Reversible peripartum cardiomyopathy in a triplet pregnancy

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Abstract

Peripartum cardiomyopathy (PPCM) is a rare form of dilated cardiomyopathy that occurs in previously healthy women in the last month of pregnancy and up to several months after delivery. The incidence of PPCM is low, but its morbidity and mortality rate are high, with a substantial risk of poor outcome of the pregnancy. Patients who have recovered from PPCM run a high risk of reoccurrence in subsequent pregnancies. In this case report we present a 32-year old female patient who developed acute heart failure (HF) associated with significantly reduced systolic function due to PPCM soon after a delivery of triplets. Treatment was immediately initiated in the intensive coronary unit with oxygen-therapy, loop diuretics, aldosterone blockers, beta blockers, angiotensin-converting enzyme (ACE) inhibitors and bromocriptine. During the follow up period, a year and a half after delivery, a complete recovery of systolic function was observed with no residual symptoms.

Key words: dilated cardiomyopathy, triplet pregnancy, peripartum period

Introduction

Peripartum cardiomyopathy (PPCM) is a rare form of cardiomyopathy characterized by the development of heart failure (HF) due to left ventricular systolic dysfunction. Since 1997, four diagnostic criteria, proposed by the American National Heart, Lung, and Blood Institute and the Office of Rare Diseases Research, are used to diagnose this condition. They are: (a) the development of HF in the last month of pregnancy or within five months of delivery; (b) the absence of a determinable etiology for HF; (c) the absence of demonstrable heart disease before the last month of pregnancy; (d) left ventricular systolic dysfunction defined by a left ventricular ejection fraction (LVEF) < 45%, fractional shortening <30%, or both. (1,2) The incidence of PPCM ranges from 1:300 to 1:15,000 live births. The published mortality rates range from 25 to 50%. (3) Etiology is unclear but it may be connected to hemodynamic and hormonal changes in pregnancy or cytokine dysfunction. (4) There are a number of factors that are associated with PPCM such as: preeclampsia, multiparity, advanced maternal age, multiple pregnancy, cocaine abuse, selenium deficiency and prolonged oral tocolytic therapy with beta adrenergic agonists. The treatment is based on the administration of standard drug therapy for acute and chronic HF that includes the use of diuretics, intravenous and oral vasodilatators, intravenous inotropes, angiotensin-converting enzyme (ACE) inhibitors or angiotensin II receptor blockers, beta-blockers, spironolactone, and digoxin. (5,6)

Case report

In this case report we present a 32-year old female patient who developed PPCM after delivering triplets. She was admitted to the gynecology and obstetrics clinic in the 32^{nd} gestational week.

Her past medical history was unremarkable. No cardiovascular disease was noticed. Physical status was within normal limits. Up to the 30^{th} week of gestation the pregnancy was uneventful. At that time shortness of breath was noticed, but this was attributed to advanced/ triplet pregnancy. In the 35^{th} gestational week elective cesarean section was performed. Three healthy babies were delivered. The early postoperative period was unremarkable. Post-delivery, arterial hypertension was

discovered and treated with urapidil and nifedipine. Five days after delivery, the patient reported shortness of breath. Oxygen saturation was reduced to 92%. Chest examination revealed bilateral crackles. A gallop rhythm with a holosystolic murmur (intensity 4/6, loudest heard at the apex) was noticed. Chest X-ray showed pulmonary interstitial edema, an increase in the cardiac silhouette and bilateral pleural effusion. Echocardiography showed a dilated left ventricle (to 62 mm), reduced systolic function of the left ventricle (LVEF-35% by Simpson measurement), an enlarged left atrium (dimensions 64×66 mm), massive mitral regurgitation, good contractility of the right ventricle (tricuspid annular plane systolic excursion-TAPSE 26 mm) and medium tricuspid regurgitation with pulmonary hypertension (pulmonary artery systolic pressure – PASP 75 mmHg). MSCT pulmonary angiography showed no thrombi in the pulmonary circulation. Initial N-terminal proBrain Natriuretic Peptide (NTproBNP) level was 15 316 pg/ml (normal values <125 pg/ml). The patient was transferred to the intensive coronary unit where she was treated with oxygen-therapy, loop diuretics, aldosterone blockers, beta blockers, ACE inhibitors, bromocriptine and other supportive therapy.

