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Case Report

A Case Report of Latent Peri-Partum Cardiomyopathy, Obscured by Hypertension

Adeel Riaz[1], Muhammad Mubbashir Sheikh[2], Hafiz Muhammad Umair[1], Muhammad Shah Nawaz ul Rehman[4], Iram Liaqat[3] and Muhammad Mubin[4*]

[1]*Department of Anesthesia DHQ Teaching Hospital Sahiwal, Pakistan*
[2]*School of Medicine, Northwestern University, Feinberg Chicago, USA*
[3]*Microbiology Laboratory, Department of Zoology, GC University, Lahore, Pakistan*
[4]*Center of Agricultural Biochemistry and Biotechnology, University of Agriculture, Faisalabad, Pakistan*

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ABSTRACT

Peripartum Cardiomyopathy (PPCM) is a pregnancy-associated cardiac disorder, which is potentially life-threatening and manifests as left ventricular dysfunction and heart failure. The disease is rather rare and in most patients cardiac function recovers well, but long-term morbidity and mortality are not uncommon. Research studies suggest pregnancy-induced hormones and mediators, which cause vascular dysfunction, trigger that peri-partum cardiomyopathy. Genetic factors are also thought to play a role in pathophysiology. Management of peri-partum cardiomyopathy is same as cardiac failure and drugs against mediators like prolactin are under investigations, but no proven disease-specific therapies had been reported. We report a case of 35-year-old female who, instead of heart failure, had hypertension as sole presenting complaint of PPCM. Complications associated with PPCM are severe progressive heart failure, arrhythmias, heart block, cardiopulmonary block and thromboembolism. Death could be caused by worsening heart failure, arrhythmias and cardiopulmonary block.

Introduction

Peri-partum Cardiomyopathy (PPCM) is an uncommon but serious cardiac disorder of unknown etiology that typically occurs in the peri-partum period, characterized by left ventricular systolic impairment and presenting with symptoms of heart failure [1]. Diagnosis is based on the following criteria, 1) heart failure towards the end of pregnancy or early postpartum period, 2) left ventricular systolic dysfunction based on echocardiography showing left ventricular ejection fraction (LVEF) <50%, fractional shortening <30% or both, 3) no previous history of heart disease prior to pregnancy, 4) no known etiology [2, 3]. Prevalence is reported to be 1 in 400 live births in Haiti, 1 in 1000 live births in South Africa, 1 in 3000-4000 live births in United States and 1 in 20000 live births in Japan [4, 5]. Risk factors associated with PPCM are hypertensive disorders of pregnancy (HDP), African American race, advanced maternal age, multiple pregnancy, obesity, smoking and genetics [2, 6, 7]. Exact pathophysiology of PPCM is not fully understood but many theories have described association of PPCM with viral myocarditis, autoimmune origin, endothelial and cardio toxic factors like soluble FLT1 (sFLT1), and cleaved prolactin [8].

Case Presentation

A 35-year-old female with history of Cesarean section 5 days back presented to the ER with signs and symptoms of intestinal perforation. Vitals were, BP= 159/98, Pulse=93/min, Temp= 98.9F, RR=20/min. She was investigated accordingly and emergency exploratory laparotomy was planned. IV lines secured and she was resuscitated with fluids and antibiotics. Patient was also catheterized and NG tube was passed for gastric decompression. Physical examination showed absent bowel sounds and did not reveal any abnormal breathing or heart sounds in chest. Patient's history revealed that pregnancy was uncomplicated during the preterm, labor/cesarean section and post-surgery period. There was no history of primary hypertension, pregnancy induced

[*]*Correspondence to: Dr M. Mubin, Centre of Agricultural Biochemistry and Biotechnology, University of Agriculture, Jail Road 38000, Faisalabad, Pakistan; Tel: 04192001612925, 923006615706; E-mail: mmubin@uaf.edu.pk*

