A 28-YEAR-OLD SLE MALE PATIENT WITH RIGHT HEART FAILURE DUE TO PULMONARY HYPERTENSION

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ABSTRACT

Right Heart Failure (RHF) is a quickly progressive syndrome with systemic congestion with impaired Right Ventricular (RV) filling and/or decreased RV outflow output. Pulmonary hypertension (PH) is most common cause of RHF. PH will increase the afterload of RV and leads to the clinical syndrome of RHF with systemic congestion and inability to adapt the output of right ventricle to peripheral demand at exercise. A 28-year-old male patient, a photographer, lived in North Aceh was admitted to the ED of Cut Meutia Hospital. He was admitted with worsening dyspnea since 2 days before, initiated after a period of fever. He also complained of fatigue, palpitations in ordinary activities and chest pain occasionally. He had history of presyncope and syncope. He denied any history of hypertension, type 2 diabetes, allergies, and active smoking. He had no congenital or family history of heart disease. Physical examination revealed full of conciousness with slightly abnormal vital sign. He had malar rash, dilated jugular vein. The apex of the heart shifts to the axilla anterior line, S1>S2 with systolic murmur (+). ECG showed complete RBBB with RAD and chest x-ray showed cardiomegaly. The transthoracic echocardiogram revealed severe TR (Tricuspid Regurgitation), dilatation of RARV (Right Atrium Right Ventricle), with severe PH (Pulmonary Hypertension). The patient was diagnosed with RHF (Right Heart Failure) caused by primary PH. The patient was placed on intravenous furosemide, spironolacton, digoxin, and sildenafil. Treatment and lifestyle modification were expected to increase the quality of life.

Keywords: pulmonary hypertension; right heart failure; quality of life, SLE

INTRODUCTION

The American Heart Association/American College of Cardiologists (AHA/ACC) provides Heart Failure (HF) as a complex clinical syndrome in the setting of functional or structural heart disorders impairing ventricular ability to fill the ejection blood.

The Heart Failure Association (HFA) on ESC guidelines defines acute RHF is quickly progressive syndrome with systemic congestion with impaired filling of RV and/or decreased the outflow output of RV. Mostly, RV failure impact to RV chamber dilatation and tricuspid regurgitation so that will increase RV afterload or preload. In the end, the basis definitions RHF is unstable hemodynamics function of cardiac and the inadequacy of pulmonary circulation.

The prevalence of RHF has significant variation on graphically. Best on ESC registry of HF, RHF incidence around 4.5% of patients with acute HF in USA and around 3% in other ESC regions. Broad categories of causes RHF can be stratified into: pulmonary hypertension (PH), diseases of the pericardium and RV/valve pathology. PH is commonest cause of RHF in global, with prevalence around 1% up to 10% in individuals with >65 years old. PH is defined by a mean pulmonary artery pressure (mPAP) ≥25 mm Hg at rest by right heart catheterization. The high mPAP is associated with pulmonary circulation structural alterations. The crucial role in RV adaptation are preload, afterload, and contractility. RV adaptation in PH represents a maladapted of ventricle at one end and an adapted of right ventricle at the other end. A maladapted of right ventricle is characterised by dilated RV with reduced SV, increased filling pressures and systolic function, whereas an adapted of RV is characterised by slightly dilated RV with preserved stroke volume (SV), normal filling pressures, and systolic function.

RVF is cause of all deaths accounts around 20%, secondary to CHF. Patients with acquired heart diseases affect by RVF because it enforce an independent effect on the prognosis of obstructive pulmonary diseases and RV hemodynamic function, correlates with rate of mortality in patients with primary PH. Main determinant of clinical outcomes in various forms of CHDs, which subjects the RV to abnormal loading conditions. RHF is progressively being recognized as a major challenge in cardiovascular problem.

