

Heart Involvement in Systemic Lupus Erythematosus

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Abstract

Pericardial effusion is considered as one of criteria to diagnose systemic lupus erythematosus (SLE) based on American Rheumatism Association (ARA) criteria. There is limited data about the incidence and characteristic pericardial effusion in SLE patients in our country. The aim of this study is to report and assess the incidence of and characteristic heart involvement in SLE. This is cross-sectional study conducted in secondary referral hospital in Tangerang county, west part of Jakarta, capital city of Indonesia. We evaluated from medical record and echocardiogram data from 2013-2015 patients diagnosed with SLE according to ARA criteria. From 33 SLE medical records reviewed, we found 13 patients (40%) with pericardial effusions. All patients were in active stage. Clinical assessment and transthoracic echocardiogram were used to diagnose pericardial effusions. Eighty percent pericardial effusion positive patients had minimal effusion. The others had moderate effusion. No tamponade patients were recorded. Ninety six percent patients were female with median age 24(13-51) years old. Three patients were reported having pulmonary arterial hypertension. One patient had thrombus in left ventricle. All patients had clinical symptoms of cardiac such as heart failure and chest pain. For the conclusion, the incidence of pericardial effusion in SLE patient was 40 percent. Eighty percent patients had minimal effusion. All patients had cardiac symptoms related.

Keywords: *pericardial effusion, systemic lupus erythematosus*

Abstrak

Efusi pericardium merupakan salah satu kriteria untuk mendiagnosis lupus eritematosus sistemik (LES) berdasarkan kriteria *American Rheumatism Association* (ARA). Hingga saat ini belum banyak data terkait insiden dan karakteristik efusi perikard pada pasien LES di negara kami. Tujuan dari studi ini adalah melaporkan dan menilai insiden dan karakteristik keterlibatan ginjal pada LES. Studi ini adalah potong lintang yang dilakukan di rumah sakit umum tipe B di daerah Tangerang, sebelah barat Jakarta, ibukota Indonesia. Kami mengevaluasi rekam medis dan data ekokardiogram pasien dari 2013-2015 yang terdiagnosis LES berdasarkan kriteria ARA. Dari 33 rekam medis SLE yang dievaluasi, ditemukan 13 pasien (40%) dengan efusi pericardium. Semua pasien pada stadium aktif. Penilaian klinis dan ekokardiogram transtorakal digunakan untuk mendiagnosis efusi perikard. Delapan puluh persen pasien dengan efusi pericardium memiliki efusi minimal. Sisanya memiliki efusi moderat. Tidak ada pasien dengan tamponade yang ditemukan. Sembilan puluh enam persen pasien adalah wanita dengan median usia 24 (13-51) tahun. Tiga pasien dilaporkan memiliki hipertensi pulmonal arteri. Satu pasien memiliki trombus di ventrikel kiri. Semua pasien memiliki gejala klinis jantung berupa gagal jantung dan nyeri dada. Simpulannya insiden efusi perikard pada pasien LES adalah 40 persen. Delapan puluh persen memiliki efusi minimal. Semua pasien memiliki gejala jantung terkait.

Kata kunci: *efusi perikard, lupus eritematosus sistemik.*

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Background

Cardiac involvement is one of major concerns in treatment of patients with Systemic Lupus Erythematosus (SLE). This involvement has been recognized since the beginning of 20th century, but newly recognized clinical entities have been detailed due to the introduction of very sensitive, non-invasive or semi-invasive cardiac imaging technique.

Pericardial effusion is considered as one of criteria to diagnose SLE based on American Rheumatism Association (ARA) criteria. There is limited data about the incidence and characteristic pericardial effusion in SLE patients in our country.

Several autoantibodies such as anti-phospholipid antibodies (aPL), anti-SSA/Ro antibodies and anti-endothelial cells antibodies, can mediate cardiac damage. These autoantibodies can directly affect heart tissue or alternatively, can trigger mechanisms able to cause heart damage; for example, aPL can contribute to cardiac damage enhancing

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atherosclerosis phenomena, causing thrombosis of coronaries arteries or starting in immune-complexes-mediated reaction and deposition at the valve level.²⁻⁴

Cardiovascular disease has recently been acknowledged as a primary cause of morbidity and mortality in SLE as well as in APS, numerous factors leading to atherosclerosis have been reported. Other cardiac manifestation, such as pericarditis, myocarditis, endocarditis and conduction disturbances are often mild and usually subclinical features are more prevalent than clinically apparent disease.²

The aim of this study is to report and assess the proportion and characteristic of pericardial effusion in SLE as a cardiac manifestation of the disease.

