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Case Series

Obstetrics and Gynaecology Section

Unusual Presentation of Peripartum Cardiomyopathy- A Case Series

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ABSTRACT

A rare condition called Peripartum Cardiomyopathy (PPCM) causes a pregnant woman's heart to weaken and expand. It occurs in the last month of pregnancy or within five months of delivery. The presented paper was a series of three cases (29-year-old women, 26-year-old women, 26-year-old women) with unusual presentations of PPCM. Cases 1 and 2 demonstrated unique PPCM presentations that included abrupt cardiogenic shock and failure symptoms and signs. The postpartum period's typical PPCM appearance is illustrated by case 3. The index patients (cases 1 and 2) had low systemic blood pressures, acute respiratory distress, and reduced cardiac output that was indicative of cardiogenic shock. An early echocardiogram was ordered as a result, and the results were suggestive of heart failure. Their varying clinical manifestations posed a significant diagnostic problem due to the heterogeneity. Even though they are uncommon, such catastrophic presentations including acute respiratory distress and low-output cardiac failure can happen. In these unusual cases, rapid pharmacological and mechanical support is required. In order to provide patients with the finest and most efficient care possible, it is crucial to understand the aetiology, clinical signs and symptoms, management, and prognosis of PPCM. Thus, physicians need to be familiar with different presentations of PPCM and always consider it with a high index of suspicion to expedite treatment for a potentially lethal condition to get a better outcome.

Keywords: Cardiac failure, Myocardiopathy, Pregnancy, Respiratory distress

INTRODUCTION

The PPCM is a rare, but life-threatening form of heart failure affecting women in the last month of pregnancy or the first five months postpartum without any previously recognisable heart disease. It is a kind of non ischaemic dilated cardiomyopathy related to pregnancy and diagnosis of exclusion following a concurrent evaluation of peripartum heart failure [1].

Because of the variety in their clinical manifestations, PPCM poses a significant diagnostic difficulty. This article also discusses the origin, clinical signs and symptoms, management, and prognosis of unusual presentation of PPCM in three patients, all of which are important to comprehend to give patients the best and most effective care possible.

CASE SERIES

Case 1

A 29-year-old women, G2P1L1, reported to the labour room at 37 weeks days of gestation with chief complaints of pain in the lower abdomen since one day, increasing in frequency. Previous LSCS was at term with no co-morbidities and uneventful past and present antenatal history, with no personal and family history of the cardiovascular disease. On examination she was found to be in early labour with doubtful scar integrity.

Emergency LSCS was done under spinal anaesthesia and the patient had atonic Postpartum Haemorrhage (PPH) immediately after surgery which was managed medically. Blood and blood products including 2 units packed cells and 1 unit FFP were transfused. During immediate postoperative period, in the recovery room, the patient developed abdominal discomfort and drowsiness with a pulse rate of 130/min (low volume), systolic BP of 60 mm Hg, and abdominal distension. Subsequently, the patient was intubated and resuscitative measures were taken. The patient was then shifted to ICU for elective postoperative ventilation and haemodynamic monitoring.

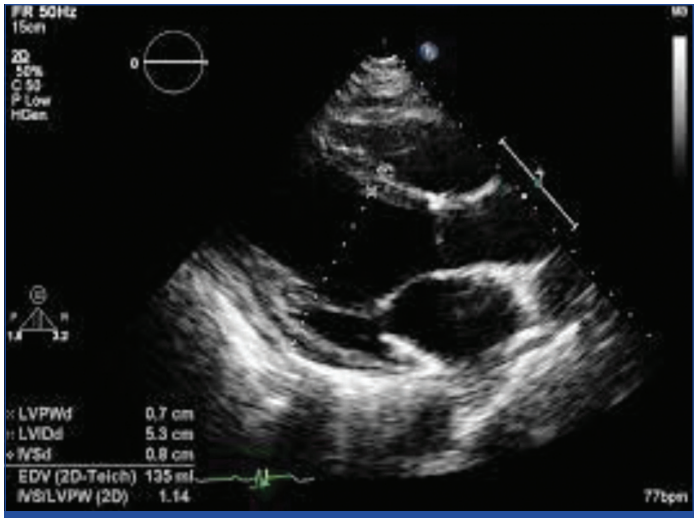
The patient again developed a second episode of hypotension with bradycardia after an hour, and was started on inotropic support. Echo showed ejection fraction of 55%, mildly dilated left atrium, normal left ventricular function, and tachycardia. Blood investigations were sent and cardiac markers were found to be raised [Table/Fig-1].

