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Woman with Acute Heart Failure et Causa Peripartum Cardiomyopathy - A Case Report

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Abstract: ***Peripartum cardiomyopathy (PPCM) is a type of dilatative cardiomyopathy, which is characterized by heart failure due to left ventricular systolic dysfunction at the end of pregnancy or several months after delivery. Incidence of PPCM is low at 0.1% in pregnancy, but the morbidity and mortality are quite high between 5-32%. In some women, clinical and echocardiography can improve return to normal conditions, but in some other women can develop into heart failure and sudden cardiac death. This case was reported a woman with PPCM came with acute heart failure after five months of childbirth. Patients had no complaints of dyspneu before and during pregnancy, and no previous history of hypertension before. One week after giving birth the patient complained of dyspneu, after getting therapy the patient never came back to control. Five months later the patient came with a more severe complaint of dyspneu, leg edema and ascites. Where the results of echocardiography showed a lower ejection fraction that is <32.68%. Changes in normal pregnancy make PPCM onset easily covered and unknown, because of similar manifestations. Prognosis of PPCM is associated with improving of ventricular function. Failure of heart size returns to normal is associated with mortality and morbidity. Early diagnosis and the latest therapies for heart failure have an important role in reducing PPCM mortality and morbidity.***

Keywords: cardiomyopathy, peripartum cardiomyopathy, dilatative cardiomyopathy

1. Introduction

Peripartum Cardiomyopathy based on the National Heart and Blood Institute and the Office of Rare Diseases of the National Institute of Healthy Heart failure caused by left ventricular systolic dysfunction that occurs during late pregnancy and five months after postpartum without other causes of heart failure and disease beforehand.(1) Incidence of PPCM was low at 0.1% in pregnancy, but the morbidity and mortality were quite high between 5-32%. The diagnosis of PPCM is often late because PPCM symptoms are not specific, especially in late pregnancy. In some women, clinical and echocardiography can improve until returning to normal conditions, but in some other women can develop into heart failure and sudden cardiac death. Therefore caution regarding the symptoms of peripartum cardiomyopathy needs to be improved. Early diagnosis and the latest therapies for heart failure have an important role in reducing PPCM mortality and morbidity.

2. Case Report

A 43-year-old woman came to the emergency room with complaints of *dyspneu*, leg edema and ascites. Complaints have become increasingly in the past five months. Patients complain of fatigue easily during activity, coughing and waking during sleep due to tightness. History of chest pain and previous *dyspneu* is denied. The patient said there had never been a history of hypertension or diabetes mellitus before, had never found a lump in the neck. The patient five months ago had a labour in a sectarian section at another hospital. During pregnancy until the last trimester there are no symptoms of pre-eclampsia, only complaints of mild *dyspneu* such as normal complaints at the end of pregnancy. Before surgery the blood pressure rose 150/90 mmHg, and occasional PVC images were obtained on electrocardiography. Then the patient is consulted with a cardiologist. Chest radiography results from cardiomegaly with pulmonary congestion. From echocardiography results were obtained; LV dilatation, segmental normokinetic LV and LV ejection fraction of 60%. After two days of treatment the patient's condition improved. The patient is then allowed to go home with oral therapy Digoxin, Furosemid, Valsartan, Spironolactone. Patient complaints improved and never came back for follow-up.

The patient came to our emergency room complaint a *dyspneu*. Her vital sign, blood pressure was 120/70 mmHg, pulse was 120 bpm regular, RR was bpm, temperature was 36.2 C, and saturation was 99% with 3 lpm nasal oxygenation. On physical examination there was increased jugular venous pressure, wet ronkhi in both lungs, limb edema and ascites. ECG images have sinus tachycardia HR bpm, and Left Axis Deviation. X-ray shows cardiomegaly with pulmonary congestive and pleural effusion. Results of laboratory tests, Leukocytes 8.000 / uL, Hemoglobin levels 15.3 g / dl, Thrombocytes 203.000/ uL, Hematocrit 45.1%. Sodium electrolyte results of 147.8 meq / L, Potassium 3.5 meq / L, Chloride 108.7 meq / L, Albumin content 3.7 g / dl, blood urea 7.9 mg / dl, and creatinine clearance 0.78 mg / dl. The latest echocardiography result was all chamber dilatation, global segmental LV hypokinetic with LV ejection fraction of 32.68%. The patient had acute heart failure therapy with 3 lpm O2 nasal oxygenation, 0.9% NaCl IVFD 1000cc / 24 hours, and a urine catheter. Pharmacological therapy is given Furosemid pump 5 mg / hour, Nitrate 5 mg orally three times a day, and Candesartan orally 4 mg twice a day. First day of treatment the patient still complained of *dyspneu* but had begun to diminish, leg edema and ascites also diminished. Monitoring urine production of 650cc / 24 hours. For therapy, diuretics are replaced with intravenous diuretics three times a day. Second day there were no complaints of dyspneu, swelling in the legs and ascites. Monitoring of urine production of 450 cc / 2 hours, patients were given additional Carvedilol oral therapy of 6.25 mg at night. On the third day the patient was as stable and improved. Patients go home after three days of hospital treatment with Furosemid home therapy 40 mg in the morning, Candesartan 4 mg twice a day, Nitrate 5

mg three times a day, Spironolactone 25 mg in the morning, Carvedilol 6.25 mg at night.

