Case report

Spontaneous Relapse of Peripartum Cardiomyopathy Recovered In Two Days ; Case Report

Abstract:

Peripartum cardiomyopathy (PPCM)is an idiopathic heart failure occurring without any determinable heart disease during the last month of pregnancy orthe first six months postpartum. PPCM may be underappreciated, as many patients stop follow-up as soon as they achieve a normal EF.We describe a case of PPCM relapse eight years after pregnancy in a 36-year-old female while experiencing stressful circumstances, in the absence of pregnancy and other causes of cardiomyopathy, yet to recover two days later. It is unclear when a patient with PPCM may be considered fully recoveredand heart failure medications safely discontinued. Restoration of ejection fraction (EF) may not represent true recovery.There are no PPCM guidelines, and limited, conflicting data existregardingthelong-term management of PPCM patientswith recovered LV function.

Comment [A1]: What is the contribution to the medical literature?

Comment [A2]: Main lessons of the investigated case?

Keywords;Peripartum cardiomyopathy, PPCM relapse,

Introduction:

Peripartum cardiomyopathy (PPCM)is a form of heart failure with reduced ejection fraction affecting mainly young women at the end of the pregnancy or first months after delivery[(1)]. Women of African American origin, multiple gestations, hypertension, pre-eclampsia, and advanced maternal age at pregnancy are considered to have an increased risk of

developing Peripartum cardiomyopathy[2]. The incidence of cardiomyopathy varies worldwide; in the US, it is estimated to be 4000 live births affecting women, while in Nigeria, the incidence is higher, and it is estimated to be 1 in 100 live births every year[3][4].The etiology of PPCM is likely to be multifactorial. Suggested mechanisms for developing PPCM have included nutritional deficiencies, viral myocarditis, and autoimmune processes. Hemodynamic stress of pregnancy has been postulated as a potential etiology[1][5].

Furthermore, recent data show that peripartum oxidative stress linked to proteolytic cleavage of prolactin into a 16 kDa subforums with potent anti-angiogenic and pro-apoptotic properties may explain the heart's microvascular damage as the onset of the myocardial disease in PPCM [6]. PPCM usually presents with congestive symptoms (Exertional Shortness of breath, Paroxysmal nocturnal dyspnea, orthopnea, dry cough, or edema) or nonspecific symptoms such as malaise and fatigue. The diagnosis of Peripartum cardiomyopathy is often challenging and requires a high degree of suspicion as symptoms are like the usual physiological symptoms encountered during normal pregnancy[3][4].Upon physical examination, there is often jugular venous distention, displaced apical impulse, and pansystolic murmur consistent with functional mitral regurgitation. Yet, diagnostic criteria must include LVEF <45%, onset of HF at or after the end of pregnancy,and the absence of any other cause of HF[3][4]. Symptomatic patients should undergo an echocardiogram as soon as possible, and appropriate medical care shouldbe initiated promptly, maintained, and followed up, as patients who appear to have cured may have concealed subclinical ventricle dysfunction [1]

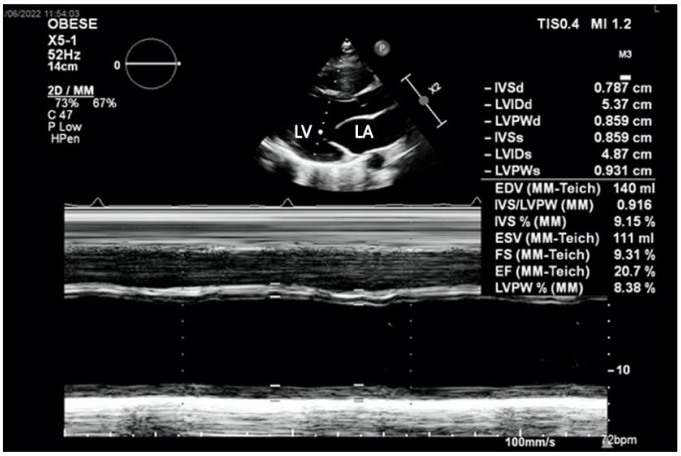
Case report:

A 36-year-old female, Gravida 2, Para2,not known to have any medical illness, was brought to the emergency room with the complaint of repeated vomiting and dizziness for one day, attributed to sudden onset of benignparoxysmal positional vertigo (BPPV).

