Editorial

http://dx.doi.org/10.4070/kcj.2013.43.11.723
Print ISSN 1738-5520 • On-line ISSN 1738-5555

kcj
Korean Circulation Journal

Cardiac Amyloidosis Determines the Prognosis of Systemic Amyloidosis; Roles and Responsibilities of Cardiologist

Ga Yeon Lee, MD and Eun-Seok Jeon, MD

Division of Cardiology, Department of Medicine, Cardiac and Vascular Center, Samsung Medical Center, Sungkyunkwan University School of Medicine, Seoul, Korea

Refer to the page 752-760

Cardiac amyloidosis is a prognostically critical condition, since it is a frequent cause of death in patients with systemic amyloidosis.[1)2)] Lee et al.[3)] reported, for the first time, describing systemic amyloidosis in Korea from the perspective of cardiologists. In this paper, the authors enrolled 129 systemic amyloidosis patients retrospectively from 1999 to 2011. They showed that cardiac involve was the major prognostic factor, and old age, elevation of cardiac troponin I (cTNI), left ventricular (LV) systolic dysfunction and diastolic dysfunction were independent prognostic factors in survival. All cardiac amyloidosis were confirmed to have cardiac involvement by well-established and uncontroversial criteria including mean left ventricular thickness over 12 mm and low voltage in electrocardiogram or pathologically-confirmed amyloid deposit in endomyocardial biopsy. These diagnostic criteria were closely related to poor outcome in systemic amyloidosis.[3)]

The diagnosis and prognosis of cardiac amyloidosis still remains to be clarified, despite the development of more sensitive tools, such as delayed gadolinium enhancement (DGE) in cardiac magnetic resonance imaging (MRI) and cardiac biomarkers. In previous studies, when cardiac MRI was performed in all AL amyloidosis patients, DGE was found in 47% of the patients with normal LV wall thickness in echocardiography.[4)] Moreover, the level of NT-proBNP proved to be a better discriminating factor than the LV thickness (>12 mm) in echocardiography.[5-8)] But, it is still not clear whether we should suspect cardiac amyloidosis in patients with normal LV thickness in echocardiography or whether early diagnosis of cardiac amyloidosis improves the prognosis of the patients.

Since AL type is the most common type of systemic amyloidosis, chemotherapy and autologous stem cell transplantation are the therapies of choice. As shown,[3)] the hematologic response in AL amyloidosis is important in improving survival.[9)] Without hematologic response, it is difficult to obtain the responses in other organs. Likewise, in this paper, cardiac amyloidosis patients with better response to chemotherapy showed better survival compared to the patients with a lesser response.[3)] Many AL amyloidosis patients achieve hematologic response with high dose of melphalan and dexamethasone therapy or autologous hematologic stem cell transplantation.[10)] Additionally, bortezomib-based chemotherapy can be applied to achieve hematologic response in patients without hematologic response through melphan-based chemotherapy or autologous stem cell transplantation. Therefore, there is a significant limitation in this retrospective study. The authors did not show data regarding the benefit of additional chemotherapy in patients who had no hematologic response. In the meantime, active assessment of chemotherapy response and effort to achieve hematologic response cannot be overemphasized in the treatment of AL cardiac amyloidosis.

To accurately assess multi-organ involvement and treat systemic amyloidosis, an interdisciplinary approach of cardiology, hemato-oncology, nephrology, pathology, and laboratory medicine and more is needed. Considering that cardiac involvement is a major prognostic factor in systemic amyloidosis, cardiologists have a great responsibility and may play a pivotal role.

Correspondence: Eun-Seok Jeon, MD, Division of Cardiology, Department of Medicine, Cardiac and Vascular Center, Samsung Medical Center, Sungkyunkwan University School of Medicine, 81 Irwon-ro, Gangnam-gu, Seoul 135-710, Korea
Tel: 82-2-3410-3419, Fax: 82-2-3410-3849
E-mail: esjeon@skku.edu

• The authors have no financial conflicts of interest.

References

1. Falk RH. Cardiac amyloidosis: a treatable disease, often overlooked. *Circulation* 2011;124:1079-85.

2. Kapoor P, Thenappan T, Singh E, Kumar S, Greipp PR. Cardiac amyloidosis: a practical approach to diagnosis and management. *Am J Med* 2011;124:1006-15.
3. Lee MH, Lee SP, Kim YJ, Sohn DW. Incidence, diagnosis and prognosis of cardiac amyloidosis. *Korean Circ J* 2013;43:752-60.
4. Syed IS, Glockner JF, Feng D, et al. Role of cardiac magnetic resonance imaging in the detection of cardiac amyloidosis. *JACC Cardiovasc Imaging* 2010;3:155-64.
5. Dispenzieri A, Gertz MA, Kyle RA, et al. Serum cardiac troponins and N-terminal pro-brain natriuretic peptide: a staging system for primary systemic amyloidosis. *J Clin Oncol* 2004;22:3751-7.
6. Kumar S, Dispenzieri A, Lacy MQ, et al. Revised prognostic staging system for light chain amyloidosis incorporating cardiac biomarkers and serum free light chain measurements. *J Clin Oncol* 2012;30:989-95.
7. Lehrke S, Steen H, Kristen AV, et al. Serum levels of NT-proBNP as surrogate for cardiac amyloid burden: new evidence from gadolinium-enhanced cardiac magnetic resonance imaging in patients with amyloidosis. *Amyloid* 2009;16:187-95.
8. Palladini G, Campana C, Klersy C, et al. Serum N-terminal pro-brain natriuretic peptide is a sensitive marker of myocardial dysfunction in AL amyloidosis. *Circulation* 2003;107:2440-5.
9. Palladini G, Dispenzieri A, Gertz MA, et al. New criteria for response to treatment in immunoglobulin light chain amyloidosis based on free light chain measurement and cardiac biomarkers: impact on survival outcomes. *J Clin Oncol* 2012;30:4541-9.
10. Dispenzieri A, Gertz MA, Buadi F. What do I need to know about immunoglobulin light chain (AL) amyloidosis? *Blood Rev* 2012;26:137-54.