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The prognosis of two distinct clinical phenotypes of SLE-PAH

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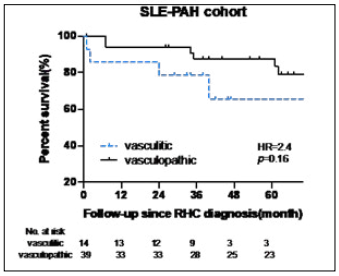
Abstract

Background: Based on the characteristics of systemic lupus erythematosus-associated pulmonary arterial hypertension (SLE-PAH), Sun et al has put forward a scoring system to distinguish two clinical phenotypes as vasculitic and vasculopathic subtypes[1]. A weighted score ≥2 suggested a vasculitic subtype by combining two factors: The time interval between SLE and PAH diagnosis <2 years and ≥2 years were 1 and 0 point; SLE Disease Activity Index (SLEDAI) >9, 5-9 and <5 were 2, 1, 0 point, respectively. While the vasculitic subtype seemed to have poorer prognosis in Sun's research, other study has shown controversial result [2].



Objectives: To find out the prognosis of two distinct clinical phenotypes of SLE-PAH.

Methods: Between 2008 and 2019, a SLE-PAH cohort confirmed by right heart catheterization (RHC) from Guangdong Provincial People's Hospital was included. Other groups of pulmonary hypertension were excluded. Based on the scoring system, patients were divided into vasculitic (weighted score≥2) and vasculopathic subtypes (weighted score<2). The endpoint was PAH-related mortality. Survival status were confirmed by clinic follow-up data or phone call.

Results: a total of 53 SLE-PAH patients were enrolled. The cases of vasculitic and vasculopathic subtype were 14 and 39, respectively. Ten endpoint events occurred. Eight attributed to PAH and the cause could not be traced in two which were still included in study. The pooled 1-, 3-, 5-year survival rates were 85.7%, 78.6%, 65.5% in vasculitic subtype, and 93.9%, 87.5%, 87.5% in vasculopathic subtype, respectively. Kaplan-Meier analysis showed vasculitic subtype tended to have a poorer prognosis than vasculopathic (p=0.16, HR 2.4, 95%CI 0.5-13.8, figure 1).

Conclusions: The prognosis of the two phenotypes of SLE-PAH was statistically indifferent while the vasculitic subtype showed a trend of worse prognosis. Further studies are needed.

Biography

Wang has graduated from Peking Union Medical College and worked in the Department of Rheumatology, Guangdong Provincial People's Hospital. She has been devoted to the clinical and basic study of connective tissue disease-associated pulmonary arterial hypertension. She has published several articles related to pulmonary arterial hypertension, presided over and participated in several relevant research projects, and made some oral presentation at national and foreign conferences.

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