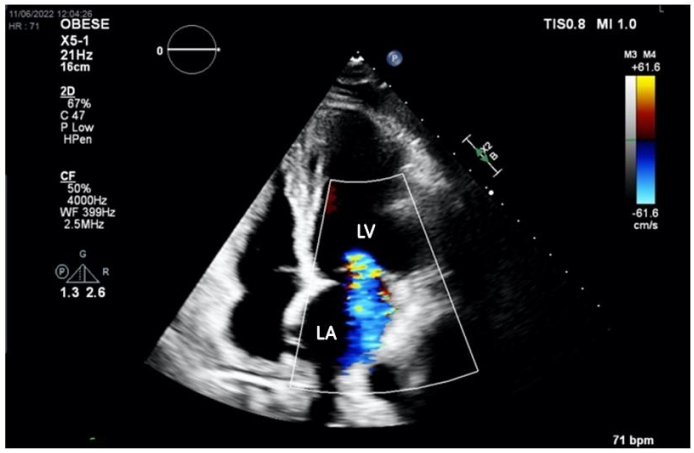
Comment [A3]: Add ethnic origin, family history, genetic information and aspects that contribute to explaining the case

Upon a detailed interview with the patient, she reported having exertional shortness of breath over the last two weeks while going through a stressful period in her life; she denies symptoms offever, cough, chest pain, palpitation, or lower limb swelling.The patient indicated that in her first pregnancy with twins eight years ago, she was told to have pericardial effusionand pregnancy-related cardiomyopathy and was instructed not to conceive in the future.She lost follow-up and conceived again three years later with no complications.Her vital signs upon admission wereblood pressure(BP)100/60 mmHg, heart rate 92 beats perminute, respiratory rate 18 per minute, oxygen saturationwas 100% on ambient air, and complete clinical examination was unremarkable.Chemistry and total blood countwere within the standard limit, while cardiac laboratory profileTroponin I 1667.9ng/L and BNP 159.8 pg/ml, in addition to a normal connective tissue disease(CTD) and viral serology work-up. Electrocardiogram(ECG) was normal, while an echocardiogram (Figure 1A) revealed mildly dilated Left ventricle (LV), severe systolic function impairment, and an estimated Ejection fraction (EF) of 25-30%, severe hypokinesia of basal to mid-LV, and Severe eccentric Mitral regurgitation (MR) with tethered posterior leaflet and a tented anterior leaflet(Figure 1B).She was started on Guide line-directed medical therapy GDMT for HF-rEF as tolerated by BP.Two days later, a repeat Echocardiogram (Figure 2A)showed marked improvement in Left ventricle function and size, near-normalEF of 45-50 % with subtle basal anterolateral wall hypokinesia,dramatic improvement in the degree of MV Regurge(Figure 2B) and MV morphology, with minimal residual eccentric MR.The decision to repeat the echocardiogram was made after the presenting symptoms of dizziness and vomiting ceased.A stress echocardiogram was scheduledtwo weeks after normalizing cardiac troponin to assess for ischemic heart disease and contractile reserve. Acardiac MRI was planned for evaluation for residual myocarditis. However, the patient failed topresent and was unable to follow up.

