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Case Report / Olgu Sunumu

Peripartum cardiomyopathy mimicking acute aortic dissection: successful salvage with extracorporeal membrane oxygenation support

Akut aort diseksiyonunu taklit eden peripartum kardiyomiyopatisi: ekstrakorporeal membran oksijen desteği ile başarılı kurtarma

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In this article, we report a case admitted with severe chest pain associated with ST segment elevation, pericardial effusion and aortic flap appearance in echocardiography at the 31 weeks of gestation, mimicking aortic dissection, but diagnosed with peripartum cardiomyopathy and successfully treated with extracorporeal membrane oxygenation after developing acute cardiogenic shock.

Keywords: Cardiogenic shock; extracorporeal membrane oxygenation; peripartum cardiomyopathy.

Bu yazıda gebeliğin 31. haftasında ekokardiyografide ST segment yükselmesi, perikardiyal efüzyon ve aort flep görünümü ile ilişkili şiddetli göğüs ağrısı ile başvuran ve aort diseksiyonunu taklit eden ancak peripartum kardiyomiyopatisi tanısı konularak, akut kardiyojenik şok sonrası ekstrakorporeal membran oksijenasyonu ile başarılı bir şekilde tedavi edilen bir olgu sunuldu.

Anahtar sözcükler: Kardiyojenik şok; ekstrakorporeal membran oksijenasyonu; peripartum kardiyomiyopatisi.

Acute onset chest pain during pregnancy is possibly related to myocardial infarction, acute aortic dissection, or pulmonary embolisms. Herein, we present the case of a female patient who was admitted to our facility with severe chest pain associated with inferior ST-segment elevation, pericardial effusion, and the appearance of an aortic flap as seen in echocardiography at 31st gestational week. This mimicked aortic dissection but was diagnosed as peripartum cardiomyopathy. The patient was treated successfully, and she delivered a healthy male baby via extracorporeal membrane oxygenation (ECMO) after developing acute cardiogenic shock.

CASE REPORT

A 29-year-old previously healthy woman presented with a sudden onset of severe chest pain and dyspnea during the 31st week of pregnancy. A physical examination was revealed a blood pressure of 120/80 mmHg, heart rate of 65 bpm, and respiratory rate of 16 breaths per minute, and an electrocardiogram (ECG) demonstrated ST-segment elevation at the inferior and lateral leads. Due to the minimal elevation in the myocardial enzyme levels, the patient was initially diagnosed as having acute coronary syndrome (ACS), and primary percutaneous angioplasty was planned. In addition, coronary angiography revealed normal coronary arteries, and transthoracic echocardiography (TTE) showed severe left ventricular dysfunction [ejection fraction (EF) 25%] and pericardial effusion. Furthermore, transesophageal echocardiography (TEE) detected a free-floating intimal flap just above the sinotubular junction (Figure 1a). Thoracic contrast-enhanced computed tomography (CT)