3. Discussion

Definition

The commonly used defiition of PPCM from the National Heart and Blood Institute and the Office of Rare Diseases of the National Institute of Healthy are heart failure caused by left ventricular systolic dysfunction that occurs during late pregnancy and 5 months after postpartum without any other cause of heart failure and other previous illnesses. [(1)] The latest definition submitted from the working group on the PPCM of the Heart Failure Association of European Society of Cardiology is simpler by removing time constraints. Peripartum cardiomyopathy is an idiopathic cardiomyopathy which shows the presence of heart failure caused by left ventricular systolic dysfunction at the end of pregnancy or several months after delivery, where no other cause of heart failure is found. This is an exclusion criterion. The left ventricle can not be dilated but the ejection fraction almost always falls below 45%. [(2)]

Epidemiology

Incidence of PPCM is quite low at 0.1% in pregnancy, but the morbidity and mortality are quite high between 5-32%. In some women, clinical and echocardiography can improve until returning to normal conditions, but in some other women can develop into heart failure and sudden cardiac death. Incidence of peripartum cardiomyopathy varies, which has been reported in Haiti: 1 in 299 live births, 1 in 2229 live births in California and 1 in 4000 live births in the United States. This wide variation is due to geographical differences and reporting patterns, besides access to echocardiography causes overestimation of this case. Some risk factors that can cause pregnant women to experience PPCM include increased maternal age, multiparity and pregnancy with preeclampsia or gestational hypertension. [(3)] Demakins et al and Brar et al found that African American women were 2.9 times more affected by PPCM. compared to white women and 7 times more than Hispanic women. Greater incidence of hypertension in African-Americans may cause this. [(4,5)]

Etiology and Patophysiology

Peripartum cardiomyopathy is a form of idiopathic primary myocardial disease associated with pregnancy. Although several etiological mechanisms have been proposed but none of them are definite etiologies. [(6)] PPCM is distinguished from other cardiomyopathies because of the occurrence associated with pregnancy. Many etiological processes are thought to be the cause of PPCM, namely viral myocarditis, abnormal immune response related to pregnancy, maladaptive response to hemodynamic stress during pregnancy, stress-activated cytokines, excessive prolactin expression, prolonged use of tocolytic and malnutrition. The European Society of Cardiology classifies PPCM as a non-familial, non-genetic form of dilatative cardiomyopathy. [(3)]

Viral myocarditis is the main mechanism thought to be the cause of PPCM and was first reported by Goulet and Melvin, who found myocarditis in myocardial biopsy of 3 women with PPCM. However clinically indistinguishable between women with or without myocarditis. [(7)]

Abnormal immune response to fetal microchimerism (fetal cells in the maternal circulation) is thought to be one of the causes of PPCM. This theory is supported because during pregnancy fetal cells are released into the mother's bloodstream and there is no rejection reaction due to natural immune suppression that occurs during pregnancy. However, after delivery, when fetal cells attach to myocardial tissue a pathological autoimmune response can occur, causing PPCM in the mother after delivery. [(7)]

During pregnancy, blood volume and cardiac output increase. Vascular smooth muscles experience relaxation so afterload decreases. This causes reversible left ventricular hypertrophy to meet the needs of the mother and baby. Temporary left ventricular dysfunction during the third trimester and the beginning of the postpartum period, can return to normal after giving birth in a normal pregnancy. PPCM is thought to occur due to a drastic reduction in ventricular function when there is a change in hemodynamics during pregnancy. [(4)]

Increased plasma concentrations of inflammatory cytokines including tumor necrosis factor α (TNF-α; C-reactive protein (CRP); and Fas / Apo-1, plasma markers of apoptosis, have been identified in PPCM. Fas / Apo-1 levels, ligands found on the surface of protein cells that play a key role in apoptosis, is higher in women with PPCM compared to healthy women. [(3)]

Hilfiker-Kleiner et al found the mechanism of new pathogenesis in PPCM, namely an increase in prolactin production. The amount of prolactin is associated with increased blood volume, decreased blood pressure, decreased angiotensin response and decreased amount of water, sodium and potassium. Research shows that PPCM in mice has specific deletions in STAT3 in cardiomyocytes. In women with PPCM, the STAT3 protein level is low in the heart and the number of cathepin D serum and 16-kD prolactin increases. [(8)]

