Conclusion: During an average observation period of four years, Aminaphte showed a good tolerability and safety profile along with sustained efficacy in patients with SSC-related RP, without disabling or serious side effects. A randomized controlled trial for Aminaphte use in the management of SSC-related RP is desirable to better assess the clinical efficacy of the drug over time.

REFERENCES:

Disclosure of Interests: ANDREA CERE: None declared, Emanuele Gotelli: None declared, Adriano Lercara: None declared, Carmen Pizzorni: None declared, Sabrina Paolino: None declared, Elisa Alessandri: None declared, Maurizio Cutolo Grant/research support from: Bristol Myers Squibb, Celgene, Pfizer, Boehringer Ingelheim, Alberto Sulli: None declared


AB0741 LUNG FUNCTIONS IN IDIOPATHIC INFLAMMATORY MYOPATHIES PATIENTS WITH INTERSTITIAL LUNG DISEASE DURING > 3 YEAR OF FOLLOW-UP.

A. Khelkovaia-Sergeeva1, L. Garzanova1, M. Starovoytova1, O. Desinova1, L. P. Ananyeva1, O. Koneva1, V.A. Nasonova Research Institute of Rheumatology, laboratory of systemic sclerosis, Moscow, Russian Federation

Background: Interstitial lung disease (ILD) is the most common internal organ manifestation of idiopathic inflammatory myopathies (IIM) that can severely affect the course of the disease.

Objectives: To describe lung functions in IIM patients with ILD during > 3 year of follow-up.

Methods: Our prospective study included 44 pts with IIM fulfilling Bohan and Peter criteria and having ILD. The mean follow-up period was 46.2±11.2 months. The mean age was 52.3±11.7 years, 34 (73%) pts were female. The median disease duration was 1.125 [0.16-18] years, 24 (54%) of pts were positive for a-Jo-1 antibody. All pts had the standard examination including a-Jo-1 antibodies (a-Jo-1) assay; forced vital capacity (FVC) and carbon monoxide diffusion capacity (DLCO) evaluation as well as high-resolution computed tomography (HRCT) scanning of the chest were performed at baseline, and 36 and more months.

Results: 61% with anti-synthetase syndrome, 17% dermatomyositis (DM), 5% amyopathic DM, 15% with overlap myositis and 2% with necrotizing myopathy were included. 70% patients had nonspecific interstitial pneumonia, 23% organizing pneumonia (OP) and 7% OP; transformed to diffuse alveolar damage. All pts received prednisone at a dose of 11.8±4.1 mg/day, immunosuppressants at inclusion received 75% pts: cyclophosphamide 54%, mycophenolate mofetil 16.9% and combination 4.5%; Ritusamim was administered in case of intolerance or inadequate response to GC and other immunosuppressive drugs in 37 pts (84%) (6-8 courses). 7 patients died, ILD progression was the cause of death in 4 cases.

Conclusion: Active immunosuppressive therapy improves lung function in IIM patients. However, some forms of ILD may have a fatal course.

Disclosure of Interests: None declared


AB0742 THE DYNAMICS OF LUNG FUNCTION IN PATIENTS WITH MIXED CONNECTIVE TISSUE DISEASE ASSOCIATED WITH INTERSTITIAL LUNG DISEASE DURING A THREE-YEAR FOLLOW-UP PERIOD

R. Shayakhmetova1, L. P. Ananyeva1, O. Koneva1, M. Starovoytova1, O. Desinova1, O. Ovsyannikova1, L. Garzanova1, A. Khelkovaia-Sergeeva1, V.A. Nasonova Research Institute of Rheumatology, laboratory of systemic sclerosis, Moscow, Russian Federation

Background: Nowadays ILD is one of the most common manifestations of systemic rheumatic diseases and one of the leading cause of death of IIM patients.

Objectives: To assess the course and outcomes of interstitial lung disease (ILD) in mixed connective tissue disease (MCTD) based on lung function after three years of follow-up.

Methods: The study included 20 patients with MCTD associated with ILD. The diagnosis was set according to Kasukawa criteria (1987). There were 19 (95%) women and 1 (5%) men; average age was 44.73±15.26 years; mean disease duration was 13.7±17.1 years. ILD was confirmed by chest HRCT. () were excluded. GCs, and immunosuppressants (IS): the average dose of prednisone was 10.2±5.5mg. To identify and assess the dynamics of ILD, there was used high-resolution computed tomography (HRCT). Their lung function was assessed by data obtained due to spirometry (in particular, forced vital capacity, FVC) and diffusing capacity of the lungs (DCL); Doppler echocardiography was also conducted.

Results: In the group of patients with MCTD there was a chronic course of the disease with a gradual addition of symptoms, musculoskeletal pathology prevailed. After three-year follow-up period there was a decrease in clinical manifestations (arthritis, myositis, proximal muscle weakness). Functional capacities of the lungs and heart did not deteriorate. DCL remained stable 67±5.15 and 66.9±13.8, respectively. Systolic pulmonary artery pressure also remained stable (31.4±5.5 and 32.8±5.2, respectively), indirectly indicating the absence of progression in pulmonary involvement. The increase of DLCO in some patients. The reduction in dose of GCs was small but statistically significant (8.6±4.58 and 7.8±6.33, respectively, p=0.007). There was a trend towards a decrease in dose of prednisone (from 10.6±5 to 8.4±5, respectively). However, these data were not clinically significant.

Disclosure of Interests: None declared