

INVESTIGATION OF IDIOPATHIC INFLAMMATORY MYOPATHIES, RELEVANCE OF NEW CLINICOSEROLOGICAL RESULTS.

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The challenge in treating idiopathic inflammatory myopathy (IIM) is to identify those patients who require early aggressive treatment. New clinicoserological classification is currently under development by myositis experts, which help us to differentiate between the clinical subgroups in context of the newly recognised myositis specific autoantibody results and careful immunohistochemical characterisation of muscle biopsy specimens. Improved classification criteria for poly-, dermatomyositis are important for clinical trials to test new drugs and will also be very important in the work to understand disease mechanisms and to develop new targeted therapies. I reviewed the detailed clinical data of 374 idiopathic inflammatory myopathy patients between 1988-2008.

I confirmed that pregnancy in female patients with idiopathic inflammatory myopathy has a higher risk for both the mother and the child in case of active disease. Almost in all of our cases when pregnancies occurred during the inactive phase of idiopathic inflammatory myopathy on low dose corticosteroid therapy the risk for the mother and fetus was minimal. So we can conclude that there is a minimal risk for the fetus and mother in pre-existed well-controlled disease. Generally it appears that the disease activity in active cases can be controlled by increasing the corticosteroid dose. Preventive monthly intravenous immunoglobulin was administered to one antisynthetase syndrome patient to prevent abortion.

Prevalence of interstitial lung disease was found to be 70,4% in patients with antisynthetase syndrome, these findings are similar to those previously reported. The ASS group without anti SS-A antibody had a more frequent association with alveolitis, presented with increased alveolar scores, and responded well to immunosuppressive therapy as usual in the treatment of myositis alone. ASS patients with coexistent anti SS-A antibodies tended to have a more severe form of ILD represented by the different HRCT pattern and increased interstitial scores at diagnosis.

I examined serological and clinical characteristics of overlap myositis compared to primary myositis. Polymyositis was found to be the most common IIM associated with another connective tissue disease. Moreover, SSc was the most common CTD associated with myositis. Prevalence of the MSA was similar to those previously reported. Prevalence of the antisynthetase syndrome was similar, anti Jo-1 antibody was the most common MSA and was found in all subgroups and predominantly in polymyositis. Anti Jo-1 positivity was a predictor of refractory disease with polycyclic disease course . requiring second line treatment options. Finally the prevalence of anti Jo-1 antibody was surprisingly high (33,3%) in rheumatoid arthritis/myositis overlap patients, however there may be limitations due to the relative low number of patients involved. I also review a case of dermatomyositis patients treated with immunosuppressive drugs, which led to the development of B cell follicular non-Hodgkin lymphoma.

Key words: myositis, pregnancy, interstitial lung disease, antibodies

Kulcsszavak: myositis, terhesség, intersticiális tüdőbetegség, autoantitestek