After prolonged hospitalization, for more than a month, echocardiography showed improvement in systolic function and a normal sized left ventricle (53 mm), with generally reduced contractility (LVEF~45%) (figure 1). A very mild mitral regurgitation was present and the pressure gradient over the tricuspid valve was 20 mmHg. NTproBNP level was now 102 pg/ml, respectively. On discharge the patient was clinically stable, without dyspnea. She was offered bisoprolol with minimal doses of furosemide, spironolactone and valsartan. Six months later, only bisoprolol was continued.

Two years after delivery the patient presented asymptomatic with a normal physical status and no echocardiographic abnormalities (figure 2).

Discussion

This case report presents a normal pregnancy with PPCM that developed

soon after delivery. By rapidly diagnosing the condition, thanks to well established criteria, and promptly initiating treatment, a good short and long term outcome was achieved .

An unremarkable history, along with symptoms of congestive heart failure, are usually present, which should lead the clinician to think of PPCM. Echocardiography, as a goal standard in cardiac imaging, and elevated natriuretic peptides can confirm the diagnosis. In our case we emphasize the triplet pregnancy as a predisposing risk factor for PPCM.

Treatment is focused on reduction of preload and afterload and improving cardiac contractility. Diuretics, beta blockers and ACE inhibitors are the main choice of treatment. (5,6) Some reports show beneficial effects of human immunoglobulins. (7) We also used bromocriptine because of its potential benefit on PCCM. (8)

Although rare, PPCM requires awareness and care, due to its high mortality and morbidity. A pregnancy aggravated by a dilated left ventricle can lead to unwanted thromboembolic events, rapid HF or sudden death. (9,10) Patients who have recovered from PPCM run a high risk for the disease in subsequent pregnancies. Preterm delivery and maternal mortality remain the highest problem. Therefore, these patients should be advised to avoid further pregnancies. If the patient insists on a new pregnancy, a thorough clinical follow-up every six months is needed until systolic function improves to at least 50%. At that point a new pregnancy could be planned with proper cautions in place. Taking into account that in twin pregnancies cardiac output is 20 percent higher than in women carrying singletons, and it peaks at 30 weeks of gestation, we can presume that in this case PPCM was mostly caused by the triplet pregnancy. (4) In our patient, a future singleton pregnancy could be considered, given the patient's full recovery and the fact that the previous pregnancy was a triplet. If cardiac function does not fully recover, future pregnancies should be avoided.

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Figure 1. Parasternal long-axis echocardiogram view (PLAX view) – reduced systolic function of a dilated left ventricle (15 days after hospitalization).

