Clinical Characteristics and Mortality Rate of Thai Elderly Onset Systemic Sclerosis

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Background: Elderly over against adult Caucasian systemic sclerosis (SSc) patients are at a greater risk of pulmonary arterial hypertension (PAH), renal impairment, cardiac disease, and muscle weakness, but a lower risk for digital ischemia. However, there has been no report comparing clinical features between Thai adult and Thai elderly onset diffuse cutaneous SSc (dcSSc).

Objectives: We aimed to identify the clinical differences and mortality rate between Thai adult and Thai elderly onset SSc.

Methods: We conducted a historical cohort study of SSc patients who were followed up at Srinagarind Hospital, Khon Kaen University, Thailand between January 2007 and December 2011. The SSc patients were 60 years and over and classified as elderly onset SSc. The clinical difference(s) between adult and elderly onset were identified. Cox regression analysis was used to estimate the probability of survival and to assess the factors associated with mortality.

Results: The medical records of 350 SSc patients were reviewed, with 53 (15.1%) cases of elderly onset SSc. According to the multivariate analysis, elderly onset SSc had higher WHO functional class, with more frequent weakness and hyperCKaemia, as well as less pulmonary fibrosis than adult onset SSc (p=0.004, 0.02, 0.02, 0.02, respectively). Among the 2,399 person-years, the incidence of mortality was 3.8 per 100 person-years with a median survival rate of 15.9 years (95%CI 12.4-17.3). The mortality rate of elderly onset SSc was 12.7 per 100 person-years; whereas, adult onset was 2.9 per 100 person-years with a median survival of 4.9 years (95%CI 3.8-7.4) and 16.1 years (95%CI 14.8-23.4), respectively. The mortality rate of elderly SSc onset was significantly higher than that of adult SSc onset (HR 5.71 (95% CI 3.54-9.20). The Cox regression analysis indicated that presence of digital ulcer and tendon friction rub had a respective HR of 7.39 (95%CI 1.28-42.60) and 37.23 (95%CI 2.10-659.09) for predicting mortality of elderly onset SSc. Neither the severity of skin tightness nor the SSc subset was associated with mortality.

Conclusion: Myopathy and limitation of physical activity are frequently observed among elderly onset SSc over against pulmonary involvement than adult onset SSc. Mortality of elderly onset SSc is 5.7 times higher, with median survival of 11 years shorter than adult onset SSc. Presence of digital ulcer and tendon friction rub is associated with high mortality rate in elderly onset SSc.

Keywords: Systemic sclerosis, Scleroderma elderly onset, Elderly