Test	Result
WBC count n/mm^3	29,000
Renal function test	Normal
Coagulation profile	Normal
Liver function test	Normal
Cardiac markers (Trop T, Trop I, CK MB)	Raised
Blood culture	No growth
Urine culture	No growth
High vaginal swab culture	No growth

[Table/Fig-1]: Blood investigation.

On postoperative day 1, a repeat echo was done, which showed an ejection fraction of 20%, biventricular dysfunction, and global hypokinesia suggestive of cardiomyopathy [Table/Fig-2].

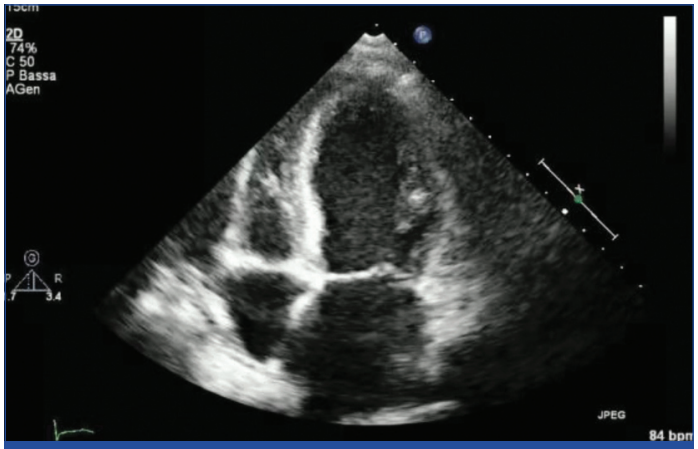
The patient was then treated with T.Digoxin 0.25 mg/day, 2 doses of Inj.Amiodarone 250 mg infusion, T.Nattokinase 2000 f.u/day, T.Ecospirin 150 mg/day, T.Ivabradine 5 mg/day, T.Trimetazidine MR 35 mg/day, Inj.Unfractionated heparin 5000 IU/day, Inj.lasix 20 mg/day, dopamine receptor agonist bromocriptine 2.5 mg/day, inotropic support with Noradrenaline DS infusion @10 mL/hour, and ventilatory support. Bacterial growth in the blood culture was present (checked on 6th postoperative day) (Initially, the patient was on Inj.Piperacillin-tazobactam and Metronidazole) and higher-order antibiotics, including Inj.Meropenem 2 gm/day in 2 divided doses,



[Table/Fig-2]: ECHO (Case 1 showing Global hypokinesia).

Inj.Clindamycin 1800 mg/day in three divided doses were started to combat the septicaemia, which the patient acquired during the due course of her stay at the ICU. The patient developed multiple ailments like gluteal bed sore, gangrene left thumb, and critical care polyradiculopathy in due course of her stay which was managed by involving a dedicated multidisciplinary specialty team.

The patient continued to be on ventilatory support for one month, and was finally weaned from oxygen support after a month. She was also under intense physiotherapy and rehabilitation training for critical care neuropathy. Serial ECHO studies were done and the ejection fraction started improving from the second week of illness and she was finally discharged on postoperative day 52. A six-month follow-up exam revealed a stable heart condition and routine echocardiography with a 65% ejection fraction [Table/Fig-3].



