Cyclophosphamide Pulse Therapy for Rapidly Progressive Interstitial Pneumonia in Dermatomyositis. A New Possibility for Rescue?

Dermatomyositis (DM) is a systemic inflammatory disorder characterized by profound lesions in muscles and skin (1). This disease is associated with pulmonary lesions which include aspiration pneumonia, infections, drug-induced pneumonia as well as interstitial lung disease (ILD) (2). ILD in DM has been reported as one of the systemic lesions of this autoimmune disease of unknown etiology (3), and recent studies especially in the Japanese literature have emphasized its importance as a prognostic factor (4-7). There is a subgroup of patients with DM and a rapidly progressive ILD who have mild or even negligible muscle symptoms (amyopathic DM), mildly increased or normal levels of muscle enzymes, and negative anti-Jo-1 autoantibody (7). These patients often show intractable respiratory failure even if treated with a high dose or pulse therapy of corticosteroids and succumb to death within one year after the onset. Intravenous cyclophosphamide pulse therapy (CY Pulse) was first described for the treatment of refractory lupus nephritis (8) and was well tolerated with beneficial effect. However, it is often difficult to evaluate its effectiveness even when the cases are successfully treated, since conventional therapy as well as pulse therapy with corticosteroids are usually performed simultaneously with or before CY pulse, as in the case reported by Shinohara and associates in this issue of the Journal (14). To address this question, it would be necessary to study a prospective trial (eg. steroid pulse along with conventional therapy versus steroids and additional CY pulse); such a study is now underway. Recent studies have also shown that cyclosporin, another potent T cell suppressant, might be useful for the control of this intractable lung disease (15). It is critical to evaluate how and when these potentially hazardous modalities should be taken. It is another important issue to clarify the immunopathogenesis of lung lesions found in this group of DM.

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Hajime TAKIZAWA, MD and Koji ITO, MD
The Department of Medicine and Physical Therapy,
University of Tokyo, School of Medicine,
7-3-1 Hongo, Bunkyo-ku, Tokyo 113

References