

Coexistence of postpartum cardiomyopathy and single coronary artery anomaly

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ABSTRACT

Postpartum cardiomyopathy (PPCM) is dilated cardiomyopathy seen in the postpartum period in women who no prior history of cardiac diseases. In this case study, coronary angiography was used to determine the etiology of the cardiomyopathy after the decompensated heart failure had regressed. A congenital single coronary artery anomaly was detected by coronary angiography. In this case, PPCM can mimic pneumonia, thus making diagnosis difficult. When there is no response to antibiotic therapy, the obstetrician must consider cardiac disease and appropriate consultation should be obtained as soon as possible. To the best of our knowledge, single coronary artery anomalies coexisting with PPCM has only been documented in one other case in the literature. An association of single coronary artery anomalies with PPCM may be random, or, alternative, might contribute to the composition of the PPCM.

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INTRODUCTION

Peripartum cardiomyopathy is idiopathic heart failure occurring in the absence of any determinable heart disease during the last month of pregnancy or the first 5 months postpartum.¹ It was first described by Demakis and colleagues in 1971. Its prevalence is higher in developing countries, e.g Nigeria, Haiti and Africa. The exact cause of postpartum cardiomyopathy (PPCM) is unknown, but various viral infections and autoantibodies have been implicated in the pathogenesis of this disease.² In addition, advanced maternal age, multiparity, African descent, twin pregnancy, pregnancy-induced hypertension and long-time miscarriage are also found to be related to PPCM, but no casual association has been shown.³ Proposed etiologies for PPCM include

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inflammation, genetic mechanisms, abnormal responses to the physiologic stress of pregnancy, autoimmune factors, viral myocarditis, nutritional deficiencies, and prolonged tocolysis. Thus, the exact cause of PPCM is unknown and the pathogenesis is probably multi-factorial.

Coronary artery anomalies are observed in 0.3-1.3% of patients undergoing diagnostic coronary angiography, in approximately 1% of routine autopsy examinations, and in 4-15% of young people who experience sudden death. In the general population, the incidence of a single coronary artery is approximately 0.024%.⁴

Coronary artery anomalies are frequently associated with other major congenital anomalies like tetralogy of Fallot and transposition of great artery anomalies. It is strongly associated with sudden death, myocardial ischemia, congestive heart failure and endocarditis. To the best of our knowledge, the coexistence of single coronary artery anomalies with PPCM has only been reported in one case in the literature. We report a rare case of PPCM with a single coronary artery anomalies.

CASE REPORT

A forty nine-year-old female patient was admitted to the cardiology clinic with complaints of dyspnea, coughing and inability to lie on her back. She had no chronic diseases, except Hashimoto's thyroiditis. She had used levothyroxine therapy for two years, and she did not smoke. She had given birth to her ninth baby by normal vaginal delivery one month previously and the labor was unproblematic. Her

complaints began two weeks after delivery when she was admitted to the gynecology and obstetrics clinic with similar complaints. She was diagnosed with pneumonia and started a course of antibiotic therapy. However, she did not respond to therapy. The patient was referred to the cardiology clinic given her dyspnea increased and orthopnea and pretibial edema developed. Upon her physical examination, dyspnea, orthopnea and generalized pretibial edema were observed, her heart rate was 100 bpm, and blood pressure was 130/80 mmHg. In the cardiac auscultation, heart sounds were rhythmic, tachycardic, S1+, S2+, S3+ and there was a 2/6 pansystolic murmur at the apex. In the lung auscultation, bilateral respiratory sounds were equal; however, she had generalized crepitant rales up to the middle zone. The anteroposterior radiograph of the chest showed increased cardiothoracic ratio and hyperdense areas in bilateral perihilar areas consistent with pulmonary venous congestion (Image 1).

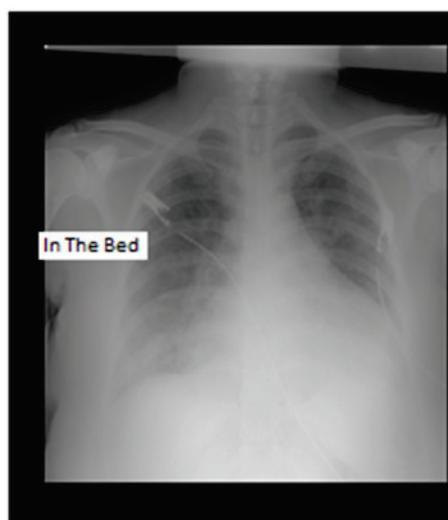


Image 1. AP chest x-ray, CTR>0.50. Hyperdense areas in bilateral perihilar region and bilateral

perihilar fullness.

The ECG showed normal sinus rhythm and there was an anteroseptal QS wave. Laboratory tests were normal. In the echocardiography, there was an ejection fraction of 0.35, the left ventricle was dilated, the left ventricle diastolic diameter was 60 mm and there was global hypokinesia, particularly in the inferior, inferoapical and septobasal areas and the lateral wall (Image 2).

