Successful management of fatal peripartum cardiomyopathy in a young pregnant woman

A case report

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Abstract

Rationale: Peripartum cardiomyopathy (PPCM) is a rare and life-threatening form of pregnancy associated myocardial disease. Patient concerns: In the present report, we describe a case of a patient with PPCM at 33 weeks of gestation with stillbirth and cardiorespiratory failure. Diagnoses: Peripartum cardiomyopathy. Interventions: The patient underwent emergency cesarean section (CS) and comprehensive medical treatments, including bromocriptine, as well as positive inotropic agents and diuretics after the CS. Outcomes: She had an uneventful recovery period, and was discharged 9 days after surgery. Her heart function was restored within 6 months after CS, and follow-up echocardiographies indicated normal heart function. Lessons: This case highlights that early diagnosis and timely termination of pregnancy are crucial in the management of PPCM. Abbreviations: ABG = arterial blood gas analysis test, CS = cesarean section, FiO2 = fraction of inspired oxygen, LVEF = left ventricular ejection fraction, PaO2 = arterial oxygen partial pressure, PPCM = peripartum cardiomyopathy, pro-BNP = pro-brain natriuretic peptide, S/D = systolic/diastolic ratio, SpO2 = oxygen saturation. Keywords: cardiovascular, cesarean section, critical care obstetrics, peripartum cardiomyopathy, stillbirth

1. Introduction

Peripartum cardiomyopathy (PPCM) is a rare form of pregnancy-associated myocardial disease characterized by left ventricular systolic dysfunction.[1] The risk factors include multiparity, advanced maternal age, multiple pregnancies, preeclampsia, gestational and pre-existing hypertension, and Afro-Caribbean race.[2] Although dyspnea and tachycardia are the most common complaints among these patients, nonspecific symptoms such as fatigue and palpitation may also be observed. Approximately half of the cases achieve spontaneous and complete recovery of left ventricular function after gestation. However, the other cases exhibit a much more progressive disease, for which intensive treatments and even heart transplantation may be needed.[3] Here, we report a serious case of PPCM that was successfully managed by a multidisciplinary team led by obstetricians.

2. Case report

An 18-year-old primigravida presented with a 2-day history of worsening dyspnea at 33 weeks of gestation. Two days before admission, the woman developed progressing shortness of breath and paroxysmal nocturnal dyspnea. Although primary examinations exhibited unremarkable results, she developed orthopnea and exhibited pink frothy sputum on the next day. Her pulse was 140 beats per minute, oxygen saturation (SpO2) was 82%, and arterial oxygen partial pressure (PaO2) was 49.1 mm Hg on arterial blood gas analysis (ABG). Moreover, fetal ultrasonography exhibited a single live fetus with a systolic/diastolic ratio (S/D) as high as 4.78. The patient was transferred via ambulance to our hospital.