Symptoms of RHF are mainly due to systemic venous congestion and/or low cardiac output. This includes exertional dyspnoea, dizziness,
fatigue, epigastric fullness, ankle swelling, and right upper abdominal discomfort or pain. Signs: an accentuated RV gallop, usually a pansystolic murmur over the tricuspid area that increased on inspiration, an enlarged tender liver, second pulmonary sound, increased jugular venous pulse (JVP), frequently present as well as ankle edema, ascites. Kussmaul's sign and increase of JVP on inspiration, can help in pointing to the cause of RHF.²,⁷

Echocardiography are important to know the causes of RV failure and known the guide treatment beside other imaging modalities. In patients presenting with severe RVF, treatment with rapidly initiation is the best way to restore hemodynamic stability and essential to prevent potentially irreversible end-organ damage. Acute treatment consists with four elements: improvement of myocardial contractility; volume optimization; restoration of perfusion pressure; and advanced options.⁷,⁸

CASE REPORT

A 28-year-old male patient, a photographer, lived in Blang Jrum, Tanah luas, North Aceh, Indonesia, was admitted to the emergency department of Cut Meutia Hospital by his family at 03.00 pm on May 30th, 2022. He was admitted with worsening dyspnea since 2 days before, initiated after a period of fever. The dyspnea was not improved with rest. He had several episodes of this symptom since 2015. The Dyspnea is not triggered by cold weather or eating certain foods. He also complained of fatigue, palpitations in ordinary activities and chest pain occasionally. He had history of presyncope and syncope. No evidence of central cyanosis before. He denied any history of hypertension, type 2 diabetes, allergies, and active smoking. He had no congenital or family history of heart disease, but had been hospitalized before and routinely visits the cardiologist since 2018.

Physical examination revealed full of consciousness with slightly abnormal vital sign; BP 110/60 mmHg , HR 115 bpm, RR 28 bpm, temperature 38.1°C, SpO2 94% on room air. He had malar rash. Thorax examination showed no chest wall deformities, symmetrical chest movement, but jugular vein was dilated. Tactile fremitus was equal bilaterally, resonant and vesicular sound over peripheral fields (no adventitious sound). The apex of the heart shifts to the axilla anterior line, S1>S2 with systolic murmur (+). No sign of hepatomegali or splenomegali

Laboratorial evaluation on May 30th and 31st presented normal CBC , normal bloodsugar level, normal RFT, increase LFT ; SGOT 52 U/L, SGPT 62 U/L, bilirubin direct 1,09 mg/dL, bilirubin indirect 4,49 mg/dL, alkaline phosphatase 269 U/L. CRP and ASTO were negative.

Figure 1. ECG Findings

12-lead electrocardiogram obtained at admission showed, ventricular rate ~95 bpm, RAD, QRS duration >12s, RSR’ pattern in V1-V3 (M-shaped) with discordant T wave changes, slurred S wave in laterala lead (I, aVL). The conclusion was complete RBBB with RAD .

Figure 2. Chest x-ray

Interpretation : Cardiomegaly with dilated pulmonary artery

The transthoracic echocardiogram revealed severe TR (Tricuspid Regurgitation), dilatation of RARV (Righ Atrium Righ Ventine), with severe PH (Pulmonary Hypertension)
The patient was diagnosed with RHF (Right Heart Failure) et causa PH primer. During hospitalization, he got O2 via nasal canul 3-4Lpm, furosemide injection/12h, spironolactone 1x25mg, Digoxin 1x0,25mg, Sildenafil 3x50mg, Lansoprazole 2x30 mg, domperidone 3x10mg, Sucralfat 3xCth II.

**DISCUSSION**

RV dysfunction (RVD) is an abnormality of RV structure or function, results in poor clinical outcomes regardless of the underlying mechanism of disease: across the spectrum of LVEF in patients with acute and chronic heart failure, post cardiac surgery, acute myocardial infarction, congenital heart disease, and PH. RHF is a clinical syndrome with HF signs and symptoms that results from RVD. RHF is arising from RV inability to support optimal circulation despite of adequate preload.1

PH is recently being the commonest cause of RHF. In the vast majority of the cases, pulmonary hypertension occurs when the LV systolic dysfunction be the cause of LV failure, and also in the setting of LV left-sided valvular heart disease, or diastolic dysfunction. CHD may be the cause of RV failure because it can increase afterload, overload, or both. Septal defects are the most common CHD that cause RV failure since the RV being volume overload because of left-to-right blood shunting.19,10,11

He was admitted with worsening dyspnea two days before, initiated after a periode of fever. The dyspnea was not improved with rest. He had several episodes of this symptom since 2015. He also complained of fatigue, palpitations in ordinary activities and chest pain occasionally. He had a history of presyncpe and syncpe. No evidence of central cyanosis before. He had no congenital or family history of heart disease, but had been hospitalized before and routinely visits the cardiologist since 2018.

