Refractory Interstitial Lung Disease of Dermatomyositis: A Proposal for a Prospective Trial for Establishing Evidence

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In this issue of Internal Medicine, Takai and colleagues reported a case of rapidly progressive interstitial lung disease (ILD) with anti Jo-1 antibody positive dermatomyositis (DM) (1). Although the initial treatment with corticosteroid, cyclophosphamide, and cyclosporine was not effective, a favorable outcome was achieved after the combined treatment of intravenous immune globulin (IVIG) with the treatment of direct hemoperfusion with polymyxin B-immobilized fiber column (PMX-DHP). This case report may encourage the physicians who struggle in treating these severe forms of rapidly progressive ILD. IVIG therapy has been accepted as a salvage treatment for refractory DM (2). However, the efficacy of the PMX-DHP treatment may be unsubstantiated with regard to evidence-based medicine.

DM is the most recognized disease entity among idiopathic inflammatory myopathies and it is characterized by the inflammatory cells infiltration in the muscles and the skin (3). ILD has been reported in up to 75% of the patients with DM and is regarded as one of the major causes of mortality (3). Although careful observation without treatment is proposed for asymptomatic patients without lung involvement, immunosuppressive therapy is typically indicated for symptomatic patients with significant physiological lung impairments. In addition, a salvage therapy, such as IVIG, is often required among the patients with acute or rapidly progressive ILD, as is often the case of amyopathic DM (the subset of DM) (3, 4).

In the last decade, many case reports, case series, and some retrospective studies (5, 6) have been published regarding the efficacy of PMX-DHP treatment for acute or rapidly progressive ILD, including DM, amyopathic DM, and idiopathic interstitial pneumonias. However, there is a possibility that some publication bias may exist; namely, ineffective cases may not have been published in the literature. Most cases reported were from Japan, and no systematic review of the PMX-DHP treatment for ILD has yet been reported to the best of our knowledge. Due to the high mortality rate of acute or rapidly progressive ILD, more trials are warranted to establish the effectiveness of these treatments. Potential treatments may include PMX-DHP treatment (1, 5, 6), immunesuppressants (7), sivelestat (8), rituximab (3, 9), recombinant human soluble thrombomodulin (10), pirfenidone (11), and/or their combinations. Acute or rapidly progressive interstitial pneumonia is regarded to appear at a higher frequency in the Japanese population compared with European and American populations (12, 13). Therefore, a prospective, large sample-sized high quality randomized trial must be conducted to clearly establish evidence for the use of these treatment modalities in these fatal interstitial lung diseases in Japan.

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References


