Abstracts

33 COMPOSITE GOALS PLUS INFLAMMATION: FURTHER RISK ASSESSMENT FOR SYSTEMIC LUPUS ERYTHEMATOSUS ASSOCIATED PULMONARY ARTERIAL HYPERTENSION IN CSTAR-PAH COHORT

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Background Chinese SLE treatment and Research Group (CSTAR) started a multi-center prospective cohort study recruiting SLE patients with pulmonary arterial hypertension (PAH) since 2006. This study aimed to investigate the validity of a multidimensional risk assessment and the prognostic value of it in SLE-associated PAH.

Methods All SLE patients were fulfilled the 1997 revised ACR criteria. PAH was diagnosed based on ESC/ERS guidelines by right heart catheterization. The outcome was all-cause mortality. Two different methods of risk categorization were applied according to baseline data, including low-risk criteria number of none to four and mean score of 1 (low-risk), 2 (intermediate-risk) or 3 (high-risk). According to first follow-up, patients were further divided into increased risk, remained risk and decreased risk group. A prediction model was used to distinguish SLE-PAH from vasculitic and vasculopathic subtype, based on the time interval between the diagnosis of SLE and PAH and Systemic Lupus Erythematosus Disease Activity Index (SLEDAI). Kaplan-Meier survival curves and Cox proportional hazards analysis were conducted.

Results 282 patients were enrolled. The 5 year survival of patients with none, one, two, three and four low risk criteria were 42.7%, 64.8%, 86.1%, 90.2% and 91.7%, respectively (HR = 0.59, 95% CI 0.44–0.78, p < 0.001). The 5 year survival of patients in low-risk, intermediate-risk and high-risk group were 92.3%, 60.4% and 50.0%, respectively (Log-rank, p = 0.001). Notably, in low-risk group, patients with vasculitic subtype had better survival than those with vasculopathic subtype (Log-rank, p = 0.044). The 5 year survival of patients with remained, decreased and increased risk were 65.4%, 88.1% and 23.8%, respectively (log-rank, p < 0.001).

Conclusions Our study, for the first time, validated the prognostic value of risk stratification strategy at baseline and follow-up visit in patients with SLE-associated PAH. Patients are recommended to have a comprehensive evaluation on PAH and SLE at baseline and every follow-up visit. The SLE disease activity and systemic manifestations predict the phenotype of SLE-associated PAH, which also affect the long-term survival and need to be involved into risk assessment of SLE-associated PAH. Improving to low-risk group can be a future treatment target for SLE-associated PAH patients in clinical practice.

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