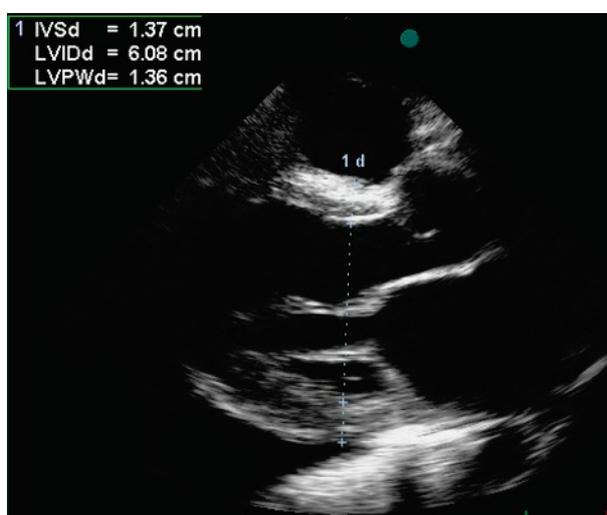


Image 2.
Postpartum cardiomyopathy.
Left ventricle diastolic diameter was 60 mm.

The aorta and bilateral atria were enlarged. Moderate mitral valve insufficiency and mild tricuspid and aortic valve insufficiency were detected by color and conventional Doppler examination. The patient, who had experienced no previous cardiac disease, was diagnosed with PPCM. After the decompensated heart failure had regressed, the patient underwent coronary angiography in order to determine the etiology of the cardiomyopathy. A congenital single

coronary artery anomaly originating from the right coronary cusp and branching to the right and left coronary arteries was detected by coronary angiography (Image 3). No occlusion was detected in the coronary arteries. In the ventriculography, generalized hypokinesia was detected in the left ventricle and the ejection fraction was found to be 38%. Ischemic heart disease was excluded.



Image 3: Coronary angiography showing the anomalous origin of the left main (LM) coronary artery from proximal right coronary artery (RCA) with subsequent retroaortic (dorsal [type D]) course to the left side.

In light of these data, a course of 80 mg parenteral furosemide, 5 mg oral perindopril, and 25 mg oral spirinolactone, digoxin and a salt-free diet was started. Fluid restriction was also recommended. Hemodynamic instability and systolic dysfunction were not observed, so the patient did not require a ventricular assist device, inotropic agents or cardiac transplantation. Dyspnea regressed and lung rales as determined by auscultation decreased about two weeks after the drug therapy.

Therefore, 12.5 mg carvedilol was added to the medical therapy. Fluid retention declined, the pretibial edema was resolved, and shortness of breath improved (clinical NYHA functional class improved from class III to class II). The effort capacity of the patient was increased and the patient was discharged with oral diuretic treatment. The patient was reevaluated by physical examination and echocardiography in the 3rd and 6th month follow ups. In the follow up examination, the shortness of breath had regressed (NYHA I), the ejection fraction was 45% in the third month and 50% in the sixth month as determined by echocardiography, and the size of the left ventricle had decreased.

DISCUSSION

PPCM is dilated cardiomyopathy emerging in the postpartum period in women who have had no previous cardiac diseases. It develops in the last prenatal month or within five months postpartum. It is defined as a disease with unexplained left ventricle systolic dysfunction and is diagnosed by echocardiography.⁵

In women who have developed PPCM, heart failure is usually accompanied by explicit fluid retention and less frequently embolic stroke, and arrhythmia may develop. Cases with poor prognosis tend to develop within the first few days postpartum. Heart failure may be severe; inotropic agents and ventricular support devices may be required. In these cases mortality varies between 3% and 60% in the acute and subacute phases. PPCM incidence is one in 1,300-15,000 live births. This wide range in incidence and mortality may be explained by

geographic differences, differences in diagnostic criteria and widespread use of echocardiography. Congestive heart failure, arrhythmia and thromboembolic events are responsible for mortality.

In our case, PPCM was considered in the differential diagnosis as heart failure symptoms accompanied by fluid retention developed two weeks after delivery. 75% of patients are diagnosed in the first month postpartum and 40% are diagnosed within the first week.⁶ It is frequently seen in women 30 years and older but also in women of all age groups. Prognosis is related to the improvement of left ventricle functions, and ventricle functions return to normal within the first six months in 30% of patients and 50% recover fully. In our patient, hemodynamic instability and systolic dysfunction were not observed, so the patient did not require a ventricular assist device, inotropic agents or cardiac transplantation. Dyspnea regressed and lung rales decreased about two weeks after initiation of drug therapy. Fluid retention declined, which resolved the pretibial edema and improved shortness of breath (clinical NYHA functional regressed from class III to class II). In the follow-up examination, the shortness of breath had regressed (NYHA I), the ejection fraction was 45% in the third month and 50% in the sixth month and the size of the left ventricle had decreased. Both clinical and echocardiographic findings had resolved at the end of the sixth months.

