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Revisiting Peripartum Cardiomyopathy: A Case Report and Review

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ABSTRACT

Peripartum cardiomyopathy (PPCM) is a rare and idiopathic disease characterized by left ventricular dilation and systolic dysfunction, that occurs late in pregnancy or during the postpartum period. Diagnosis is typically achieved through transthoracic echocardiography and cardiac magnetic resonance imaging (MRI). Management includes standard heart failure therapies such as beta-blockers, ACE inhibitors, and diuretics, with the addition of bromocriptine to target the prolactin pathway involved in PPCM pathophysiology. This report presents a case of a 31-year-old female with a history of acute lymphoblastic leukemia who, following an uncomplicated pregnancy, presented with acute heart failure symptoms. Initial evaluations suggested pulmonary embolism, but further diagnostic workup, including echocardiography and cardiac MRI, confirmed PPCM. The patient was treated with diuretics, ACE inhibitors, and beta blockers. Significant improvement was observed, with complete resolution and normal cardiac function at the 6-month follow-up.

KEYWORDS: Peripartum cardiomyopathy, heart failure, cardiac magnetic resonance, echocardiography, pregnancy, cardiomyopathy.

ARTICLE DETAILS

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I. INTRODUCTION

Peripartum cardiomyopathy (PPCM) is a rare, idiopathic disease characterized by dilation and systolic dysfunction of the left ventricle (LVEF <45%, LVFS <30%, or both), occurring in late pregnancy or the postpartum period, manifesting during the last month of pregnancy (up to 42 days before delivery) or within the first 5 months postpartum, without any other identifiable cause of heart failure. (1, 2)

Its incidence is variable, with a heterogeneous distribution and a higher incidence in countries such as Nigeria, Haiti, and China. (3)

Clinical presentation is characterized by signs and symptoms of heart failure, such as exertional dyspnea, orthopnea, paroxysmal nocturnal dyspnea and peripheral edema. There are different imaging approaches for diagnosis of PPCM, being the most useful transthoracic echocardiography and cardiac magnetic resonance.

The management of PPCM primarily involves standard heart failure therapies, including beta-blockers, ACE inhibitors, and diuretics, aimed at reducing symptoms and improving cardiac function. However, a unique aspect of PPCM treatment is the use of bromocriptine, which targets the prolactin pathway implicated in the disease's pathophysiology.

II. CASE REPORT

A 31-year-old female with a history of acute lymphoblastic leukemia (ALL) at age 7, received chemotherapy with remission, followed by cardiology monitoring, and was discharged without evidence of chemotherapy-induced cardiomyopathy.

After her first pregnancy, which was uncomplicated and lasted 38 weeks of gestation, she presented at the emergency room with sudden resting dyspnea, diaphoresis, palpitations and nausea on the third day of physiological puerperium. Physical examination revealed heart sounds with S3, bilateral crackles, and peripheral edema.

The electrocardiogram showed sinus tachycardia with left bundle branch block, and cardiac biomarkers were negative.

She was hospitalized with a presumptive diagnosis of pulmonary embolism, and D-dimer was 1103 ng/ml. Angiotomography showed no evidence of pulmonary artery occlusion. NT-proBNP was 1958 pg/ml, and subsequent measurement was 5422 pg/ml.

Transthoracic echocardiogram (figure 1) revealed a dilated left ventricle (LV) with an ejection fraction (EF) of 41%, global hypokinesia, global longitudinal strain (GLS) -12.1%, mildly dilated right ventricle (RV), moderate central mitral regurgitation, and slightly dilated atria.

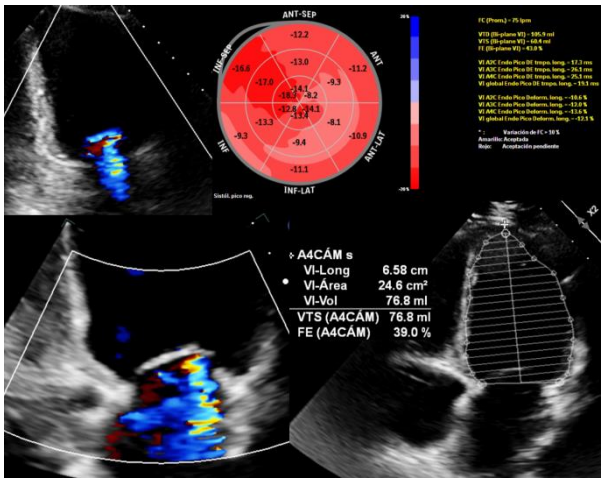

Figure 1. Transthoracic echocardiography reveals systolic dysfunction with an ejection fraction (EF) of 39%, a global longitudinal strain (GLS) of 12.1%, and mild mitral regurgitation.