Her medical history was unremarkable. She did not have a history of hypertension, congenital heart disease, myocarditis, valvular heart disease, myocardiopathy, or autoimmune disease. She did not have any siblings. During the gestation period, the patient was asymptomatic, until the current episode. She did not have hypertension or other cardiovascular diseases, and she did not have any abortions or induced labor, and had no history of medication use before gestation. She had no known allergies, and did not smoke, drink alcohol, or use illicit drugs. Her parents did not have hypertension or other cardiovascular diseases, and she did not have any siblings. During the gestation period, the patient was asymptomatic, until the current episode. She did not have hypertension, vaginal bleeding, fever, or chills before admission.
Noninvasive ventilator support (Fig. 1). Her SpO2 increased to 90% refractory respiratory failure that could not be managed via intubation and mechanical ventilation was initiated after showing a PaO2 of 48 mm Hg on supplemental oxygen via a nasal cannula. Fetal Doppler ultrasonography indicated a concurrent stillbirth. Oxygen (40%) was administered via a simple face mask at a flow rate of 8 L/min. Moreover, cedilanid (0.4 mg), torasemide (20 mg), and morphine (10 mg) were administered intravenously. The obstetrician on call suggested that the dead fetus should be removed immediately after the patient’s vital signs stabilize. However, the patient’s condition progressively deteriorated. She was intubated and mechanical ventilation was initiated after refractory respiratory failure that could not be managed via noninvasive ventilator support (Fig. 1). Her SpO2 increased to 90% after intubation under high positive end expiratory pressure and fraction of inspired oxygen (FiO2). Moreover, laboratory tests indicated pro-brain natriuretic peptide (pro-BNP) levels of 14000 pg/mL (normal range <133 pg/mL) and BNP levels of 2919.1 pg/mL (normal range 0–87 pg/mL), whereas transthoracic echocardiography indicated impairment of left ventricular systolic function, with an estimated left ventricular ejection fraction (LVEF) of 40% and mild pulmonary hypertension. Urgent discussions were conducted by the multidisciplinary team. Long-standing heart diseases, pregnancy-induced hypertension, Takotsubo cardiomyopathy, and pulmonary embolism were all ruled out based on her medical history and lack of characteristic signs and specific laboratory findings. As she exhibited rapidly deteriorating heart failure, she was clinically diagnosed with PPCM, and emergency cesarean section (CS) was strongly recommended by the obstetrician, amid conservative suggestions such as extracorporeal membrane oxygenation, suggested by the intensivist. Emergency CS was performed under general anesthesia 21 hours after admission. A dead infant weighing 1000 g was delivered, and the woman was transferred to the surgical intensive care unit. Her respiratory failure markedly improved after the CS (Fig. 1), and her BNP levels decreased to 323.3 pg/mL on the next day. She received bromocriptine (5 mg q.d.), digoxin (0.125 mg q.d.), and furosemide (40 mg q.d.) after the CS, and was discharged 9 days after surgery. She continued to receive bromocriptine (5 mg q.d.) and losartan (50 mg q.d.) for 3 months after discharge. Echocardiography at 3 months after discharge showed a LVEF of 51%, with mild mitral and tricuspid regurgitation, and her LVEF improved to 62% at 6 months postoperatively. She appeared to be in good condition during the follow-up visits.

Informed consent was obtained from the patient for publication of this case report.

3. Discussion

Peripartum cardiomyopathy is a life-threatening disease characterized by left ventricular dysfunction during the pregnancy or early postpartum period in patients without a history of cardiovascular disease. The diagnostic criteria are as follows: heart failure occurring in the last month of pregnancy or in the first 5 months after delivery; no determinable cause of cardiac failure; no demonstrable heart disease before the last month of pregnancy; and echocardiographic features of left ventricular dysfunction and an ejection fraction of <45%.4 Our case met all the diagnostic criteria.

Obstetricians may play an important role in the early diagnosis of PPCM. PPCM is a special form of heart failure with indeterminate pathogenesis, and has a wide spectrum of diverse clinical manifestations. Some of the early symptoms resemble benign complaints of hemodynamic stress during pregnancy, which could be responsible for the low rate of early diagnosis. Hence, most cases of PPCM are detected and treated by cardiologists for arrhythmia or heart failure at postpartum. Moreover, the rate of full recovery of left ventricle function is lower in patients with more progressive heart failure, thus highlighting the impact of timely diagnosis.10 With the increasing awareness of PPCM, we believe that obstetricians can play a critical role in the early diagnosis of PPCM during check-ups. In the present case, the patient was not diagnosed with PPCM during the early stage, and the baby could not be saved. The decision to terminate the pregnancy, and the timing, is critical in the management of PPCM. Emergent CS should be performed when the patients’ condition worsens, despite comprehensive conservative treatment. In the present case, we attempted to stabilize the patient’s condition to facilitate elective CS, but her condition worsened instead. Furthermore, in cases where the fetus is alive,
urgent CS should be performed to preserve the fetus. However, the likelihood of healthy babies from future pregnancies is low in PPCM patients, as PPCM recurrence in subsequent pregnancies is reportedly as high as 46%, particularly among women with LVEF <55% before pregnancy.[6]

In conclusion, we present a serious case of PPCM, wherein the patient was successfully treated via a multidisciplinary approach involving emergent CS combined with medical therapies. The findings clearly show that early diagnosis and timely termination of pregnancy are crucial in the management of PPCM.

Author contributions
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Validation: Meiqi Zhang.
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