中文題目：系統性硬皮症合併間質性肺炎患者之治療與預後：台灣單一醫學中心之經驗

英文題目：Treatment and Prognosis of Interstitial Lung Disease in Systemic Sclerosis: A Single-center Experience in Taiwan

作　　者：顏憶寧[1]，劉志偉[1,2]，廖顯宗[1,2]，陳瑋昇[1,2]，賴建志[1,2]，陳明翰[1,2*]，蔡長祐[1,2*]

服務單位：[1]臺北榮民總醫院內科部過敏免疫風濕科，[2]國立陽明大學醫學系內科學科

Objective

Systemic sclerosis(SSc) is an autoimmune disease with diverse clinical manifestations including Raynaud's phenomenon, esophageal dysmotility, sclerodactyly, and also complications such as interstitial lung disease (ILD) and pulmonary arterial hypertension. However, there are few data concerning the prognosis and clinical outcome of Asian SSc patients with ILD. In this study, we aim to evaluate the clinical outcomes of SSc patients with ILD at Taipei Veterans General Hospital.

Methods

We retrospectively investigated patients who was diagnosed with SSc complicated with ILD at our hospital between 2007 to 2017. Their clinical profiles, including pulmonary function test results, high-resolution computed tomography (HRCT) pattern, echocardiography results, and clinical outcomes were recorded.

Results

Twenty-nine patients were enrolled in this study. The mean age of the onset of SSc was 45.8 years, and 24 patients (83%) were female. Malignancy was noted in 8 patients (27.6%), including 5 cases of breast cancer, 2 cases of colorectal cancer, and 1 case of lung cancer. Pulmonary hypertension was diagnosed by echocardiography in 14 patients (60.9%). The mean duration from the diagnosis of SSc to the development of ILD confirmed by HRCT scan was 9.5 years, while 10 patients had ILD diagnosed at the same time as SSc. Fourteen patients (48.3%) were diagnosed with usual interstitial pneumonia (UIP) pattern, 13 (44.8%) with non-specific interstitial pneumonia (NSIP) pattern, and 6 (6.9%) with mixed pattern. During a mean follow-up of 31.9 months, 6 patients (20.7%) expired. The causes of death were pneumonia (5 cases) and heart failure (1 case). More importantly, most of them (5 out of 6 patients, 83.3%) expired within 24 months after the diagnosis of ILD.

Conclusion

ILD is a common complication in SSc patients, and associated with significant mortality. Further studies are required to outline a generalized and efficient treatment regimen for these patients.