Increased of PVR and mPAP ≥ 20 mmHg progressively due to obstructive modifications in the pulmonary vasculature. Initial vasoconstriction of the pulmonary vasculature leads to increased in the size and function of muscles of peripheral arteries and hypertrophy of arteries muscles. This situation might be affect to the right ventricular function and pump.12 The ordinary RV function is relating among preload, contractility, afterload, ventricular coupling and heart rhythm. The main pathophysiologic mechanism for RV failure is increased afterload of pulmonary and cardiac origin.13,14

Prolonged isovolumic contraction may be caused by RV afterload which can lead to increased myocardial wall stress. RV hypertrophy and displacement of the interventricular septum toward the left ventricle are the result of increased myocardial wall pressure. Eventually lead to left ventricular failure and ultimately heart failure.

Acute increase of RV afterload will increase wall tension which results in dilatation of the RV chamber and impaired systolic and diastolic function. RV dilatation due to increased RV afterload can cause tricuspid regurgitation. This condition leading to decreases of RV cardiac output and LV preload. Increased RV wall tension accompanied by decreased of systemic cardiac output and perfusion pressures alters the balance between myocardial oxygen supply and demand, resulting in ischemia and infarction. Increased of RV afterload in patients with cardiac problems may present with dyspnea, dizziness, fatigue, and syncope. Patients with low cardiac output may present with tachycardia.11,14

There is many kind of dyspnea like orthopnea and paroxysmal nocturnal dyspnea (PND). Orthopnea is a condition of difficulty breathing in the lying position, relieved by sitting or standing. PND is a condition of breathlessness that wakes the patient from sleep which generally occurs after 1-2 hour of sleep and usually relieved in upright position. There is redistribution of blood volume from the lower limbs and splanchnic beds to the lungs in the supine position.15

Congestion of pulmonary during supine position leading to orthopnea. There is a significant decrease in vital capacity and lung compliance due to orthopnea in patients whose additional volume can’t be pumped out by the LV. Furthermore, in patients with CHF, circulation of the pulmonary may be...
overloaded, and excess fluid may be reabsorbed from dependent parts of the body. PND and orthopnea may be caused by similar mechanism. The failing LV is suddenly not able to match the output of a normal function of RV; this consequences in congestion of the pulmonary. Decreased response of the respiratory center and decreased adrenergic activity in the myocardium during sleep may be additional mechanisms in PND.\textsuperscript{15}

The patient skin is so sensitive with sunlight, and we found malar rash and a lump on Patient finger joint of the left hand. This symptoms lead to SLE and RHD. Systemic autoimmune lupus erythematosus (SLE) is a systemic autoimmune disease with multisystemic involvement. The clinical features of SLE may vary from mild symptoms with only mucocutaneous involvement to severe life-threatening symptoms with multiorgan disorders. The most common clinical sign in patients with SLE is mucocutaneous involvement with incidence rate more than 80%. Malar rash or butterfly rash are typical skin lesions in SLE. Rheumatic heart disease (RHD) is the result of valvular damage due to abnormal reaction of immune system to Streptococcus pyogenes infection or group A streptococcus that causes acute rheumatic fever. In the end the differential diagnose can be ruled out by laboratory result.\textsuperscript{16,17}

Physical examination revealed dilated jugular vein. The apex of the heart shifts to the axilla anterior line, S1>S2 with systolic murmur (+). No sign of hepatomegali or splenomegali. One way to measure the vena cava pressure is to measure the jugular venous pressure. One sign of failing right is an increase in JVP. In right heart failure will cause blockage of blood in the right ventricle and will also continue to blockage in the right atrium and vena cava, so it will show an increase from the jugular vein. An increase in JVP will be seen with jugular venous distention, where the JVP will appear at neck level, much higher than the neck.\textsuperscript{18}

The apex shifts to the later, indicating that the apex of the heart (the top of the heart, which is at the bottom left of the heart) is shifted to the side. This indicates an enlarged heart so that the top of the heart becomes displaced.\textsuperscript{19}