The use of prolonged tocolytic is the use of sympathomimetic agents for more than 4 weeks. Lamper et al found an association between the use of tocolytic therapy and the development of pulmonary edema in pregnant women and the relationship between the use of beta sympathomimetics and PPCM. [(9)]

Clinical Manifestation and Diagnosis

Changes in normal pregnancy such as increased blood volume, increased metabolic requirements, mild anemia, changes in vascular resistance associated with mild ventricular dilatation and increased cardiac output. All of these conditions that make PPCM onset can be easily masked and unknown, because of its similar. Women with PPCM are often found to be congested, tired during activity, dizziness, chest pain, cough, limb edema, increased jugular veins, pulmonary crackles, and heart sounds S3. Symptom severity varies from New York Association Functional Class I to IV. However classes III and IV are the most frequent. Life-threatening complications include refractory heart

failure, cardogenic shock, severe ventricular arrhythmias, multiorgan failure, thromboembolism, and death. [10]

In this case the patient came to the emergency room with complaints of dyspeu, swelling of the legs, fatigue easily when active and coughing and tightness if lying down. Patients come already in a state of increasingly severe heart failure which is found signs of left heart congestion and right heart. On physical examination there was increased jugular venous pressure, wet ronkhi in both lungs, limb edema and ascites. The patient came already in a state of heart failure class NYHA III-IV.

The diagnosis of PPCM is often late because PPCM symptoms are not specific, especially in late pregnancy. PPCM should be considered in peripartum women with signs and symptoms of heart failure or patients who are long recovering. Family history needs to be cultivated further to find out familial patterns in cardiomyopathy. Some non-specific electrocardiograms in PPCM show non-specific abnormalities, prolonged QT-intervals, QRS dilation, left ventricular hypertrophy and atrial fibrillation. Some laboratory tests are also important but not specific to PPCM. [11]

In this case, at early post-partum complaint of the patient was only mild *dyspneu*, and foot swelling was also considered a normal complaint in the postpartum mother. This makes the patient ignore the complaint. Awareness of signs and symptoms and adherence to therapy can affect the patient's prognosis. At the beginning of echocardiographic complaints showed LV dilatation, segmental LV normokinetic and 60% Ejection Fraction. Five months without therapy and monitoring of jatung patients in more severe conditions. Where can we find all chamber dilatation, global segmental LV hypokinetic with Ejection Fraction <32.68%.

Recent studies regarding biomarkers that may be specific to PPCM include 16-kDa prolactin, microRNA-146a, soluble-like tyrosine kinase-1 has been found, although the diagnostic value in clinical practice still requires verification. BNP or NT-pro BNP levels (N-terminal pro-brain natriuretic peptide). Plasma BNP or NT-pro BNP concentrations have high sensitivity to inclusion, and high specificity to exclude heart failure. Despite significant hemodynamic stress, BNP or NT pro-BNP does not increase during normal pregnancy. BNP and NT pro-BNP increased not significantly in preeclampsia while in PPCM it increased significantly. [11]

Chest X-rays should be performed on patients with suspected PPCM. This X-ray can help if there is pulmonary edema, cardiomegaly, pulmonary congestion and pleural effusion. Echocardiography is a non-invasive examination and a serial evaluation examination is needed in pregnant women. In women with PPCM consistently found heart failure, namely a decrease in ejection fraction, global dilatation, and thinning of the heart wall. [3] Echocardiographic criteria in PPCM namely LV end diastolic dimension <2.7 cm / m2 and M-mode Fractional shortening <30% or left ventricular ejection fraction <45%. Echocardiography can be used to exclude multiple differential diagnoses. [10] Cardiac magnetic resonance imaging (MRI) can be a complement to echocardiography, especially with suboptimal echocardiographic features.

Management

PPCM management is similar to other forms of heart failure. However, special attention is needed to the safety of women and the excretion of drugs or metabolites during labor until the process of breastfeeding after delivery. The main goal of managing heart failure is to improve hemodynamic status, reduce signs and symptoms, and optimize long-term outcomes. Reducing preload with vasodilators such as important nitrates which are mostly safe in pregnancy and breastfeeding. Diuretics are also important for reducing preload, but attention must be given to reducing intavascular volume, thereby reducing the blood supply to the uterus and also to the fetus. Management will focus on reducing preload and afterload and increasing inotropic. Management is generally divided into management when patients with compensated heart failure and acute heart failure. Ideal management is hidralazine, nitrate, digoxin, and diuretics. Angiotensin-converting enzyme inhibitors (ACE-i) are contraindicated during labor because of their teratogenicity but this therapy is the main PPCM therapy after delivery for decreased afterload. Aldosterone antagonists are effective when ACE is not tolerated but cannot be given during pregnancy. [3]