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was also performed, with the suspicious results causing us to suspect aortic dissection (Figure 1b). Furthermore, thoracic CT was inconclusive regarding the presence of acute aortic dissection. The patient became hypotensive (blood pressure 70/45 mmHg) and tachypneic (respiratory rate of 35 per minute during her follow-up), and her peripheral perfusion became progressively disturbed due to persistent prolonged capillary filling time. Because of the patient's ongoing clinical deterioration, emergency surgery for the aortic dissection was scheduled. As a first step, the 31-week pregnancy was terminated via a cesarean section by the obstetrician just before the aortic surgery, and her abdomen was left open because of her progressively worsening hemodynamic status. After intubation, the male premature infant was transferred to the neonatal intensive care unit (ICU) where he was given the care needed for him to successfully survive. Next, a median sternotomy was performed, and almost 200 ml of serous material was aspirated. The patient's aorta was completely normal except for the thick fibrous material surrounding it; however, her heart was dilated and globally hypokinetic. Despite inotropic support, the patient was still acidotic and had a low cardiac output; therefore, an intraaortic balloon pump (IABP) was inserted as an supportive therapy. Unfortunately, the patient's clinical situation worsened during the closure of the chest, and her heart arrested suddenly. Thus, we made the decision to use a venoarterial ECMO device for circulatory support (QUADROX PLS, MAQUET Cardiopulmonary AG, Hirrlingen, Germany). Under open cardiac massage, the ascending aorta was cannulated at the arterial line while the right atrium was cannulated at the venous line. Afterwards, the venoarterial ECMO was initiated. At first, the blood flow through the device was increased to achieve a satisfactory mean arterial blood pressure and oxygenation rate (2.8-3 lt/min). Following this, her hemodynamic parameters rapidly improved, so we progressively decreased her use of inotropic medications. The abdomen was then reexamined for hemostasis and then closed. Due to central cannulation, the sternum was draped and left open for delayed closure. Her blood gases stabilized [pH 7.36, partial pressure of oxygen in the blood (PaO_2) 102 mmHg, partial pressure of carbon dioxide in the blood ($PaCO_2$) 40.4 mmHg, bicarbonate (HCO_3-) 22.3 mmol/l, base excess 1.8 in fraction of inspired oxygen (FiO_2) 0.4]. After 48 hours of ECMO support and satisfactory improvement in the patient's hemodynamics, she was successfully weaned from the ECMO, and her chest was closed. On the fourth postoperative day, the IAPB was withdrawn, and the patient was extubated on the fifth postoperative day. Her control TTE revealed an EF of 35% and improved left ventricular wall motion compared with the preoperative findings. Due to her uneventful clinical recovery, she was discharged from hospital on the ninth postoperative day after another TTE showed persistent improvement in the global contractile pattern with an EF of 58%. The patient and her baby are now doing well.

DISCUSSION

Peripartum cardiomyopathy (PPCM) is seen in one out of every 15,000 deliveries, with mortality rates ranging between 7 and 50%.[1] It is a type of dilated cardiomyopathy in which deterioration in cardiac function occurs, especially between the last trimester and up to the fifth postpartum month. Moreover, PPCM is a diagnosis of exclusion because patients have no prior history of heart disease and there are no other known possible reasons for cardiac failure.[1,2] Both PPCM and stress-induced cardiomyopathy are recently defined pathologies with unknown

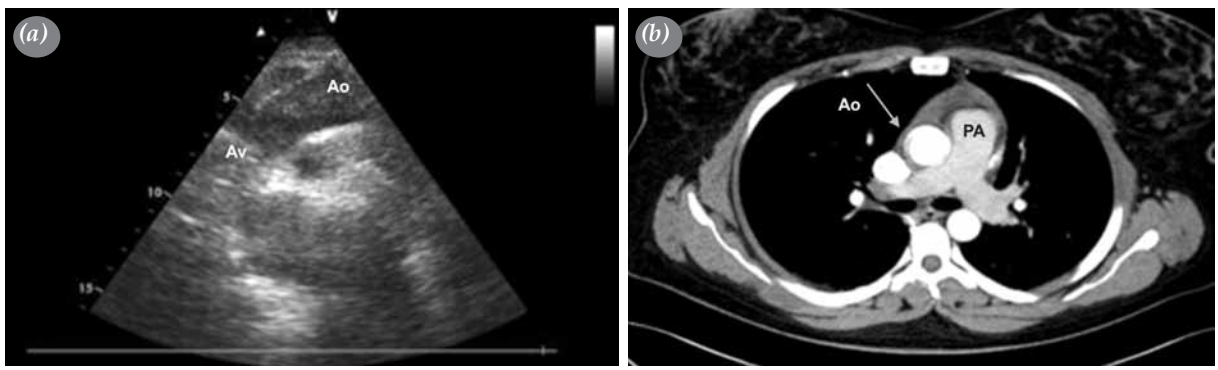

Figure 1. (a) Transesophageal echocardiographic view showing the aortic flap (arrow) and **(b)** contrast-enhanced thoracic computed tomography suggesting the presence of a suspicious intimal flap in the ascending aorta. Ao: Ascending aorta; Av: Aortic valve; PA: Pulmonary artery.

etiologies. They are often associated with ST-segment elevation, slightly elevated cardiac enzymes, and apical akinesia and are often mistaken for acute myocardial infarction.[3] The etiology of PPCM and stress-induced cardiomyopathy remains unknown, but PPCM is a disease of the late gestational period or early hemoptysis. Furthermore, PPCM patients rarely present with cardiogenic shock.