Laboratory evaluation on May 30\textsuperscript{th} and 31\textsuperscript{st} presented normal CBC, normal bloodsugar level, normal RFT, increase LFT; SGOT 52 U/L, SGPT 62 U/L, bilirubin direct 1.09 mg/dL, bilirubin indirect 4.49 mg/dL, alkaline phosphate 269 U/L. CRP and ASTO were negative. Blood tests are recommended to rule out anemia. RFT to assess renal function before starting therapy. One of the signs of heart failure is impaired systemic perfusion and is usually also caused by failure of the pumping function of the heart. The main pathophysiology underlying impaired liver function is passive blockage from a low increase in cardiac output and due to failure of perfusion. Decreased cardiac output results in less than optimal blood perfusion to the liver resulting in hypoxia which, if chronic, can result in atrophy of hepatocyte cells. This situation results in increased levels of liver enzymes (SGOT, SGPT), and can cause hemodynamic abnormalities, coagulation disorders, impaired liver function and impaired albumin synthesis function.\textsuperscript{20}

CRP is elevated in acute rheumatic fever, Rheumatoid arthritis, Acute Myocardial Infarction, Postoperative infections, Bacterial infections, Viral infections, Crohn's disease, Reiter's syndrome, Vasculitis syndrome, Lupus Erythematosus, Tissue necrosis or trauma.\textsuperscript{1,5} Drugs that can lower CRP levels such as colchicines and statins. In clinical applications, CRP is a predictor of cardiovascular events, particularly coronary heart disease and is stronger than LDL. CRP levels indicate the presence of atherosclerosis which is a risk factor for CHD. This patient had a negative result, which means that there was no agglutination during the CRP examination.\textsuperscript{21,22}

One of the tests to detect group A hemolytic streptococcal infection is to use anti-streptolysin O (ASTO). ASTO is one of the antibodies that are often tested to establish the diagnosis of rheumatic heart disease, ASTO can also be useful in diagnosing streptococcal infection and its complications in evaluating effective treatment. If there is agglutination, then positive (+) Streptolysin ASTO, but if there is no agglutination, negative (-) Streptolysin ASTO. This patient had a negative result, which means there was no agglutination during the ASTO examination.\textsuperscript{23}

12-lead electrocardiogram obtained at admission showed, complete RBBB with RAD. An ECG should be performed in all patients with clinical suspicion of heart failure. Pulmonary hypertension (PH) is the most common of RHF. Abnormalities on the ECG are often found in severe degrees of PH. However, the absence of abnormalities on the ECG does not mean the diagnosis of PH has been ruled out. ECG abnormalities that can be found include P
pulmonary, right axis deviation, right ventricular hypertrophy, right ventricular stretch, right bundle branch block, and QTc prolongation. Strains in the right ventricle have the highest sensitivity rates. Prolongation of the QTc and widening of the QRS complex indicate disease severity. Supraventricular arrhythmias such as atrial flutter and atrial fibrillation can be found at an advanced stage. RAD or right axis deviation is a shift in the axis of the heart that deviates to the right. This condition often occurs when the right heart chamber enlarges, such as pulmonary embolism, obstructive pulmonary disease, heart muscle conditions, heart failure and other conditions. RVH defined by an abnormal enlargement of the cardiac muscle surrounding of the RV. It is found towards the lower-end of the heart and it gets blood from the proper chamber and pumps blood to the lungs. Since RVH is an broadening of muscle it emerges when the muscle is required to work harder. Subsequently, the most causes of RVH are pathologies of frameworks related to the correct atrium such as the pulmonary supply route, the tricuspid valve or the aviation routes. 

Chest X-Ray showed Cardiomegaly with pulmonary artery dilatation. The chest X-ray often shows cardiomegaly (cardiothoracic ratio (CTR) > 50%), especially if the heart failure is chronic. Cardiomegaly can be caused by left or right ventricular dilatation, LVH, or occasionally a pericardial effusion. The degree of cardiomegaly is not correlated with left ventricular function.

Echocardiogram revealed severe TR, dilatation of RARV, with severe PH (Pulmonary Hypertension). Echocardiographic examination is not only performed to assess estimated pulmonary artery systolic pressure, but also can assess signs of RV and LV dysfunction. The probability of PH based on transthoracic echocardiography using the combination of several parameters, where the TR velocity (TRVmax) is the main variable. Other parameters such as RV size, interventricular septal function, fluctuations of the inferior vena cava (IVC) on the respiratory cycle, systolic area of the right atrial, pattern of systolic flow velocity against early diastolic pulmonary regurgitant velocity (PVAcct), and diameter of the pulmonary arteries.

