Faktor Prognostik Progresivitas Penyakit Paru Interstisial terkait Sklerosis Sistemik Berdasarkan High Resolution Computed Tomography Toraks dan Modified Rodnan Skin Score = Prognostic Factors for Progression of Systemic Sclerosis-associated Interstitial Lung Disease Based on Chest High Resolution Computed Tomography and Modified Rodnan Skin Score

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Abstrak

Latar Belakang: Penyakit paru interstisial (ILD) merupakan salah satu manifestasi sklerosis sistemik (SSc) pada paru dan faktor mortalitas utama SSc. SSc-ILD meningkatkan angka mortalitas 5 tahun pasien SSc sebesar 3 kali lipat. Hampir dua pertiga pasien SSc-ILD dengan kelainan minimal pada high resolution computed tomography (HRCT) toraks memperlihatkan progresivitas signifikan dalam 2 tahun. Model prediksi progresivitas SSc-ILD yang tersedia, yakni GAP (gender, age, and lung physiology) dan SADL (smoking history, age, and diffusion capacity of the lung), terbukti memiliki nilai prognostik yang baik. Model prognostik yang melibatkan parameter HRCT toraks dan Modified Rodnan Skin Score (mRSS) diharapkan dapat membantu seleksi pasien SSc-ILD yang memerlukan pemantauan ketat atau terapi dini untuk mencegah progresivitas.
Metode: Studi ini melibatkan pasien SSc-ILD yang menjalani pemeriksaan HRCT toraks awal dan evaluasi di Rumah Sakit Umum Pusat Nasional Dokter Cipto Mangunkusumo pada periode Januari 2016 hingga Desember 2021. Dilakukan volumetri kuantitatif menggunakan piranti lunak 3DSlicer® pada HRCT toraks awal untuk menghasilkan persentase volume paru abnormal, high attenuation area (HAA), dan low attenuation area (LAA) yang selanjutnya dianalisa sebagai faktor prognostik. Pola ILD pada HRCT toraks awal dan nilai mRSS masing-masing subyek diidentifikasi dan dianalisa sebagai faktor prognostik progresivitas SSc-ILD. Progresivitas SSc-ILD dikategorikan menjadi progresif dan non-progresif berdasarkan selisih persentase volume paru abnormal antara HRCT toraks awal dan evaluasi.
Hasil: Perbedaan rerata yang bermakna ditemukan pada volume paru abnormal, volume HAA, dan volume LAA, nilai mRSS antara SSc-ILD progresif dan non-progresif ($p < 0,001$). Berdasarkan receiver operating characteristic curve, ditetapkan nilai titik potong dari masing-masing variabel. Nilai titik potong persentase volume paru abnormal ditetapkan sebesar 32,82% dengan nilai sensitivitas 100% dan spesifisitas 93,8%. Nilai titik potong persentase volume HAA ditetapkan sebesar 19,76% dengan nilai sensitivitas 93,8% dan spesifisitas 93,8%. Nilai titik potong persentase volume LAA ditetapkan sebesar 9,89% dengan nilai sensitivitas 62,5% dan spesifisitas 62,5%. Nilai titik potong mRSS ditetapkan sebesar 18,5 dengan sensitivitas 93,8% dan spesifisitas 100%. Tidak ada perbedaan proporsi pola ILD antara kedua kelompok tersebut (p 0,220).
Kesimpulan: Volume paru abnormal > 32,82%, volume HAA > 19,76%, volume LAA > 9,89%, dan/atau nilai mRSS > 18,5 merupakan prediktor progresivitas SSc-ILD. Hasil volumetri kuantitatif abnormalitas paru pada HRCT toraks dan nilai mRSS merupakan faktor prognostik progresivitas SSc-ILD yang mudah diperoleh dan diaplikasikan dalam praktik klinis sehari-hari.
.....Background: Interstitial pulmonary disease (ILD) is one of the manifestations of systemic sclerosis (SSc)

in the lungs and the main mortality factor of SSc. SSc-ILD multiplies the 5-year mortality rate of SSc patients by 3 times. Nearly two-thirds of SSc-ILD patients with minimal abnormalities in chest high resolution computed tomography (HRCT) showed significant progressivity within 2 years. The available prediction models of SSc-ILD progression, namely GAP (gender, age, and lung physiology) and SADL (smoking history, age, and diffusion capacity of the lungs), have been proven to demonstrate excellent prognostic values. Prognostic models involving chest HRCT parameters and Modified Rodnan Skin Score (mRSS) are expected to aid the selection of SSc-ILD patients who require close monitoring or early therapy to prevent progression.
Method: This study involved SSc-ILD patients who underwent initial and follow-up chest HRCT examination and evaluation at the National Central General Hospital of Doctor Cipto Mangunkusumo in the period from January 2016 to December 2021. Quantitative volumetric measurement was performed using 3DSlicer® software on the initial chest HRCT to yield abnormal pulmonary volume, high attenuation area (HAA) volume, and low attenuation area (LAA) volume percentage which were subsequently analyzed as prognostic factors. ILD patterns in the initial chest HRCT and mRSS values of each subject were identified and analyzed as prognostic factors of SSc-ILD progression. The progression of SSc-ILD is classified into progressive and non-progressive based on the abnormal pulmonary volume percentage difference between the initial and follow-up chest HRCT.
Result: Significant mean differences were found in abnormal lung volume percentage, HAA volume percentage, LAA volume percentage, and mRSS values between progressive and non-progressive SSc-ILD groups ($p < 0.001$). Based on the receiver operating characteristic curve, the cut-off point value of each variable is determined. The cut-off point value of the percentage of abnormal pulmonary volume was set at 32.82% with a sensitivity value of 100% and a specificity of 93.8%. The cut point value of the HAA volume percentage was set at 19.76% with a sensitivity value of 93.8% and a specificity of 93.8%. The LAA volume percentage cut point value was set at 9.89% with a sensitivity value of 62.5% and a specificity of 62.5%. The mRSS cut-off value was set at 18.5 with a sensitivity of 93.8% and a specificity of 100%. There was no significant in the proportion of ILD patterns between the two groups (p 0.220).
Conclusion: Abnormal lung volume > 32.82%, HAA volume > 19.76%, LAA volume > 9.89%, and/or mRSS value > 18.5 are predictors of SSc-ILD progression. Quantitative volumetric results of pulmonary abnormalities in chest HRCT and mRSS values are prognostic factors of SSc-ILD progression that are easily obtained and applied in daily clinical practice.