

**Interstitial Lung Disease  
in  
Polymyositis and Dermatomyositis**

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**AKADEMISK AVHANDLING**

som för avläggande av medicine doktorsexamen vid Karolinska Institutet  
offentligen försvaras i Thoraxklinikernas föreläsningssal N2:U1  
Karolinska Universitetssjukhuset, Solna  
fredagen den 8 december 2006, kl 09.00



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**Stockholm 2006**

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## Abstract

Polymyositis and dermatomyositis are rare disease entities affecting skeletal muscle and other organs such as the lungs. Interstitial lung disease (ILD) is increasingly recognized as a serious complication of poly-/dermatomyositis. The reported prevalence of pulmonary involvement varies widely due to use of different clinical, radiological, functional and pathological criteria. The etiology and pathogenesis of myositis as well as myositis-associated ILD are still unclear and it is not known when during the course of disease ILD develops. Infiltration of T cells and macrophages in the muscle tissue suggests an important role of T cell-mediated immunity in the pathogenesis of the diseases. It is still unknown which antigen these cells recognize and which cytokines are important in the inflammatory process. The role of autoantibodies in disease mechanisms of myositis is also not clear.

In order to establish the prevalence, characteristics and the course of myositis associated ILD, as well as putatively relevant pathogenetic factors we investigated an unselected group of patients with newly diagnosed poly-/dermatomyositis using chest radiography/ high resolution tomography and pulmonary function tests. Furthermore, we investigated T cell receptor (TCR) V gene usage in bronchoalveolar lavage (BAL), muscle biopsy and peripheral blood T cells by using T cell specific monoclonal antibodies. Moreover, we analyzed the relationship between presence of ILD-related autoantibodies, genotype and balance between serum levels of cytokines suggested to be involved in the disease (tumor necrosis factor (TNF), interleukin (IL)-10).

ILD, defined by radiographic changes and/or restrictive ventilatory impairment, was recorded in up to 79% of the patients. The number of patients with ILD had been even higher with the use of bronchoscopy and BAL, as alveolitis was also evident in patients without evidence of ILD through radiographic examinations or lung function tests. Arthritis and positive anti-Jo-1 autoantibodies were more common in ILD-patients than in patients without ILD. The course of myositis-associated ILD varied. In most cases pulmonary function tests stabilized, improved or even normalized after initiation of immunosuppressive therapy. A common targeted antigen in muscle and lung tissue was suggested by a restricted TCR BV gene usage in the lungs and muscle. The presence of anti-Jo-1 antibodies and anti-Ro52 antibodies was associated with higher TNF/IL-10 ratios in myositis patients and this ratio seemed to have a genetic basis, thus suggesting a role of genes as a predisposing factor for ILD.

In conclusion, ILD is a common manifestation of myositis. We propose that all newly diagnosed patients, regardless of pulmonary symptoms, should be screened for ILD by physical examination, chest radiographic examination, lung function tests and screening for anti-Jo-1 antibodies in order to identify patients with ILD early in the course of disease, when it is likely that the clinical course may improve by immunosuppressive treatment. Restricted TCR BV usage in the lungs and muscle make it important to include the lungs in the search for etiology of myositis. Patients with anti-Jo-1 antibodies have a high risk for developing ILD. This could be genetically determined, mediated through altered cytokine production. An increased knowledge concerning the pathogenesis will hopefully make it possible to develop more selective therapies for myositis patients.

**Keywords:** dermatomyositis, polymyositis, interstitial lung disease

**ISBN:** 91-7357-002-8

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