<table>
<thead>
<tr>
<th>TR V Max (m/s)</th>
<th>PH sign (minimum 2)</th>
<th>Probability</th>
</tr>
</thead>
<tbody>
<tr>
<td>≤2.8/immeasurable</td>
<td>there is not</td>
<td>Low</td>
</tr>
<tr>
<td>≤2.8/immeasurable</td>
<td>there is</td>
<td>Currently</td>
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<tr>
<td>2.9 – 3.4</td>
<td>there is not</td>
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<tr>
<td>2.9 – 3.4</td>
<td>there is</td>
<td></td>
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<tr>
<td>&gt;3.4</td>
<td>there is</td>
<td>High</td>
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Echocardiographic signs of suspected PH are used to assess the probability of PH as an additional examination. This patient had tricuspid regurgitation. Tricuspid regurgitation is a disorder of the closure of the tricuspid valve. The valves open when the atria contract to pump blood into the ventricles, and close when the ventricles contraction to prevent blood from the flow back into atrial. If the tricuspid valve is open, it does not close tightly allowing blood to flow back into the atria. When blood flows back, the right atrium becomes more active and enlarges.

The patient was diagnosed with RHF (Righ Heart Failure) et causa PH primer. Recommend of the Guideline about risk stratifying by functional class. Treatment should be goal oriented order to achieve low-risk status for the patient. Phosphodiesterase type 5 (PDE-5) inhibitors such as sildenafil have been shown to improve symptoms and hemodynamic parameters of patients with PAH. The rapid onset (15 minutes), peak effects (60 minutes) and a 4-6 hour onset of action of Sildenafil increase of cardiac output and RV contractility, decrease of mPAP and PVR. Sildenafil recommendations according to WHO functional class II-III is IA.

Diuretics are used in volume management in patients with PAH is the several classes. This patient uses a combination of furosemide and spironolactone. Loop diuretics (furosemide) are most commonly used because they are effective in inhibiting reabsorption of sodium. Spironolactone is a commonly used aldosterone antagonist diuretic and is often prescribed in combination with loop diuretics to achieve improved diuresis. Digoxin has been shown to improve cardiac output or RV ejection fraction acutely in idiopathic PAH and RVD.
Right heart failure is the largest contributor to morbidity and mortality in patients with right heart failure, such as pulmonary hypertension. \(^{27}\) Tricuspid regurgitation as in this patient is caused by right ventricular pressure overload. \(^{32}\) Because the presence of signs of right heart failure, progression of symptoms and presence of syncope, estimated 1-year mortality exceeds 10% in this patient. \(^{30}\)

If at the time of examination found hemodynamically stable, then therapy can be in the form of medication and can be considered sports exercises such as walking, cycling and treadmill. \(^{33}\) There is no limit to the style, frequency, intensity and duration of the exercise. All types of vaccines can be used for patients such as COVID-19, Pneumococcal and influenza vaccines. Perhaps psychosocial support, genetic counseling and medication for travel can be considered. \(^{34}\) Organ transplantation in patients with refractory chronic RHF may be considered if the cause of chronic RHF is reversible and comorbidity assessments have been ruled out. \(^{1}\) A shortage of donor organs can increase patient mortality while waiting for a scheduled transplant. \(^{30}\) Unfortunately, lung transplants are not available in Indonesia. In terms of diagnosis, the most important autoantibody to detect is ANA (antinuclear antibody). This test is used to check for the presence of certain antibodies in the blood which most people with SLE have. About 98% of people with lupus have a positive result on the ANA test, making it the most sensitive method of confirming the diagnosis. In addition to treatment with drugs, patients need to be educated to make some lifestyle changes to minimize recurrence and improve quality of life. Some important things that need to be conveyed are: avoiding excessive physical activity, avoiding smoking, avoiding changes in weather due to the inflammatory process, avoiding stress and physical trauma, avoiding direct sun exposure, especially at 10:00 to 15:00, wearing closed clothing and sunscreen SPV30PA+++ minimum 30 minutes before leaving the house, avoid exposure to UV lights, avoid using contraceptive drugs or other drugs that contain estrogen hormone occasionally. deThe patient was diagnosed with RHF (Right Heart Failure) et causa PH primer.

REFERENCES
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