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The Relationship Between Modified Rodnan Skin Score (mRSS) With Pulmonal Hypertension and Lung Fibrosis in Systemic Sclerosis Patients

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ABSTRACT

Background: Systemic sclerosis (SSc) is a connective tissue disease, which affects the skin, blood vessels, heart, lungs, kidneys, gastrointestinal tract and musculoskeletal system. The manifestations in blood vessels include pulmonary hypertension which can be seen from echocardiography, while the manifestations to the lungs in the form of pulmonary fibrosis are examined by high-resolution computed tomography (HRCT). Modified Rodnan Skin Score (mRSS) a method for measuring skin thickness in SSc patients. **Objective:** This study aims to determine the relationship between mRSS and pulmonary hypertension and pulmonary fibrosis in SSc patients. **Methods and Materials:** Research with a cross sectional approach at Dr Kariadi Hospital. There were 23 study subjects with SSc patients having their mRSS measured by a rheumatologist and an HRCT examination by a radiologist to see the percent of lung damage and echocardiographic examination by a cardiologist to measure the Tricuspid Regurgitation Velocity Maximum (TRV Max) as a marker of pulmonary hypertension. Normality test using Saphiro-Wilk. The Spearman rank correlation test was used to analyze the relationship between the mRSS score, pulmonary hypertension and pulmonary fibrosis. **Results:** The proportion of study subjects with a mean age of 40.39 years, women, and 4.43 years of illness. The mean mRSS score was 17.43. Examination of pulmonary hypertension from echocardiography found most of the light category 95.6%, heavy category 4.4%, while the HRCT examination found pulmonary fibrosis of 86.9%, normal 13.1%. There are significant relationship between mRSS and pulmonary fibrosis ($r = 0.485$, $p = 0.019$), and significant relationship between pulmonary hypertension ($r = 0.63$, $p = 0.001$). **Conclusion:** There is a significant relationship between mRSS, pulmonary fibrosis and pulmonary hypertension.

1. Introduction

Systemic sclerosis (SSc) is an autoimmune disease characterized by microvascular damage, resulting from innate and adaptive immune dysregulation, and fibrosis of multiple organs. Pathological changes in the lungs, gastrointestinal tract, kidneys and heart determine the clinical

outcome. The degree of involvement of the skin and the rate at which it develops reflect the severity of complications of the visceral organs.¹

The prevalence of SSc in the United States is 276 to 300 cases per million and the incidence is about 20 cases per million per year. Women are generally

affected more than men and tend to be more severe. Systemic sclerosis has the highest mortality rate among connective tissue diseases with a 10-year survival of 55% risk for pulmonary hypertension (PH) with disease onset in the elderly (> 65 years).²

Pulmonary hypertension (PH) and pulmonary fibrosis are diseases that can increase the mortality rate for patients with SSc. Modified Rodnan total Skin Score (mRSS) is one of the reliable methods for measuring skin thickness in SSc patients. mRSS is a semi quantitative method that is often associated with lung damage.^{3,4} The aim of this study was to prove the relationship between the high mRSS of SSc patients with the incidence of pulmonary hypertension and the degree of pulmonary fibrosis.

2. Method

This cross sectional study involved 23 SSc patients at Dr Kariadi Hospital who underwent mRSS examination by rheumatologists and performed HRCT examination by radiologists to see the percent of lung damage and echocardiography examination to measure TRV Max as a marker of pulmonary hypertension. Normality test using Shapiro-Wilk. The Spearman rank correlation test was used to analyze the relationship between the mRSS score and pulmonary hypertension and pulmonary fibrosis.

3. Result

The proportion of study subjects with a mean age of 40.39 years, mostly women (95.7%), length of illness was 4.43 years. The mean mRSS score was 17.43. Examination of pulmonary hypertension from echocardiography found most of the light category 95.6%, the heavy category 4.4%, while the HRCT examination found pulmonary fibrosis of 86.9%, normal 13.1%. There is a significant relationship between mRSS and pulmonary fibrosis ($r = 0.485$, $p = 0.019$), and a significant relationship between pulmonary hypertension ($r = 0.63$, $p = 0.001$)

4. Discussion

This study shows that there is a significant relationship between mRSS and the incidence of

pulmonary hypertension. Similar findings were reported by Kumar et al studied the prevalence of pulmonary hypertension in SSc ($n = 100$) and found 32% had pulmonary hypertension on echocardiography.⁵ Patients with SSc-PH tend to have other organ involvement such as kidney, left heart failure, diastolic dysfunction and pericardial effusion. The mechanisms of PH in SSc patients are mainly intimal proliferation, medial hyperplasia, and adventitial fibrosis leading to obliterative vasculopathy.⁶

This study also showed a significant relationship between mRSS and pulmonary fibrosis. Hafez et al found that 25 of 30 samples (83%) of pulmonary fibrosis from HRCT examination were higher than samples without pulmonary fibrosis manifestations.⁷ The close association between skin sclerosis and pulmonary fibrosis in SSc patients is demonstrated by several aspects of clinical experience. Skin sclerosis and pulmonary SSc-fibrosis develop in the early years in the course of SSc disease. The pathohistological features of skin and pulmonary involvement in SSc patients are very similar, inflammatory cell invasion is seen in the early stages, and the proliferation and degeneration of collagen fibers in the late stages.^{8,9}

This study has several limitations. First, it is only done in one hospital, so the results cannot be generalized to other hospitals. Second, a future prospective cohort study design to look at the outcome of therapy and the development of complications from sclerosis to the visceral organs. Third, mRSS, echocardiography and HRCT examinations were not carried out in close proximity so that it could cause bias, the fourth right heart catheterization examination as the gold standard to ensure pulmonary hypertension could not be done in this study.

5. Conclusion

There is a significant correlation between the mRSS score, pulmonary hypertension and the degree of lung damage.

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