Although its etiology is not fully known, many risk factors are thought to play a role in PPCM, including advanced maternal age, grand multiparity, preeclampsia, long-standing tocolytic

therapy, black ethnicity, twin pregnancy and also viral infections, autoimmunity and selenium deficiency.⁷ Our case was a woman of advanced age who had not had any previous cardiac diseases and who was admitted with symptoms of heart failure. Grand multiparity and advanced age may have played a role in the development of PPCM in this case.

In the pathophysiological development of PPCM, inflammatory agents (TNF- α , interferon- γ , interleukin-6, C-reactive protein), myocarditis, oxidative stress, fetal chimerism and genetic associations play an important role. With regards to the causal role of fetal chimerism, during pregnancy fetal cells escape into the maternal circulation but are normally destroyed by the maternal immune system. When the maternal immune system is weakened, chimeric cells invade and settle in the maternal heart. Maternal antibodies directly attached to the chimeric cells result in dilated cardiomyopathy. Consistent with this proposed mechanism, high titers of antibodies against the cardiac myosin cells have not been identified in healthy pregnant women or idiopathic dilated cardiomyopathy. Also, there is a strong relationship between high titers of antibodies and multiple and subsequent pregnancies.³ In our case, the patient had given birth to her ninth baby, thereby supporting the relationship with subsequent pregnancies. In addition, our patient had Hashimoto's thyroiditis, an autoimmune condition. In our opinion, there might be a relationship between autoimmune antibodies and PPCM.

Even after full recovery of left ventricular function, subsequent pregnancies carry a risk of relapse of

PPCM. A study in Haiti followed 99 patients, 15 of whom became pregnant again. Eight of the women who became pregnant again experienced worsening heart failure and long-term systolic dysfunction.⁸ We believe that patients with PPCM need close follow-up due to the risk of recurrence in subsequent pregnancy.

Patients with PPCM have the classical symptoms of cardiomyopathy: abruptly deteriorating dyspnea, orthopnea, coughing and tachycardia. Making a diagnosis may be difficult as these symptoms may also be seen in pulmonary thromboembolism, pneumonia, amniotic fluid embolism and asthma in pregnancy. Our case had dyspnea and coughing and was first diagnosed as pneumonia by the gynecology and obstetrics clinic. A course of antibiotics was started and she was referred to the cardiology clinic as no response to therapy could be obtained and the symptoms deteriorated. Thus, PPCM should certainly be considered in cases that have newly developing similar symptoms in the postpartum period. A cardiological evaluation should be made in order to make a diagnosis. Diagnosis is made upon echocardiography. Diagnosis may be made by showing that the ejection fraction is below 45%, systolic dysfunction and/or fractional shortening is below 30%, and/or diastolic diameter is above 2.7 cm/m². In our case, the diastolic diameter was seen to increase (60 mm) (Image 2) and systolic functions globally decreased as seen by echocardiography and ventriculography. The disease has no specific treatment. Symptomatic treatment methods include fluid and sodium restriction, inotropic and

respiratory support and medical treatment for heart failure (ACE inhibitors, beta-blockers and diuretics).⁹ In our patient, we used both medical treatment methods, including beta-blockers, diuretics and digoxin and also symptomatic treatment methods of fluid and sodium restriction. Hemodynamic instability and systolic dysfunction were not observed so the patient did not require a ventricular assist device, inotropic agents (dopamine, dobutamine) or cardiac transplantation.

Single coronary artery anomalies are usually asymptomatic but according to the severity of the atherosclerosis, myocardial ischemia, syncope and sudden death may also occur.

When we searched for single coronary artery anomalies and PPCM in PubMed, we found only one case report. This was an anomalous origin of the left main coronary artery from the pulmonary trunk masquerading as peripartum cardiomyopathy. Daniels C. et al, presented a 16-year-old girl who was diagnosed with peripartum cardiomyopathy with congestive heart failure. Coronary angiography showed anomalous origin of the left main coronary artery from the pulmonary trunk.¹⁰

Right and left coronary arteries normally originate from ostia which are normally located in the Valsalva sinuses of the aorta. In our case, the left coronary artery was seen to have originated from the right sinus Valsalva together with the right coronary artery from a single sinus. The patient was followed up with medical therapy as the course of the left coronary artery was normal. Most coronary artery anomaly cases do not show symptoms

and they are usually detected during coronary angiography or incidentally during autopsies.¹⁰ Despite the absence of significant hemodynamic impairments in most coronary artery anomaly cases, there are also the cases which have resulted in myocardial ischemia and sudden death.¹¹ Our case is interesting due to the coexistence of PPCM and single coronary artery anomaly.

An association of single coronary artery anomalies with PPCM may be random or it might contribute to the composition of the PPCM. During coronary angiography performed to determine the etiology of the PPCM, single coronary artery anomalies should be kept in mind. As in this case, PPCM can mimic pneumonia and this situation makes the diagnosis difficult. When there is no response to antibiotics, the obstetrician must suspect cardiac disease and cardiological consultation should be done as soon as possible.

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