Our patient was at the 31st week of pregnancy, and she presented were a sudden onset of severe chest pain and ST-segment elevation on ECG. Our initial working diagnoses were ACS or acute aortic syndrome (AAS). Normal coronary angiography excluded the diagnosis of primary coronary pathology, and a suspicious view of an intimal flap in the aorta, the presence of pericardial effusion, and an inconclusive thoracic CT made us focus more on the differential diagnosis of acute aortic dissection. Her progressive clinical deterioration mandated both the saving of the premature infant via a rapid cesarean section and a median sternotomy in order to instantly rule out acute aortic dissection and relieve the pericardial effusion.

Acute aortic dissection is uncommon in pregnancy. The symptoms may be highly variable and can mimic other common conditions such as ACS. The European Society of Cardiology (ESC) guidelines recommend that aortic dissection along with myocardial infarction, a pulmonary embolism, and preeclampsia should be considered for any pregnant woman presenting with acute chest pain. Hence, a high index of suspicion must be maintained, especially when symptoms suggest this possibility. In our patient, TEE showed a highly suspicious intimal flap in the ascending aorta, and a CT scan did not definitively exclude the presence of aortic dissection because of the appearance of the thickened pericardium adhering to the ascending aorta. These findings were also consistent with what we found during the operation. Both CT and TEE are highly accurate in the diagnosis of aortic dissection. When CT is the initial diagnostic test, TEE is also needed to evaluate aortic regurgitation, and when TEE is initially performed, CT is needed to evaluate the extension of the dissection. Therefore, both are required for an accurate diagnosis. Although TEE and CT are highly sensitive and specific for aortic dissection, neither is 100% accurate. The potential causes for false positive results in CT scans are abnormal venous structures around the aorta (i.e., a low-lying innominate vein that can mimic an aortic flap), a large contrast bolus administered at the time of the procedure, or the presence of a thickened pericardium adhering to the ascending aorta.[4] Therefore, the accuracy of CT should not be overestimated in the diagnosis of acute aortic dissection.

The operative findings of a normal aorta and pericardial effusion brought to mind the possibility of PPCM. Hence, we decided to support the heart more aggressively and inserted the IABP and used the ECMO device because the patient had previously been young and healthy. In addition, survivors of PPCM usually recover, but they may occasionally need a heart transplant. In either case, ECMO can serve as a bridge to recovery or as a bridge to the transplant.

Extracorporeal membrane oxygenation was first used to support the postoperative period in congenital heart surgery in the 1950s. In the 1970s, it began to be used for respiratory problems in neonates. The technique has widened in parallel to improvements in technology, and nowadays, it serves as a well-known, short-term cardiopulmonary support system.[5,6] By draining the venous blood, removing the CO2, and then adding O_2 through an artificial lung, ECMO is achieved. The blood is then returned to circulation via a vein or artery. The functions of the heart and lungs are either totally or partially replaced by the ECMO system, which is able to support the circulation and respiration for up to 30 days. This provides enough time for the heart and lungs to recover.

Postcardiotomy cardiogenic shock and acute myocardial infarction related to cardiogenic shock are the predominant indications for short-term ECMO usage. Our experience along with a few other case reports in the literature that focused on PPCM indicate that it is rare to treat PPCM using ECMO,[7-9] but our case also indicates the need for an aggressive treatment modality in PPCM patients.

Declaration of conflicting interests

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