Treatment with diuretic medications led to an initial improvement in acute heart failure. Subsequently, therapy for heart failure with mildly reduced ejection fraction was initiated, using angiotensin-converting enzyme inhibitors and beta blockers.

A posterior cardiac MRI (figure 2) reported LV EF of 45% and diffuse intramyocardial late gadolinium enhancement, confirming the diagnosis of peripartum cardiomyopathy.

After stabilization and up-titration to maximal tolerated dosage, the patient was discharged with optimal medical therapy.

At the 6-month follow-up, echocardiography showed an LV EF of 65%, with normal global and segmental LV wall motion, GLS -20.3%, normal RV and atrial volumes, and no valvular disease. Clinically the patient was asymptomatic with total recovery of her previous

III. DISCUSSION

Peripartum cardiomyopathy is an idiopathic condition recognized as a significant cause of heart failure, leading to substantial mortality and morbidity. This makes it a critical concern for the healthcare system.

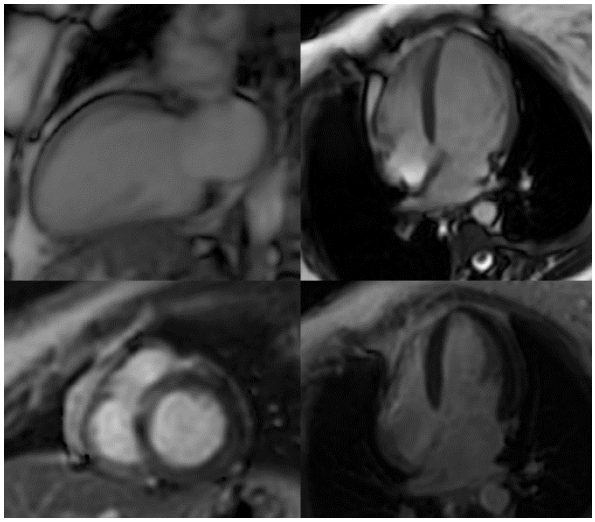

Figure 2. Cardiac magnetic resonance (CMR) reveals left ventricular dilation with diffuse intramyocardial late gadolinium enhancement

Despite its still underreported incidence and prevalence, it is important to recognize peripartum cardiomyopathy as a potentially fatal but treatable condition.

In international registries from 2004 to 2011, which included 32,219 women aged 15 to 54 years, an increasing trend in the incidence of this condition was observed, rising from 8.5 per 10,000 live births in 2004 to 11.8 per 10,000 live births in 2011. This increase has been associated with factors such as higher maternal age, a greater frequency of multiple pregnancies and preeclampsia, comorbidities such as diabetes mellitus, systemic arterial hypertension, and obesity, as well as greater visibility of the disease, which has helped reduce underdiagnosis of this condition. (4)

There are various risk factors that have been associated with peripartum cardiomyopathy, such as: hypertension (OR: 6.41; IC: 4.81-8.44; p= <0.0001), eclampsia (OR: 5.93; IC: 2.88-10.9; p= <0.0001), anemia (OR: 4.89; IC: 3.95-6.03; p=<0.0001), substance abuse (OR: 4.12; IC: 2.71-6.04; p=<0.0001), autoimmune diseases (OR: 3.61; IC: 1.42-7.43; p=0.002), multiple pregnancies (OR: 2.88; IC: 2.07-3.92; p=<0.0001), asthma (OR: 2.23; IC: 1.53-3.15; p=<0.0001), preeclampsia (OR: 1.99; IC: 1.78-2.69; p= <0.0001), smoking (OR: 1.43; IC: 0.89-2.22; p=0.12), obesity (OR: 1.42; IC: 0.84-2.24; p=0.16), diabetes mellitus (OR: 1.42; IC: 0.73-2.49; p=0.26). (5)