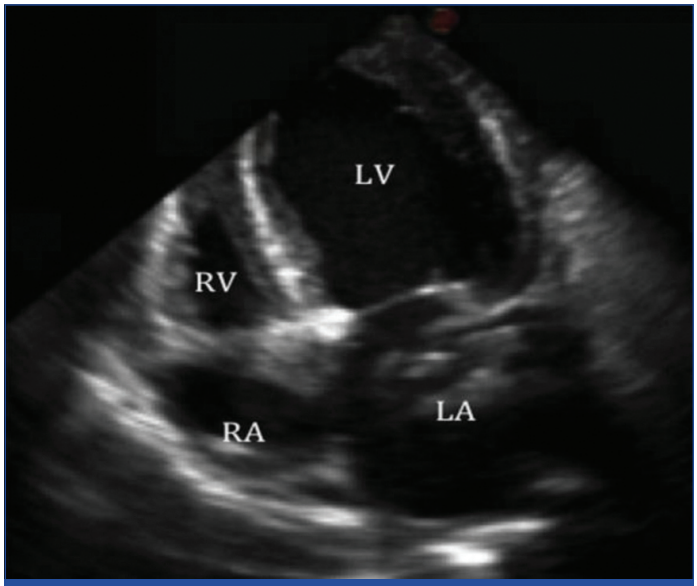
[Table/Fig-3]: A six-month follow-up exam revealed a stable heart condition.

Case 2

A 26-year-old women G2P1L1, reported to labour room with chief complaints of continuous high-grade fever for the past one day and was admitted in view of acute febrile illness. Previous LSCS was at 36 weeks of gestation. She was known to have subclinical hypothyroidism and was taking 75 mcg thyroxine, and oral iron therapy. She had an anterior wall fibroid which was diagnosed incidentally during a routine antenatal scan.

After admission, she got labour pains and was taken up for emergency repeat LSCS under spinal anaesthesia in view of doubtful scar integrity. It was a transverse lie with dorso inferior and the baby was delivered by breech. A sessile 7×5×4 cm subserosal fibroid was present at the upper flap of the uterine incision near the right angle necessitating myomectomy for the uterine incision closure. Uterine atonicity was noted and attempted medical management, but suddenly the patient developed hypotension and desaturation. Hence, she was intubated, converted to general anaesthesia, and proceeded to surgical management including step-wise devascularisation and B-Lynch compression sutures.

The patient was then shifted to ICU postoperatively for haemodynamic stabilisation. In the ICU, she again had the second episode of hypotension needing inotropic and ventilatory support with no evidence of vaginal or intraperitoneal bleeding. A Cardiologist's opinion was sought and bedside ECHO showed global hypokinesia, moderate to severe LV dysfunction with an ejection fraction of 30% [Table/Fig-4]. Trop T was positive with elevated CK MB (45 IU/L) and CK total (170 IU/L) enzymes suggestive of PPCM.



[Table/Fig-4]: ECHO showing global hypokinesia and moderate to severe LV dysfunction condition.

The patient was then started on antifailure drugs, Bromocriptine 2.5 mg/day, Digoxin 0.25 mg/day, and Nattokinase 2000 f.u/day. Ventilatory and inotropic support was continued. On the first two postoperative days, the patient was found to have deranged LFT, RFT, and coagulation profile, blood products were transfused accordingly [Table/Fig-5].

Tests		Values
LFT	Bilirubin (T)	2.3 mg/dL (Raised)
	Bilirubin (Indirect)	1.5 mg/dL (Raised)
	Bilirubin (Direct)	0.8 mg/dL (Raised)
	ALT	129 U/L (Raised)
	AST	150 U/L (Raised)
	ALP	315 U/L (Raised)
	GGT	50 U/L (Normal)
	LDH	150 U/L (Normal)
RFT	Blood urea	52 mg/dL (Raised)
	Serum creatinine	1.6 mg/dL (Raised)
Coagulation parameters	Platelet	90,000 (Reduced)
	BT	3'10'' (Normal)
	CT	6'10'' (Normal)
	PT/INR	16/1 (Normal)
	APTT	42 (Raised)

[Table/Fig-5]: Biochemical estimation.

Later from the 3rd postoperative day, the patient gradually improved clinically and by ECHO findings. She was extubated and completely taken-off from inotropic support on postoperative day 6. Serial ECHO studies were done and ejection fraction improved to 50% and discharged on postoperative day 12. Follow-up examination at three and six months showed a stable cardiac status and normal echocardiogram with ejection fraction of 60%.

