. However beta blockers were used in relatively less number of patients (71.9%). These figures are similar to a study by Amos et al., in which diuretics were used in 85%, ACEI in 90%, digoxin in 75% and beta blockers in 69%.[12] Although the etiology of PPCM is not known, many investigators believe that it may be related to myocarditis or an abnormal immune response to pregnancy.[5,31] This injury would lead to remodeling and subsequent left ventricular dilatation. Both ACEI and beta blockers have been shown to prevent remodeling and reduce left ventricular dimensions.

The reported prognosis of PPCM varies in the literature, but prognosis is currently encouraging with advanced management. Mortality rates of up to roughly 50% have been reported in the literature.[7,8,11,15,19,23,26] Mortality of 12.5% in this series is comparable to 15%, reported in a series of 100 patients from South Africa.[30] In another series of 55 patients, Amos et al. reported no deaths, but 10% of the patients required heart transplantation[12]. While comparing the mortality with other series we must consider the fact that cardiac transplantation or left ventricular assist device was not available for the population studied in this series.

Follow up echocardiogram was not available to compare with the base line, however symptom improvement was noted in 59.4%.

CONCLUSION

PPCM usually affects young and multi-gravida women. About one third of patients have their symptom onset in the last month of pregnancy while two thirds after the delivery. Shortness of breath is the most common symptom. It has a high mortality, however more than half of the patients improve with time.

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