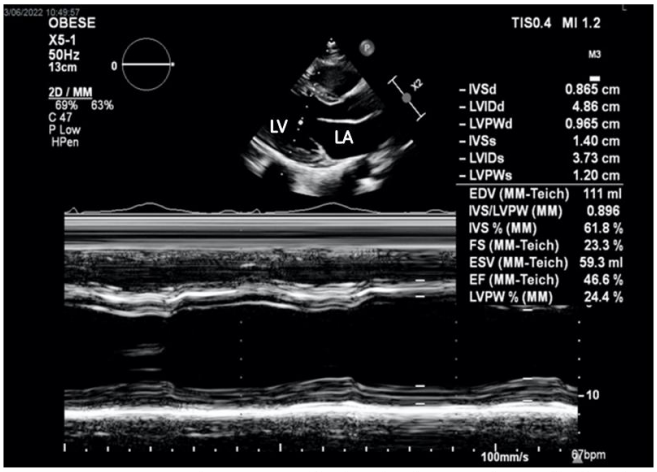
Comment [A4]: Explain the diagnostic reasoning, according to the results obtained from the interventions carried out



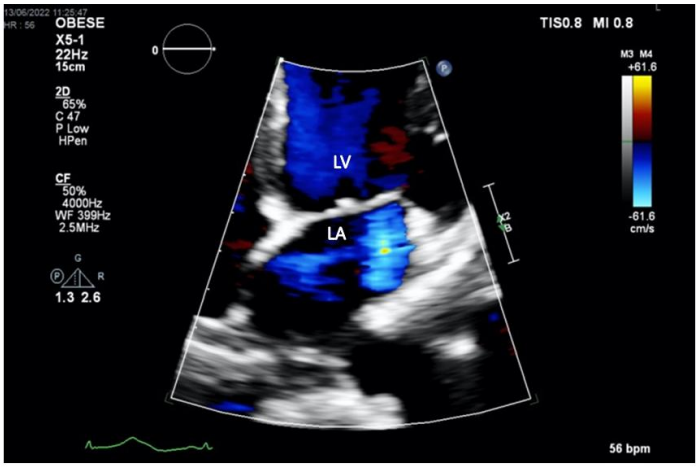
(Figure 1A): Echocardiogram showing mildly dilated LV, severe systolic function impairment..



(Figure 1B): Apical 4C view-color doppler on Mitral valve showing severe eccentric Mitral regurgitation.



(Figure 2A): PLAX view - M-mode showing improved systolic function, and dimension EF ~50 %.



(Figure 2B): Apical 4C View-color doppler of Mitral valve showing improvement in the degree of MR with minimal residual eccentric MR.

Discussion:

Peripartum cardiomyopathy is a type of heart failure that onset during pregnancy or early postpartum[3]. The leading cause behind PPCM is challenging to demonstrate. Still, there are potential causes, including genetic factors myocarditis, pathological immune response to pregnancy, pathological response to hemodynamic changes of pregnancy, hormonal abnormalities, angiogenic imbalance, stress-activated cytokines, and nutritional deficiencies [1][3].Physical examination often reveals jugular venous distention, displaced apical impulse, presence of S3, pansystolic murmur consistent with functional mitral regurgitation, pulmonary rales, or peripheral edema [3][4]. The key to diagnosing PPCM is the markdown of myocardial function detected in Echocardiography LVEF <45%, and there may or may not be ventricular dilatation [1][3]. An echocardiogram not only detects decreasing LVEF but also evaluates LV and right ventricular dilatation, functional mitral and/or tricuspid regurgitation, pulmonary hypertension, biatrial enlargement, and Intracardiac thrombus [1][3]. However, Cardiac MRI using T2-weighted spin-echo sequences enables the precise diagnosis of myocarditis, necrosis, and LV thrombi and yields accurate ventricular volume measurements[7][8]. Although echocardiography provides a gross measure of function, it has a limited ability to detect subtle changes, and some variability in measurements is expected. There are three reasons for the mitral regurgitation: 1) Leaflet abnormalities caused by a variety of mechanisms, including myxomatous degeneration and endocarditis, 2) Papillary muscle and chordate tendinae dysfunction and rupture, and 3) Functional regurgitation secondary to LV annular dilation due to LV dilation or ischemic- related papillary muscle displacement [9]. Dilated left ventricle or left atrial chamber causes leaflet structural abnormalities, which will cause functional mitral regurgitation[10].Our case did not reveal any evidence of leaflet, papillary muscle, or chordae tendinae abnormality. A trans-thoracic echocardiogram revealed depressed left ventricle function, left ventricle dilation, and severe mitral regurgitation without leaflet abnormality. Two days later, the echocardiogram was repeated, showing improved left ventricle function and mild mitral regurgitation. It is believed to be functional mitral regurgitation because of valve tethering coupled with annular and left ventricle dilation. This distinction case improvement in left ventricle function, dilatation, and mitral regurgitation further exemplified the recovery.