Case 3

A 26-year-old women primigravida at 38 weeks of gestation reported to the labour room in early labour, progressed spontaneously. She was a known case of gestational diabetes mellitus, under medical nutrition therapy. She was delivered by emergency caesarean section under general anaesthesia for foetal distress. The patient had major atonic PPH, managed with Intramuscular Inj.Methergine 0.2 mg, Intramuscular Inj.Prostodin 250 mcg, Oxytocin 15 IU infusion, Rectal misoprostol 800 mcg, and surgically by bilateral uterine artery ligation and Hayman compression sutures. Blood and blood products were transfused. She was then shifted to ICU for postoperative ventilation and monitoring. The echocardiogram showed a normal ejection fraction of 62% and the patient was extubated the next day.

The patient then developed skin necrosis over the wound site for which wound debridement and secondary suturing was done on 5th day of surgery. She then had a normal recovery and was discharged on the 11th day of surgery.

On the 16th day of surgery, the patient then presented with breathlessness at rest, sweating, and shoulder pain to the emergency department. On examination, there was tachycardia and tachypnea with normal blood pressure and oxygen saturation. ECG showed sinus tachycardia and echocardiogram showed global hypokinesia of the left ventricle, moderate left ventricular systolic dysfunction with an ejection fraction of 43%, dilated left ventricle, grade 2 mitral regurgitation, and grade 3 diastolic dysfunction. She was then treated with T. Digoxin 0.25 mg/day, T.Ivabradine 5 mg/day, and Bromocriptine 2.5 mg/day and recovered completely after three months with a normal ejection fraction of 60%. On follow-up after three months, she had stable cardiac status with normal ejection fraction and mild mitral regurgitation.

DISCUSSION

Incidence and aetiology: A rare and potentially fatal syndrome known as PPCM is linked to high rates of maternal and newborn morbidity and mortality [1,2]. Cases 1 and 2 demonstrate unique PPCM presentations that included abrupt cardiogenic shock and failure symptoms and signs. A similar presentation was stated by Abdulraheem E et al., in which severe PPCM was complicated by COVID-19 infection, where the patient required emergent intubation, sedation, and mechanical ventilation for acute hypoxemic respiratory failure attributed to cardiogenic shock [1]. When there is no other heart failure causes, it is defined by a decline in left ventricular systolic performance at the end of pregnancy or in the postpartum period [3-5].

Its true incidence is not known, ranging from 1 in 2,400 people to 15,000 people in western world [1,3]. There are regional differences, with larger incidences observed in parts of Africa [4]. Different population demographics, the strictness of the definition, and underreporting brought on by ignorance or misinterpretation are likely to be the causes of the incidence's wide variation [5].

Aetiology is still unknown. It is postulated that it may be due to nutritional deficiencies, small vessel coronary artery abnormality, hormonal effects, toxaemia, maternal immunological response to foetal antigen or myocarditis. Recurrence is common in subsequent pregnancies, and as a familial foundation for the condition has been shown [6], family planning may be impacted. Although the aetiology of PPCM remains unclear, a number of potential risk factors have been proposed.

Risk factors and clinical presentation: Horne D et al., study shows that the risk factors are multiparity, advanced maternal age, multifoetal gestation, preeclampsia, African descent, maternal cocaine abuse, selenium deficiency, long-term (more than four weeks) oral tocolytic therapy with beta adrenergic agonists which was in par with the present case series [7]. Case 2 patient was presented with subclinical hypothyroidism and case 3 with gestational diabetes Mellitus. Despite these risk factors, the illness can still affect patients, and this is the subject of future research that could lead to the development of novel therapeutic strategies. Congestive heart failure symptoms and signs, as well as chest pain, are clinical manifestations of PPCM. However, these symptoms and indications also appear in a wide range of different illnesses, from healthy pregnancy to pulmonary emboli and upper respiratory infections. In fact, this frequently results in missing or delayed diagnoses of PPCM and underestimating the condition's prevalence [8].

