Clinical manifestations, lymphocyte subsets, and complications of patients with common variable immunodeficiency in Taiwan

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Objective: To analyze the clinical features, lymphocyte subsets, and complications of patients with common variable immunodeficiency (CVID).

Methods: In this retrospective study, twelve patients with CVID diagnosed between 1993 and 2007 were enrolled. Clinical features as well as complications were recorded. Lymphocyte subsets in the peripheral blood (PB) were analyzed using flow cytometry.

Results: Eight patients (66.7%) were male and four patients (33.3%) were female and the mean age at onset was 38.7 ± 9.4 years. The mean levels of serum immunoglobulin (Ig) G, IgA and IgM were 277.8 ± 201.3 mg/dL, 25.6 ± 17.4 mg/dL, and 34.0 ± 17.2 mg/dL, respectively. Markedly decreased percentage of B cells was noted in 7 (87.5%) patients with CVID and elevated percentage of CD8+ T cell was found in 6 (75%) patients after analysis of lymphocyte subsets. Eleven of 12 had recurrent infection, including pneumonia, sinusitis, urinary tract infection and septicemia. Six of 12 had chronic lung disease, such as bronchiectasis. Three patients had autoimmune phenomenon and 4 had tumor.

Conclusion: Our CVID patients had a higher risk of recurrent infection, chronic lung disease, tumor, and autoimmune phenomenon, including Sjögren’s syndrome and antiphospholipid antibody syndrome (APS).

Key words: Common variable immunodeficiency, clinical manifestation, complications, lymphocyte subsets

Introduction

Common variable immunodeficiency (CVID) is a primary immunodeficiency characterized by deficient antibody production [1-5]. The pathophysiologic defect underlying CVID is unknown. Patients typically experience recurrent sinopulmonary infections, chronic diarrhea, enhanced risk of malignancy, granulomatous disease, and autoimmune phenomenon [1-5].

Early diagnosis of patients with CVID, appropriate treatment with antibiotics for infections, and replacement with gamma-globulins prevent long-term complications of this disease and dramatically improve the life quality and life expectancy of these patients [6].

To our knowledge, no case series analysis of CVID has been conducted in Taiwan. The aim of this study was to investigate the clinical manifestations, lymphocyte subsets in the peripheral blood (PB) and complications of 12 patients with CVID.

Methods

Patients

Twelve patients, aged from 21 to 70 years with the diagnosis of CVID were enrolled at Taichung Veterans General Hospital between 1993 and 2007. CVID was diagnosed using the World Health Organization criteria.