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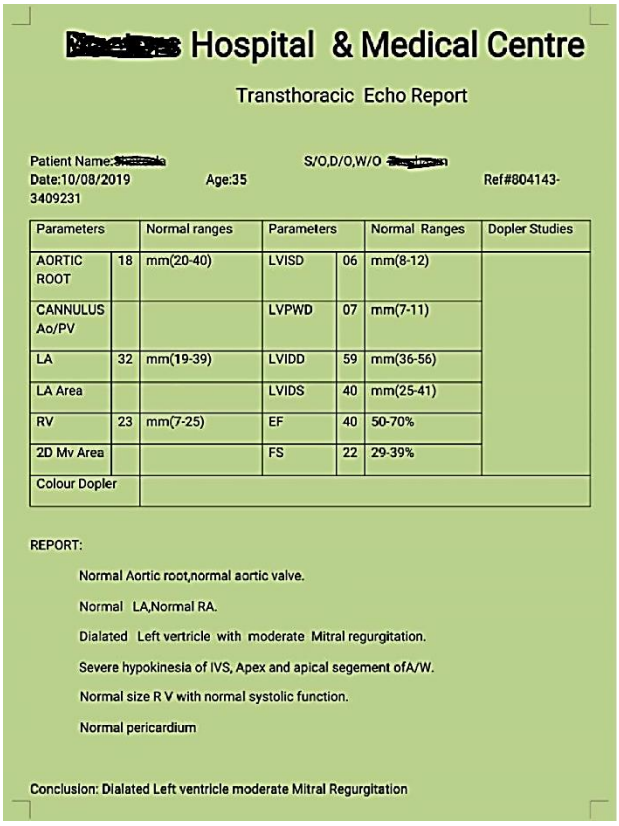
hypertension, pre-eclampsia, eclampsia, diabetes mellitus or any cardiac issues. She had no proteinuria or edema during antenatal visits.

Patient was shifted to operation theatre and given general anaesthesia as planned, cardiac monitor with EKG leads attached for continuous vital monitoring. Surgery was started and abdomen was opened when suddenly the blood pressure of patient started to fall rapidly. So much so that patient became blood pressure-less/pulseless and oxygen saturation fell below 20%. Urgent resuscitation efforts started with chest compressions and IV Adrenaline. Patient responded to resuscitation efforts and was put on cardiac support with continuous slow IV adrenaline infusion. Meanwhile the patient's chest revealed generalized crackles. Fluid was restricted, Furosemide given for diuresis and patient responded well. Surgery was continued as blood pressure stabilized and at the end of surgery patient was off cardiac support. After surgery the patient was in a very stable condition with blood pressure returning close to baseline readings of 153/96. Patient was shifted to ICU for nursing care with clear instructions of fluid restriction.



[redacted] Hospital & Medical Centre

Transthoracic Echo Report

Patient Name:[redacted] S/O,D/O,W/O [redacted]
Date:10/08/2019 Age:35 Ref#804143-3409231

Parameters		Normal ranges	Parameters		Normal Ranges	Dopler Studies
AORTIC ROOT	18	mm(20-40)	LVISD	06	mm(8-12)	
CANNULUS Ao/PV			LVPWD	07	mm(7-11)	
LA	32	mm(19-39)	LVIDD	59	mm(36-56)	
LA Area			LVIDS	40	mm(25-41)	
RV	23	mm(7-25)	EF	40	50-70%	
2D Mv Area			FS	22	29-39%	
Colour Dopler						

REPORT:

Normal Aortic root,normal aortic valve.

Normal LA,Normal RA.

Dialated Left vertricle with moderate Mitral regurgitation.

Severe hypokinesia of IVS, Apex and apical segement ofA/W.

Normal size R V with normal systolic function.

Normal pericardium

Conclusion: Dialated Left ventricle moderate Mitral Regurgitation

Figure 1: Figure shows the findings of Cardiac Echocardiogram, showing reduced ejection fraction of 40%, fractional shortening 22%, mitral regurgitation, left ventricular dilation, hypokinesia of interventricular septum, apex and apical segments of A/W.

