Scleroderma renal crisis: a rare complication of systemic sclerosis with poor prognosis – experience in a medical center in central Taiwan

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Objective: Scleroderma renal crisis (SRC) is the most life-threatening complication of systemic sclerosis (SSc), which is characterized by malignant hypertension and rapidly progressive renal failure. In this study, we investigate the incidence, clinical features, treatment and outcome of our SRC patients.

Patients and Methods: We reviewed the records of 161 patients with SSc, seen from January 1991 to December 2005 in the Department of Rheumatology, Taichung Veterans General Hospital, Taiwan. Tests for anti-nuclear antibodies (ANA) in sera were screened against Hep-2 cells by indirect immunofluorescence assay. Quantification of autoantibodies to extractable nuclear antigen, including anti-Scl-70 antibody (Scl-70) and anti-centromere antibody, were tested with the AtheNa Multi-Lyte ANA test system. Data collected from the records included gender, age at onset, age at diagnosis, clinical manifestations, laboratory data, treatment and outcome. Fisher’s exact test was used to compare differences in categorical variables.

Results: Seventy-eight (48.4%) patients were limited type SSc and 83 (51.6%) patients were diffuse type SSc. Four (2.5%) patients of a total of 161 patients had SRC. All of our SRC patients were diffuse type with positive Scl-70. SRC occurred at 37.7 ± 55.72 months (range from 1-120 months) after the diagnosis of SSc. The duration between Raynaud’s phenomenon to disease onset was 3.75 ± 2.06 months. There were significant difference at myocardial involvement (p<0.05) and pericardial effusion (p<0.05) between the SRC and Non-SRC diffuse SSc patients. Two patients died within four months after the SRC, one patient required permanent dialysis and one patient partly regained renal function.

Conclusion: SRC is a rare complication in Taiwanese SSc patients and has a poor prognosis. Whether early administration of angiotensin converting enzyme inhibitor might prevent or ameliorate the onset of SRC needs further investigation.

Key words: Scleroderma renal crisis, anti-Scl-70 antibody, complications, prognosis

Introduction

Systemic sclerosis (SSc) is a systemic autoimmune disease characterized by diffuse fibrotic and degenerative change in vessels, skin and internal organs [1]. Kidney involvement, as manifested by renal failure, was first described in SSc in 1863 [1]. The single most life-