Methods

This is cross-sectional study conducted in secondary referral hospital in Tangerang county, west part of Jakarta, capital city of Indonesia. We evaluated from medical record and echocardiogram data from 2013-2015 patients diagnosed with SLE according to American Rheumatology Association (ARA) criteria. Incomplete medical record will be excluded. Pericardial effusion was diagnosed using the echocardiogram criteria. All echocardiogram data with SLE diagnosed were all included to this study. Characteristic subject and echocardiogram data were presented. All clinical characteristic data were reviewed related to echocardiogram data.

Results

From 33 SLE medical records reviewed, we found 13 patients (40%) with pericardial effusions. Ninety six percent patients were female with median age 24 (13-51) years old. All patients were in active stage with positive Anti-Nuclear Antibody (ANA) and decreased one of complement₃ (C₃) or C₄. Clinical assessment and transthoracic echocardiogram were used to diagnose pericardial effusions. Eighty percent pericardial effusion positive patients had minimal pericardial effusion based on echocardiography criteria. The rest had moderate pericardial effusion. No tamponade patients were recorded. Three patients were reported having pulmonary arterial hypertension (PAH) with tricuspid regurgitation. One patient had thrombus in left ventricle. All patients had clinical symptoms of heart failure and or typical chest pain.

Discussion

The Heart is frequently involved in SLE with the prevalence of cardiac involvement to be >50%. In the past, cardiac manifestations were severe, often leading to death and they were frequently found in post-mortem examinations. In our study the prevalence of heart involvement was 13%. Of 13% patients, there were found pericardial effusion. Some of patients had comorbidity of other cardiac. Nowadays, cardiac manifestations are often mild and asymptomatic and they can be recognized by echocardiography.²⁻⁴ In our study, all of patients with positive pericardial effusion had symptoms of heart failure.

The most common types of pericardial involvement with systemic inflammatory diseases are acute (or recurrent) pericarditis and asymptomatic pericardial effusions. Pericardial involvement in systemic inflammatory disease is thought to be immune-mediated, although concomitant infection may play a role in some cases. Systemic inflammatory diseases may be either "etiologic" or "permissive" of increased susceptibility to an unrelated primary cause (eg, viral).⁵⁻⁷

While some patients may manifest with the typical signs and symptoms of pericarditis (fever, pleuritic chest pain) or a hemodynamically significant effusion (dyspnea, fatigue, hypotension), many are incidentally found to have pericardial involvement during other testing (electrocardiography, echocardiography, computed tomography of the chest, or magnetic resonance imaging of the heart). Symptoms of pericardial involvement in systemic inflammatory disease may be insidious or may present suddenly with rapid progression. Cardiac tamponade and constrictive pericarditis are possible, but rare, complications. In our study, there were no patients found having signs of pericarditis.⁵⁻⁷

Myocarditis is the most characteristic features of myocardial involvement in SLE. Clinical detection of myocarditis ranges from 3 to 15%, although it appears to be much more common in autopsy studies suggesting the largely subclinical nature of lupus-associated myocarditis. Sign and symptoms are similar to those of myocarditis due to other causes (dyspnoe, tachycardia, arrhythmias) and they can progress to ventricular dysfunction, dilated CM and heart failure. The echocardiographic studies cannot definitely diagnose myocarditis but global hypokinesis, in the absence of other known causes, is strongly suggestive.²

Heart valve abnormalities (vegetations and/or thickening) are the most frequent cardiac manifestations of PAS. These alterations were known as Libman-Sacks endocarditis, a verrucous endocarditis of valve leaflets, papillary muscles and the mural endocardium. Heart valve lesions are frequently reported in patients with APS with or without SLE. Valvular disease is mild and asymptomatic only rarely do aPL positive patients develop valve disease severe enough to require surgical treatment. The valvular abnormalities resulting from Libman-Sacks lesions may predispose patients to bacterial endocarditis, so prophylactic antibiotics should be used for dental or surgical procedures with an increased risk of transient bacteremia.²

In SLE patients, the role of traditional and non-traditional risk factors for atherosclerosis is still debated. Some studies have shown that traditional cardiovascular risk factors are also more predictive in SLE patients than in age and sex-matched healthy subjects. In older age hypercholesterolemia and hypertension were three most common predictors of CAD. Moreover, corticosteroids therapy seems to increase the serum concentration of cholesterol, lipoproteins and triglyceride.⁸⁻¹⁰

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Other non-traditional risk factors associated with the auto-immune-inflammatory pathogenesis of the disease or with immunosuppressive therapy must also be taken into account: among these SLE-related risk factors, besides cumulative dosage and/or length of corticosteroid therapy, disease duration, high score of activity or damage could contribute to the development of atherosclerosis plaque.⁸⁻¹⁰

Conclusion

The incidence of pericardial effusion in SLE patient was 40 percent. Eighty percent patients had minimal effusion. All patients had cardiac symptoms related.

Acknowledgement

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Conflict of interest

None