Beta adrenergic antagonists such as metoprolol and carvedilol can be given to PPCM and can increase life expectancy. Beta blockers should not be given in the early stages of PPCM because they can reduce perfusion in acute heart failure. Digoxin as an inotropic agent is also safe to use during pregnancy, is recommended for pregnant women with left ventricular systolic dysfunction and an ejection fraction of less than 40% with a failure. The following guidelines for compensated heart failure on PPCM. The following guidelines for the management of compensated heart failure in PPCM. [3]

Table 1: Management of peripartum cardiomyopathy with compensated heart failure[3]

Non Pharmaceutical Therapies Low-sodium diet: limit of 2 g sodium per day Fluid restriction: 2 L/day Light daily activity: if tolerated (eg, walking)
Antepartum Management of PPCM **Beta blocker** : Carvedilol, Metoprolol **Vasodilator**: Hydralazine **Digoksin**. **Thiazide diuretic**: Hidroklorotiazide
Postpartum Management of PPCM **Angiotensin-converting enzyme (ACE) inhibitor** Captopril, Enalapril, Ramipril, Lisinopril **Angiotensin-receptor blocker (bila ACE-I tidak mampu menoleransi)** Candesartan, Valsartan **Vasodilator** Hydralazine, Isosorbid Dinitrat **Aldosterone antagonist** Spironolacton, Eplerenone **Beta blocker**Carvedilol, Metoprolol **Warfarin** (jika fraksi ejeksi <35%)

Management of PPCM patients with acute heart failure starts from the airway, breathing and cycles. Assessment of the airway is very important because of excessive intravascular volume pregnancy. Respiratory needs to be assisted with additional oxygenation to reduce the signs and symptoms of hypoxemia and measure continuous oxygen saturation. Blood pressure monitoring and fetal monitoring need to be done. Pharmacologically can be given intravenous diuretics with attention, vasodilators and inotropic agents. Furosemide is given with consideration of creatinine clearance and nitroglycerin given according to clinical status and blood pressure.

In this case, patient had acute heart failure therapy with 3 lpm O2 nasal oxygenation, 0.9% NaCl IVFD 1000cc / 24 hours, and a urine catheter. Pharmacological therapy is given Furosemid pump 5 mg / hour, Nitrate 5 mg orally three times a day, and Candesartan orally 4 mg twice a day. First day of treatment the patient still complained of dyspneu but had begun to diminish, leg edema and ascites also diminished. Monitoring urine production of 650cc/24 hours. Furosemid pump was replaced with intravenous diuretics three times a day. The second day monitoring of urine production 450 cc/24 hours, patients were given additional Carvedilol oral therapy of 6.25 mg at night. Patients discharge after three days of hospitalized with take home therapy Furosemid 40 mg in the morning, Candesartan 4 mg twice a day, Nitrate 5 mg three times a day, Spironolactone 25 mg in the morning, Carvedilol 6.25 mg at night. This patient showed a good therapeutic response and clinical symptoms disappeared for 3 days of treatment.

Prognosis

The prognosis of PPCM is positively associated with improving ventricular function. Failure of heart size to return to normal size is associated with mortality and morbidity. Women with persistent ventricular dysfunction are more difficult to survive and return to normal heart function compared to women with increased left ventricular function. Sliwa et al said ejection fraction is a strong pedicler of outcomes in women with PPCM. [(12)] Abboud et al. reported 50% of women with PPCM returned to normal ventricular function within 6 months after giving birth. (13) Medical therapy according to the ACCF / AHA guidelines should continue if the heart function had not returned to normal. Early diagnosis and the latest therapy for heart failure have an important role in reducing PPCM mortality and morbidity. Attention must also be given to subsequent pregnancies in patients with improved PPCM. Improvement of left ventricular function is a predictor for subsequent pregnancy. [(14)]

Resume

PPCM occurs in women at the end of pregnancy or a few months after giving birth, which previously had no other heart disease. Changes during normal pregnancy are almost the same as the initial onset of PPCM signs and symptoms so that the diagnosis becomes late. The patient can ignore the initial complaint until the complaint becomes more severe. As in these patients, the initial mild complaint with ejection fraction was still good at 60%, in five months the complaint became more severe until the ejection fraction <32.68%. Patients come already in a state of acute heart failure, both left and right heart congestive. The prognosis of PPCM is positively associated with improving ventricular function. Women with persistent ventricular dysfunction are more difficult to survive and return to normal heart function compared to women with increased left ventricular function. Early diagnosis, serial examination, and the latest therapy for heart failure are expected to play an important role in reducing PPCM mortality and morbidity.

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