Prognostic factors of survival in patients with dermatomyositis and polymyositis

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Background: To determine the incidence, characteristics, and prognostic factors of mortality in patients with polymyositis (PM) and dermatomyositis (DM).

Materials and Methods: Medical records of 151 PM/DM patients treated at Chang Gung Memorial Hospital between 2000 and 2007 were retrospectively reviewed.

Results: Twenty-five (16.6%) of the 151 PM/DM patients had associated cancer. Thirty-two (21.2%) of the 151 PM/DM patients had interstitial lung disease (ILD). During follow-up, 30 (19.9%) patients died. Overall patient cumulative survival rates were 81.0, 77.6, and 74.6% at 1, 2, and 5 years, respectively. In univariate analysis, old age at PM/DM onset, cancer, ILD, DM, diabetes mellitus, low creatine phosphokinase (CPK) level, and use of intravenous immunoglobulin were associated with increased mortality (p=0.018, 0.011, <0.001, 0.002, 0.001, 0.009, and 0.007, respectively). Multivariate analysis excluding anti-Jo-1 antibody was performed after adjusting for CPK level. Only ILD (OR = 12.93, 95% CI = 3.97-42.13, p<0.001) and cancer (OR = 4.10, 95% CI = 1.21-13.91, p=0.023) were associated with mortality. If the anti-Jo-1 antibody was included in multivariate analysis (n = 80), then ILD (OR = 15.40, 95% CI = 2.68-88.02, p=0.002) and DM (OR = 12.56, 95% CI = 1.21-130.62, p=0.034) were associated with increased mortality.

Conclusion: This study underlines the high mortality rate that associated with PM/DM. Survival time was significantly shorter for patients with ILD, cancer or DM than those without.

Key words: Prognosis, survival analysis, polymyositis, dermatomyositis

Introduction

Polymyositis (PM) and dermatomyositis (DM) are inflammatory connective tissue diseases with unknown etiologies, predominantly involving the muscles and skin, but also producing pulmonary manifestations and may be associated with malignancies [1-5]. DM is a multisystem inflammatory disorder primarily affecting proximal skeletal muscles and the skin, and is characterized by typical heliotrope rashes and Gottron's papules. Previous reports found several factors associated with a high mortality rate for patients with PM/DM, such as interstitial lung disease (ILD), old age, and cardiac involvement. [6-11,19,23]. Notably, PM- or DM-associated malignancies also contribute to poor prognosis. The aim of this study was to define both clinical and biochemical features in patients died of PM/DM, and to identify predictors of PM/DM mortality.