The overexpression of the STAT3 factor gives resistance to cardiotoxicity and is activated during pregnancy and postpartum. In murine models lacking STAT3 activity in cardiomyocytes, increased oxidative stress was observed, which stimulates the release of cathepsin D. Cathepsin D cleaves 23 kDa prolactin into a 16 kDa fragment that induces endothelial apoptosis, capillary rupture, and vasoconstriction. (6)

At the end of gestation, the placenta secretes sFlt1 (soluble Fms-like tyrosine kinase 1), an antiangiogenic protein that sequesters VEGF (vascular endothelial growth factor) and PIGF (placental growth factor) and is considered

the major inducer of hypertension, endothelial dysfunction, and preeclampsia. The administration of sFlt1 to nulliparous animals caused cardiomyopathy, even in the absence of pregnancy, suggesting that it is a key factor in the development of PPCM. The placental secretion of sFlt1 in preeclampsia and twin pregnancies may explain the epidemiological relationship. (7)

The two-hit model is a pathophysiological theory that describes vascular damage caused by antivascular hormonal effects in late pregnancy and early postpartum, which induce heart failure in women with a genetic predisposition (alteration of the TTN gene in up to 65% of patients with PPCM, as well as other minor associations in genes such as FLNC, DSP and BAG3). (3)

The echocardiogram may show varying degrees of left ventricular (LV) dilation, LV systolic dysfunction, right ventricular (RV) dilation, dilation of both atria, mitral and tricuspid regurgitation, and pulmonary hypertension.

Cardiac magnetic resonance allows tissue characterization with high spatial resolution, diagnostic precision and reproducibility. It is safe during pregnancy and breastfeeding. PPCM does not have a specific scarring pattern, but it can be useful in excluding other cardiomyopathies and is characterized by systolic dysfunction with non-ischemic late enhancement.

Additionally, imaging can identify poor prognostic factors: severe left ventricular (LV) systolic dysfunction, right ventricular (RV) systolic dysfunction, intracavitary thrombus, LV end-diastolic diameter >60 mm, low tissue Doppler velocities, myocardial edema (increased T2 index, EGE index, T2 mapping), late gadolinium enhancement (presence, extent, and persistence), abnormal T1 mapping indices, microvascular disease, and lack of contractile reserve on stress imaging. (8)

Various therapeutic resources are available, ranging from medications to ventricular assist devices or transplantation.

If the patient presents with signs of hemodynamic instability, the use of vasopressors or inotropes is recommended (levosimendan and milrinone are preferred over dobutamine because it is associated with worst outcomes). If vasodilator therapy is required, nitroglycerin is preferred, and if there is refractoriness to initial management, the use of ventricular assist devices may be recommended until shock resolution or as a bridge to transplantation. (9)

During the last trimester, the use of ACE inhibitors, aldosterone antagonists and ARNI is contraindicated, and the use of loop diuretics, vasodilators (hydralazine, amlodipine) and beta blockers (beta 1 selective – metoprolol) is recommended for symptomatic management, and if anticoagulation is required (recommended in LVEF <35% or with evidence of intracavitary thrombus or thrombotic events), the use of warfarin or LMWH (low mass weight heparina) is indicated. (10)

The only targeted treatment available for PPCM is bromocriptine, but it is not included in the essential drug list of the Mexican health system pharmacies.

The use of Bromocriptine 2.5 mg every 12 hours for 2 weeks, followed by 2.5 mg every 24 hours for 6 weeks, has been described. It is currently a recommended as specific medication for patients with PPCM by complementing the standard treatment. (2) (11)

Most studies report only LVEF monitoring at 6 months of follow-up, with recovery rates varying by race and region between 20-60%, with a greater probability of recovery if the VTD of the left ventricle is <60 mm and has an LVEF >30%. (9)

IV. CONCLUSIONS

Peripartum cardiomyopathy remains a rare but critical cause of heart failure in the peripartum period, demanding heightened clinical awareness due to its potentially fatal outcomes. Early recognition and timely intervention are essential to improve prognosis, as demonstrated in this case report. The successful recovery of the patient, with complete normalization of cardiac function within six months, underscores the efficacy of standard heart failure therapies, including beta-blockers, ACE inhibitors, and diuretics.

While the use of bromocriptine represents a targeted approach in PPCM by addressing the prolactin pathway, this case highlights that favorable outcomes can also be achieved without it, especially in settings where it may not be readily available. Moreover, the ability to continue breastfeeding without compromising treatment emphasizes the importance of personalized patient care.

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