Young Fellow and Best Abstract Presentations

Title: A Retrospective Study on the Clinical Features and Outcomes of Patients with Systemic AL Amyloidosis in A Single Center in Hong Kong

Presenter: Dr Wai-Pan LAU, Hong Kong

Objective: To retrospectively investigate the clinical features, current treatment regimens, clinical outcome and prognostic factors in patients with systemic amyloid light chain (AL) amyloidosis in our center in Hong Kong.

Methods: The medical records of patients who had visited to Queen Elizabeth Hospital and had been diagnosed with systemic AL amyloidosis from 2006 to 2019 were reviewed. Patient's characteristics including the presenting symptom, age, gender, blood pressure, major organ function (including cardiac and kidney), free light chain level, cardiac enzyme, bone marrow status, treatment received and its response were analyzed.

Results: A total 44 patients was evaluated in this cohort analysis. The median age was 66.2 with male predominate in this study. Proteinuria was found in 31% of patients as initially presentation, followed by 20% of them with clinically limbs edema and 15.9% of them with shortness of breath. The most affected organ by AL amyloidosis was kidney which accounted for 63.6%, followed by 56.8% of them with cardiac involvement. The median overall survival was 45 months. The median overall survival in subgroup with cardiac amyloidosis was 17.0 months, compared to 82.0 months in subgroup with non-cardiac involvement (P value = 0.03). Four associated prognostic factors for overall survival were identified in this study. Age >/=65, difference in free light chain>180 mg/L, systolic blood pressure </=100mmHg at diagnosis and elevated serum HsTroponin I /Troponin I level predict poor prognosis. Overall response rate of 84% was seen in bortezomib based treatment, compared to 42.9% in non- bortezomib treatment arm as front line treatment of amyloidosis.

Conclusion: The clinical features and the outcome of patients with systemic AL amyloidosis in Hong Kong are similar to the data from international centers. Diagnosis of the disease could be changeling in some of the cases due to the non-specific sign and symptom as initial presentation. Early recognition of disease and rapid initiation of treatment is essential to reduce the irreversible organ damage by light chain amyloid and further improve the outcome of the disease.