

Cardiology 2018 : D-Dimer: A Novel Predictor of Survival in Patients with Cardiac Light Chain Amyloidosis - Xian Cheng - First Affiliated Hospital of Nanjing Medical University

Xian Cheng,

First Affiliated Hospital of Nanjing Medical University, China.

Introduction

Light-chain (AL) amyloidosis is a form of systemic amyloidosis that is characterized by extra-cellular deposition of pathologic insoluble β fibrillar immunoglobulin light chains in virtually every organ except the central nervous system. AL amyloidosis is usually associated with plasma cell dyscrasia, in which a malignant proliferation of plasma cells secretes an unstable light chain that is prone to misfolding. AL amyloidosis is a relatively rare disease, occurring with an incidence of approximately 10 patients per million per year; however, cardiac involvement has been reported in approximately 70% of cases, resulting in rapidly progressive heart failure (HF) and a very poor prognosis, especially in untreated patients. The progression of cardiac AL amyloidosis involves myocardial hypertrophy and decreased myocardial compliance. Causes of death include congestive HF, ventricular tachyarrhythmia, bradyarrhythmia, and severe hypotension. The median survival time from diagnosis of cardiac AL amyloidosis ranges from 6 to 24 months. Prompt treatment is essential, as 25-30% of cardiac AL amyloidosis patients will die within the first year of diagnosis. There remains an unmet need for non-invasive tools that allow early diagnosis of cardiac AL amyloidosis. Currently, diagnosis is challenging, as the gold standard is endomyocardial biopsy (EMB), which is an invasive procedure with risks associated with sampling. Biomarkers such as N-terminal pro b-type natriuretic peptide (NT-proBNP) and cardiac muscle troponin T have been shown to add clinically useful information for the management of patients with AL amyloidosis. Therefore, the presence of clinical factors and/or biomarkers of cardiac pathology may play a critical role in identifying cardiac AL amyloidosis and determining the severity of cardiac involvement. The objective of this study was to identify independent risk factors and predictors of survival in patients with cardiac AL amyloidosis.

Methods: This study included 26 patients with cardiac AL amyloidosis who were diagnosed by biopsy between October 2009 and January 2016. All the patients were followed up until August 26th, 2016. Baseline clinical data including clinical symptoms, laboratory data, and echocardiographic findings were recorded. Univariate and multivariate Cox proportional hazard regression analyses were performed to identify risk factors for all-cause mortality. The Kaplan-Meier method and log-rank test were used to compare survival times

Results: In univariate and multivariate analysis, N-terminal pro b-type natriuretic peptide (NT-proBNP) and D-dimer were independent risk factors for all-cause mortality in patients with cardiac AL amyloidosis. The cutoff value of NT-proBNP for 6-month all-cause mortality was 4509.5 ng/L (sensitivity 73.3%, specificity 77.8%, area under curve (AUC) 67%, 95% confidence interval (CI) 0.442-0.899). The cut-off value of D-dimer for 6-month all-cause mortality was 1.22 mg/L (sensitivity 60%, specificity 90%, AUC 70%, 95% CI: 0.489-0.911). Patients with NT-proBNP or D-dimer levels above the cut-off value had a higher all-cause mortality rate compared to patients below.

Conclusion: D-dimer may be an important biomarker of prognosis in cardiac AL amyloidosis patients.

Biography:

Xian Cheng is currently working in First Affiliated Hospital of Nanjing Medical University, China. She has published more than 25 papers in reputed journals and has been serving as an editorial board member of reputed

E-mail: djxu@njmu.edu.cn