OBJECTIVE: To determine the response to treatment and the long-term outcome of patients with the antisynthetase syndrome associated with anti-Jo-1-antibodies. PATIENTS AND METHODS: A total of 12 patients with histologically proven myositis and anti-Jo-1-autoantibodies were evaluated over a mean follow-up period of 66.4 months. In all patients neuromuscular function tests, electromyographic examinations, pulmonary function tests and high-resolution-computed tomography of the lungs were performed regularly. RESULTS: Muscle function improved in all patients with treatment, and a complete clinical response was achieved in 5 patients. Pulmonary function worsened in 1 patient, who died from respiratory failure, but normalised in 4 patients. Arthropathy progressed despite improvement of myositis and pulmonary status in 2 patients. Discontinuation of treatment was facilitated in 1 patient, although long-term therapy was required in 10 patients. In 2 patients with refractory disease, treatment with intravenous immunoglobulins was successful. Severe side effects of treatment occurred in 7 patients and overall mortality rate was one of 12 (8 %). CONCLUSION: The antisynthetase syndrome associated with anti-Jo-1-antibodies requires long-term immunosuppressive therapy in most patients. Whereas a complete clinical response of muscular...
symptoms is frequent, continued deterioration of the pulmonary system may occur despite immunosuppressive treatment, and may lead to fatal outcome. An interdisciplinary therapeutic approach is necessary for best possible results in these patients.