Physical examination: Tachycardia, increased jugular venous pressure, pulmonary rales, and peripheral oedema are among the symptoms of heart failure that are frequently discovered during a physical examination. Chest radiography frequently reveals heart enlargement and pulmonary venous congestion, while an ECG normally exhibits sinus tachycardia with no discernible alterations [8]. The index patients (Case 1 and Case 2) did not have any of these manifestations but had low systemic blood pressures, severe respiratory distress, and low cardiac output suggestive of cardiogenic shock. This prompted an early echocardiography which was suggestive of cardiac failure. Such catastrophic presentations though unusual can occur, with severe respiratory distress and low-output cardiac failure, necessitating immediate pharmacological and mechanical support [4]. Hence, the differential diagnosis includes other pulmonary causes like acute pulmonary oedema, and cardiac causes including myocardial infarction or Takotsubo cardiomyopathy. Differential diagnosis in the peripartum period is briefed in [Table/Fig-6].

Diagnosis: Diagnosis of PPCM includes four criteria:

1. Development of cardiac failure in the last month of pregnancy or within five months after delivery;
2. Absence of an identifiable cause for cardiac failure;
3. Absence of recognisable heart disease prior to the last month of pregnancy;
4. Left ventricular systolic dysfunction demonstrated by classic echo cardio graphic criteria such as depressed shortening fraction or ejection fraction [9,10].

Variables	PPCM	Pre-existing CMP, valve disease or congenital heart disease	Pregnancy associated myocardial infarction	Pulmonary embolism/amniotic fluid embolism	Takotsubo (Stress induced cardiomyopathy)
History/ Presentation/ onset	Most commonly postpartum onset of dyspnoea	Earlier onset (during second trimester) sometimes family history	Retrosternal chest pain, abdominal discomfort, nausea	Pleuritic chest pain Infection	Dyspnea following stress of varying severity
ECHO	Left and/or right ventricular dysfunction	Evidence of pre-existing valve disease or congenital defect	Regional hypokinesis/ akinesis	RV dysfunction, elevated RV pressure, McConnell's sign	Transient hypokinesis, akinesis or dyskinesis of the LV mid-segments with or without apical involvement.
Biomarkers	Elevated natriuretic peptides	Elevated natriuretic peptides	Elevated Troponin	Elevated D-Dimer, Troponin and natriuretic peptides	Elevated Troponin possibly. Elevated natriuretic peptides
Other tests	MRI	MRI and genetic tests	Coronary angiography	CT scan or V/Q scintigraphy, Coronary angiography	MRI

[Table/Fig-6]: Differential diagnosis of acute dyspnea and collapse in peripartum period.

Management: The precise management approach for PPCM relies on each unique clinical instance however it primarily depends on the patient's stability. As in first two cases, acute heart failure during pregnancy is treated in the same manner as acute heart failure at any other stage of life [9]. This comprises beta blockers, nitrates, diuretics, inotropic support, and ventilatory support. Mechanical circulatory support, such as a Ventricular Assist Device (VAD), should be considered for individuals whose conditions worsen despite receiving the best medical care. As in third case, the management of patients with stable heart failure in PPCM comprises antifailure medications while avoiding ACE inhibitors and ARB during the prenatal period due to their toxicity [9]. In PPCM, bromocriptine is recommended in acute and stable cardiac failure cases.

Follow-up and prognosis: It is unknown when to stop taking these meds, but atleast a year should pass before stopping. The last resort is frequently heart transplantation, if medical treatments are unsuccessful. Appreciatively, the rate that necessitated transplantation has dropped between 4% and 7% in recent years [11]. Positive long-term survival rates and reasonable transplant success rates are present [12].

A normalisation of the ejection fraction occurs in roughly 50% of patients. Regardless of recovery, a second pregnancy is typically not advised for these patients due to the high danger for both mother and baby. PPCM recurs in more than 30% of subsequent pregnancies [13].

CONCLUSION(S)

The PPCM is a very uncommon condition that can have fatal effects. Early detection and efficient treatment can raise the likelihood of full recovery with a healthy heart, as was the case in our instances, while also lowering mortality and morbidity rates. Cases 1 and 2 illustrate unusual presentations of PPCM with symptoms and signs of acute cardiogenic shock and failure. Case 3 illustrates the typical presentation of PPCM in the postpartum period. Thus, physicians need to be familiar with different presentations of PPCM and always consider it with a high index of suspicion to expedite treatment for a potentially lethal condition for a better outcome.

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