SPECIFICITY OF ANTI-TRNA SYNTHETASE AUTOANTIBODIES CORRELATED WITH CLINICAL COURSE AND PROGNOSIS OF MYOSITIS-ASSOCIATED INTERSTITIAL LUNG DISEASE

Vasushi Kondo, Yoko Nakagome, Kazuki Hirano, Yasushi Koyama, Sho Sasaki, Katayoshi Kuriyabashi, Yuto Izumi, Chihyo Yamada, Shinji Sato. Tokai University School of Medicine, Division of Rheumatology, Department of Internal Medicine, Isehara-shi, Japan

Background: Interstitial lung disease (ILD) is associated with mortality among patients with idiopathic inflammatory myopathy (IMI). Anti-aminocarboxyl-RNA synthetase (anti-ARS) autoantibodies have been identified highly specific for IMI and coincidence of ILD. It has been identified that non-Jo1 positive anti-ARS patients may have decreased survival rate. However, clonal characteristics and prognosis in myositis-associated ILD with an individual anti-ARS autoantibody have not been clarified.

Objectives: To compare the characteristics and cumulative survival of myositis-associated ILD with different positivity of anti-ARS antibodies.

Methods: We investigated retrospectively all consecutive patients with myositis-associated ILD evaluated from 1999-2017. All autoantibodies were screened by using RNA and protein immunoprecipitation assays. Demographic data, laboratory findings and chest computed tomography (CT) images were obtained.

Results: Among 105 patients with myositis-associated ILD, enrolled, 47 had anti-Jo1 antibody and 58 had non-Jo1 antibody: anti-EJ (n=20), anti-PL12 (n=12), anti-PL7 (n=11), anti-KS (n=8) and anti-OJ (n=7). The diagnosis at first visit were polymyositis (PM; n=28), dermatomyositis (DM; n=42), clinically amyopathic dermatomyositis (CADM; n=3) and antisynthetase syndrome (ASS; n=31). Most common causes of death in this study were exacerbation of ILD (n=10), 71.4%. Anti-Jo1 positive patients had better ILD prognosis than non-Jo1 patients (log rank test, p = 0.03). The prevalence of rapid progressive ILD (RP-ILD) and upper lung field lesions in CT scans were significantly higher in anti-Jo1 positive patients (p=0.02 and p=0.05, respectively). Although the prognosis of ILD was not significantly different among 6 anti-ARS antibodies, anti-EJ positive patients had poor ILD prognosis than non-Jo1 patients (log rank test, p = 0.03). The prevalence of rapid progressive ILD (RP-ILD) and upper lung field lesions in CT scans were significantly higher in anti-EJ positive patients (p=0.017 and p=0.005, respectively). The diagnosis at first visit were polymyositis (PM; n=28), dermatomyositis (DM; n=42), clinically amyopathic dermatomyositis (CADM; n=3) and antisynthetase syndrome (ASS; n=31). Most common causes of death in this study were exacerbation of ILD (n=10), 71.4%. Anti-Jo1 positive patients had better ILD prognosis than non-Jo1 patients (log rank test, p = 0.03). The prevalence of rapid progressive ILD (RP-ILD) and upper lung field lesions in CT scans were significantly higher in anti-Jo1 positive patients (p=0.02 and p=0.05, respectively). Although the prognosis of ILD was not significantly different among 6 anti-ARS antibodies, anti-EJ positive patients had poor ILD prognosis than non-Jo1 patients (log rank test, p = 0.03). The prevalence of rapid progressive ILD (RP-ILD) and upper lung field lesions in CT scans were significantly higher in anti-Jo1 positive patients (p=0.017 and p=0.005, respectively).

Conclusion: Our data confirms that the characteristics and outcomes of myositis-associated ILD are distinct by anti-ARS autoantibodies and highlighted differences of characteristics and prognosis in myositis-associated ILD with individual anti-ARS autoantibody have not been clarified.

disclosure of Interests: None declared, Etsuko Iwata and Masui for their assistance.

Acknowledgement: The authors would particularly like to thank Etsuko Iwata and Masui for their assistance.

Disclosure of Interests: None declared


THE EFFECTS OF BOSENTAN FOR TREATMENT OF DIGITAL ULCER IN KOREAN PATIENTS WITH SYSTEMIC SCLEROSIS: PROSPECTIVE, MULTICENTER, OPEN-LABEL TRIAL

Kyungh Ann Lee1, Hyun-Sook Kim2, Bo Young Kim2, Yun Sung Kim2, Sung Jae Choi1, Jung Ran Choi1, 1The Soochunhyang University Seoul Hospital, Internal Medicine, Seoul, Korea, Rep. of (South Korea); 2Gangneung Asan Hospital, Internal Medicine, Gangneung, Korea, Rep. of (South Korea); 3Chosun University Hospital, Internal Medicine, Gwangju, Korea, Rep. of (South Korea); 4Korea University College of Medicine, Internal Medicine, Seoul, Korea, Rep. of (South Korea); 5Pohang St. Mary’s Hospital, Internal Medicine, Pohang, Korea, Rep. of (South Korea)

Background: Bosentan is an orally administered dual endothelin1 (ET-1) receptor antagonist approved in the EU for reducing the number of new digital ulcers in patients with systemic sclerosis (SSc) and ongoing digital ulcer (DU). Oral bosentan therapy was beneficial and generally well tolerated in patients with DU associated with SSc. A total of 3 RCTs were identified. The first two were high quality (RAPIDS-1 and RAPIDS-2) with 122 and 188 patients and treatment durations of 16 and 24 weeks, respectively. Both studies showed that, compared with placebo, bosentan significantly reduced the number of new ulcers; however, it did not have an effect on the healing of preexisting ulcers and pain.

Objectives: This study evaluated the efficacy and safety of bosentan in DU secondary to SSc in Korean patients.

Methods: From December 2013 to December 2017, a prospective, multicenter, open-label trial was performed to investigate the complete healing of DU and tolerability of bosentan in Korean patients with SSc. The SSc was described in a single-center cohort. Moreover, an association of anti-RNAP3 with breast cancer, particularly when synchronous with SSc onset, was also demonstrated.

Disclosure of Interests: Maria Grazia Lazzaroni: None declared, Mariam Ciavai: None declared, Eugenia Bertoldo: None declared, Franco Franceschini: None declared, Angela Tincani: Consultant for: UCB, Pfizer, Abbvie, BMS, Sanofi, Roche, GSK, AlphaSigma, Lilly, Janssen, Cellgene, Novartis, Paolo Airò: None declared