Next morning one of the nurses by mistake started fluid infusion and the patient once again went to cardiac depression and shock. But this time patient was managed robustly by stopping the infusion, injecting adrenaline and furosemide. As soon the patient was stable, Cardiac Echocardiogram was performed that revealed reduced ejection fraction of 40%, fractional shortening 22%, mitral regurgitation, left ventricular dilation, hypokinesia of interventricular septum, apex and apical segments of A/W. Figure 1 shows the echocardiographic findings. Other labs for differential diagnosis including cardiac enzymes were normal. EKG showed normal sinus rhythm. A diagnosis of peri-partum cardiomyopathy with a typical presentation of hypertension was made based on the above-mentioned criteria. Cardiology facility was involved and patient was managed conservatively with fluid restriction, diuretics and continuous vital monitoring. On fifth day patient was stable, oral free, ambulatory, and going to toilet without any difficulty. She was shifted back to parent ward and later on discharged home with follow up with cardiology in 1 week. Patient was followed for 6 months on for heart failure medications and her blood pressure normalized in the range of 118-133/75-92. Ejection fraction was 55%-60% on follow up echocardiography. She is in good health without any sequel.

Discussion

Historically risk factors for PPCM occur mostly in old black women but this trend is changing towards young women, favored by this case report, as the patient is young Asian female. Other risk factors are hypertensive disorders of pregnancy like eclampsia; multi-fetal pregnancy, obesity etc. The unique feature in this patient was that she did not have any presenting features like dyspnea or symptoms of heart failure before she was given intravenous fluid. The only notable finding was hypertension at presentation, which was thought to be due to stress of peritonitis. This hypertension also confounded the diagnosis of PPCM during surgery in the first place. But it was later described that hypertension was related to PPCM and it was the only presenting feature. This is a unique finding never described before for PPCM. Although hypertensive disorders of pregnancy are strongly associated with PPCM, this patient did not have any history of hypertension before the presentation for intestinal obstruction, no history of hypertension in antenatal period as well [9]. Hence a diagnosis of PPCM with atypical presentation of hypertension was made after further findings of echocardiography and diagnostic criteria described above.

Many theories have been proposed to describe the pathophysiology of PPCM postulating that it might be caused cardiovascular stress of pregnancy due to fluid retention, others suggest inflammatory response to pregnancy, yet others say autoimmune response to fetal cells and other mediators that lodge in the myocardium. Histologic findings of cardiac biopsy also show inflammatory process in many patients with PPCM. Diagnosis is made on the criteria mentioned above. Echocardiography shows decrease in ejection fraction, hypokinesia. Chest X-ray shows pulmonary congestion, increased cardiac size and sometimes pulmonary effusions. ECG is typically non-revealing. Other investigations including BNP are neither helpful nor necessary. But cardiac enzymes and pre-eclampsia must be ruled out. Treatment is the same as that of heart failure including diuretics, beta-blockers, digoxin and lifestyle modifications of fluid and salt restriction [10, 11]. ACE inhibitors and angiotensin receptors blockers can be used post-partum but contraindicated in pregnancy [12]. Due to the implication of prolactin components in the pathogenesis of the disease, prolactin antagonist Bromocriptine is being investigated as a treatment with favorable results [13, 14]. Medical treatment is continued for at least one year and then can be adjusted based on the symptoms. Failure of medical treatment warrants heart transplant as last option [15].

Conclusion

Majority of PPCM cases present in patients with normal blood pressures but hypertensive disorders of pregnancy are most important independent

risk factors that may aggravate the heart failure associated with this disease. PPCM presents with slowly progressive dyspnea, lower extremity edema and features of worsening heart failure. It may present with hypertension only, and heart failure may ensue after external stress, like intravenous fluids, is applied to already compensated cardiac function, triggering sudden collapse of the patient.

Conflicts of Interest

None.

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