P-10
Malignancies associated with idiopathic inflammatory myopathies: a 35-years retrospective study

R. Neri, V. Iacopetti, S. Barsotti, A. d'Ascanio, A. Tavoni[1], M. Mosca, G. Iacopetti, S. Bombardieri
Internal Medicine, AOU Pisa, UO Reumatology, Italy; [1] Internal Medicine, AOU Pisa, UO Immunoallergology Clinic, Pisa, Italy

An increased incidence of malignancy in patients with poly/dermatomyositis (PM/DM) has been reported; several points remain unclear: incidence and the predictive factors for the presence of cancer and its prognosis.

Aim of the study was to evaluate the frequency of malignancy among myositis patients diagnosed in our Unit in a 35 years follow-up (from 1975 to 2010). We compared epidemiological, clinical, serological and survival data of the patients with cancer associated myositis (CAM) with the findings of primary myositis, trying to define predictive factors for malignancies development.

162 primary PM/DM patients were enrolled by our Rheumatology Unit. Diagnosis of PM/DM was based on Bohan and Peter criteria (1975). Myositis was considered tumor-associated if diagnosed within 2 years before or after the diagnosis of cancer. Concerning the comparison of CAM and primary myositis, we analyzed: age, sex, systemic symptoms (Raynaud's phenomenon, dysphagia, dyspnea, fever, arthritis, dysphonia), creatinekinase (CK) levels at onset, autoantobodies, number of deaths, survival rates.

Out of 162 patients with myositis, cancer was present in 18/72 DM patients (25%) and in 9/90 PM patients (10%). In 10 cases the malignancy and myositis appeared simultaneously; in 6 cases the tumor was diagnosed before and in 11 after the onset of myositis; cancer types more frequent were breast and ovarian carcinoma. Patients with CAM were older than those without cancer (59 yr vs. 51yr) $p < 0.01$. There was a female predominance in primary myositis group (M/F = 30/51 in PM and 13/41 in DM) and in CAM-DM (6/12) while CAM-PM patients were equally shared.Dysphagia was more frequent in CAM than in primary myositis patients (37% vs. 17,5%) $p < 0.035$; in the CAM subgroup was more present in DM than PM (44% vs. 14%) $p < 0.006$. Prevalence of other extramuscolar features was similar in the 2 subgroups.

In patients with CAM, CK medium levels were lower in than in primary myositis (3013 U/L vs. 2412 U/L); positive ENA were significantly more present in primary myositis group (20% vs. 7%). AntiJO1 antibodies were detectable in 6% of PM/DM patients but not in cancer group.

22/135 (16%) with primary myositis and 15/27 patients with CAM (55%) died; in this second group deaths were in the first two years from diagnosis and in 13/15 patients were caused by cancer.

The survival rate of primary PM/DM at 5 years was significantly higher (87% and 74%) compared to the CAM patients, where survival at 2 years was 56% for PM and 44% for DM.

The risk of cancer is significantly higher in older female patients with DM. Some clinical and serological features can be considered predictive risk factors for malignancy, although without statistical significance: dysphagia, lower CK levels, lower prevalence of anti-ENA antibodies.

The overall survival rates were considerably worse in CAM; early discovery of malignancy is crucial and examinations for detections of an underlying malignancy are important in the management of patients with PM/DM.

P-11
Rehabilitation of patients with neuromuscular diseases in the region of the Bay of Kotor (Montenegro)

R. Ognjenovic, K. Tomcuk
Hospital "Vaso Cuckovic" Risan, Montenegro

The aim of this paper is to demonstrate the ability of professional teams from medical institutions in Bay of Kotor (Montenegro) to carry out the rehabilitation of patients with neuromuscular diseases in the optimal way.

A multi-year clinical course with adequate rehabilitation of two patients with neuromuscular disease is presented. In the first case, the course of the disease with rehabilitation of a boy diagnosed with Duchenne muscular dystrophy followed from his fifth to eleventh year of life is presented.

Another patient (with spinal muscular atrophy type 3) was followed for 3 years.

Muscular dystrophy, as a group of muscle diseases characterized by progressive weakness and atrophy of the muscles responsible for body movements, fortunately, is not a common disease in the region of the Bay of Kotor.

In first case an initial muscle weakness was noticed at age 3. Diagnostics(high levels of serum creatine phosphokinase,genetical testing which confirmed the deletion of dystrophin gene,EMG, biopsy, and a confirmation that the mother is the carrier) was made in his fifth year.

Abnormality was noticed during running, jumping and standing up (with Gowers maneuver). Other development of weakness in lower extremities, joint contractures, scoliosis. Until the age of 10 the patient had been sent out annually to a stationary physical therapy professionally led with education of the mother. He also took adequate doses of prednisolone intermittently. Since year 11 he was wheelchair-bound.

A patient with Kugelberg Welander spinal muscular atrophy (SMA) type 3 noticed the first symptoms (fatigue of gaiter muscles during walking) in age 12. After the diagnostics the only possible treatment was started -physical therapy (in hospital and at home).He still walks independently. Other neuromuscular diseases are sporadic and very rarely seen in our region.

In neuromuscular diseases maximizing the use of rehabilitation procedures with all medical supplies is necessary, so that the patient would be able to move independently for longer.

To the satisfaction of professional teams (pediatricians, neurologists,physiatrists, orthopedists, physical therapists and medical technicians) in Montenegro, rehabilitation of patients with neuromuscular diseases in recent years is being organized better than before, despite bad socio-economic conditions. It has implemented in the special institutions located on the coast.