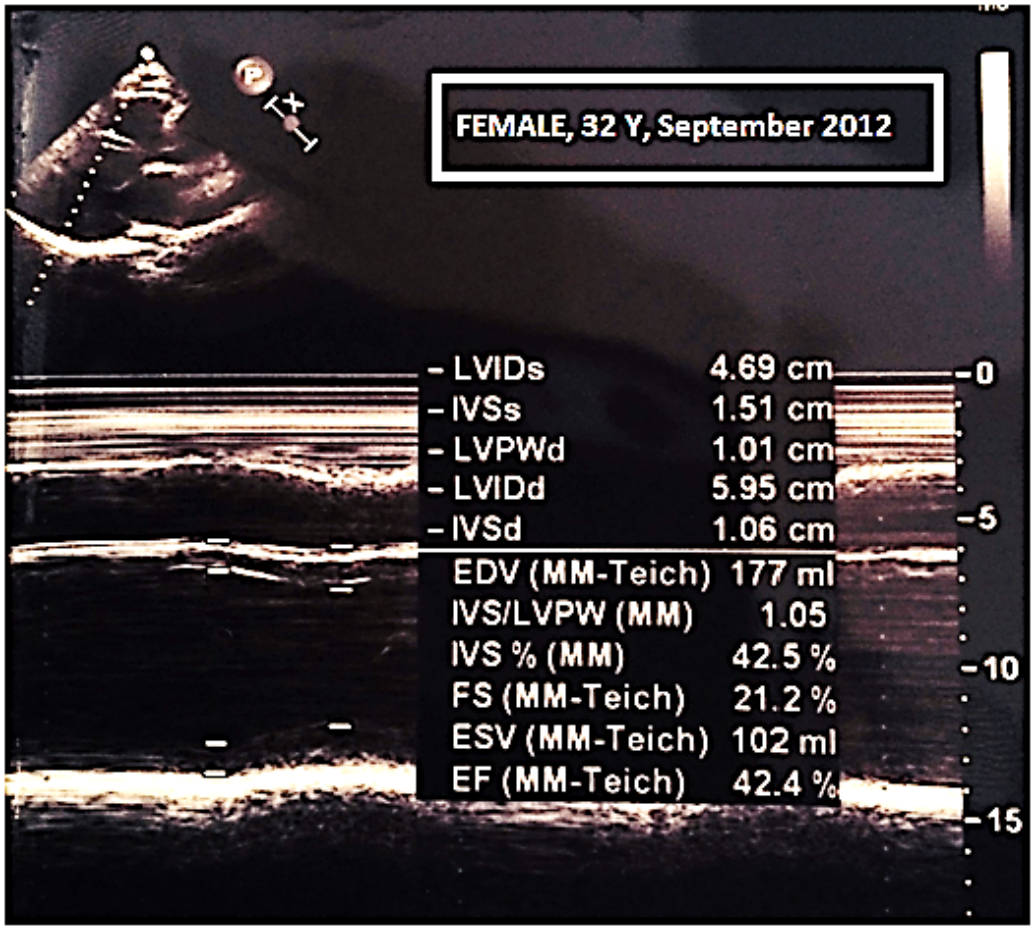
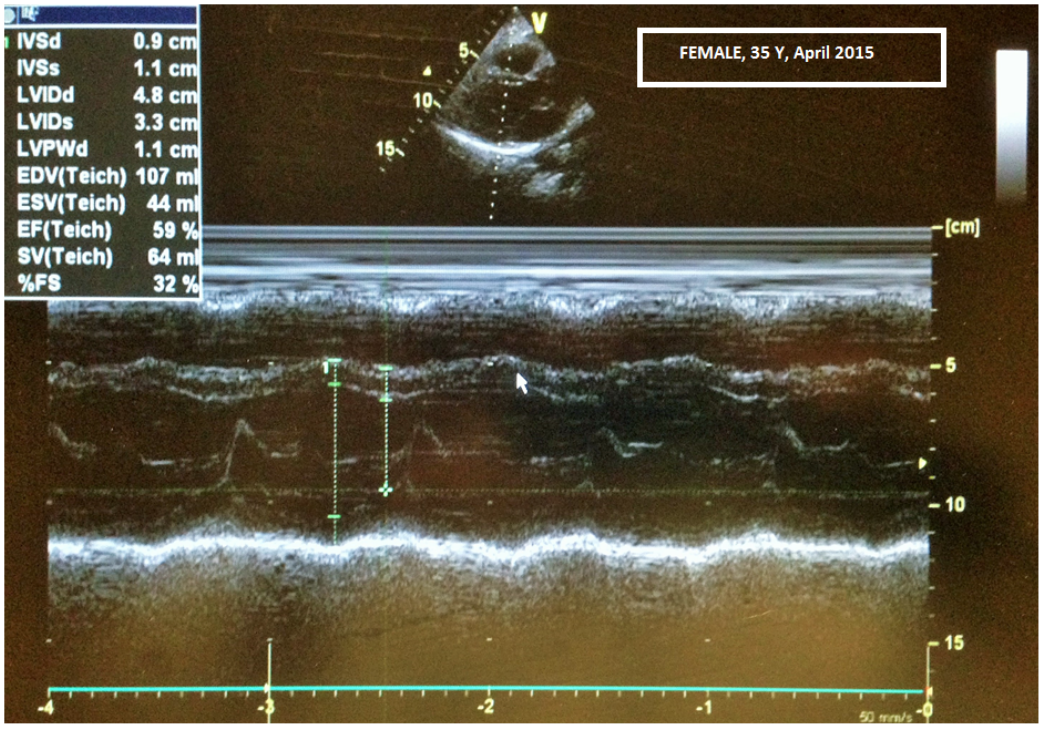


Figure 2. Parasternal long-axis echocardiogram view (PLAX view) – normal systolic function of a normal sized left ventricle (two years after hospitalization).



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