An incomplete myocardial recovery condition considerably impacts the recurrence of PPCM[3]. This case illustrated a sudden cardiac function deterioration after PPCM recovery; in our case, she had negative viral and CTD work-up, which was probably triggered by stress two weeks earlier and revealed by an acute presentation of BPPV associated with vomiting.PPCM patients who appear to have cured may have concealed subclinical ventricle dysfunction [1]. Consequently, position them for future risk of relapse [1].

Additionally, Women with PPCM have the chance of relapse after recovery during subsequent or without pregnancy [11][12].In about 20% of cases, subsequent pregnancies showed a risk of recurrence.Therefore women are advised to avoid pregnancy in the future [13][14][15].Despite EF recovery, 20% of women, compared with 54% with persistent LV dysfunction, will have aggravation in the following pregnancies [5].PPCM patients risk heart failure relapse in the subsequent pregnancy[6]. Thus, Patients who desire to get pregnant in the future should wait for at least five years after the initial ejection fraction has normalized, and they should undergo an echocardiogram at Six weeks and six months postpartum and annually after that[2][7][8][16]. Although current studies have shown the inverse relationship between the level of systolic left ventricular function after the recovery from heart failure and the risk of relapse in subsequent pregnancies, 50% is the relapse rate of heart failure in the next pregnancy for patients who were not fully recovered from PPCM[9]. A study defined PPCM "recovery" as an LVEF of at least 0.55. The ejection fraction range between 0.50 and 0.54 can be accompanied by mildly persistent left ventricular dilatation and ventricle remodeling, contributing to a higher relapse rate[9]. It is advisable to continue with the treatment for the acute stage of PPCM following American Heart Association Guidelines to prevent any harmful damage with close echocardiography monitoring. An echocardiogram provides a gross measurement of LV function; it does not provide dependable results if the patient is fully recovered. Adding other modalities should be considered for further validation, such as exercise stress echocardiography to assess PPCM is a piece of practical information on how the heart may respond under the stress of pregnancy, labor, and delivery [9]. It would also be advisable to continue GDMT for PPCM to avoid relapse; in the TRED-HF (Withdrawal of pharmacological treatment for heart failure in patients with recovered dilated cardiomyopathy) piolet study, patients with dilated cardiomyopathy relapsed six months after discontinuing heart failure medications compared with those continuing treatment (44% vs. 0%), including twoPPCM patients, one of which

relapsed[17].Our case and other reported cases require solid guidelines addressed specifically for PPCM.

Comment [A5]: What were the limitations in the case studied? add

Conclusion:

The course with PPCM in women can be unpredictable, and even women who seem to recover may still be at risk for future relapse due to untreated subclinical dysfunction. No evidence supports continuing on HfrEF treatment, andthere are no consensus recommendations about medication duration.; however, these patients exemplify the significance of maintaining long-term management and periodic monitoring. Even when their EF on echocardiography has normalized, other imaging techniques, such as cardiac magnetic resonance imaging, may determine the extent of myocardial healing and risk of relapse.

Comment [A6]: Main lessons of the investigated case

CONSENT

Written informed consent was obtained from the patient for publication.

ETHICAL APPROVAL

The study was approved by King Abdullah International Medical Research Center (KAIMRC).

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